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PONTINE TEGMENTUM HEMATOMA

A CASE REPORT WITH THE "ONE-AND-A-HALF "SYNDROME WITHOUT PYRAMIDAL TRACT DEFICIT

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SUMMARY - The author reports the case of a 54-year-old male patient with a pontine hematoma and with the one-and-a-half syndrome, cerebellar ataxia and no signs of pyramidal tract involvement. The absence of involvement of the pyramidal tract in the case reported herein is likely due to variation in the vascular anatomy of the pons. The pathophysiologic mechanisms of the one-and-a-half syndrome and of the clinical findings recorded are discussed.

KEY WORDS: pontine hemorrhage, one-and-a-half syndrome, pyramidal tract.

Hematoma tegmentar pontino: relato de caso com a síndrome "one-and-a-half" e sem déficit piramidal

RESUMO - O autor relata o caso de um paciente de sexo masculino de 54 anos com hematoma pontino que se manifestou por síndrome "one-and-a-half" e ataxia cerebelar mas sem sinais de acometimento do trato piramidal. A ausência de acometimento do trato piramidal é decorrente possivelmente de uma variação da anatomia vascular do segmento pontino do tronco cerebral. Os achados clínicos do caso e os mecanismos fisiopatológicos da síndrome "one-and-a-half" são discutidos.

PALAVRAS-CHAVE: hemorragia pontina, síndrome one-and-a-half, trato piramidal.

The "one-and-a-half" syndrome is a clinical disorder of the horizontal conjugated ocular movements with horizontal gaze palsy in one direction and an internuclear ophthalmoplegia (INO) in the opposite direction. The full blown syndrome yields a total paralysis of one eye which remains fixed in the midline position (One) for all lateral movements and the other eye is only able to abduct exhibiting horizontal jerk nystagmus in abduction (Half). The syndrome is the result of damage to structures lying within the pontine tegmentum, namely, the paramedian pontine reticular formation, the abducens nuclei, and the internuclear fibers of the medial longitudinal fasciculus. The most common reported causes of this syndrome comprise injuries to the brain stem caused by strokes, multiple sclerosis, hemorrhages, gliomas, arteriovenous malformations, basilar artery aneurysm, metastatic melanoma, surgical procedures in the posterior fossa, ependymoma of the fourth ventricle, astrocytoma of the cerebellum, and head trauma[3,8,14,19].The clinical presentation includes diplopia, visual blurring, oscillopsia, "quivering" of the eyes. The neurological exam may show in addition to the classical signs of this syndrome, gazed-evoked upbeat vertical nystagmus, horizontal and horizonto-rotatory ipsilateral gaze nystagmus, spontaneous contralateral gaze nystagmus, skew deviation, convergence changes, exotropia, and orthotropia[3,19]. The ocular changes can be accompanied

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by other neurological signs: Horner syndrome, cerebellar ataxia, pyramidal tract changes, cranial nerves changes (II, V, VII, VIII, IX, XII)[4,14,17,19]. In the cases of pontine hemorrhagic strokes, the extension and localization of the hemorrhagic lesion determines both the extent of the clinical picture and the frequent association of gaze, cerebellar, and pyramidal tract signs.

This paper is meant to document a patient with peculiar clinical findings uncommonly reported in the literature.

CASE REPORT

CJF, a 54-year-old male was admitted to the emergency ward because of a headache sudden in onset, dizziness, nausea and vomiting, dysartria, right gaze diplopia and unsteady gait. Previous medical history was unremarkable. The neurological examination revealed both eyes resting below horizontal meridian in the primary position, bilateral vertical nystagmus and miotic isocoric pupils. There was a right peripheral facial palsy and decreased pain sensation on the left side of the face. Cerebellar testing yielded an ataxic gait with a falling tendency to the left and an index-nose dysmetria of the left arm. No involvement of the pyramidal tract was recorded in the neurological exam. Other than these findings, his physical exam was unremarkable and his blood pressure was normal. Twenty-four hours after admission, a palsy of the horizontal conjugated gaze to the right and an adduction palsy of the right eye were recorded. Convergence was impaired. Pupils were light-responsive bilaterally. Dysmetria became bilateral in nature and the patient could not stand up or walk without assistance. No motor deficit was recorded and the Babinski's sign was not registered bilaterally. A CAT scan and MRI disclosed a hemorrhage located on the right paramedian pontine tegumentum (Fig 1). Ten months after the stroke, the patient still showed some incapacitation to work due to ataxia and diplopia. The neurological exam showed only an abduction palsy of the right eye.

COMMENTS

1 - One-and-a-half syndrome

A review of the organization of the brain stem anatomic structures and mechanisms responsible for the control of horizontal ocular movements is useful at this point.

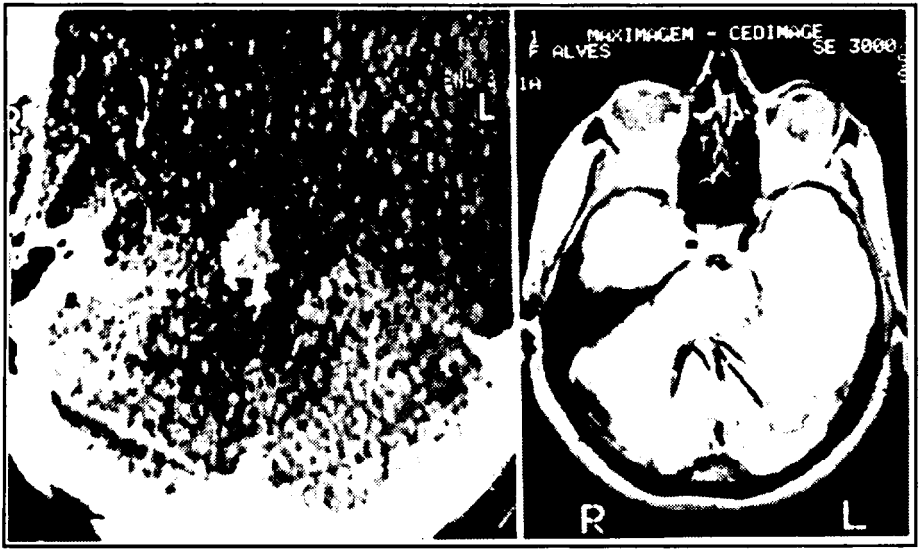

Fig 1. Patient CJF. CAT scan disclose a hemorrhage located on the right paramedian pontine tegmentum. MRI showing the same lesion one month later.

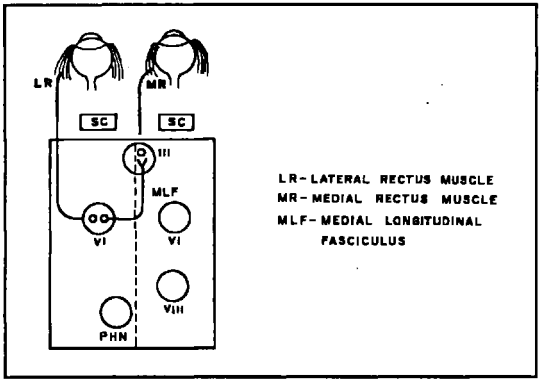

Fig 2. Diagram of structures involved in horizontal conjugate gaze[14].

The main structures involved in the organization of the horizontal eye movements are the abducens nuclei (VI cranial nerve), the internuclear fibers of the medial longitudinal fasciculus (MLF) and the paramedian pontine reticular formation (PPRF)[4,6,9,10,14,17,19] (Fig 2).

The abducens nucleus is located in the lateral region of the median pontine protuberance[5]. The VI nerve nucleus contains motor neurons for the lateral rectus muscle. It also contains excitatory interneurons that give forth fibers that decussate at the same topographic level of their parent cell bodies. These excitatory fibers ascend through the contralateral MLF terminating on cell bodies of neurons of the oculomotor nucleus that control the contralateral medial rectus muscle.

The medial portions of the nucleus reticularis magnocellularis have been denominated "paramedian pontine reticular formation" (PPRF). The PPRF is situated rostrally and ventrally to the abducens nucleus[19]. The PPRF contains two distinct types of neurons associated with eye movements. The first one is the phasic type of neuron that becomes activated during the voluntary saccadic eye movements and during the quick phase of nystagmus. The second type, the tonic neurons are activated during pursuit movements and during the slow nystagmus phase. The PPRF is connected to the ipsilateral abducens nucleus by means of a direct excitatory pathway. No direct inhibitory pathway from the PPRF projects to the contralateral abducens nucleus. The inhibition of the contralateral abducens nucleus is contrived by the gigantocellular tegmental field (FTG) that lies caudal to the PPRF[3,17,19] (Fig 3).

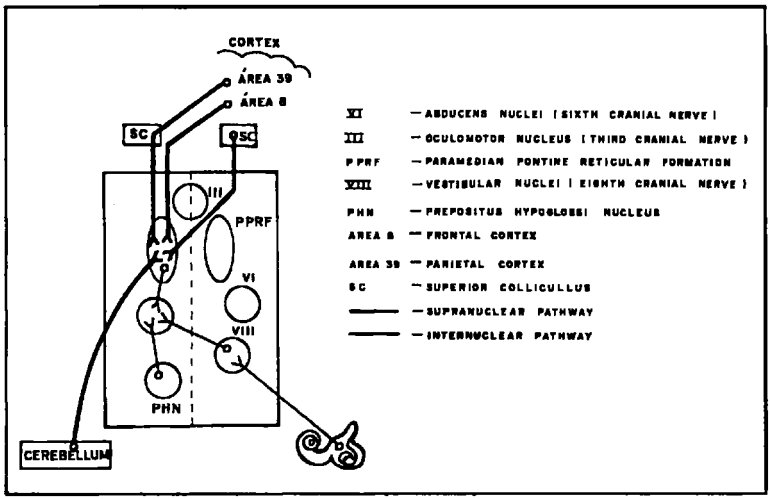

Fig 3. Diagram of brain stem pathways for horizontal eye movements[19].

In addition to the pathways mentioned, the abducens nucleus is linked to other excitatory and inhibitory pathways. The contralateral vestibular nuclei (these nuclei are responsible for the vestibulo-ocular reflexes) and the ipsilateral prepositus hypoglossi nucleus (involved with foveal pursuit) constitute the excitatory pathways to abducens nucleus[6,10,17,19]. The ipsilateral vestibular nuclei and the contralateral propositus hypoglossi nucleus constitute the inhibitory pathways to the abducens nucleus[6,10,17,19].

The PPRF is the premotor activating center for all ipsilateral horizontal ocular movements[10,14] with the exception of reflex ocular movements[10,17]. Its main afferent inputs are:

- Fibers from Brodman's area 8 in the frontal lobe. This area is responsible for voluntary conjugated ocular movements. These corticofugal fibers traverse the anterior limb of the internal capsule and bifurcate at the diencephalon in two separate bundles. One bundle is called dorsal transthalamic and this bundle is predominantly uncrossed. The other bundle is more ventral and crossed and it is called peduncular-capsular pathway. The uncrossed transthalamic pathway terminates in the pretectum area, superior colliculus, and periaqueductal gray matter. The ventral pedunculo-capsular pathway decussates at the topographic level of the troclear nucleus and terminate on the contralateral PPRF[1,3,9,18].
- Fibers from the superior colliculus. These fibers contour the periaquecductal gray matter laterally, decussate in the Meynert's decussation and descend through the medial tectospinal fasciculus terminating in the contralateral PPRF[10].
- Fibers from the posterior ipsilateral cerebellar vermis. These cerebellofugal fibers are not related to the generation of saccadic eye movements but to its coordination[3,10].
- Fibers from the posterior parietal cortex (Brodman's area 39) and from adjacent intraparietal sulcus. These fibers are involved in visually-guided saccadic eye movements[3,10].

Palsy of the horizontal conjugated ocular movements

There are four theoretical distinct possibilities to account for horizontal conjugated gaze palsy: A) a damage to both the abducens as well as PPRF nuclei; B) a damage to the ipsilateral abducens nucleus alone; C) a damage to the ipsilateral PPRF alone; D) a damage to the nerve roots of the ipsilateral abducens nucleus and damage to the contralateral MLF[17,19]. Damage to the abducens nucleus alone produces the arrest of all horizontal eye movements, voluntary and non-voluntary (reflex movements).

Unilateral damage to the PPRF produces two remarkable findings. With PPRF lesions rostral to the abducens nucleus, there is ipsilateral paralysis of the horizontal conjugated gaze (saccades and pursuit). This paralysis of saccades and pursuit movements is dissociated from vestibular-induced reflex horizontal eye movements where this type of movements are well preserved. On other hand, damage to PPRF at the level of the abducens nucleus produces ipsilateral abolishment of saccadic and pursuit voluntary eye movements as well as reflex eye movements[17,19]. A judgement based on clinical grounds to make out between damage of the abducens nucleus and the caudal portion of the PPRF may be difficult. Damage to the more rostral segment of the PPRF usually leave the reflex eye movements unimpaired. The presence of a peripheral facial palsy ipsilateral to the clinically determined PPRF damage strongly suggests an involvement of the VI nerve[17,19].

Palsy of the lateral gaze can also be caused by damage to the abducens motor fibers to the ipsilateral lateral rectus muscle and damage to the MLF fibers to the contralateral medial rectus muscle. In this case, there should exist two distinct anatomic damaged sites to account for palsy of the lateral gaze. A distinction based on clinical grounds between a damage to the nerve roots supplying the medial and lateral rectus muscles and an isolated damage to abducens nucleus can be made from the follow-up.

The persistence of lateral gaze palsy (medial and lateral rectus muscle) is consistent with one anatomic site of damage, the abducens nucleus. On the other hand, a desconjugate gaze palsy at any

time in the follow-up is consistent with damage to the nerve roots. In the case reported herein, there was a persistence of only the right eye abduction paresis. This finding is more suggestive of damage to the motor neurons rather than a damage to the abducens nucleus[17,19].

Internuclear Ophthalmoplegia - INO

It occurs as a consequence of a damage to the ipsilateral MLF. Clinically, the syndrome is characterized by an adduction paralysis of the ipsilateral eye on the attempted gaze to the contralateral gaze; a horizontal jerk nystagmus in the contralateral abducting eye, and typically convergence is intact if the lesion does not extend to the mesencephalon[19]. Internuclear ophthalmoplegia in its complete form does impair to some extent the horizontal pursuit eye movements but it definitely does not abolish them[19].

II - Anatomo-clinical correlation

Brain stem hemorrhage has been extensively reported in the literature. Several reports[2,4,11-13,15] specifically focus on the different pontine lesions and their clinical manifestations, etiology factors, treatment and prognosis. The clinical presentation of a pontine hemorrhage is extremely variable and it depends on the site of the hematoma and its extent[2,4,13,18]. Involvement of cranial nerve function in association with cerebellar and pyramidal tract dysfunction is the rule in the clinical picture[2,4,11-13,15]. The case reported herein showed clear signs of cerebellar, ocular movement, V and VII cranial nerve dysfunction. However, no pyramidal tract dysfunction was recorded throughout the clinical picture. This is an uncommon finding reported only once in the literature reviewed by the author[16]. The etiologic factors more commonly reported in the literature are, namely, hypertensive hemorrhages and arteriovenous malformations[12,15]. This patient the author reports, did not present hypertension either upon admission or during follow-up. Additionally, the lack of intraventricular bleeding and the favorable outcome on follow-up lead the author to believe that an arteriovenous malformation is the underlying etiology of the pontine bleeding in this case.

Specific clinical syndromes have been reported to exist in accordance to the extension of the brain stem involved by the hematoma[2,4,11-13]. Different clinical manifestations can result either from destruction of structures or from the compressive effects exerted by the bleeding upon brain stem nuclei and fibers. Masiyana et all. reported a direct relationship between the size of the hematoma and prognosis. Subjects displaying CAT-determined hematomas that did not exceed 1/4 of the overall diameter of the mid-pons exhibited a favorable prognosis[13].

Aleksic and Budzilovich[2], Caplan and Goodwin[4], Gillilan[7] gracefully discuss the anatomic correlation between brain stem blood vessels and different diagnostic possibilities. According to these authors, the distribution of the arteries, mainly under the intrinsic circulation viewpoint, composes distinct topographic zones that in turn may generate distinct clinical syndromes. The case reported herein displayed a CAT scan and MRI-determined hemorrhage located on the right paramedian pontine tegumentum (Fig 1). Involvement of the median perforating branches of the superficial arteries of the median line, that is, anterior spinal, vertebral and basilar arteries was likely. The pyramidal tract is irrigated mainly by median and paramedian branches along its course within the brain stem[4,7]. The involvement of the pyramidal tract may be produced either directly by an impairment of its arterial circulation or indirectly by its compression. In the case reported herein, there was absolutely no clear signs of pyramidal tract involvement in the pons, a finding usually not reported in the literature.

The total absence of pyramidal tract involvement herein can only be explained by a variation in the vascular anatomy of the pons.

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