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Chiasmal syndrome

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ABSTRACT

BACKGROUND

Chiasmal syndrome is the constellation of signs and symptoms that are associated with lesions of the optic chiasm. Pituitary adenoma is its single most common cause.

METHODS

A MEDLINE search of the biomedical literature was conducted to uncover papers on topics related to chiasmal syndrome published since 1989.

RESULTS

The MEDLINE search retrieved a total of 163 citations. Each citation was screened for relevance. Copies of each relevant paper were obtained and their references were examined for any publications that the original search may have missed and for older publications that continue to be of importance.

CONCLUSIONS

This review article discusses the etiology, diagnosis, and management of chiasmal syndrome.

KEY WORDS

chiasmal syndrome, optic chiasm, pituitary adenoma, bitemporal hemianopsia

Trevino R. Chiasmal syndrome. J Am Optom Assoc 1995; 66:559-75.

Chiasmal syndrome is the constellation of signs and symptoms that are associated with lesions of the optic chiasm. The most common cause of chiasmal syndrome is tumor. One-fourth of all brain tumors occur in the chiasmal region and many of these lesions will produce visual symptoms.[1] Pituitary adenomas constitute more than half of the tumors responsible for chiasmal syndrome and are the single most common cause of this disease. Non-compressive causes of chiasmal syndrome—such as inflammation and ischemia—are quite rare. This paper will review the etiology, diagnosis and management of chiasmal syndrome.

There has been a major shift in the prevalence of visual symptoms in patients presenting with chiasmal syndrome over the past several decades. Classic papers on chiasmal disease emphasized vision loss as a presenting symptom.[1] More recently, however, endocrine dysfunction, specifically the hyperprolactinemia syndrome, has become the most common presenting finding.[2] This change is attributed to the development of laboratory assays for the pituitary hormone prolactin (PRL)—PRL was discovered as a human hormone in 1971—and to advances in radiologic imaging techniques that have permitted the detection of microadenomas (tumors less than 10 mm in diameter) that were not detectable on plain skull x-rays. Before the development of radioimmunoassays and the neuroimaging techniques of magnetic resonance imaging (MRI) and computed tomography (CT), patients with prolactin-secreting microadenomas would develop the typical clinical syndromes of gonadal dysfunction and galactorrhea/gynecomastia, but their causes were not understood. Only after a tumor had grown large enough to become visible on plain skull x-rays would it be discovered, and at that point it was often producing vision loss. The prolactin radioimmunoassay and CT scanner have made the diagnosis of many pituitary adenomas possible at a very early stage, long before the development of vision loss. Today, fewer than 10 percent of patients presenting with pituitary adenoma have visual field defects.[2]

Clinical manifestations of chiasmal syndrome

The hallmark of chiasmal syndrome is a bitemporal pattern of vision loss. Slow-growing neoplasms produce a gradual painless loss of vision that may go unnoticed by the patient for months or years. Vision loss is often, but not always, accompanied by headache. Indeed, it is often the headache, not the vision loss, which leads the patient to seek care. The subtlety of the early manifestations of chiasmal syndrome often results in a protract-

ed delay in discovering the lesion.[1] Diagnosis is further confounded in some patients with advanced bitemporal defects by the presence of peculiar sensory phenomena that may mimic binocular vision disorders.[4] These difficulties contribute to making the delayed diagnosis of chiasmal syndrome a relatively frequent cause of malpractice actions.[5]

Symptoms of chiasmal syndrome

The most common symptoms of chiasmal syndrome are decreased acuity, headache, diplopia, and loss of stereopsis (Table 1).

Vision loss

Chiasmal compression by a mass lesion will cause slowly progressive loss of visual function. Not infrequently, however, fluctuations in vision may produce symptoms of acute vision loss.[6] Asymmetry of vision loss is common, and one eye may show advanced deficits, including acuity loss, while only temporal depression is found in the visual field of the fellow eye.[7] Because chiasmal disease is usually caused by large mass lesions that distort the entire chiasm and adjacent optic nerves, depression of central acuity in one or both eyes is the rule; a pure bitemporal hemianopsia with intact acuity is rare.[8]

Signs and symptoms of Chiasmal Syndrome

symptoms	signs
vision Loss	visual field defects
headache	optic atrophy
diplopia	papilledema
loss of depth perception	ocular motor paresis
amenorrhea/impotence	see-saw nystagmus
galactorrhea/gynecomastia	CSF rhinorrhea

CSF= cerebrospinal fluid

Table 1

Visual symptoms are usually vague or nonexistent until acuity is affected. Decreased acuity, not the realization of a peripheral field constriction, prompts the patient to seek care. The unilateral presenting symptoms and inadequate visual field assessment are the chief causes of misdiagnosis of chiasmal lesions.[3,5,9]

Headache

Headache is a very common symptom in patients with pituitary adenoma and other intracranial tumors. Studies find that about 45 percent of patients with pituitary adenoma will report this symptom, and it is the presenting complaint in many of these patients.[2] The headaches are often localized to the brow or periorbital region. In rare cases, a severe headache associated with rapidly progressive bilateral vision loss and diplopia may signal the rapid enlargement of a pituitary tumor, known as apoplexy, a major neurosurgical emergency.[10] Because headache is such a common symptom of chiasmal syndrome, it has been suggested that a careful visual field examination be included as part of the work-up of all headache patients.[2]

Diplopia

Chiasmal syndrome is associated with both paretic and nonparetic forms of diplopia. Palsies of the third, fourth, and sixth cranial nerve may occur as a result of lateral extension of a tumor into the cavernous sinus. Nonparetic diplopia results from the "hemifield slide phenomena" in patients with a complete bitemporal hemianopsia or large, dense bitemporal scotomas.[4] In such patients, there is loss of fusion between the two remaining nasal hemifields. Vertical and horizontal slippage of the hemifields will produce symptoms of intermittent diplopia or of vertical steps in horizontal lines.

Loss of depth perception

Complete bitemporal hemianopsia can produce alterations in depth perception due to "post-fixational blindness."[4] Common symptoms include difficulty with precision tasks demanding anteroposterior orientation such as cutting finger nails, threading needles, and operating precision tools. In these patients, convergence results in the crossing of the two blind temporal hemifields, producing a completely blind triangular area of the binocular visual field that has its apex at the point of fixation.[4,11] Objects beyond the fixation point fall on nonfunctioning nasal retina and thus disappear.

Endocrine dysfunction

The most common presenting complaints in patients with pituitary tumors are related to metabolic changes caused by excess secretion of hormone by the tumor.[2] The most frequently cited symptoms are amenorrhea, impotence, galactorrhea, and gyneco-

ed delay in discovering the lesion.[3] Diagnosis is further confounded in some patients with advanced bitemporal defects by the presence of peculiar sensory phenomena that may mimic binocular vision disorders.[4] These difficulties contribute to making the delayed diagnosis of chiasmal syndrome a relatively frequent cause of malpractice actions.[5]

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mastia. Less common are the development of acromegalic or Cushingoid features.

Signs of chiasmal syndrome

Signs of chiasmal syndrome include visual field loss, optic atrophy and papilledema, oculmotor nerve pareses, and nystagmus (Table 1).

Visual field defects

Chiasmal lesions produce characteristic visual field defects. The localizing value of these defects makes perimetry a key test in the clinical diagnosis of chiasmal syndrome. Normal anatomical variation in the location of the chiasm over the sella will affect whether a pituitary tumor produces a junctional, bitemporal, or homonymous hemianopic field defect.

The average vertical distance between the optic chiasm and the pituitary fossa is 10 mm (Fig. 1). Therefore, a pituitary tumor must have a substantial suprasellar extension before it produces visual loss. This requirement is why small pituitary tumors, so-called microadenomas, never produce visual field defects. The optic chiasm is located directly over the pituitary gland in 80 percent of normal individuals. It is anterior to the pituitary ("prefixed") in 9 percent of the population, and posteriorly displaced ("postfixed") in 11 percent.[12] Pituitary tumors will tend to produce optic tract lesions in prefixed chiasms and optic nerve involvement in postfixed chiasms. The inferior nasal fibers are the first to decussate at the chiasm. A few of these fibers will loop anteriorly into the terminal portion of the contralateral optic nerve (Wilbrand's knee) before turning posteriorly to continue through the optic chiasm and into the optic tract. Lesions of the distal optic nerve can therefore produce a superior temporal visual field defect in the contralateral eye.[13]

Perichiasmal lesions produce four major patterns of vision loss: unilateral central scotoma, junctional scotoma, bitemporal hemianopsia, and incongruous homonymous hemianopsia. The central scotoma is the most common perimetric finding in prechiasmal compressive lesions.[8] Pituitary adenomas may produce optic nerve compression when the optic nerve is postfixed. Lesions at the junction of the optic nerve and chiasm can produce an ipsilateral central scotoma and a contralateral supratemporal visual field depression—the so-called junctional scotoma (Figs. 2 and 3). Bitemporal hemianopsia is the hallmark of chiasmal disease and represents the most common visual abnormality in patients with pituitary adenomas[14] (Figs. 4 and 5). The defects may be peripheral, central, or a combination of both with or without "splitting" of the macula. The defects may be absolute or relative, and when relative they may only be identifiable with quantitative perimetry. Many patients with bitemporal field defects have normal visual acuity. Loss of visual acuity in one or both eyes is usually, but not always, associated with other visual deficits, such as loss of color vision. With pituitary adenomas, vision loss usually occurs first in the superior temporal quadrants, and suprachiasmal lesions will usually, but not always, produce inferior visual field defects. Pituitary adenomas in patients with a prefixed optic chiasm may produce an incongruous homonymous hemianopsia due to compression of the optic tract.

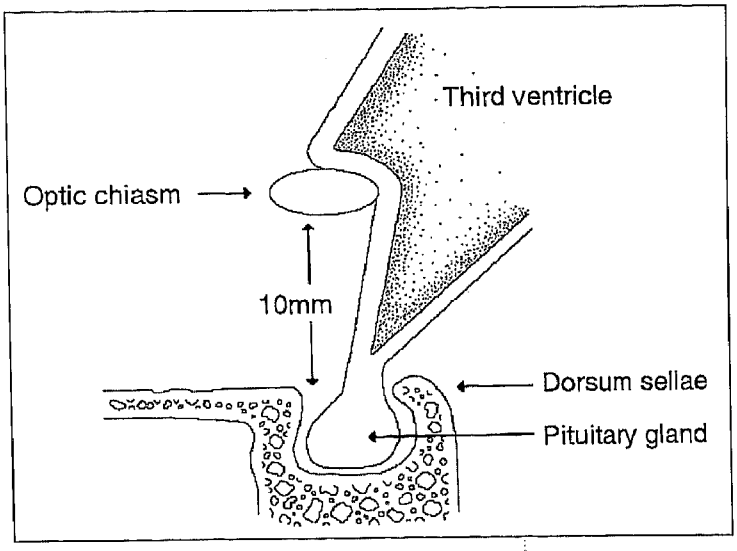

Figure 1

A sagittal view of the optic chiasm, pituitary gland, and adjacent structures.

Although bitemporal hemianopsia is the hallmark of chiasmal syndrome, there are a few conditions unrelated to chiasmal disease that can produce a bitemporal field defect. Such "pseudo-bitemporal hemianopsias" are the result of bilateral retinal or optic nerve disease (Table 2). These conditions do not generally pose a very great diagnostic challenge since the bitemporal scotomas will usually only vaguely resemble a true bitemporal hemianopsia—for instance, they rarely respect the midline—and their source is usually readily identifiable on ophthalmoscopy (Figs. 6, 7, and 8).

Optic atrophy and papilledema

Lesions of the optic nerves, optic chiasm, or optic tract will eventually produce optic atrophy. Chiasmal lesions producing a bitemporal hemianopsia would be expected to produce atrophy of nerve fibers from

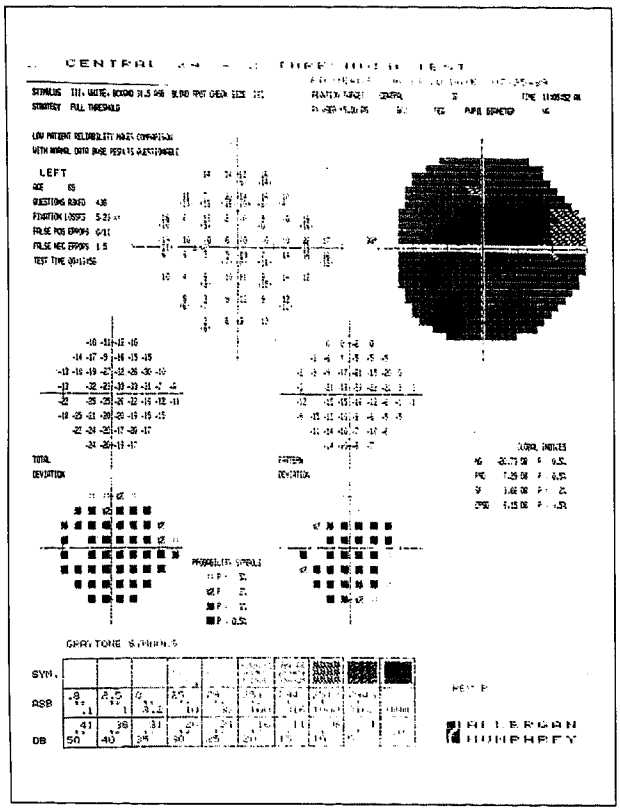

Figure 2

Junctional scotoma, left eye. This 69-year-old man presented with vision loss in his left eye of 4 weeks' duration. Vision was 20/25 OD and finger counting OS. An afferent pupillary defect was present on the left side. CT scan revealed a pituitary adenoma.

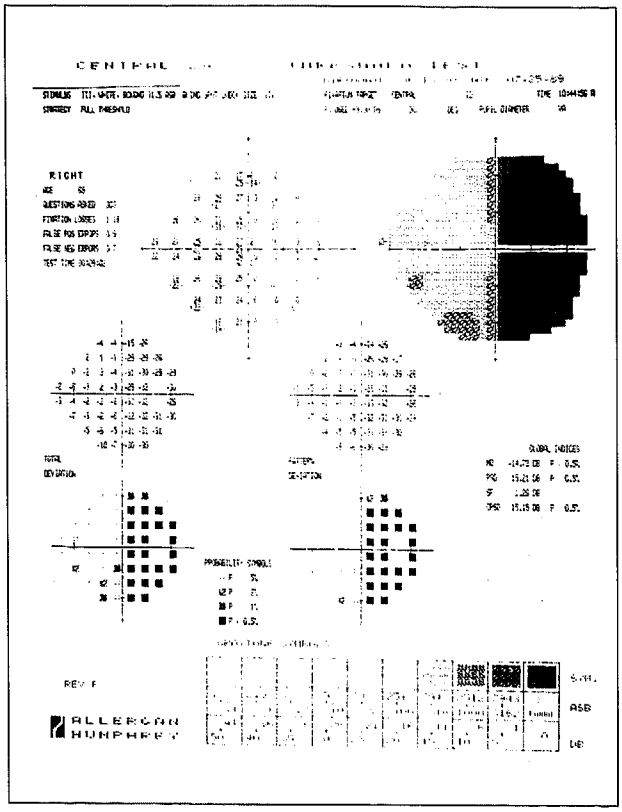

Figure 3

Junctional scotoma, right eye, of the patient described in Figure 2.

ganglion cells located nasal to the fovea in each eye, producing a horizontal band of atrophy in each eye (so-called "bow tie" atrophy).[15] In practice, however, disc pallor is usually mild or absent in patients with pituitary adenoma.[7,14] Furthermore, pituitary adenomas are almost never associated with papilledema.[11] On the other hand, large suprasellar tumors, especially meningiomas[16] and craniopharyngiomas,[11] will often cause papilledema.

Oculomotor pareses

Oculomotor pareses are relatively rare, found in no more than 5 percent of patients with pituitary adenoma.[1,2,17] They are almost always associated with other manifestations of the tumor, but can occur in isolation.[18] Palsies of the third, fourth, and sixth cranial nerve may occur as a result of lateral extension of a tumor into the cavernous sinus. The oculomotor nerve is most often affected. The paresis is rarely complete, and the pupil may or may not be affected.

See-saw nystagmus

Patients with suprasellar tumors, such as craniopharyngiomas, may develop this rare form of dissociated nystagmus due to compression of the midbrain with damage to the interstitial nucleus of Cajal or its connections. See-saw nystagmus is characterized by synchronous, alternating elevation and intorsion of one eye and depression and extorsion of the fellow eye.[19-21]

Cerebrospinal fluid rhinorrhea

Pituitary adenomas that extend down into the sphenoid sinus may produce discharge of cerebrospinal fluid (CSF) from the nose.[11] The amount of fluid that is discharged varies considerably. It may be intermittent and is sometimes influenced by the posture of the head, occurring, for instance, only when the head is bent forward. Affected patients may also experience other nasal symptoms such as nasal obstruction, epistaxis, and blood-stained sputum.[11]

Differential diagnosis of chiasmal syndrome

The major cause of chiasmal disease is tumor, the most common being a pituitary adenoma. Other common tumors of the chiasmal region include gliomas, meningiomas, and craniopharyngiomas. Compression of the optic chiasm may also be caused by aneurysm of adjacent blood vessels and by mucoceles or abscesses caused by intracranial infection. Noncompressive disease of the optic chiasm is relatively rare, and may be classified as being ischemic or nonischemic in nature. The differential diagnosis of chiasmal disease is summarized in Table 3. Several specific causes of chiasmal syndrome are discussed below.

Pituitary adenoma

Pituitary adenomas are benign, slow-growing tumors. Their prevalence has been placed at 14.7 cases per 100,000 population per year.[22] They account for about 10 to 25 percent of all intracranial tumors that present clinically, and have been found in up to 27 percent of adults in unselected autopsies.[23] Pituitary adenomas most frequently appear in patients 30 to 45 years of age.[24] There are no apparent significant racial or gender differences in their prevalence.

The traditional cellular classification of pituitary adenoma (e.g., chromophobic, basophilic) has given way to a functional system based on the hormone the tumor is secreting. Approximately 75 percent of pituitary adenomas are secretory and 25 percent are nonfunctioning tumors that do not secrete hormones.[23] The most common secretary adenoma is the prolactin-secreting adenoma (also called prolactinoma), which accounts for at least 25 percent of cases. Adenomas that release growth hormone (GH), gonadotropic hormones, or adrenocorticotropic hormone (ACTH) each comprise another 10 percent of adenomas. Tumors secreting multiple hormones make up another 10 percent of cases. Adenomas that secrete thryoid-stimulating hormone (TSH) are rare, making up less than 1 percent of all cases of pituitary adenoma. The hormone being secreted, if any, will determine the signs and symptoms that the tumor will produce.

Because nonsecreting adenomas usually produce only nonspecific symptoms such as headache prior to the development of vision disturbances, they are often not detected until they have grown large enough to produce chiasmal disease. In fact, nonsecreting tumors are more frequently associated with visual field defects than any other type of pituitary adenoma.[2] Large adenomas (both secretory and nonsecretory) may cause partial or complete hypopituitarism by compression of adjacent normal gland or interruption of the pituitary stalk that connects the gland with the hypothalamus.[25] Manifestations of hypopituitarism depend on the specific hormones that are lacking, but common findings include signs and symptoms of gonadotropin deficiency (amenorrhea, impotence); hypothyroidism (fatigue, cold intolerance); ACTH deficiency (fatigue, weight loss); and vasopressin deficiency (diabetes insipidus).

Prolactin-secreting adenomas

Prolactin-secreting adenomas produce the hyperprolactinemia syndrome. In women, this syndrome takes the form of galactorrhea, or the production and release of breast milk. The discharge may be copious, or expressible only manually. Often galactorrhea is accompanied by amenorrhea, arrestation of the menstrual cycle. Most women with the galactorrhea-amenorrhea syndrome do not have neurologic or visual symptoms or signs because the tumor is still relatively small when discovered.[26] In men, hyperprolactinemia leads to impotence, decreased libido, and infertility. Other findings include gynecomastia (breast enlargement), obesity, and hypogonadism. Men who develop these problems may wait a considerable time before seeking medical help.[25] By then the tumor has often grown large enough to produce headache, vision loss, or even cerebrospinal fluid rhinorrhea. Men with prolactinomas thus tend to have more severe clinical manifestations than women.

Growth hormone adenoma

Acromegaly and gigantism are the result of growth hormone overproduction. The classic features of acromegaly include marked enlargement of the hands and feet with thickening of the overlying skin, and coarsening of facial features characterized by enlargement of the mandible and abnormally prominent supraorbital rims.[25] These changes take place so gradually that gross distortion of appearance may occur before the condition is recognized. If the epiphyses of the long bones have not yet closed, the patient will develop a marked increase in the length of the bones, resulting in gigantism. Not infrequently, pituitary tumors that secrete growth hormone secrete other hormones as well, such as prolactin or thyroid-stimulating hormone.

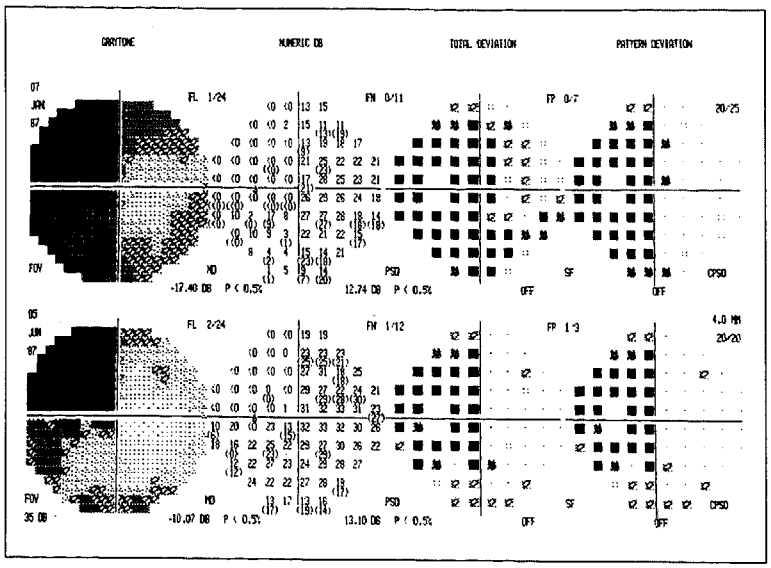

Figure 4

Bitemporal hemianopsia, left eye, in a 45-year-old fork lift driver who presented with complaints of loss of depth perception and transient vertical diplopia of gradual onset over a year. The top visual field was performed at the time of diagnosis. The bottom visual field reveals some visual recovery at 3 months following successful transsphenoidal surgery for a pituitary adenoma.

of affected women.[27] These tumors are often very large at the time of diagnosis. About half of affected individuals will have visual field defects.[27] There are two instances in which adenomas may be associated with hypothyroidism. First, large adenomas that are not secreting TSH may destroy sufficient normal pituitary tissue to produce a secondary hypothyroidism. Also, chronic primary hypothyroidism can produce a "reactive enlargement" of the pituitary gland, due to loss of the negative feedback mechanism in which hormones released by the thyroid gland suppress TSH secretion by the pituitary. Longstanding, untreated hypothyroidism is therefore a rare cause of chiasmal disease.[28]

ACTH-secreting adenoma

Cushing's disease (also known as pituitary-dependent Cushing's syndrome) develops in patients with ACTH-secreting pituitary adenomas.[23] Many other conditions will also result in ACTH (cortisol) excess, such as tumors of the adrenal cortex, and all will produce the characteristic clinical features of Cushing's syndrome. Typical findings include truncal obesity, with wasting of the limbs and moon facies. These patients may also suffer from hypertension, backache due to osteoporosis, and mental disorders. These tumors rarely grow large enough to affect vision. In fact, in 60 percent of cases the distinctive features of Cushing's disease leads to recognition of the syndrome before the adenoma has had time to grow large enough to produce sellar abnormalities that are detectable on CT scanning.[23]

Thyroid-stimulating hormone adenoma

Pituitary adenomas may be associated with both hyperthyroidism and hypothyroidism.[11] Hyperthyroidism due to a TSH-producing pituitary tumor is uncommon, but is becoming increasingly recognized with the development of sensitive TSH immunoradiometric assays.[27] These patients develop all the usual clinical manifestations of hyperthyroidism, including a goiter. Ophthalmopathy rarely occurs in these patients. TSH tumors are frequently multihormonal, with acromegaly being present in 22 percent of cases and amenorrhea/galactorrhea present in 20 percent

Gonadotropic hormone adenoma

Adenomas that arise from the gonadotroph cells are among the most common adenomas of the pituitary, but they were not recognized until recently because they secrete inefficiently, and the secreted products — follicle-stimulating hormone and luteinizing hormone (FSH and LH) — do not cause a recognizable clinical syndrome.[29] Consequently, gonadotroph adenomas are usually not detected until they become large enough to produce vision loss or, less commonly, other neurologic symptoms. Unlike most other pituitary macroadenomas (tumors greater than 10 mm in diameter), gonadotroph macroadenomas do not produce a dramatic elevation in the serum concentrations of its hormonal products.[29] Macropro-lactinomas, for example, typically produce serum prolactin (PRL) concentrations 100 to 1,000 times normal. Gonadotroph macroadenomas, on the other hand, cause only up to a 10-fold elevation in the serum concentrations of its secretory products, and often they are not elevated at all. Ironically, many patients with gonadotroph adenomas actually develop LH deficiency due to compression of normal gonadotroph cells by the adenoma and lack of secretion of adequate amounts of LH by the tumor.[29] The result in men is decreased energy and libido and in women is amenorrhea. There are no known symptoms of excessive secretion of FSH or LH in men or women.[29]

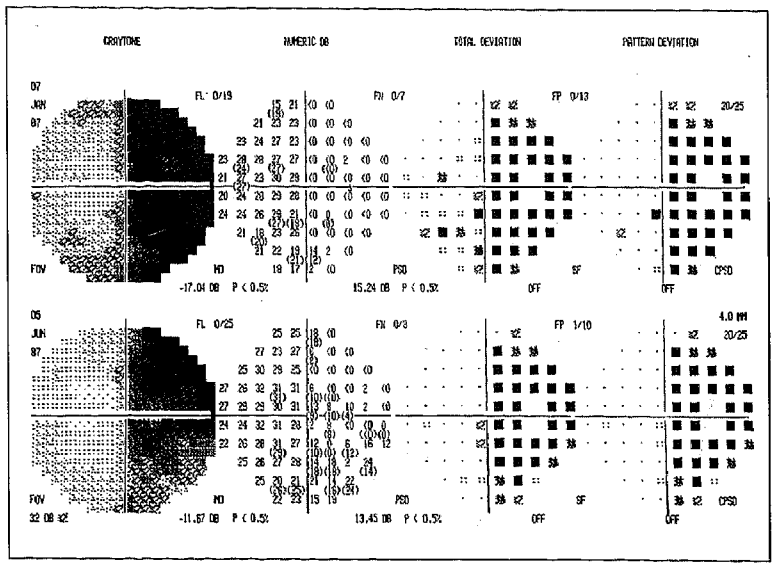

Figure 5

Bitemporal hemianopsia, right eye, of the patient described in Figure 4.

Pituitary apoplexy is the spontaneous, rapid expansion of a pituitary tumor due to either infarction of or hemorrhage into the tumor.[8] It is estimated from histological studies that about 18 percent of pituitary adenomas undergo acute or subacute changes.[30] Often the apoplexy is subclinical, but it is a potentially catastrophic event. In a typical case, a patient who may or may not be known to harbor a pituitary adenoma suddenly develops a severe headache.[11] It may be generalized or retrobulbar in location, and may be associated with neck stiffness or pain. Within a few hours the patient develops double vision due to unilateral or bilateral ophthalmoplegia. The third, fourth, or sixth nerves may be affected. Headache, double vision, and subarachnoid hemorrhage have been called the classic triad of pituitary apoplexy.[10] Many patients will experience progressive loss of vision that may be mild or severe. Total blindness can occur over a period of minutes, hours, or days. A progressive deterioration in level of consciousness leading to coma and death may ensue. Radiation therapy, trauma, and pregnancy have been implicated as precipitating pituitary apoplexy.[8] Corticosteroid replacement therapy is the first therapeutic measure for apoplexy.[8] Neurosurgical decompression is performed in patients with vision loss or mental impairment.

The diagnosis of pituitary adenoma requires neuroimaging studies and tests of endocrine function. CT scanning or magnetic resonance imaging will identify most pituitary adenomas regardless of their size. An enlarged sella turcica that contains an intrasellar mass with a suprasellar component is virtually pathognomonic of pituitary adenoma[11] (Figs. 9 and 10). The widespread availability and superb sensitivity of CT and MRI have made plain skull x-rays obsolete in the evaluation of patients with suspected pituitary adenomas.

The therapeutic options for patients with pituitary adenomas include observation without intervention, surgery, medication, and radiation therapy. Transsphenoidal microsurgical removal of pituitary tumors has become the surgical procedure of choice, and is very successful in restoring vision and normalizing pituitary function. The surgery may be followed by a course of radiation therapy to reduce the risk of recurrence. Radiotherapy is rarely used as a primary treatment,[31] except in cases where proton beam therapy or some other form of stereotactic radiosurgery is being performed.[32] Bromocriptine (Parlodel), an ergot derivative and dopamine agonist, is the medicine used most often in the treatment of pituitary adenomas. Although the exact mechanism of action is unknown, it inhibits prolactin secretion by the pituitary and will rapidly shrink prolactin-secreting adenomas.[33] Bromocriptine can produce improvement in visual symptoms in as little as 72 hours.[34] Bromocriptine may be used as the primary mode of therapy for micro- and macroprolactinomas.[26] It can be used before surgery for shrinkage of the tumor and as a postoperative therapy for patients with incomplete removal of the tumor and persistent elevation of serum prolactin levels. Bromocriptine's side effects are usually mild and consist of nausea, vomiting, faintness, and hypotension. Because bromocriptine is not a cure—it is "tumorstatic" rather than "tumorcidal"—patients must take this medication for life. If the patient discontinues the medication, as women do during pregnancy because of possible teratogenic effects, the size of the tumor may rapidly increase.[35] Many women with microprolactinomas have uneventful pregnancies; however, they should be monitored closely for signs of tumor enlargement. Watchful waiting may be appropriate for microadenomas that are asymptomatic or when present in women with amenorrhea who are not interested in becoming pregnant.[23]

Other tumors

Other tumors that are frequently associated with chiasmal syndrome are craniopharyngioma (20 to 25 percent of cases), meningioma (10 percent), and glioma (7 percent). These tumors are a much less common cause of chiasmal disease than pituitary adenomas (50 to 55 percent).

Craniopharyngioma

This tumor is the second most common cause of chiasmal syndrome and constitutes 3 percent of all intracranial tumors.[11] The tumor arises from nests of squamous epithelial cells that lie between the anterior and posterior lobes of the pituitary gland. These tumors are usually admixtures of solid cellular components and variable-sized cysts which may contain degenerated blood, necrotic tissue, calcified debris, and even fully developed teeth. Craniopharyngiomas occur most frequently in children under the age of 15.[24] There is a bimodal age incidence, with a large peak in the first decade and then a much smaller peak in the years 50 to 70. Craniopharyngiomas are always located, at least in part, in the suprasellar cistern, where they may compress the intracranial portion of the anterior visual system, the hypothalamus, and anterior communicating and intracranial portion of the internal carotid arteries, and occasionally the ventral brainstem. Compression of the third ventricle frequently results in obstructed flow of CSF and increased intracranial pressure. In children, initial symptoms are usually related to the tumor's effect on the hypothalamus and pituitary, and include hydrocephalus, retarded growth, disturbances in heat regulation, and diabetes insipidus. Vision loss is usually profound but not appreciated until other abnormalities become apparent.[11]

Differential diagnosis of Bitemporal Hemianopsia

More common	Chiasmal lesions
	Tilted optic discs
	Overhanging redundant upper lid tissue
Less common	Bilateral cecocentral scotomas
	Nasal sector retinitis pigmentosa
	Enlarged blind spots
	Bilateral medullation of nasal nerve fibers
	Chloroquine maculopathy
	Glaucoma

Table 2

Progressive loss of vision is often the first symptom of craniopharyngioma in adults. Although the visual symptoms and signs of disease are similar to those caused by other suprasellar lesions, a substantial number of patients present with a central scotoma rather than with bitemporal hemianopsia.[36] The clinician must therefore consider the possibility of a suprasellar lesion in any patient with optic neuropathy, even when a "classic" chiasmal field defect is absent. Vision loss is very slowly progressive over months to years; however, there may be fluctuations in visual function which can present clinically as acute loss of vision with spontaneous recovery, simulating optic neuritis. Acute onset of symptoms may also signal the development of a chemical arachnoiditis caused by the leakage of fluid from cysts within the tumor into the subarachnoid space.[11] Other symptoms of craniopharyngioma include dementia and changes related to the tumor's effect on the hypothalamus and pituitary, such as decreased libido, impotence, galactorrhea, amenorrhea, reduced energy, weight gain, and diabetes insipidus.

The diagnosis of craniopharyngioma is made on the basis of neuroimaging studies. The cardinal features of craniopharyngioma are areas of calcification and cyst formation.[37] The treatment of craniopharyngioma is almost always surgical, often followed by a course of radiation therapy. Options range from total excision to simple aspiration of cyst contents. Visual prognosis depends primarily on severity of vision loss at presentation and degree of manipulation of the optic nerves and chiasm at the time of surgery. Although the prognosis is not as good as that for pituitary adenoma or suprasellar meningioma, many patients will experience impressive recovery of vision following surgery.[36]

Meningiomas

Meningiomas in the region of the optic foramen, medial sphenoidal ridge and tuberculum sella can produce prechiasmal or chiasmal compression, as can large subfrontal meningiomas that extend posteriorly. Meningiomas most frequently occur in middle-aged females. The earliest and most consistent symptom of a suprasellar meningioma is unilateral or markedly asymmetric vision loss,[24] but nonspecific headaches are also common.[16] Vision loss is typi-

Other tumors

Other tumors that are frequently associated with chiasmal syndrome are craniopharyngioma (20 to 25 percent of cases), meningioma (10 percent), and glioma (7 percent). These tumors are a much less common cause of chiasmal disease than pituitary adenomas (50 to 55 percent).

Craniopharyngioma

This tumor is the second most common cause of chiasmal syndrome and constitutes 3 percent of all intracranial tumors.[11] The tumor arises from nests of squamous epithelial cells that lie between the anterior and posterior lobes of the pituitary gland. These tumors are usually admixtures of solid cellular components and variable-sized cysts which may contain degenerated blood, necrotic tissue, calcified debris, and even fully developed teeth. Craniopharyngiomas occur most frequently in children under the age of 15.[24] There is a bimodal age incidence, with a large peak in the first decade and then a much smaller peak in the years 50 to 70. Craniopharyngiomas are always located, at least in part, in the suprasellar cistern, where they may compress the intracranial portion of the anterior visual system, the hypothalamus, and anterior communicating and intracranial portion of the internal carotid arteries, and occasionally the ventral brainstem. Compression of the third ventricle frequently results in obstructed flow of CSF and increased intracranial pressure. In children, initial symptoms are usually related to the tumor's effect on the hypothalamus and pituitary, and include hydrocephalus, retarded growth, disturbances in heat regulation, and diabetes insipidus. Vision loss is usually profound but not appreciated until other abnormalities become apparent.[11]

Progressive loss of vision is often the first symptom of craniopharyngioma in adults. Although the visual symptoms and signs of disease are similar to those caused by other suprasellar lesions, a substantial number of patients present with a central scotoma rather than with bitemporal hemianopsia.[36] The clinician must therefore consider the possibility of a suprasellar lesion in any patient with optic neuropathy, even when a "classic" chiasmal field defect is absent. Vision loss is very slowly progressive over months to years; however, there may be fluctuations in visual function which can present clinically as acute loss of vision with spontaneous recovery, simulating optic neuritis. Acute onset of symptoms may also signal the development of a chemical arachnoiditis caused by the leakage of fluid from cysts within the tumor into the subarachnoid space.[11] Other symptoms of craniopharyngioma include dementia and changes related to the tumor's effect on the hypothalamus and pituitary, such as decreased libido, impotence, galactorrhea, amenorrhea, reduced energy, weight gain, and diabetes insipidus.

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Differential diagnosis of Bitemporal Hemianopsia

More common	Chiasmal lesions
	Tilted optic discs
	Overhanging redundant upper lid tissue
Less common	Bilateral cecocentral scotomas
	Nasal sector retinitis pigmentosa
	Enlarged blind spots
	Bilateral medullation of nasal nerve fibers
	Chloroquine maculopathy
	Glaucoma

Table 2

cally unilateral initially, but as the tumor slowly grows over a period of 1 or more years it will eventually produce chiasmal or contralateral optic nerve involvement.[38] The average duration of visual symptoms at the time of diagnosis is about 2 to 3 years.[16] Although meningiomas are slow growing tumors, fluctuations in vision can produce symptoms of acute vision loss with spontaneous recovery that mimic optic neuritis. Meningiomas can even produce symptoms of periocular pain made worse by eye movement.[16] As with prolactinomas, tumor growth may accelerate during pregnancy.[16] Ophthalmoscopy will reveal either normal optic discs (early diagnosis), disc pallor (late diagnosis with optic atrophy), or papilledema (increased intracranial pressure due to obstruction of CSF circulation). Diagnosis is made by neuroimaging. Although not pathognomonic, finding hyperostosis and blistering of surrounding bone on CT scan is a sensitive indicator of meningioma.[39] The preferred treatment is surgical removal. Visual outcome is highly dependent on duration of symptoms at the time of surgery.[39] Lengthy duration of acuity loss and severe visual deficit, however, do not preclude postoperative recovery of vision.[38]

Optic gliomas

Optic gliomas are primary astrocytic tumors that infiltrate the optic nerves, chiasm, hypothalamus, optic tract, and third ventricle.

Gliomas usually occur in children, with about 80 percent occurring before age 10 and over 90 percent occurring before age 20.[23] They are benign and often nonprogressive.[40] Chiasmal gliomas in children can be classified as anterior, affecting the optic nerves and chiasm (so-called optico-chiasmatic gliomas), or posterior, with involvement of the optic tracts or hypothalamus (so-called optico-hypothalamic gliomas). The former is associated with a more favorable prognosis, whereas the latter is more aggressive and less responsive to therapy.[41,42] The typical patient with a chiasmal glioma is 4 to 8 years old and presents for evaluation of poor vision or strabismus. Vision loss may be unilateral or bilateral. Chiasmal gliomas do not usually produce a bitemporal pattern of vision loss. Central scotomas occur in 70 percent of cases.[43] Associated findings may include headache, proptosis (with orbital involvement), optic atrophy, and nystagmus. With posterior tumors, diabetes insipidus or other hypothalamic signs and symptoms may be evident. Approximately 28 percent of patients with optic gliomas will have neurofibromatosis.[41] The diagnosis of optic glioma is usually made neuroradiologically. Treatment options include surgical resection, radiation, chemotherapy, and watchful waiting.

Differential diagnosis of Chiasmal Syndrome

compressive disease

tumor

pituitary adenoma • craniopharyngioma
meningioma • optic glioma
dysgerminoma

other causes of chiasmal compression

lymphocytic hypophysitis • aneurysms
metastatic diseases • mucocele
arachnoidal and epithelial cysts

noncompressive disease

ischemia

arteriosclerosis • radionecrosis
optochiasmal arachnoiditis • arteritis
pituitary apoplexy • chiasmal apoplexy
perichiasmatic hemorrhage
compression of vascular supply by adjacent tumors

nonischemic

inflammation
• multiple sclerosis • sarcoidosis • tuberculosis

toxins
• phenprazine (Catron) • muslin gauze
• ethchlorvynol (Placidyl)

trauma
• intracranial surgery • nonsurgical trauma

traction
• empty sella syndrome • hydrocephalus

congenital dysplasia
• anterior forebrain malformations

Table 3

There does not at this time appear to be a consensus among experts as to a preferred treatment for chiasmal gliomas.[44] Surgical resection of chiasmal gliomas neither improves vision nor prolongs life.[7] The role of radiation or chemotherapy as a primary mode of treatment is controversial.[31,45,46] Given the indolent nature of many optic gliomas, nonintervention is often the best treatment option.[40]

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Other causes of chiasmal compression

Rare causes of chiasmal compression are pituitary infiltration, vascular abnormalities, and sphenoethmoidal mucoceles.

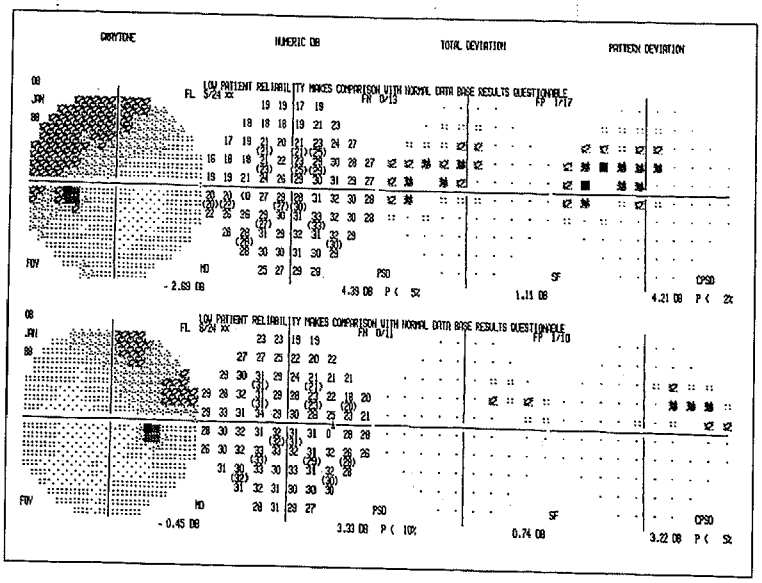


Figure 6

Pseudo-bitemporal hemianopsia due to tilted optic discs in a 49-year-old asymptomatic female. Perimetry reveals bilateral superior-temporal depression. Top: left eye. Bottom: right eye.

Lymphocytic hypophysitis

This disease is a rare, idiopathic, lymphocytic infiltration of the pituitary gland that usually occurs during pregnancy.[47,48] Lymphocytic infiltration of the pituitary results in depression of hormone secretion and swelling of the gland. Patients may present clinically with symptoms of hypopituitarism or vision loss. The condition may be treated with steroids or partial hypophysectomy. An autoimmune etiology is suspected.

Aneurysms

Aneurysms of the intracranial portion of the internal carotid,[49,50] the origins of the middle or anterior cerebral,[51] and of the anterior communicating arteries[52] can produce vision loss as the result of compressing the optic nerves and chiasm. Large aneurysms in this region can mimic a suprasellar neoplasm,[53] including hyperprolactinemia due to compression of the pituitary stalk.[50] Damage to the pituitary stalk, which connects the gland to the hypothalamus, results in hyperprolactinemia because of interruption of the normal inhibitory influence exercised by the hypothalamus on secretion of prolactin from the pituitary. In some patients vision loss represents the only neurologic complication of the aneurysm.[50] Diagnosis is made by MRI and cerebral arteriography. The traditional treatment is carotid ligation in the neck. Sclerotic anterior cerebral[54] and intracranial internal carotid[55] arteries may also act as mass lesions that compress the optic nerve and chiasm, but rarely give rise to a bitemporal hemianopsia.

Mucocele

Mucocele of the posterior ethmoid and sphenoid sinuses is characterized by chronic headache and dysfunction of one or more of the cranial nerves that pass through the orbital apex.[56] Involvement of one or both optic nerves is more common than chiasmal involvement. Diagnosis is made on the basis of radiologic findings. Treatment is endonasal decompression.

Noncompressive causes of chiasmal disease

The vast majority of chiasmal disease is caused by neoplastic lesions. There are, however, several nontumoral causes of chiasmal syndrome (Table 3). Most of these nonneoplastic conditions present clinically with an acute onset of symptoms that is distinctly different from the slowly progressive vision loss associated with tumor growth.

Ischemia of the optic chiasm can produce abrupt onset of bitemporal visual loss. Specific etiologies of ischemic chiasmal syndrome (ICS) include:

- vascular insufficiency secondary to atherosclerotic plaque formation[57]
- optochiasmal arachnoiditis[58]
- arteritis due to various disease processes,[59] especially giant-cell arteritis
- pituitary apoplexy[10]
- chiasmal radionecrosis following radiation therapy[31]
- chiasmal apoplexy[60]—hemorrhage within the chiasm itself
- compression of chiasmal vascular supply by adjacent tumors[61]
- perichiasmatic hemorrhage from intrasellar and anterior communicating artery aneurysms.[62]

Nonischemic noncompressive causes of chiasmal disease include inflammation, toxins, trauma, and traction. Multiple sclerosis is a common cause of optic nerve disease, and less commonly will produce chiasmal visual field defects.[63] Sarcoidosis can manifest itself as a disease of the central nervous system capable of producing granulomas anywhere within the brain, including the optic nerves and chiasm.[64] In fact, neurosarcoidosis is one of the chief nontumoral causes of chiasmal syndrome.[7] The toxic effects of various substances have been implicated as a rare cause of bitemporal visual loss, including pheniprazine (Catron),[65] an antidepressant that is no longer commercially available in North America or Europe, ethchlorvynol (Placidyl),[66] an oral hypnotic used in the treatment of insomnia, and muslin gauze,[67] used in intracranial surgery. Closed head trauma is a well-recognized cause of chiasmal syndrome.[68] Prolapse of the optic chiasm into the sella turcica—the so-called empty sella syndrome (ESS) — is a rare cause of chiasmal disease. ESS results from extension of the subarachnoid space into the pituitary fossa through an incompetent diaphragma sella.[19] Primary ESS occurs spontaneously, and is associated with pseudotumor cerebri. Secondary ESS follows radiation therapy or surgery for pituitary tumor.

Clinical evaluation of chiasmal syndrome

The evaluation of patients for suspected chiasmal syndrome requires both thorough examination and special testing (Table 4). Screening patients with unexplained unilateral or bilateral vision loss[70] or chronic headache[2] with perimetry will help to avoid misdiagnosis of chiasmal lesions that present with visual disturbances or headache as the initial clinical manifestation. Patients seen on consultation with known chiasmal lesions require a comprehensive eye examination, including automated threshold perimetry,[9] to determine the extent of any vision loss. The findings should be conveyed to the referring physician, and the patient provided with appropriate follow-up.

History

Chiasmal syndrome can occur at any age, with glioma and craniopharyngioma as the primary causes in children and pituitary adenoma and meningioma as the most common causes in adults. There is no known predilection for race. Ninety percent of patients who present with microprolactinomas are women and 60 percent of patients with macroprolactinomas are men.[25] Prolactinomas in women will usually present clinically at an earlier stage, with symptoms related to the galactorrhea-amenorrhea syndrome. Although sexual dysfunction will occur in most men with prolactinomas, this is the presenting complaint in only 15 percent or less of cases. Delay in seeking medical help explains the larger tumors reported in men.[25]

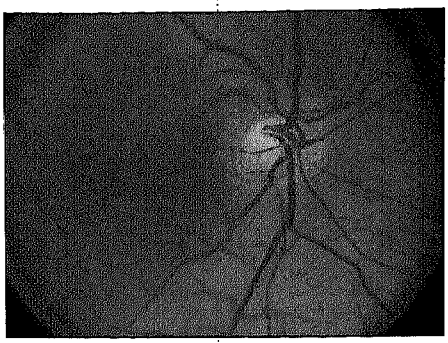


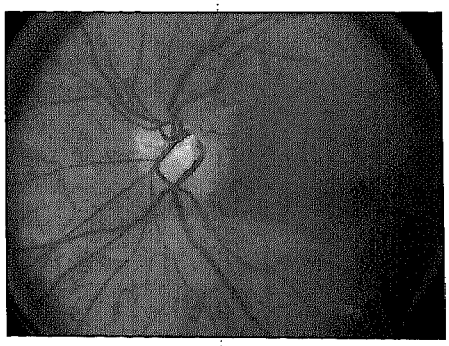
Figure 7

Fundus photograph, right eye, of patient described in Figure 6 depicting inferior nasal tilting of the optic disc and increased visibility of the choroid infranasal to the nerve head.

The evaluation of patients with chronic headache always includes a careful history.[71] Patients with prolactinomas may experience frontal or periorbital headaches, and this type of headache is often the presenting complaint.[2] Because these individuals often suffer gonadal dysfunction but do not usually volunteer this information to the eye care practitioner, it has been suggested that the eye care practitioner include questions about reproductive and sexual dysfunction as part of the headache history.[2]

Pupils and color vision

Because chiasmal vision loss is often asymmetric, an afferent pupillary defect (APD) can usually be found, but such is not always the case.[5] Indeed, the absence of an APD may give the clinician a false sense of security that the vision loss is not due to an afferent lesion, only for the clinician to later discover a lesion of the optic chiasm. Color vision has been reported to be a more sensitive indicator of afferent system damage than visual acuity in patients with chiasmal lesions.[36] When the lesion involves the distal optic nerve (junctional syndrome), associated findings will include reduced color vision and an afferent pupillary defect on the affected side.[11] Lesions of the optic tract will not produce loss of visual acuity or color vision, but can create an afferent pupillary defect in the eye ipsilateral to the hemianopsia (contralateral to the lesion).[11]



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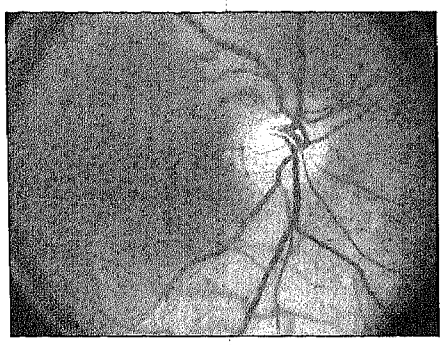

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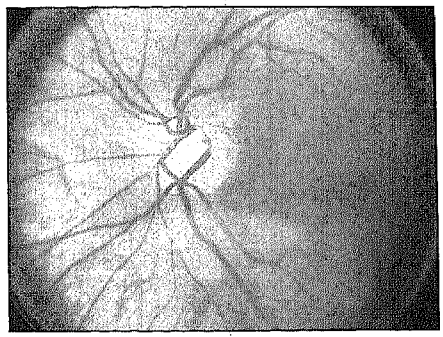

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Fundus photograph, left eye, of patient described in Figure 6 depicting inferior nasal tilting of the optic disc and increased visibility of the choroid infranasal to the nerve head.

Binocular vision and ocular motor function

Chiasmal syndrome may be associated with symptoms of diplopia due to either oculomotor paresis or decompensated phoria (hemifield slide phenomenon). Whenever the symptom of diplopia is encountered a careful search for its origin is warranted.[8] Evaluation of such patients may include ocular motor fields, cover testing, and tests of binocular vision such as fusional vergences.

Complete bitemporal hemianopsia can produce loss of binocular sensory fusion with symptoms of intermittent diplopia and difficulties performing near tasks.[4] Clearly, these patients will perform poorly on tests of binocular function, such as fusional vergences. Taken together with a complaint of chronic headache, it is easy to see how such patients could be mistakenly diagnosed as having a binocular vision disorder rather than a chiasmal lesion. The loss of visual acuity that is usually associated with advanced bitemporal defects should alert the clinician to the possibility of neurologic disease.[8]

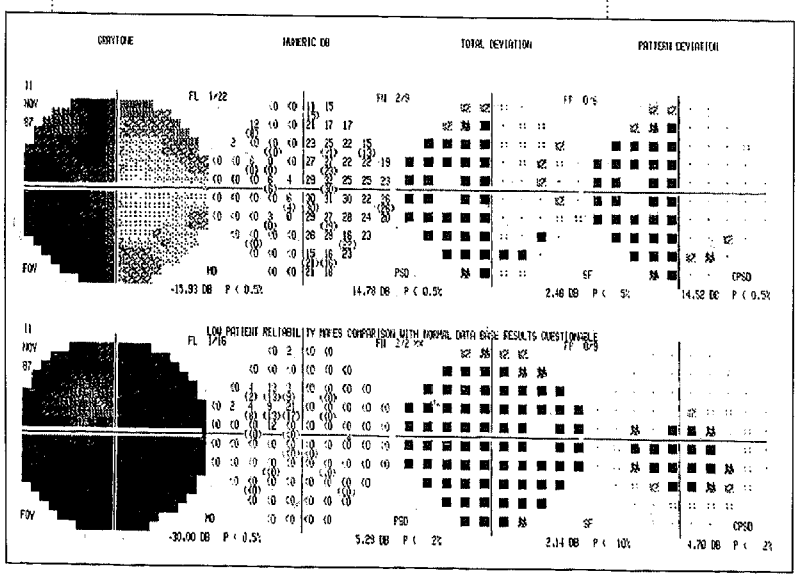

Figure 9

A 53-year-old male presented with symptoms of gradually progressive vision loss in his right eye of 6 months' duration. Visual acuity was 20/400 OD and 20/25 OS. Perimetry revealed a temporal hemianopsia in the left eye (top) and a superior-nasal island of vision in the right eye (bottom).

Ophthalmoscopy

Optic disc pallor and papilledema should be sought in any patient suspected of harboring an intracranial tumor. Ophthalmoscopy is best performed through a dilated pupil;[72] however, pupil dilation should be avoided whenever pupillary status needs to be monitored as a vital sign, as would be the case in any patient with suspected pituitary apoplexy.

Perimetry

The single most important ophthalmic test in the evaluation and monitoring of chiasmal syndrome is plotting the visual field. Perimetry yields valuable information on the presence and extent of vision loss from chiasmal and perichiasmal lesions. It is also valuable in charting visual recovery following treatment of the offending lesion.

Confrontation testing of the visual fields, while a useful adjunct, is not a substitute for perimetry. Although the sensitivity of confrontation testing can be improved through the use of red-colored targets, such as mydriatic bottle caps,[73] field defects can be missed even when they are producing significant loss of visual acuity.[5]

Automated threshold perimetry offers many benefits over conventional manual testing methods,[9] but both methods are equally effective in detecting field loss from chiasmal compression. One recent study of patients with pituitary adenomas found that in about 80 percent (275/345) of eyes tested with both Goldmann and automated perimetry, the visual field results were similarly evaluated as normal or abnormal by both methods.[74] The remaining 20 percent of eyes were approximately equally divided between fields that were judged to be abnormal by one method but normal by the other.

Visual evoked potentials

Recent studies have demonstrated that visual evoked potentials (VEPs) are effective in evaluating and monitoring vision loss in patients with chiasmal lesions.[75,76] Good correlation between the presence of a visual field defect and abnormal VEPs makes VEP testing useful in cases where reliable visual fields are unattainable. It has also been suggested that the VEP is superior to routine perimetry in detecting compressive chiasmal lesions because it can reveal subclinical visual loss before visual field defects become manifest.[75]

Neuroimaging

Magnetic resonance imaging is generally superior to computed tomography in evaluating the pituitary and parasellar region.[77-79] Radiologic studies should be focused on the optic chiasm and pituitary fossa. One should communicate clinical information about the case at hand directly to the neuroradiologist so that the very best techniques and most appropriate studies and sections may be obtained.[80] Knowing the region of interest, the radiologist can direct a "coned-down" field of view and a thin-section study through the area. The high cost of these studies (often over $1,000 each) dictates that they only be ordered when the expectation of finding a lesion is reasonably high.[81]

Laboratory studies

Laboratory evaluation in chiasmal syndrome is primarily directed at assessing endocrine function in patients with pituitary tumors (Table 5) and is best directed by an endocrinologist or experienced internist. Intrasellar mass lesions detected by neuroimaging should be evaluated by measurement of serum concentrations of pituitary hormones to determine if the lesion is of pituitary or nonpituitary origin and, if pituitary, the cell of origin.[29]

Pituitary adenomas may lead to hormonal over- or underproduction. The most common hormones produced by pituitary tumors are prolactin and growth hormone.[25] Large pituitary tumors may cause partial or complete hypopituitarism by compression of adjacent normal gland or by damaging the pituitary stalk that connects the gland to the hypothalamus.[25]

Approximately 2 percent of pituitary adenomas are associated with multiple endocrine neoplasia syndrome—type 1 (MEN 1).[22] These patients, in addition to any form of pituitary adenoma, have parathyroid and pancreatic adenomas, which may also contribute to the initial clinical presentation. Therefore, patients with pituitary adenomas should be evaluated prior to therapy for abnormalities of calcium or glucose metabolism.[22]

Management of chiasmal syndrome

Chiasmal syndrome may arise from a wide variety of disorders. The optometric evaluation of patients with chiasmal syndrome is directed at determining the degree of any vision loss and initiating an investigation into the cause of the syndrome (if it has not already been done). The treatment of chiasmal syndrome is dictated by the underlying cause. The role of the eye care practitioner in the management of patients with chiasmal syndrome is to monitor the patient's visual status, including any recovery that may occur following medical or surgical treatment of the offending lesion.[8]

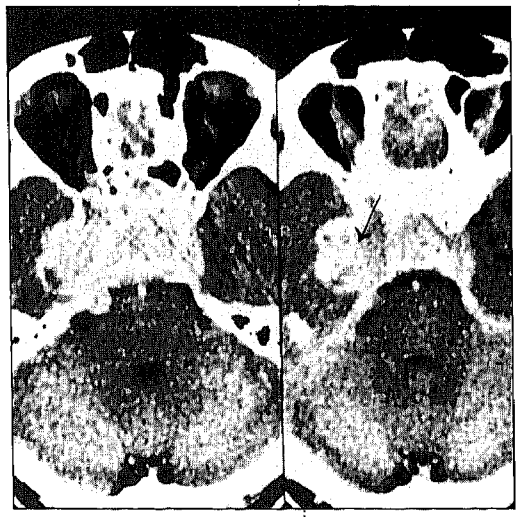

Figure 10

CT scan of the patient described in Figure 9 reveals a large pituitary adenoma that extends into the suprasellar cistern superiorly, the right middle cranial fossa laterally (arrow), and the sphenoidal sinus inferiorly.

Several investigators have reported visual improvement in about 75 percent of cases with preoperative vision loss undergoing transsphenoidal surgical treatment for pituitary adenoma.[17,82-84] Cohen et al.[84] reported postoperative improvement in visual acuity in 79 percent (154/200) of eyes and of visual fields in 74 percent (146/200) of eyes following transsphenoidal surgery for pituitary adenoma. They found that the visual outcomes were better in younger patients, those with shorter duration of symptoms, and those with lesser degrees of preoperative visual acuity (but not visual field) loss. The severity of preoperative visual field defects did not affect the postoperative outcome; even patients with severe field loss often experienced dramatic postoperative recovery. el-Azouzi[83] reported that approximately 50 percent (15/32) of patients with preoperative field loss recovered normal visual fields on Goldmann perimetry following transsphenoid surgery. Of eyes with preoperative vision loss, Sullivan[82] reported improvement in visual acuity in 74 percent (45/61) of eyes and improvement in visual fields in 68 percent (59/87) of eyes. Normal visual fields were recovered in 44 percent (38/87) of eyes. He found that the degree of preoperative acuity or visual field loss were not predictive of postoperative visual outcome, but that optic disc pallor was associated with a poorer prognosis for visual recovery. Visual recovery following surgical decompression of the chiasm is usually very rapid, with much of the recovery in function occurring within the first week following surgery.[85] Rapid visual recovery also has been reported following medical treatment of pituitary adenoma with bromocriptine (Parlodel).[33,34] Visual recovery following surgical removal of other parachiasmal tumors is not always as promising as it

Evaluation for suspected Chiasmal Syndrome

History	
	History of vision loss/visual disturbances
	Headache?
	Sexual dysfunction or other endocrine disturbances?
Pupils/Color vision	
	Afferent pupillary defect?
	Dyschrometopsia?
Binocular vision/Oculomotor function	
	Oculomotor paresis?
	Decompensated phoria?
Ophthalmoscopy	
	Optic atrophy or papilledema?
Automated threshold perimetry	
	Evidence of chiasmal visual loss?
	Substitute VEP if perimetry unattainable or unreliable
Neuroimaging	
	MRI of parasellar region if evidence of chiasmal lesion
Laboratory studies	
	Hyperprolactinemia or other endocrine imbalance?

Table 4

is with pituitary adenomas.[34] In fact, Repka et al.[36] suggest that patients with vision loss due to craniopharyngioma be advised that there is a high likelihood that the vision loss will be permanent.

Perimetry should be performed within a few days following surgery to confirm stable or improving vision. If vision is worse than the preoperative level, neuroimaging should be performed immediately to rule out a hematoma or other lesion that may be compressing the chiasm.[8] Periodic monitoring of the visual fields thereafter is recommended (Table 6).

Patients treated with bromocriptine require regular surveillance by ocular, endocrine, and neuroimaging studies.[34] Annual eye examinations, including formal perimetry, are recommended for microadenomas.[8] Monthly examinations are recommended for larger tumors and during pregnancy. Rare cases of continued tumor growth following initiation of bromocriptine therapy have been reported.[80] Poor compliance with therapy is suspected in such cases. The importance of compliance with therapy should be reinforced at each visit.

Vision loss following apparently successful treatment of a parachiasmal lesion with initial return of visual function may be due to tumor recurrence, empty sella syndrome, radiation injury syndrome, or chiasmal arachnoiditis.[8]

Laboratory tests for detection of Pituitary Adenoma

tests for anterior pituitary hormones

agent	test
growth hormone (GH)	serum GH and somatomedin C levels • glucose suppression of GH
prolactin (PRL)	serum prolactin level • chlorpromazine- or TRH- provocative tests
ACTH	plot diurnal plasma ACTH levels • dexamethasone supression of ACTH
thyrotropin	serum TSH, T4, and α-subunit levels • calculate molar ratio of α-subunit to TSH
gonadotropins	serum FSH, LH and testosterone • TRH stimulation of FSH

tests for posterior pituitary function

agent	test
vasopressin	serum vasopression level • urine and serum osmolarity

tests for men 1 syndrome

condition	test
hyperinsulinism	serum glucose level • glucose tolerance test
hyperparathyroidism	serum calcium and phosphate levels

Table 5

Evaluation for suspected Chiasmal Syndrome

History	
	History of vision loss/visual disturbances
	Headache?
	Sexual dysfunction or other endocrine disturbances?
Pupils/Color vision	
	Afferent pupillary defect?
	Dyschromatopsia?
Binocular vision/Oculomotor function	
	Oculomotor paresis?
	Decompensated phoria?
Ophthalmoscopy	
	Optic atrophy or papilledema?
Automated threshold perimetry	
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Table 5

Tumor recurrence

Recurrence rates vary in accordance with a number of factors, including tumor type and size at surgery, whether total surgical excision was achieved, whether adjunctive therapy (such as radiation) was employed, and the length of follow-up. Generally, recurrence of pituitary adenomas is relatively low, with 10-year follow-up studies indicating a 10 to 20 percent recurrence rate after initially successful surgery.[22] On the other hand, craniopharyngiomas are notoriously more difficult to totally remove, but radiotherapy following surgery can reduce recurrence to under 30 percent.[24]

Empty sella syndrome

Vision loss secondary to herniation of the chiasm into an enlarged empty sella following pituitary surgery is a rare occurrence.[8] Diagnosis is made by magnetic resonance imaging, which will reveal intrasellar herniation of the chiasm. The degree of herniation does not always correlate with the severity of vision loss.[87]

Radionecrosis

The hazards of radiation therapy are well known.[31] Radionecrosis is directly related to the dosage of the radiation. The threshold for brain necrosis is 6000 rads delivered in 30 fractions of 200 rads over a 6-week period.[31] The peak incidence of vision loss occurs 8 to 13 months after completion of radiotherapy. Radiation necrosis can present a difficult diagnostic problem because its signs and symptoms often mimic recurrence of the tumor. Radionecrosis of the anterior visual pathways usually presents as painless loss of vision in one eye, frequently followed by involvement of the fellow eye. Most visual field abnormalities are central scotomas and nerve fiber bundle defects typical of optic nerve involvement, but chiasmal or optic tract patterns may occur. In general, vision loss is progressive and irreversible. No effective treatment has been found.

Optochiasmal arachnoiditis

Optochiasmal arachnoiditis is a localized inflammatory process at the base of the brain affecting the chiasm, optic nerves, and the surrounding meninges.[11] Causes of this condition include basal meningitis, trauma (including surgery), intracranial tumor, empty sella syndrome, systemic disease, and occurrence as an isolated phenomenon.[11] Vision loss is usually bilateral but often asymmetric. Thickening of the chiasm may be found on MR imaging.[67] Vision frequently improves with high-dose oral steroids, but surgical lysis of inflammatory arachnoid adhesions may be required.

Schedule for follow-up visual field testing*

treatment	visual field follow-up
surgery alone	immediately postoperatively 4 to 6 weeks postoperatively at 4-month intervals for 1 year yearly for 5 years then every 2 years
surgery and radiation	immediately postoperatively at completion of radiotherapy at 6-month intervals for 1 year yearly for 5 years then every 2 years
Bromocriptine alone	every 6 months for 1 year, then yearly more closely for larger tumors monthly during pregnancy

Table 6

*Adapted from: Burde RM, Savino PJ, Trobe JD. Clinical decisions in neuro-ophthalmology. St Louis: C.V. Mosby, 1985.

Conclusion

Chiasmal syndrome is the constellation of signs and symptoms that are associated with lesions of the optic chiasm. Pituitary adenoma is the single most common cause of this disease. Visual symptoms are usually vague or nonexistent until acuity is affected. Diagnosis of chiasmal lesions is often delayed because the presenting symptoms are frequently unilateral and adequate visual field testing is not performed. These difficulties contribute to making the delayed diagnosis of chiasmal syndrome a relatively frequent cause of malpractice actions. To avoid misdiagnosis of chiasmal lesions, patients who present with unexplained unilateral or bilateral vision loss or chronic headache should receive perimetry. Confrontation testing is inadequate for detecting early bitemporal loss. The high cost of neuroimaging studies dictates that they only be ordered when the expectation of finding a lesion is reasonably high. The role of the eye care practitioner in the management of patients with chiasmal syndrome is to monitor the patient's visual status, including any recovery that may occur following medical or surgical treatment of the offending lesion.

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surgery alone	immediately postoperatively
	4 to 6 weeks postoperatively
	at 4-month intervals for 1 year
	yearly for 5 years
	then every 2 years
surgery and radiation	immediately postoperatively
	at completion of radiotherapy
	at 3-month intervals for 1 year
	yearly for 5 years
	then every 2 years
Bromocriptine alone	every 6 months for 1 year, then yearly
	more closely for larger tumors
	monthly during pregnancy

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