Neuro-opthalmological aspect of chiasmal-and-sellar disorders

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This article reviews the causes, pathogenesis, and major manifestations of chiasmal syndrome. In addition, it reviews the methods for diagnosing the condition, and the course of the syndrome in patients with various chiasmal-and-sellar disorders such as pituitary adenoma, craniopharyngioma, tuberculum sellae meningioma, and skull base aneurysm. The principle neuro-ophthalmological symptoms include decreased visual acuity, bitemporal visual field defects, and development of descending optic nerve atrophy. Early detection of ophthalmological symptoms is of critical importance for timely neurosurgical treatment.

Any influence that modifies the entire structure influences a substructure, and vice versa; therefore, the eye is affected by any disease of the body, and the body is affected by any disease of the eye.
Paul Bert, a French physiologist

Chiasmal-and-sellar disorders are numerous and include pituitary adenoma, craniopharyngioma, meningiomas arising from the tuberculum sellae, sphenoid wing and olfactory groove, chiasmal and third ventricular gliomas, third ventricular hydrocephalus, optochiasmatic arachnoiditis, multiple sclerosis, cranial trauma, posterior cranial fossa tumors, skull base aneurysm, intoxications, etc. [1-3].

The chiasm lies 10 mm above the sella turcica, which contains the pituitary gland. It is 12 to 18 mm in diameter and contains the decussation of nasal retinal nerve fibers. Fibers arising from the nasal half of each retina cross in the optic chiasm. Posteriorly, the chiasm abuts the hypothalamus and the anterior portion of the third ventricle; it lies within the circle of Willis. The anatomic position of the chiasm is described as prefixed, normal, or postfixed based on the relative position of the chiasm (anterior, above, or posterior) to the sella. In the majority of cases (80%) the chiasm is situated overlying the diaphragma sellae [4]. The individual variation in relative position contributes to clinical variation in the presentation of neuro-ophthalmological chiasmal syndromes.

A pathological process in the chiasmal and sellar region results in the development of neuro-ophthalmic symptoms of chiasmal syndrome (CS) with decreased visual acuity, visual field defects and development of descending optic nerve atrophy. Other symptoms that may develop in CS include diplopia, impaired color perception, impaired stereoscopic vision, photophobia and a number of generalized symptoms like headache and hormonal abnormalities [5-8].

Ophthalmic symptoms have a prognostic value since the disease may lead to a total loss of visual function. The primary pathogenetic mechanism of CS is local compression of the chiasm proper from pathology focus which, in turn, leads to optic nerve fiber atrophy.

Changes in optic nerve fibers arise not only at the compression site, but also at distant sites. The chiasm may be displaced and compressed toward the vessels of the circle of Willis. Sometimes CS is apt to be present as a distant symptom when the tumor is in the brain. A major pathogenetic component of the process is ventricular system expansion or brain displacement. In case of ventricular system expansion, the expanding (and, commonly, subtentorial) mass causes obstructive hydrocephalus. In addition, the primary lesion extends to the floor of the third ventricle and then compresses the optic chiasm or intracranial portion of the optic nerve, which is followed by the development of optic nerve atrophy. Another pathogenetic component is progressive lesion growth and brain displacement resulting in brain compression against the skull base and indirect chiasmal compression [9].

We classify chiasmal lesions in five categories according to the topography of the lesion and pattern of visual field loss: intrachiasmal, anterior suprachiasmal, posterior suprachiasmal, perichiasmal and intrachiasmal.

1. Infrachiasmal lesions

The pathological process (most commonly, pituitary adenoma) is located in the sella turcica. Visual field abnormalities (typical bitemporal hemianopia and

descending optic nerve atrophy) develop as the lesion size increases to greater than 15 mm.

2. Anterior suprachiasmal lesions

The lesion is manifested by inferior temporal hemianopia and unilateral optic nerve lesion. Common causes include sphenoid wing mass, tuberculum sellae meningiomas, frontal lobe gliomas, and aneurysms of the anterior communicating artery or anterior cerebral artery).

3. Posterior suprachiasmal lesions

The lesion is accompanied by bitemporal hemianopia frequently beginning in the inferior quadrants. The papillomacular bundle lesion causes a central or paracentral scotoma, and spread of the pathological process to the optic tract causes homonymous hemianopia. The most common causes of posterior suprachiasmal lesions are craniopharyngioma, osteoma, and third ventricle expansion due to inflammation, hydrocephalus or tumor.

4. Perichiasmal lesions

These lesions are most commonly caused by perichiasmal adhesive meningitis resulting from syphilis, bacterial infection or trauma. Variable visual field defects are observed in chiasmal arachnoiditis.

5. Intrachiasmal lesions

These may develop from a chiasmal tumor, demyelination, Devic's disease, or craniocerebral injury. In children, intrachiasmal lesions may result from chiasmal gliomas. Bitemporal hemianoptic scotomas are characteristic of these lesions.

It is visual impairment that is the most common reason for the initial visit of patients with undiagnosed CS to the ophthalmologist's office, although visual impairment is not an early symptom of the disease but becomes evident when the process is spreading from the supracellar region to the chiasm. Therefore, it is the ophthalmologist who is fully responsible for making the correct diagnosis and, correspondingly, for initiating timely treatment.

Detection of the focus of the disease in the retrobulbar region is of great practical importance since in such cases atrophy is frequently caused by neurosurgical disease and thus requires not ophthalmological, but neurosurgical care.

Patients with supposed CS should undergo a comprehensive eye examination including visual acuity, pupil responses, ocular motility, IOP measurement, anterior biomicroscopy, direct ophthalmoscopy, color vision testing and careful visual field testing [4, 10]. The main ophthalmologist's task in these patients is to detect early signs of compressive optic neuropathy before descending optic nerve atrophy develops.

Conducting careful visual field studies in the early disease is critically important. Color vision studies may reveal the earliest manifestations [11, 12].

In spite of decreased visual acuity and visual field loss, the fundus may show no changes for a long time, and it is the early phase of the pathological process when most diagnostic errors can be discovered.

Detecting subclinical neuro-ophthalmological signs of CS with optic coherence tomography (OCT) is important. Peripapillary retinal nerve fiber layer (RNFL) defects are the earliest sign of CS and a number of neurodegenerative disorders (Alzheimer's disease, multiple sclerosis, Parkinson's disease and cerebellar ataxia). Peripapillary RNFL thinning precedes visual field defects. In most patients with CS, nerve fiber thinning had been observed for more than 3-5 years before visual field changes occurred [13, 14].

RNFL and ganglion cell layer thinning may be also observed in pituitary microadenoma in the absence of chiasmal compression. This may be explained by two factors. First, the presence of initial chiasmal compression that was too subtle to be reflected by a change in neuroimaging measures. Second, the pituitary gland may secrete vasoactive peptide resulting in ischemia and slow axoplasmic flow [4, 15].

The field defect seen in a classical CS with normally located chiasm is a bitemporal hemianopia. Initially, a central relative or absolute scotoma or partial (relative or absolute) bitemporal hemianopia is revealed. Quadrant, bitemporal central or paracentral scotomata may be found. Vision is generally lost first in either or both superior temporal quadrants (the chiasm is compressed from below) [11, 12, 16, 17].

Unilateral central defects (scotomata) with contralateral temporal hemianopia may develop when the chiasm is pre- or postfixed [4].

Homonymous tractus hemianopia is infrequently found in a CS, and has some key signs that make it different from homonymous central hemianopia: sudden mono- or bilateral visual acuity loss, marked asymmetry of visual field defects and fundus changes in the form of atrophy of the optic nerve. The surest indication of the homonymous tractus hemianopia is the hemianopic pupillary response to light: if a light is directed at the nonfunctioning part of the retina, there is absence of pupillary reaction, but if the light is directed at the functioning part of the retina, there is pupillary response. Pupillary response is always preserved in central hemianopias [2, 18]. Visual field defects are first found with regard to green and/or red objects only with the preserved perception of "white" in the visual system. Hemichromatopsia is an intermediate form in the development of absolute hemianopia.

It should be noted that, in patients with CS, visual field changes are found later than peripapillary RNFL and retinal ganglion cell layer thinning. Tieger et al [14] believes that the automatic perimetry sensitivity is not high enough to reliably reflect early signs of a CS which can be detected with OCT imaging. In addition, the potential of visual evoked potentials for diagnosing CS is being investigated [19].

It is important to keep in mind the anatomical feature of the arrangement of the cella turcica, the floor of which is covered with the dura mater. The latter contains pain-sensitive nerve endings (mostly of the first trigeminal

nerve division), and irritation of these endings due to tumor-induced inflammation or compression may result in headache. The lateral walls of the pituitary fossa are formed by the cavernous sinuses which contain the internal carotid arteries and the sympathetic plexus inside, and a number of cranial nerves (CN) (the CN III, CN IV and CN VI as well as the first division of the CN V) in the external wall. Involvement of specific sites of this region may cause CN III, CN IV and CN VI pareses together with neuralgic pain disturbances over the region supplied by the ophthalmic branch of the trigeminal nerve (the so called syndrome of the external wall of the cavernous sinus). This is sometimes accompanied by temporary diplopia, anisocoria and mild ptosis. Oculomotor abnormalities are found in 1.4-4.5% of cases with syndrome of the external wall of the cavernous sinus. Most commonly, oculomotor nerve lesion occurs, which begins with ptosis followed by limitation of the eye movements in upgaze and/or downgaze. Pupil responses are preserved or slightly reduced infrequently. Complete ophthalmoplegia is rare, and most commonly occurs in tumoral hemorrhage [2, 6].

Cavernous sinus compression can result in impaired orbital outflow leading to mild exophthalmos and conjunctival and lid edema. Lesion of the internal carotid artery sympathetic plexus may lead to Horner's syndrome (ptosis, miosis, and enophthalmos) [2].

Pituital adenoma is the most common brain tumor causing the choroidal syndrome in adults, and accounts for 15% of the cases of intracranial tunors in the adult population [20]. Tumors smaller than 10 millimeters are called microadenomas; they are located within the sella and manifested by endocrine abnormalities (reduced sexual potency, menstrual cycle abnormalities and acromegalia). Ophthalmological abnormalities arise when there is tumor expansion beyond the sella turcica. Ophthalmological symptoms depend on the direction of growth of the tumor. Suprasellar extension of the pituitary adenoma and intracranial portions of the optic nerves produce a CS [21]. The tumor initially compresses the central chiasm containing decussating nasal retinal fibers from below the optic chiasm. This is followed by the development of a bitemporal hemianopia. Decreased visual acuity is found when the tumor affects the intracranial portion of the optic nerve, and the papillomacular bundles become involved in the pathological process. Apart from bitemporal visual field defects, a central or paracentral scotoma may be also found [1, 16, 22].

In the presence of bitemporal hemianopia, optic nerve pallor is a predicting factor for postoperative visual function [23].

Important accompanying symptoms of pituitary adenoma include headache radiating to the eye and the forehead and temporal regions. As patients may have a variety of endocrine abnormalities including irregular menstrual cycle, galactorrhea, acromegalia, dwarfism, diabetes insipidus, obesity and sexual disorders, careful taking of not only ophthalmological, but also general medical history is warranted. Complaints of headache, sexual disorders, increased thirst, and abnormal weight gain or loss, together visual complaints and finding of optic nerve atrophy of unclear etiology should prompt consideration of a neurosurgical disease, namely, endocrine pituitary tumors. Unfortunately, rather often patients with the initial manifestations of CS are misdiagnosed with a variety of eye disorders including glaucoma, retinal degeneration, and optic neuropathy, and undergo long-lasting treatments which do not afford symptom relief. Moreover, massive vascular therapy worsens patient's condition and accelerates tumor growth.

The neuroophthalmological picture in pituitary apoplexy is characterized by a sudden reduction or loss of vision, sometimes with acute temporal hemianopia in the presence of marked headache. CN III, CN IV and/or CN VI lesions resulting in ptosis, mydriasis and limitation of the eye movements may occur if the neoplasm is located close to the site of the cavernous sinus [24, 25].

Craniopharyngioma, which is more common in children and adolescents, presents a special clinical picture. This benign tumor causes sudden changes in the chiasm and intracranial portions of optic nerves [26]. An atypical CS involving bitemporal hemianopia with crowded optic dics or secondary optic nerve atrophy is characteristic for craniopharyngiomas. In patients with craniopharyngioma, endocrine abnormalities in the form of loss of pituitary function (short stature and/or impaired sexual function) precede loss of visual function.

In tuberculum sellae meningiomas, the tumor is located anteriorly to the chiasm, and causes inferior chiasmal compression, which is frequently asymmetric. Choroidal syndrome is the most common symptom of the disease in the absence of endocrine abnormalities and hyperprolactinemia. Visual field defects usually begin in the superior temporal quadrants [20, 27].

Sphenoid wing meningiomas are parasellar tumors (a medial third of the sphenoid wing is directly adjacent to the sella turcica). Therefore, they exert chiasmal changes that are more apparent in the chiasmal portion adjacent to tumor gtowth. These tumors frequently raise intracranial pressure and cause crowding of the optic disc [3, 21].

Chiasmal gliomas are rare and occur more frequently in children aged from 4 to 12 years. The symptoms include CS and hypothalamic symptoms. The syndrome develops slowly and usually unevenly throughout the course of the disease. Hypothalamic symptoms are manifested by obesity and polyuria. Intracranial pressure, if raised, causes additional symptoms (headache and vomiting) [28].

Intracranial aneurysms may cause neuroophthalmological symptoms due to the chiasm being in anatomical proximity to the circle of Willis. Paraclinoid (or paraophthalmic) aneurysms arise from the segment of the internal carotid artery (ICA) between the distal dural ring and the origin of the posterior communicating artery. These aneurysms are difficult to diagnose since they are manifested only by a gradual decrease in visual acuity

and the development of CS. Aneurysm rupture resulting in subarachnoid hemorrhage may be clinically manifested by sudden headache, neurological deficit and decrease in visual acuity [20, 29, 30].

Chiasmal neuritis (chiasmal inflammation) results in acute loss of visual acuity and bitemporal visual field defects. Inflammation may be categorized as infectious, non-infectious and idiopathic. Chiasmal inflammation may be due to an infectious disease such as tuberculosis, parotitis, Epstein-Barr disease, or Lyme disease. Non-infectious inflammation may be due to sarcoidosis or systemic lupus erythematosus, and may occur in patients taking ribavarin, interferon, isoniazid or ethambutol for long periods [4, 31].

In multiple sclerosis, chiasmal lesions are manifested when demyelinating foci are located at the chiasm and optic tracts. Demyelinating process in different anatomical structures is morphologically similar, and the clinical picture depends on the location of plaques. That is why symptoms of retrobulbar neuritis and CS may be present in a patient with multiple sclerosis [2].

Traumatic CS develops following a head injury causing injury to the frontal bone and fracture of the anterior skull base. The degree of visual acuity loss and visual field defects is variable [4, 24].

Chiasmal syndrome as a manifestation of a chiasmal-and- sellar disorder requires surgical treatment in most cases. Neuro-ophthalmological studies allow predicting postoperative visual function. A study of Danesh-Meyer et al [13, 32, 33] has established a clinical marker that correlates strongly with the degree of visual recovery after surgical intervention in patients with significant visual loss as a result of chiasmal compression. The degree of reversibility of visual dysfunction with compression of the anterior visual pathway is related to the loss of RNFL thickness, as measured by the OCT. The study [32] demonstrated that there is an increasing probability of improvement to near normal visual function with increasing RNFL thickness up to approximately 85 μm, after which there is no further improvement in visual function.

Ganglion cell complex (GCC) thinning can occur before visual function loss. OCT analysis of the ganglion cell layer may be useful for the prognosis of visual function recovery from surgical or medical decompression of the chiasm [14]. Early surgical decompression before significant RNFL and GCC thinning may lead to better postoperative visual function. Therefore, given that CS may be a manifestation of any of a variety of underlying diseases, thorough assessment taking into account differential diagnoses is essential.

Most patients with early CS first present to ophthalmologists. Variable visual field defects (bitemporal contraction or scotomata) are observed, but temporal segments tend to be affected. Unfortunately, ophthalmologists do not always take a complete general medical history with attention to endocrine and sexual disorders, neurological changes and traumatic history. This may be the reason for the fact many patients with undiagnosed chiasmal syndrome come to neurosurgeons only after being misdiagnosed and mistreated for glaucoma, cataract or retinal degeneration with medications contraindicated in chiasmal tumors. This leads to decreased visual acuity, progression of the pathological process, and irreversible loss of sight. Chiasmal syndrome early detection of by ophthalmologists and timely referral to relevant specialists will facilitate improved visual function outcomes and quality of life for patients.

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