NEURO-OPHTHALMOLOGICAL AND PUPILLARY SYNDROMES IN OPHTHALMOLOGY PRACTICE

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Abstract: *The article discusses various neuro-ophthalmological syndromes including pupillary abnormalities identified in ambulatory setting and that are often difficult to interpret. Pupillary abnormalities are uncommon. Some of these abnormalities are discovered incidentally, for example the tonic pupil (Holmes-Adie syndrome). Documentation of changes is important, not necessarily for the assessment of patient dissatisfaction, but to prevent misinterpretation of the problem in the future. Regarding the patient in critical condition, recognition of cerebral herniation syndrome, particularly uncal and transtentorial herniation is decisive. Pupillary abnormalities within the efferent system of the neuro-ophthalmological clinical syndromes are caused by parasympathetic, sympathetic lesions, physiological conditions, congenital anomalies, others.*

Pupillary syndrome

Argyll Robertson described the pupilary syndrome in 1869. It represents the isolated paralysis of the photomotor reflex. This sign must meet the following criteria in order to be considered real: the disorder must be permanent and the pupillary immobility to be invariable, irrespective of light intensity or duration of obscuration; the existence of dissociation of pupillary reactions, that is the abolition of the direct and consensual photomotor reflex and the conservation of the syncynetic constriction to accommodation and convergence; the disorder should be bilateral (possibly unilateral); the pupil, more often in case of miosis, to be normal; the retina and the common oculomotor nerve should be normal. The definition should be strict, otherwise there will be designated a whole range of pupillary abnormalities, with some degree of dissociation to light. Pupils are not necessarily fixed to light, although they become fixed in the subsequent stages.(1,2) What is essential is the dissociation between photomotor reflexes and convergence accommodation.

Preservation of the normal acuity should be included in the criteria to exclude many conditions in which a depression of light sensitivity, for example due to optic nerve disease, to lead to light – near dissociation.(1,3)

The condition is pathognomonic for neurosyphilis. The cases are now rare, although recent flourishing of primary syphilis cases suggests that its incidence could increase in the future. The place the most likely of a causal lesion is one which affects reflex fibers to light, registered in the oculomotor nucleus. Lesions are most commonly placed in the pretectal area.(4)

At ocular level, the two pupils are small, irregular, are slowly shrinking, strong (punctifom) upon accommodation and convergence; bilateral ocular damage, asymmetrical; small pupils in irregular miosis, spinal miosis, preservation of the accommodative reflex (normal constriction of the pupil in convergence); near/distance dissociation; very difficult dilatation; direct and consensual photomotor reflex to light is diminished; convergence reflex is preserved.(5,6)

From the *neurological* point of view, the most frequent cause is neurosyphilis: tertiary syphilis, heredotabes associated with tendinous areflexia, progressive general paralysis (PGP); rarely, luetic meningitis, encephalitis, disseminated sclerosis, tumours of the cap peduncle, syringomyelia; syringobulbia, chronic alcoholism, Parinaud's syndrome, psychiatric disorders.(7,8)

Systemically, diabetes can be encountered.

Tonic pupil

Patients with a tonic pupil tend to have differences in pupil's size. Sometimes, the patient notices a problem in bright light, a consequence of the failure of the constriction reflex. It is usually unilateral and tends to predominate in young and middle-age women. The damaged pupil reacts incompletely to light and dark, so this is higher than the collateral one in the light, but it may be lower in the dark because of the intact dilation reflex of the undamaged eyeball. The cardinal feature is the almost tonic reaction. Whenever there is a slightly depressed reaction in a normal pupil, it is essential to closely observe the reaction. It should be possible to diagnose the problem without using pharmacological agents. Accommodation component of the reaction to near is also tonic. Most cases occur spontaneously, but sometimes the status evolves from a state of acute iridoplegia. Such a progression has been seen in a patient with neurosarcoidosis. The phenomenon is thought to be a consequence of aberrant regeneration of fibbers accommodation before the ciliary ganglion damage. In many cases, the condition was not recognized, either by the patient or by the physician. In some cases, deep tendon reflexes are depressed (considered Holmes-Adie syndrome) due to the disruption of the synaptic transmission in the monosynaptic reflex arc of the spinal cord.(3)

It is produced by the paralysis of the peripheral nervous system with the damage of the second neuron and of the postganglionic innervation for the pupillary sphincter and ciliary muscle. It is more common in women of 20-40 years old.

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In terms of symptoms, it is asymptomatic in 20% of cases; blurred vision due to accommodation paresis, difficulty in reading, photophobia, ciliary spasm and pain in the forehead when working at close distances; accommodation can lead to astigmatism.(4)

At *ocular* level, it is unilateral in 80% of cases. The pupil registers reduced tonic constriction to light and near, slow redilatation after accommodation, irregular dilated pupil, the pupil can become mitotic in time, in the old forms, after prolonged near activity, the pupil becomes smaller than in the healthy eye and it stays as such for long, because the reactions are slow and tonic. The photomotor reflex is absent; at the slit lamp - sphincter sectoral paralysis; vermicular movements on pupillary edge of the iris, pharmacological test to 0.125% pilocarpine: the normal pupil does not shrink; Adie's pupil leads to myosis.(5,7,8)

Neurologically, one can notice a decrease of tendon reflexes associated with tonic pupil, autonomous innervation dysfunction.

Systemically, arterial hypertension is recorded.

The differential diagnosis is made taking into account infections (syphilis, shingles, measles); giant cell arteritis; cataract surgery; iatrogenic in (Laser photocoagulation, alcohol-based retrobulbar injections, cryotherapy); alcoholism; diabetes; amyloidosis; amyloidosis; cancer associated dysautonomia; migraine. Some symptoms may resolve spontaneously, but 4% may become bilateral.(5,6)

The treatment consists of anti-glare lenses for patients with photophobia, optical correction for near, 0.5% pilocarpine for the ciliary spasm.

Oculosympathetic paralysis

It occurs in Horner syndrome, when the ocular sympathetic fibers reach the eye, after a cross which begins in the hypothalamus. It is caused by the paralysis of the sympathetic nervous system.(3)

The causes can be central with protoneuron damage in: cerebral pathology, syringomyelia, Wallenberg marrow lateral syndrome, spinal tumours.(9) There also may be preganglionic causes with second neuron damage: Pancoast tumour (apical lung cancer) aneurysms, carotid, aortic dissection, aortic, cervical spinal injuries;(5,6) neuron III postganglionic damage: facial vascular algia, internal carotid dissection, nasopharyngeal tumours, otitis media, cavernous sinus tumours;(7) congenital Horner syndrome: obstetrical injury with brachial plexus lesion, perinatal thoracic surgery, congenital tumour, neuroblastoma, lymphoma, tumour of cervical ganglion.(9)

At *ocular* level, the characteristic features of Horner syndrome (the consequence of the interruption of these fibers) are discreet ptosis (1-2 mm) with slightly lower eyelid lift).(9,3) Ptosis is reduced temporarily. Enophthalmia is not confirmed if measurement of the formal globe position is made.(9,3) The conjunctival vessels may be dilated for a short period of time immediately after the onset.

The following may also occur: miosis, anisocoria accentuated in darkness; normal reaction to light and convergence; delay in dilatation when the lights go out; homolateral facial anhydrosis if the lesion is located before the upper cervical ganglion; iris pigmentation reduced on the affected side;(9,8) iris heterocromia (in the old congenital area); hyperactive accommodation; ocular hypotonia, excessive secretion of tears.

Although the pharmacological tests have been suggested in order to distinguish pre- and postganglionic syndrome cases, the results are not consistently necessary and the distinction is rarely decisive in the clinical practice. If necessary, the condition can be confirmed by instillation of 4% cocaine eye drops. Normal pupil dilates, while Horner pupil fails to do so. Sometimes, there may be some difficulties in distinguishing miosis of Horner syndrome from a physiological anisocoria. If in doubt, take the patient into a dark room. Horner pupil fails to dilate in darkness, so anisocoria becomes more apparent. In the case of physiological anisocoria, both pupils dilate, so the degree of anisocoria remains unchanged.

4% cocaine test confirmed the diagnosis: normal - pupila dilates, Horner pupil does not dilates.

Hydroxyamphetamine 1% tissue allows to distinguish between preganglionic lesions from those postganglionic.

Preganglionic ilesions - both pupils dilate; postganglionic lesions - Horner pupil does not dilate.

1 ‰ adrenaline test for the pre / postganglionic differentiation: - preganglionic – the pupils do not dilate, postganglionic - Horner pupil dilates.(9)

From the *neurological* point of view, the following symptoms can occur: sweating on the face, finding it difficult to emphasize in the clinical practice; Horner syndrome can be founded as part of neurological disorders (e.g. lateral medullary syndrome), but it is sometimes founded in isolation. The question is whether an isolated Horner syndrome deserves investigation. Of course, an X-ray package is reasonable in such circumstances.(6,9)

Although isolated Horner syndrome has been described as presenting syringomyelia, anamnesis also indicates that the patient had occipital headache episodes, as well. A painful Horner syndrome may be the only manifestation of extracranial carotid artery dissection and justifies vascular imaging. Other neurological symptoms include: anhydrosis at face, neck and ipsilateral level; transient increase in facial temperature, hemifacial atrophy.(3)

Uncal and transtentorial herniation

Brain hernia is a side effect of the very large intracranial pressure that occurs when part of the brain is squeezed over structures in the skull. The brain can move on structures such as falx cerebri, cerebelli Tentorium, and even through the foramen magnum of the skull (through which the spinal cord is connected to the brain). Hernias can be caused by a number of factors that determine a mass effect and increased intracranial pressure (ICP). Tthese include traumatic brain injury, intracranial hemorrhage, or brain tumors.(10)

Hernias can also occur in the absence of large ICP when mass injuries, such as hematomas occur at the boundaries of some brain compartments. In such cases, local pressure is increased in the place where hernia occurs, but this pressure is not transmitted to the rest of the brain, and hence, there is no ICP increase.(10)

As hernia puts extreme pressure on certain parts of the brain and, therefore, suppresses the blood supply to various parts of the brain, it is often fatal. Therefore, extreme measures are taken in the hospital to prevent this condition by reducing intracranial pressure or decompression (leakage) of a hematoma that exerts local pressure on one side of the brain.(10)

There are two major classes of hernia: supratentorial and infratentorial. Supratentorial hernia is structurally above the tentorial notch, the infratentorial one being below.(10)

At *ocular* level, the earliest papillary sign in uncal herniation is a pupillary dilation with damaged response to direct light. The sign may be evident when the patient is slightly sleepy. The pupil is widely dilated and fixed. This stage may last for a few hours before the appearance of other neurological signs.(3) In later stages of uncal herniation, ophthalmoplegia occurs.(3)

Before diagnosing an early uncal herniation syndrome,

it is vital to check whether pupillary asymmetry has not been recorded in the past and that mydriatic drops have not been used in order to facilitate ophthalmoscopy.(3)

In uncal herniation, a common subtype of transtentorial herniation, the inner part of the temporal lobe, the uncus can be pushed so much that it moves to the Tentorium and exerts pressure on the brainstem, especially on the midbrain. Tentorium is a structure inside the skull made up of dura mater of the meninges. The tissue can be removed from the cerebral cortex in a process called decortication.(10)

The uncus compresses the oculomotor nerve, which can affect the parasympathetic nerve afferent to the eyes on the lateral side of the nerve, causing pupil dilation and not the contraction in response to light as it should. Pupils dilatation often precedes the somatic motor effects of the cranial nerve III compression, which show "down and out" eye deviation as a result of the loss of innervation of all ocular muscle motility with the exception of the lateral right (innervated by cranial nerve VI) and superior oblique (innervated by cranial nerve IV). Symptoms appear in this order, as the parasympathetic fibers are surrounding the motor fibers of the cranial nerve III, and therefore they are the first to be compressed.(10)

Central herniation syndrome (transtentorial) produces different signs. Although this syndrome occurs more likely with medially placed injuries, lateral masses may also occur.(2) Transtentorial herniation represents the moving down of the midline brain structures through the tentorial notch, putting pressure on the underlying structures, including the brainstem. This is a life-threatening situation due to the cranial nerves pressure, with symptoms including dilated pupils, nonreactive, ptosis, and a low level of consciousness. Regarding the transtentorial herniation, the pupils remain equal, although miotic, and, at least initially, resistant to the response to light.

The earliest *ocular* sign in transtentorial hernia tends to be the paralysis of eye movement upwards giving a characteristic appearance of "sunset eyes". Also, in these patients, often as a terminal complication, diabetes may occur, as a result of pituitary stalk compression. Subsequently, the pupils become fixed and the horizontal eye movements are damaged.(10)

Ipsilateral compression of the posterior cerebral artery will lead to the primary visual cortex ipsilateral ischemia and contralateral visual field deficits in both eyes (contralateral homonymous hemianopia).(10)

The *neurological* signs include the rapid decline in the level of consciousness.(3) Sometimes, an ipsilateral hemiplegia due to the compression of the cerebral peduncle on the contralateral tentorial edge may appear.(3)

Another important finding is the so-called the Kernohan's notch, resulting from the contralateral compression of the cerebral crus containing descending corticospinal fibers and some fibers of the corticobulbar tract. This leads to ipsilateral hemiparesis. Because the cortical tract predominantly innervates the flexor muscles, leg extension can be also seen. At the same time with increasing pressure and hernia progression, there will be a distortion of the brainstem which will lead to Duret hemorrhage (breaking of small vessels of the parenchyma) in the median area from the midbrain and stem. The rupture of these vessels causes linear or flamed bleeding. Damaged brainstem can lead to decorticate posture, respiratory centre depression and death. Other possibilities resulting from brainstem distortion refer to lethargy, low heart rate and pupil dilation. Uncal herniation can advance to central herniation.(10)

A complication of Uncal herniation is bleeding. This goes to the midbrain and stem, which were compressed to the damage of the reticular formation damage. If untreated, death will result.(10)

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