BILATERAL INTERNUCLEAR OPHTHALMOPLEGIA AND CLIVUS FRACTURE FOLLOWING HEAD INJURY

Case report

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ABSTRACT - Internuclear ophthalmoplegia is a remarkable finding, particularly in patients victims of head injury. The medial longitudinal fasciculus, which is believed to be lesioned in cases of internuclear ophthalmoplegia, has an unique brain stem position and the mechanism involved in brain stem contusions implies a maximal intensity of shearing forces on the skull base. We describe a very rare association of bilateral ophthalmoplegia and clivus fracture following head injury, without further neurological signs. The patient history, his physical examination and the image investigation provide additional evidence to some of the mechanisms of injury proposed to explain post-traumatic internuclear ophthalmoplegia.

KEY WORDS: head injury, internuclear ophthalmoplegia, skull base, clivus fracture.

Oftalmoplegia internuclear bilateral e fratura de clivus após traumatismo cranioencefálico: relato de caso

RESUMO - Oftalmoplegia internuclear é um interessante achado clínico, sobretudo em pacientes vítimas de trauma cranioencefálico. O fascículo longitudinal medial, cuja lesão é responsável pelas alterações observadas na oftalmoplegia internuclear, localiza-se no interior do tronco encefálico. O acúmulo de forças de impacto geradas pelo traumatismo craniano na base do crânio é responsável pela formação de contusões no tronco encefálico. Descrevemos um caso em que se observa rara associação entre oftalmoplegia internuclear bilateral e fratura de clivus em um paciente com traumatismo cranioencefálico leve, sem outras anormalidades neurológicas. A história, o exame físico e a investigação imagenológica do paciente proporcionam evidências adicionais para alguns dos mecanismos propostos para explicar oftamoplegia pós-traumática.

PALAVRAS-CHAVE: traumatismo cranienecefálico, oftalmoplegia internuclear, base do crânio, fratura de clivus.

After head injury, ocular motor impairments occur in 3 to 7% of patients. Third, fourth and sixth nerve palsies are believed to account for the majority of cases, while skew deviation, nystagmus, supranuclear palsies and defective convergence are less often encountered¹. In acute trauma, the presence of an adduction deficit should raise the suspicion of internuclear ophthamoplegia (INO)². Classically described by impaired adduction on attempted lateral gaze, followed by horizontal nystagmus in the abducting eye with normal convergence, INO is caused by a lesion of the medial longitudinal fasciculus (MLF). The most common cause of bilateral INO is multiple sclerosis and unilateral INO is often due to vascular disease3-5. Other causes of INO include syphilis, brain stem neoplastic lesions, arteriovenous malformations myasthenia gravis, Arnold-Chiari malformation, and nutritional and metabolic encephalopaties^{1,4,6}. Head injury is a much rarer cause and frequently yields a rich clinical picture with a constellation of brain stem related signs among which INO is an outstanding feature.

Basilar skull fractures are difficult to diagnose with standard radiographic studies and are probably underdiagnosed in cases of head injury^{7,8}. Computed tomography (CT) with bone window settings and bone algorithms has made the diagnosis of these types of fracture more precise. Clivus fractures are nonetheless still difficult to diagnose because the clivus, as defined by the basioccipital bone and body of the sphenoid bony extends from the tuberculum sella to the foramen magno⁷, is located deep to the anatomical center of the skull base. Moreover, there is not a specific clinical finding associated with clival fracture.

Fracture of the clivus and INO have never or very scantly been described in association¹.

We report a case of this type and discuss mechanisms of injury.

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CASE

A 36 year-old patient was admitted for medical evaluation after have been involved in a one-vehicle bicycle accident, which rendered him unconscious for some minutes. On the emergency room, he was found to be awake and confused (Glasgow Coma Score 12), with a remarkable bilateral oculo-hematoma. Pupillary responses were normal and visual acuity was intact. Divergent strabismus was present, with a bilateral palsy for eye adduction on attempted lateral gaze with horizontal nystagmus of the abducting eye (Fig1). Convergence was normal albeit the altered level of consciousness made the examination difficult. Vertical gaze was not impaired. Neither motor nor sensory deficits were found. There were signs of ethanol intoxication. Investigation involved a radiographic study of the skull and cervical spine, which were normal. Cranial CT and magnetic resonance image (MRI) revealed a bilateral frontopolar contusion, a ponto-mesencephalic contusion, and diffuse pneumoencephalus (Fig 2). The bone window CT scan depicted a longitudinal fracture of the clivus (Fig 3).

The patient maintained an altered level of consciousness (Glasgow Coma Score 13 or 14) for 10 days after the admission. When the patient finally regained consciousness, he complained diplopia and headache. By that time, physical examination was not altered from admission, except for resolution of the oculo-hematoma. Twenty days after the admission the patient was discharged home without any improvement on extra-ocular movements. He was finally seen one month after the accident in the outpatient clinic, when he referred some improvement of his double vision. Physical examination revealed a slight amelioration of the strabismus.

DISCUSSION

INO is a rare complication of head injury. It may be directly caused by a brain stem lesion due to the impact, or indirectly result from the effect of mass occupying lesions and subsequent herniation^{9,10}. Most often bilateral, post-traumatic INO frequently yields complaints of diplopia in patients with mildly



Fig 1. Divergent strabismus without other cranial nerves abnormalities.

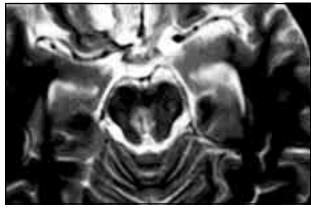


Fig 2. T2 weighted MR image showing region of increased sign within the mesencephalus.



Fig 3. Bone window CT scan showing a longitudinal trace of skull fracture.

impaired or intact level of consciousness¹. Brain stem signs are often conspicuous and INO tends to be a remarkable finding of a very rich clinical picture¹¹. Isolated INO is uncommon and it has been reported following mild head injury^{1,2}.

Some mechanisms have been proposed to explain the cause of INO after head injury. Lesion to the MLF is believed to cause INO. Located at the periventricular portion of the pons and midbrain, the MLF may be particularly more susceptible to the shearing forces generated by the trauma because these forces are known to be maximal near the ventricular system. A pressure gradient is formed after head injury and the forces applied to the skull tend to concentrate in the brain stem resulting in nerve fibers stretching and tearing around the ventricular system or in the white-gray matter transition¹². Another reason that explains the MLF vulnerability is the fact that the forces that are produced in the supratentorial space may cause a temporary downward displacement. The posterior portion of the brain stem is particularly more susceptible to displacements than the anterior portion because the latter is maintained fixed by the small perforators of the basilar artery^{10,12}. Finally, the

brain stem movement generated by the trauma may stretch the basilar artery perforating branches resulting in ischemia of the MLF, which is located in a vascular watershed zone of these arteries^{11,13}.

The clivus is described as the entire thickness of the basioccipital bone and the body of the sphenoid bone, extending from the tuberculum sella to the foramen magnum⁷. Due to its unique location at the very center of the skull, clivus fractures are difficult to be diagnosed8. They are generally recognized in the setting of some findings such as cerebrospinal fluid (CSF) leaks, cranial nerve palsies⁷ or oculo-hematoma. With standard radiographs, clivus fractures are believed to be underdiagnosed8, what has been overcome by bone window CT scan studies7. Three types of skull fractures have been reported: 1.longitudinal, which means that the fracture trace extends from the tuberculum sella to the foramen magnum; 2. transverse, near the spheno-occipital synchondrosis, from one carotid canal to the other; and 3.oblique, from the dorsum sella to the contralateral petroclival fissure. Regardless of the fracture type, the striking majority of patients have severe or moderate head injury, with an elevated number of unfavorable outcomes⁷. Among the survivors, multiple cranial nerve palsies, CSF leaks or diabetes insipidus are the most frequent sequelae. The most common mechanism of injury consists of motor vehicle accidents. The mechanical forces believed to be responsible for basilar fractures involving the clivus are the sum of outbending forces located far from the local of the impact that exceed the elastic capacity of the skull and result in a fracture line that runs through the clivus^{7,14}. These forces most often result from crushing the head in the lateral direction¹⁵. More precisely, the forces produced in an anterolateral impact that run through the lateral wall of the orbita or the sphenoid wing, or forces produced in a posterolateral impact and transmitted through the petrous ridge result in transverse or oblique fractures of the clival complex, while an occipital blow or an axial loading with trapping of the clivus between the vertebral column and the petrous bones result in longitudinal fractures¹⁶. The neurological injuries in patients with longitudinal fractures are usually quite severe due to the amount of energy transmitted to the brain stem⁷. Vertebral or basilar arteries entrapment inside the fracture¹⁷⁻¹⁹ or traumatic aneurysms²⁰ of the posterior circulation are other serious complications or longitudinal fractures. Cranial nerve palsies, in contrast, are more frequently seen in the setting of a transverse or oblique fracture due to the proximity of the cavernous sinus, the carotid canals and the nerves III through VI.

Internuclear ophthalmoplegia is a rare and remarkable finding in patients with head trauma. It has been scantly reported to occur in isolation, without other cranial nerves or long tracts signs, because the unique position of the medial longitudinal fasciculus within the brain stem. Moreover, the association between skull base fractures and internuclear ophthalmoplegia is a rare finding, especially in cases of moderate head injury. The present description of such association provides a new clue to one of the pathologic mechanism that may be involved in the formation of the pontine-mesencephalic lesion, which is the sum in the skull base of the elastic forces applied to the skull and the shearing forces applied to brain. As previously stated, this may be particularly important in cases of longitudinal fractures, when a considerable amount of energy is transmitted to the brain stem and the clivus. As a consequence, the observation of a clivus fracture, particularly those longitudinally oriented, should raise the suspicion of potentially life threatening brain stem lesions.

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