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RETROPULSION AND VERTIGO IN THE CHIARI MALFORMATION

Case report

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ABSTRACT - We describe a rare case of a 30 year-old woman with intense vertiginous sensation, lack of body balance and a tendency to fall backwards, making it necessary for two people to sustain her. The magnetic resonance imaging of the craniocervical junction evidenced tonsilar herniation at the inferior level of C1, and during the operation performed in sitting position, we observed crowding of the cerebellar tonsils at the level of C3. After the osteo-dural-neural decompression, the symptomatology remitted on the same day of the operation.

KEY WORDS: vertigo, retropulsion, Chiari malformation, cerebellar tonsils, posterior cranial fossa.

Retropulsão e vertigem na malformação de Chiari: relato de caso

RESUMO - Descrevemos um caso raro de mulher de 30 anos com intensa sensação vertiginosa, desequilíbrio do corpo e tendência à queda para trás, sendo necessário o auxílio de duas pessoas para ampará-la. A ressonância nuclear magnética da junção craniovertebral evidenciou herniação tonsilar ao nível da borda inferior de C1 e, durante a operação, em posição sentada, foi observado o deslocamento craniocaudal das tonsilas cerebelares ao nível de C3. Após a descompressão ósteo-duro-neural, houve regressão da sintomatologia, no dia da operação.

PALAVRAS-CHAVE: vertigem, retropulsão, malformação de Chiari, tonsilas cerebelares, fossa craniana posterior.

The neural dysgenesis, named afterwards as Chiari malformation (CM), was initially described by Cleland[1] (1883) and later by Chiari (1891)[2], that noticed in many hydrocephalic patients that the cerebellar tonsils migrated into the spinal canal. Chiari[2] also found at the necropsy of a 6 month-old infant an anatomic change in which the pons, medulla oblonga and the fourth ventricle were displaced down, to the level of the fifth cervical vertebra, in the spinal canal. Arnold[3] (1894) described a case of a lumbo-sacral myelomeningocele in which the cerebellar tonsils were found dislocated caudally to the mid cervical canal. This description is identical to the malformation described by Chiari[2]. Chiari[4] (1894) reported the anomalies of the hindbrain found in 63 cases of hydrocephalus and defined the spectrum of anomalies which is now recognized as Chiari malformation types I, II, III and IV.

In the original description, type I was characterized by downward displacement of the cerebellar tonsils and the medial portions of the inferior cerebellar lobes which accompanied the medulla oblonga into the cervical spinal canal. The type II showed downward displacement of the cerebellar tonsils, vermis and, at least, a part of lengthened fourth ventricle into the cervical spinal canal. In the type III, the hydrocephalic cerebellum, pons and medulla oblonga were inside a cervical meningocele (hydroencephaloceles cerebellaris cervicalis), through a spina bifida of the first three cervical vertebrae. Finally, in the type IV, there was hypoplasia of the cerebellum without herniation of cerebellar structures into the spinal canal.

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Otherwise, Schwalbe and Gredig[5], pupils of Arnold, considered that the anomalies previously described by Cleland, Chiari and Arnold consisted of a specific anatomical syndrome they designated "Arnold-Chiari" malformation. However, Carmel et al.[6] proposed the denomination Cleland-Chiari malformation, considering that Arnolds description was identical to the anomalies previously described by Chiari. Unfortunately the description of Cleland was forgotten in the world literature, while the concept "Chiari Malformation" was accepted in the same literature. It is a malformation of frequent occurrence, especially associated to the basilar impression (BI) and syringomyelia (SM). In northeast Brazil, the association of BI and CM presents high incidence, as observed in the studies of Canelas[7] and Canelas et al.[8], Caetano de Barros[9], Gonçalves da Silva[10], Taricco[11], Arruda[12,13], Carneiro Filho[14], Gonçalves da Silva et al.[15,16]. The symptomatology is characterized by a cerebellar syndrome, nuchal pain, vertigo and horizontal, downbeat or rotatory nystagmus, among others.

The publication of this case is based on the rareness of the clinical picture characterized by retropulsion and vertigo, which is not found in the consulted literature.

CASE

A 30 year-old woman was assaulted by a vertiginous crisis of great intensity, lack of body balance and tendency to fall backwards, needing double support in order not to fall. She mentioned a shock sensation when attempting to move her head forward or backwards, which began in the cervical region, spread down to her feet and, right after that, up to the head. The neurological exam evidenced a marked postural instability - the patient could not remain in orthostatic position without the help of two people - suppression of the nauseous reflex and discrete hypopallesthesia in the inferior members. Nystagmus was not verified. The magnetic resonance image (MRI) evidenced the herniation of the cerebellar tonsils (Fig 1). She was submitted to posterior fossa surgery, through the osteo-dural-neural decompression, technique used by Gonçalves da Silva[12] and Gonçalves da Silva et Holanda[13], characterized by an large craniectomy, dissection of the cerebellar tonsils, which reached C3 level (the caudal limit of the laminectomy) (Fig 2) and the regional arteries, large opening of the fourth ventricle, intrapial aspiration of the cerebellar tonsils, suture of the residual pial sacs to the lateral duramater in ascending position (Fig 3) and, finally, a dural grafting was performed with the use of bovine pericardium (Fig 4). In the present study, the cerebellar tonsils were loose and did not show any adhesions to the circumscribed tissues. The post-operative MRI revealed absence of the herniated cerebellar tonsils and formation of great cisterna magna (Fig 5).

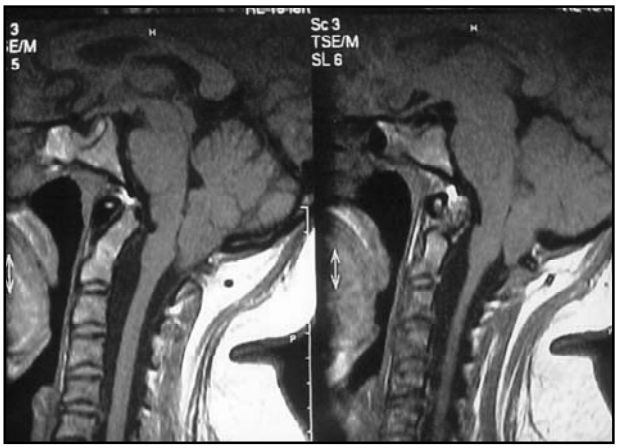

Fig 1. Herniated cerebellar tonsils up to C 1.

DISCUSSION

Among all symptoms and clinical signs observed in the CM, nuchal pain and the vestibular syndromes seem to be the dominant.

Brandt[17] described the several directions of falls in vestibular diseases: as lateral, in the vestibular neuritis and the Wallenberg syndrome; forward, in the benign paroxysmal positioning vertigo, and fore-and-aft in the bilateral vestibulopathy. The author, however, did not mention the vestibular syndrome in the CM. Paul et al.[18], in a 71-case CM casuistry, verified neck pain in 69% and lack of balance in 40% of his patients. Williams[19] related, in a 54-case casuistry, 64.8% of migraine and 91.1% of vertigo. Caetano de Barros[9], among 21 patients, mentioned neck pain in 57.1% and vertigo in 23.8%. Gonçalves da Silva[15], in a casuistry of 245 BI and/or CM and SM cases, observed neck pain in 53% and vertigo in 54.6%.

Some studies were mentioned in the literature, concerning rare signs and/or symptoms associated with the CM. Rullan[20] published a case of CM with bilateral laryngeal paralysis. Gol and Hellbusch[21] registered two cases of superior members paralysis. The authors admitted these clinical aspects were caused by traction of the cervical roots, which were initially directed upwards and then downwards, towards the neural foramina. Thomas and Boyle[22] related two cases of CM associated basilar migraine, admitting that some patients with this type of migraine could be CM carriers. Hudgins[23] related accesses of paroxysmal rage in two children with CM, presumably caused by menin-

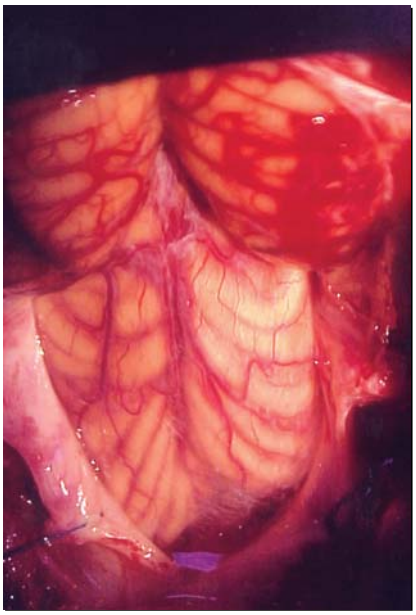

Fig 2. Dissection of the cerebellar tonsils with herniation up to C3, during the operation in sitting position.

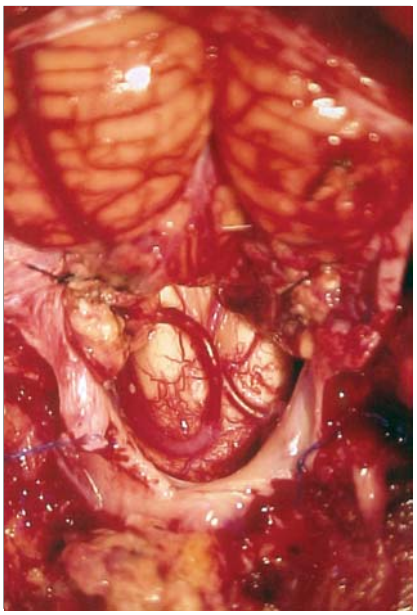

Fig 3. Tonsilectomy and large opening of the fourth ventricle and fixation of the residual pial sac to the lateral dura-mater in cranial position.

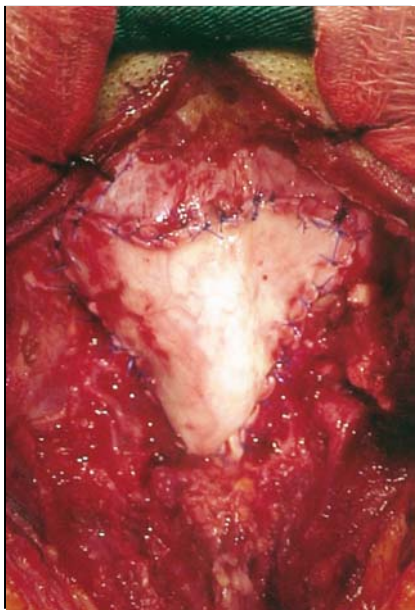

Fig 4. Dura- mater graft.

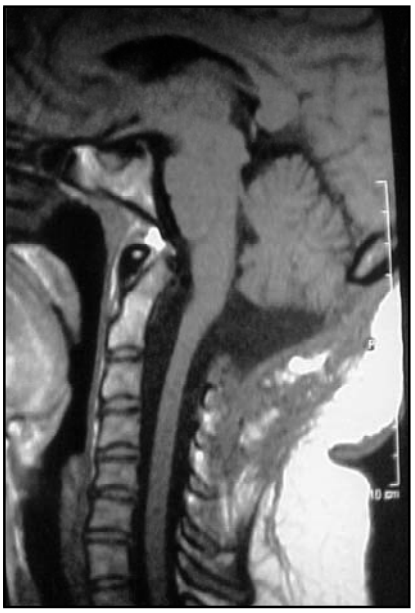

Fig 5. Postoperative MRI showing the recently created cisterna magna.

geal irritation. In both of them the choleric attacks regressed after the operation.

In the presently related case, the acute symptomatology of retropulsion and vertigo presents itself as a rarity in the clinical aspects of CM. Possibly, the thick cerebellar tonsils, functioning as an expansive process, could compress the vermis, which could likely be the cause of the retropulsion. DeJong[24] mentioned that in the cerebellar vermis and medial line lesions, the patient may not be able to remain erect, falling both forward and backwards.

On the other hand, the descending and ascending shock sensation could originate from the process of friction of the cerebellar tonsils on the cerebellum, medulla and especially the spinal cord, provoking irritation of the dorsal funiculi on the occasion of front or back flexion of the head.

As for the surgical treatment, we opted for large craniectomy of the posterior fossa, taking under consideration that the posterior fossa, in BI and/or CM and SM cases, is notoriously reduced in its volume, as described by Ackermann[25], Nyland and Krogness[26], Marin-Padilla[27], Marin-Padilla and Marin-Padilla[28], Schady et al.[29] and Vega et al.[30], among others. The large craniectomy involves the formation of great cisterna magna, a fundamental condition for the brainstem and cerebellum to migrate cranially. Let us cite as an illustration the studies of Badie et al.[31], which demonstrated the smaller size of the posterior fossa in the CM and that it increased in volume after the decompression. Milhorat et al.[32] verified a decrease of 13.4 ml in the volume of the posterior fossa and 40% (10.8 ml) in the cerebrospinal fluid volume of this region. Sahuquillo et al.[33] compared the results obtained in 10 cases in which a reduced craniectomy was performed with other 10 that were subjected to extensive craniectomy. Cranial migration of the cere-

bellum and brainstem was observed in all the later patients, while in the cases where reduced craniectomy was performed there was caudal migration in 7 patients.

The dissection of the cerebellar tonsils and of the regional arteries, especially the posterior inferior cerebellar arteries, as well as the large opening of the fourth ventricle and intrapial aspiration of the cerebellar tonsils, as demonstrated by Batzdorf[34], take part in the decompression process of the posterior fossa, therefore facilitating the circulation of cerebrospinal fluid from the fourth ventricle to the recently created cisterna magna, and so, preventing the reappearance of the craniospinal pressure dissociation.

Of further importance is the observation that even though the tonsils were located at the level of C1 on the preoperative MRI, intraoperative exploration, with the patient in sitting position reveled crowding of the tonsils at the level of C3. Probably the cerebellar tonsils have the tendency to migrate downwards on the orthostatic position. In the future, the introduction of MRI carried out in the upright position, will enable us to detect a difference on the topography of the cerebellar tonsils, between the dorsal position placement and the ortostatic position.

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