A Rare Cause of Cervical Pain in Elderly: Chiari Malformation Type I Associated with Basilar Invagination

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Abstract

Cervical pain is common in daily practice especially in the elderly in whom the etiologies are largely dominated by degenerative lesions. However, the practitioner should keep in mind secondary causes and search for them according to the clinical context. We report a case of cervical pain in a 67-year-old man whose etiology was a Chiari malformation type I associated with a basilar invagination.

Keywords: Cervical pain; Elderly; Chiari malformation type I; Basilar invagination; Brain MRI; Spinal cord MRI

Introduction

Cervical pain is a frequent reason for consultation in daily practice, particularly in the elderly [1]. Its etiologies are dominated by degenerative lesions of the cervical spine. It is important, especially in the elderly, not to be misled by images of osteoarthritis on X-rays and to know how to complete the lesion assessment at the slightest doubt in search of a secondary cause that may be traumatic, tumoral, infectious, metabolic and more rarely malformative [2]. We report a case of cervical pain in a 67 year-old man whose cause was a Chiari malformation type I associated with a basilar invagination.

Patient and Observation

A 67-year-old patient, a shopkeeper with no particular medical or surgical history and no ethylotoxic habitus had consulted in rheumatology for upper cervical pain with limitation of cervical movements and the occurrence of a sensation of instability when walking. On clinical examination, the patient had a good general condition with normal vital signs, Cervical and upper thoracic pain are classically found, associated with occipital headaches, visual variations such as basilar invagination which is defined as a developmental abnormality of the craniovertebral junction in adults [3]. Arnold-Chiari malformation is a congenital anomaly defined as a herniation of the contents of the posterior cerebral fossa through the foramen magnum. Type I is characterized by a downward herniation of the cerebellar tonsils and is defined as a displacement of the cerebellar tonsils more than 5 mm below the foramen magnum [4]. It is frequently accompanied by a wide range of additional anatomical variations such as basilar invagination which is defined as a developmental abnormality of the craniocervical junction in which the odontoid process extends abnormally upward and backward into the foramen magnum. [3,5,6].

The incidence of Chiari malformation type I is estimated at 1/1000 births [5], its association with basilar invagination varies from 12% to 35% according to the series [7,8]. The age of onset is usually between the third and fourth decade; there is a slight female predominance [4]; the advanced age in our case may have contributed to the misdiagnosis, however Zhuo-wei Lei et al report an extreme of 64 years, which is close to the case described in our observation [9].

The duration of symptom progression averages 60 months in adults [10]; and 15% to 30% of adult patients with Chiari malformation type I would be asymptomatic [11]. The symptomatology of Chiari malformation is polymorphic, with clinical signs usually discrete.

Discussion

Basilar invagination and Chiari malformation are the most common malformations of the craniocervical junction in adults [3]. Arnold-Chiari malformation is a congenital anomaly defined as a herniation of the contents of the posterior cerebral fossa through the foramen magnum. Type I is characterized by a downward herniation of the cerebellar tonsils and is defined as a displacement of the cerebellar tonsils more than 5 mm below the foramen magnum [4]. It is frequently accompanied by a wide range of additional anatomical variations such as basilar invagination which is defined as a developmental abnormality of the craniocervical junction in which the odontoid process extends abnormally upward and backward into the foramen magnum. [3,5,6].

The patient was rapidly referred to neurosurgery, a sub occipital craniectomy was performed. The immediate postoperative course was simple with a clear regression of the cervical pain.

Figure 1: Brain MRI Sagittal T2 weighted image (A) and axial T1 weighted image (B) showing a postion of the cerebellar tonsils through the foramen magnum with a tapered and pointed appearance (blue arrows) indicating a Chiari malformation type I.

Figure 2: Brain MRI Sagittal T2 weighted image (A) and CT scan of the brain in the bone window (B) in sagittal sections showing a high position of the odontoid, which is procident to the base of the skull (yellow arrows), the red line represents the Chamberlain line.

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al disturbances, instability when walking, vertiginous syndrome, sleep apnea, or signs of damage to the lower cranial pairs resulting in hypoaesthesia of the face, trigeminal neuralgia, hypoacusis, dysphagia, dysphonia, false alimentary routes, lingual atrophy and/or sternocleidomastoid atrophy or veil paralysis [4]. Basilar invagination is clinically manifested by a shortening of the neck, with asymmetry of the skull and/or face, painful limitation of cervical rachis movements. Neurologically, it may be responsible for damage to the mixed nerves, a pyramidal syndrome and deep sensitivity disorders [3]. This symptomatology corroborates that of our patient who presented both cervical pain with limitation of cervical spine movements and a cerebellar syndrome.

The diagnostic of certainty is based in all cases on MRI, which shows the descent of the cerebellar tonsils at a distance of at least 5 mm below the foramen magnum [3]. Basilar invagination is demonstrated on CT scans of the skull base in the bone window or on standard lateral views of the cervical spine, by studying the position of the odontoid in relation to the skull base using three reference lines: Chamberlain’s line, Mc Gregor’s line and Mc Rae’s line. In current practice, Chamberlain’s line is the most commonly used; it extends from the posterior part of the hard palate to the opisthion, which is the midpoint of the posterior border of the foramen magnum. The tip of the odontoid process is usually found below or on Chamberlain’s line. It is considered normal for the odontoid process to extend no more than 2.5 mm beyond this line. Basilar invagination can also be visualized on MRI [3,4].

Treatment is surgical, consisting of decompression of the posterior cerebral fossa with corrective surgery to limit the effect of the bone impaction of the axis on the brainstem; a regression of the algesic symptoms is generally observed after the surgery [12].

Conclusion

Cervical pain is a symptom and a very frequent reason for consultation in daily practice, of which the degenerative mechanical causes are by far the most frequent. In the elderly, given the high frequency of osteoarthritis and the often non-specific nature of the initial cervical symptoms, it is important to remain vigilant, with a systematic and rigorous clinical examination and a complete paraclinical work-up in order not to ignore symptomatic cervical pain, the prognosis of which could be pejorative if the appropriate treatment is not implemented.

Conflicts of Interest

We declare not to have any conflicts.

References