SYNDROMES INVOLVING THE CRANIAL NERVES III, IV, VI

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Keywords: *cranial nerves, syndromes, peduncular lesions, cavernous lodge, anterior and middle cranial base, orbital apex, upper cerebellar artery*

Abstract: *Several cranial nerves may be affected by the same pathologic process. In this situation, the main clinical problem is to determine whether the lesion is located inside or outside the cerebral trunk. The lesions extending on the surface of the cerebral trunk are characterized by damage to the adjacent cranial nerves and by later and easier damage to the long sensory and motor pathways and to the segmental structures that are located inside the cerebral trunk. The opposite of these phenomena proves the existence of intrabulbar, intrapontine and intra-mesencephalic lesions. The extrabulbar damage produces more likely bone erosion or widening of the cranial nerve access way. The intrabulbar damage affecting the cranial nerves often produces a cross sensory or motor paralysis (signs of cranial nerves on one side of the body and signs of tracts on the opposite side). Due to their anatomical relationships, multiple paralyses of cranial nerves form a number of different syndromes.*

The third pair of cranial nerves

Peduncular syndromes

The Weber's syndrome consists of homolateral paralysis of the oculomotor nerve with controllateral hemiplegia, occurring during pedunculated lesions, bridge, marrow. It occurs in hemorrhage, softening, tumours, traumas.(1)

At ocular level, the following can be described: complete III homolateral ptosis-paralysis, fixed, dilated pupil. The association of a homonymous hemianopia shows the involvement of the lateral geniculate body, the optic band or damage to the posterior cerebral artery.(2)

Systemically: contralateral-paralysis of the face and tongue, polyuria polydipsia.(1,3,4)

The Benedict's syndrome (of the cerebral peduncle skull-cap is described by hyperkinesia, ataxia, paresis, tremor-paralysis III. It is produced by lesions of the red (lower) nucleus with nerve III damage by tumour infiltration (nasopharyngeal tumours, pontine glioma), haemorrhages with potentially nerves VI, VII damage.(1)

At ocular level, there occur: common oculomotor paralysis (III) homolaterally associated with damage to the movement of convergence, elevation and depression of the eye, diplopia, palpebral ptosis, midriasis, cycloplegia, divergent strabismus.(2)

Systemically, there occur unilateral hyperkinesia - contralaterally: extremities hemiparesis, hemi-hypoaesthesia - ataxia, hemitremor when resting and intentionally.(1,2)

The Claude syndrome (lower red nucleus syndrome) is characterised by ataxia, hemianesthesia, oculomotor nerve palsy.

It is a paramedical mesencephalic lesion triggered in the middle brain. Often, the terminal branches of the paramedial arteries supply the lower part of the red nucleus.(2)

In terms of ocular symtpomes, one can mention the ipsilateral paralysis of the oculomotor and trochlear nerves (III, IV). Systemically, the following can be mentioned: controllateral limb ataxia (occasionally), contralateral haemyneaesthesia, slow trembling.(1)

The Nothnagel syndrome: It is a lesion occurring in the peduncle under decussation and is characterized by total paralysis of the nerve III and cerebellar ataxia.(2)

The cavernous sinus

The lateral wall of the cavernous sinus extends inferiorly from the tentorial margin and continues with the dura that covers the Meckel's cave and middle cranial fossa. The oculomotor and trochlear nerves penetrate the cavernous sinus roof. The carotid artery leaves the cavernous sinus at the level of the medial face of the anterior cliniod. The cavernous sinus extends from the upper orbital fissure up to the tip of the temporal bone. The pericavernous venous plexus extends up to the level of the round and oval foramen.(5)

The oculomotor (III), trochlear (IV), abducens (VI) and ophthalmic (V1) nerves converge anteriorly to the upper orbital fissure. Nerves III, IV and V1 are located at the wall of the cavernous sinus, in this order, from top to bottom. The nerve VI is deeply intracavernous located in the immediate vicinity of the intracavernous segment of the internal carotid artery (ICA). The venous space in the cavernous sinus communicates posteriorly with the superior and inferior petrosal sinus, basilar sinus, superior ophthalmic vein.(5)

The posterior syndrome of the cavernous fossa (Jefferson) or of the anterior broken hole (Bonnet) is another syndrome that associates a late damage to the third pair of nerves, respectively the pairs IV and VI, with the predominance of the Vth pair, which is totally involved. Causes of this syndrome are carotid aneurysm, tumours of the cavum or of the Gasser's lymph node, cholesteatoma of the bone tip.(1)

The middle syndrome of the cavernous fossa.

The origin of this syndrome refers to disorders of the carotid artery (aneurysms, thrombosis), or some sellar or parasellar tumours.(1)

Upper and middle skull base syndromes

The Forster-Kennedy syndrome: ipsilateral anosmia, ipsilateral optic atrophy, controlateral papillary edema.(5)

The superior orbital fissure syndrome (Rochon-Duvigneaud) consists of decreased sensitivity V, emeda in the upper eyelid, ophthalmoplegia, possible optic nerve involvement.

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Article received on 27.04.2017 and accepted for publication on 25.08.2017

ACTA MEDICA TRANSILVANICA September 2017;22(3):48-50

The etiology may be inflammatory, traumatic, tumour or vascular, sphenoid meningioma, carotid aneurysm, arachnoiditis.

At ocular level, the following can be noticed: -mild exphthalmia, palpebral ptosis, complete or partial ophthalmoplegia III, V, VI, decreased visual acuity, hipo/anesthesia in the V1 area, abolished photomotor reflex, abolished corneal reflex, papilledema, atrophy of the optic nerve.

Systemically, there occur decrease of frontal and ocular nasociliary sensitivity.(2)

The Jacob and Rallet orbital apex syndrome consists of signs and symptoms of the sphenoid fossa syndrome to which the optic nerve damage is also added, as well as the progressive blindness, deep exophthalmia, possibly pulsatile.(5)

The Tolosa Hunt syndrome mainly affects the cranial nerves III, IV, ophthalmic ram V, VI in the lateral wall of the cavernous sinus. It has as cause an aneurysm or cavernous sinus thrombosis, invasive tumours of the cavernous sinus and pituitary fossa, benign granulomas. It is clinically similar to the cavernous sinus syndrome. At ocular level, it is characterized by ophthalmoplegia and systemically, by ipsilateral trigeminal neuralgia.(2,5,6)

The Gradenigo Syndrome involves damage to the cranial nerves V and VI. The site of the lesion is at the top of the bony bone. The etiology of this syndrome is a bony osteitis or a tumour at the level of the internal auditory canal. At ocular level, it is characterised by ipsilateral abducens paralysis and systemically, by trigeminal neuralgia.(2,5,7)

The Raeder paratrrigeminal syndrome. The symptoms of this syndrome are the neuralgia in the optic nerve area (VI), the Claude-Bernard-Horner syndrome, ipsilateral nerve VI paresis.(5)

The Jaccoud syndrome is a petro-sphenoid spread syndrome; II, III, IV, V, VI nerves are damaged in the retrosphaloidal space by large middle skull tumours. It is mainly characterized by ophthalmic neuralgia and nerve III paresis.(5,6)

The Foix syndrome is a syndrome of the cavernous sinus characterized by damage to nerves III, IV, ophthalmic branch of nerve V and nerve VI at the level of the sphenoidal fossa, by an invasive sphenoid bone tumour or aneurysm, unilateral opthalmoplegia, neuralgia in the ophthalmic (V1) nerve, exophtalmia.(5.6)

Pathology of anterior and middle skull base:

The pathology of the anterior and middle skull base can be tumoral, characterized by pituitary adenoma, craniopharyngioma, optic/hypothalamic nerve glioma, esthesioneuroblastoma (olfactory neuroblastoma), tumours with rhinopharyngeal origin (nasopharyngeal carcinoma), tumours with orbit extension.(5) The vascular pathology is based on anterior communicating artery (ACoA) aneurysms, Anterior communicating artery (ACom) aneurysm, internal carotid artery (ICA) aneurysms, intracavernous internal carotid artery (ICA) aneurysms, ophthalmic artery aneurysms, pituitary artery aneurysms, distal anterior cerebral artery aneurysms, carotid-cavernous fistula.(5)

The fourth pair of cranial nerves

The Guillain Syndrome (upper cerebellar artery syndrome or vascular syndrome of the upper portion of the protuberant protrusion) consists of the paralysis of the fourth controllateral pair, the syringomyelic type dissociated hemianesthesia and the homolateral kinetic cerebral hemisyndrome.(1)

The Jefferson Syndrome (cavernous fossa posterior syndrome) and Rochon-Duvigneaud syndrome (syndrome of the sphenoid fossa) also involve the participation of the fourth pair of cranial nerves.(1)

The Jacod syndrome (Jacod, Petrosfenoid Carrefour) The orbital apex syndrome, constantly involves the paralysis of the 6th pair of cranial nerves. The Jacod syndrome is characterized by trigeminal neuralgia, enlargement of cervical lymph nodes, ophthalmoplegia, trigeminal neuralgia, optic atrophy. Orbital apex syndrome, also known as Jacod syndrome, is a collection of cranial nerve deficiencies associated with a mass lesion near the top of the orbit. This syndrome is a separate entity from the Rochon-Duvigneaud syndrome, which occurs as a result of a lesion immediately anterior to the orbital apex. Most commonly, the optic nerve is involved.(2,6,8)

The lesion involves the nerve II up to the nerve VI. The most common causes are large tumours in the cranial or temporal fossa that compress the nerves. More common is the malignant nasopharyngeal tumour originating in the lateropharyal area. The nerves that pass through the oval foramen, the round foramen, and the sphenoid fissure give rise to ophthalmoplegia. The optic nerve may also be involved.(2,4)

At ocular level, this syndrome is characterized by unilateral blindness and ophthalmoplegia with facial hemiplegia or trigeminal neuralgia; unilateral blindness, depending on optic nerve involvement, trigeminal neuralgia (ophthalmic branch) and nerve III paresis, Optic atrophy if nerve II becomes involved.

Systemically, trigeminal neuralgia occurs, as well as unilateral or bilateral enlargement of cervical lymph nodes.

The cavernous sinus syndrome consists of multiple paralyses of the cranial nerves III, IV, V, VI, Horner' pupil.

The fascicular (nuclear) syndrome is produced by hemorrhage, infarction, demyelination, trauma. At ocular lebel, Horner's pupil can be noticed controlaterally.(2,9)

The subarachnoid space syndrome is produced by forceps, tumours (pinealoma, meningioma), meningitis, neurosurgical trauma.(2)

Orbital syndromes: there can be seen multiple paralyses of cranial nerves III, IV, V, VI; Horner's pupil; proposis, chemosis.(2)

The sixth pair of cranial nerves

The Millard-Gubler's syndrome translates the involvement of the sixth pair in the portion that intersects the pyramid fasciculus not decussated yet. It is characterized by a cross, contralateral limb paralysis, and facial homolateral paralysis, as a result of damage to the nerves VI and VII and the cortico-spinal tract. It usually occurs as a result of a pontine infarction, with the involvement of the antero-inferior cerebellar artery. The paralysis of the oculomotor nerve is particular in the case of nuclear paralysis, as being parcellar because of its nucleus (which is made up of several cellular groups).(1)

At ocular level, there occur diplopia, internal strabismus, external right paralysis (frequently bilateral) and systemically, we can mention face paralysis, controllateral arm and foot hemiplegia.(2)

The Foville Syndrome (Peduncular Syndrome)

The causes that lead to the occurrence of this syndrome are: vaginal tumours, haemorrhages, multifocal sclerosis, unilateral obstruction of paramedian branches.

At ocular level, there can be seen: paralysis of nerve VI, of the nucleus and of the supranuclear field with the paralysis of conjugated movements on the lesion side, inability of eye abduction and deviation of eye, convergence is preserved. Nystagmus with large oscillations can also occur/

Systemically, peripheral paralysis of the facial nerve, contralateral hemiplegia.(2)

The Gradenig syndrome. In this syndrome, the nerves V, VI, VII, VIII are damaged at the level of the apex of the bony

bone through petrositis, tumours, acoustic neuromas.

Some of the possible causes are traumatisms/meningitis/cerebellar-cholesteatoma angle tumours. At ocular level, there occur ipsilateral paralysis of nerve VI (the involvement of nerve VI at the passage through the intracranial Dorello canal), internal right homolateral spasm, transient involvement of nerves III and IV, diplopia, pain in the Vth ophthalmic area with photophobia, lacrimation, reduction of corneal sensitivity occasionally and optic nerve involvement.

Systemically, there can occur ear infections with hearing impairment, mastoiditis, possible nerve VII palsy, possible signs of meningitis, trigeminal neuralgia, ipsilateral abducens paresis.(10)

Diagnosis of cranial nerves

The diagnosis of these conditions is based on anamnesis, clinical examination and imaging investigations - CT, brain MRI.

Treatment of cranial nerve syndromes

Treatment, progression and prognosis in various cranial nervous disorders depend on the nature of the pain that has led to the damage to the nerve or nerves.

REFERENCES

1. Arseni C, David M, Chiliman M, Glăvan I. I, Maretsis M; Neurooftalmologie; Editura didactică şi pedagogicăBucureşti; 1981; p. 30-36.
2. Dumitrache M. Sindroame şi boli Sistemice cu manifestări oculare; Editura Medicală Bucureşti; 2014. p. 304-310,419-422.
3. Dumitrache M. Tratat de Oftalmologie, Editura Univesitară Carol Davila, Bucureşti. 2012;(1):214-217.
4. www.mediculmeu.com›Endocrinologie si metabolism›Afectiunile nervilor cranieni Paraliziile multiple de nervi cranieni: Afectiunile nervilor cranieni. Accessed on 12.01.2017.
5. www.neurochirurgie4.ro/images/pdf/2014/etaj-anterior.pdf. Accessed on 23.01.2017.
6. www.romedic.ro/sindroame ale nervilor cranieni. Accessed on 4 Feb 2017.
7. Bălaşa R, Pascu I. Sindroamele neurologice majore, University Press, Târgu Mureş; 2006.
8. Bălaşa R. Major Neurological Syndromes, University Press, Târgu Mureş: 2012.
9. Bălaşa R. Nervii cranieni, University Press, Târgu Mureş: 2010.
10. Miller NR, Newman NJ, Biousse V et al (Eds.) - Walsh and Hyot's Clinical Neoroophtalmology: the essentials, Lippincott Williams & Wilkins, Philadelphia: 1999.