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A very unusual case of atlantoaxial dislocation

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Atlantoaxial dislocation is a rare disorder of the craniocervical junction characterized by C1-C2 instability and loss of normal articulation. Causes may be traumatic, inflammatory, congenital or multifactorial. Congenital malformations are usually early recognized. This is the case of an elderly, tetraparetic patient affected by a previously undetected atlantoaxial dislocation associated with very rare anomalies such as os odontoideum and malformation of C1. CT/MR scans showed also severe foramen magnum stenosis and myelopathy. The role of a superimposed traumatic injury of the upper cervical spine is unclear. In patients harboring congenital deformities of the craniocervical junction even a minor head-neck trauma can precipitate the onset of severe-to-catastrophic neurologic syndrome. The great importance of timely detection and management of craniocervical anomalies is discussed.

Key words: Atlantoaxial joint, Dislocation, Atlas, Os odontoideum, Cervical spine

INTRODUCTION

Atlantoaxial dislocation (AAD) is a pathologic condition caused by C1-C2 instability and consequent loss of normal articulation. Causes for loss of stability may be traumatic, inflammatory (rheumatoid arthritis), congenital or multifactorial. Pure traumatic atlantoaxial dislocations, which means in the absence of predisposing risk factors, are very rare. Congenital deformities of the craniocervical joint, which can occur in genetic and non genetic disorders (Down syndrome, Goldenhar syndrome, Ehler-Danlos and Marfan syndrome, skeletal dysplasias, mucopolysaccharidosis, osseous abnormalities), are usually early diagnosed. We are reporting the unusual case of an elderly patient affected by a previously undetected symptomatic AAD associated with rare disorders such as dystopic os odontoideum (OO) and unfused anterior and posterior arch of C1. The great importance of timely detection and management of patients with craniocervical junction abnormalities is also discussed.

CASE REPORT

This 78 year-old-woman was admitted to the department of internal medicine for a subocclusive intestinal syndrome. Besides congestive cardiomyopathy, pleural effusion and moderate cognitive impairment, mild tetraparesis and marked head movement restriction were pointed out. Chronic neck pain was reported. Our patient had lived for a long time in a catholic assisted living community and had no relatives. We had information about a few falls occurring in the past followed by the application of a cervical collar for a short period of time and progressive decline in mobility.

Static CT/RM imaging studies of the cervical spine showed a severe AAD associated with dystopic OO, congenital malformation of C1 (unfused anterior arch and hypoplasia of the posterior arch), severe foramen magnum/upper cervical spine stenosis and myelopathy (Figs. 1-2). Evoked potential studies were moderately abnormal. Craniometric values – Skull Basal Angle and Clivus Canal Angle – were within normal limits [1]. Since

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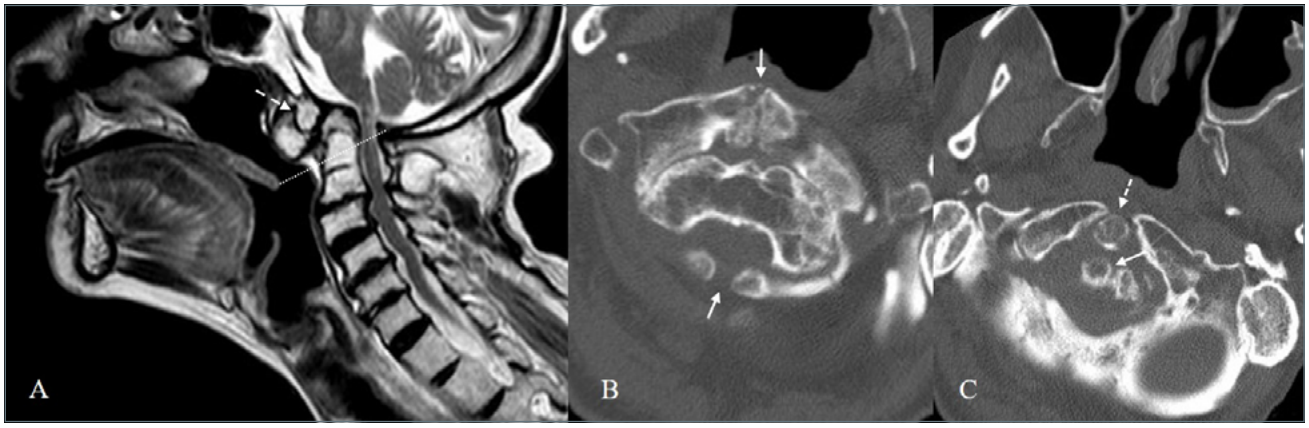

Figure 1. A. Foramen magnum stenosis, parenchymal enhancement of the medulla oblongata and upper cervical myelopathy. Increased atlanto-dental interval (6 mm). Dystopic os odontoideum (⇢) and basilar invagination (---- Chamberlain line). **B**. Unfused anterior and posterior arch of C1. Anterior arch is hypertrophic. **C**. Odontoid process (→) and os odontoideum (⇢).

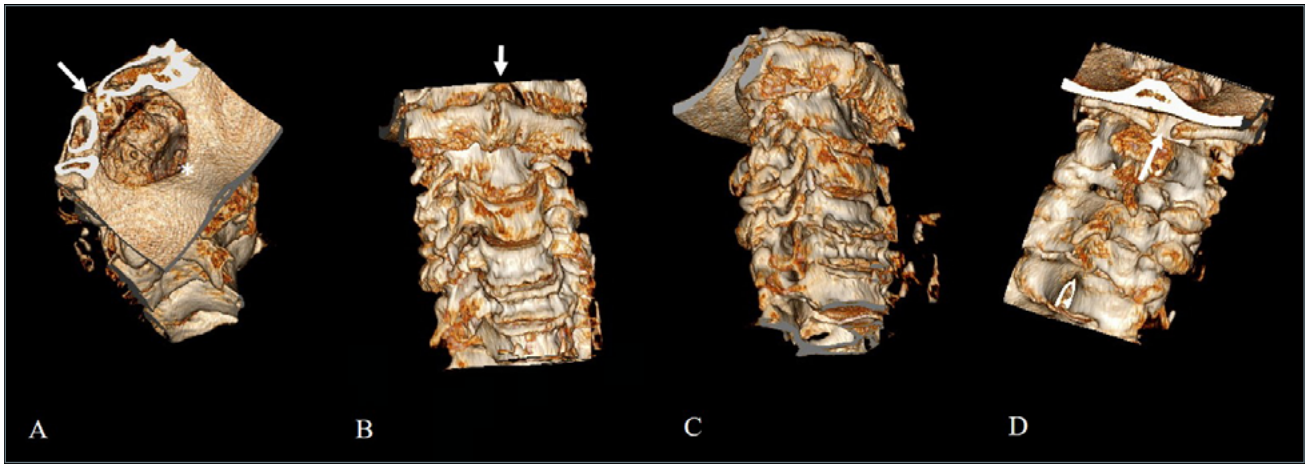

Figure 2. A. Dystopic os odontoideum (→). Severe foramen magnum stenosis (*). **B**. Os odontoideum (→). Unfused anterior arch of C1 which is also hypertrophic. **C**. Note the significant separation of the odontoid from the anterior arch of C1. **D**. Hypoplasia of the posterior arch of the atlas (type A) (→).

a poor clinical condition precluded surgical procedures, external immobilization with neck brace was applied. The patient died for sepsis two months after hospitalization.

DISCUSSION

Late diagnosis of a severe symptomatic malformation of the atlantoaxial joint is very unusual considering advances in medical care, innovation in healthcare technology, and heightened public awareness.

In this patient AAD is part of a complex congenital malformation of the craniocervical joint; however, the possible role of a superimposed traumatic injury of the upper cervical spine remains unclear. Unfortunately, earlier radiological studies are lacking and if they had been performed, it is very likely that the malformation was overlooked or improperly interpreted.

Etiology of OO, which is generally early diagnosed, is still controversial. For this poorly understood condition both traumatic and congenital causes have been hypothesized by several authors [2,3]. OO has been classified in two anatomical types – orthotopic and dystopic – based on the position of the dens tip [4]: orthotopic OO lies in the location of the normal odontoid process and moves with C2 and the anterior arch of the atlas; dystopic OO is diplaced and generally fused to the basion or to the anterior ring of C1. The natural history of OO is variable. Although it can be associated with atlantoaxial instability, predictors of patient deterioration have not yet been clearly identified [4]. Coexistence of OO

and bipartite atlas is quite rare. Midline anterior cleft of the atlas is a very rare congenital anomaly; it is present in 0,1-0,3% of population [5 6] whereas the incidence of posterior arch anomalies ranges from 0,7 to 4% [5 7 8]; posterior and anterior arch anomalies can sometimes coexist [9]. Congenital bipartition of the atlas in the absence of other malformations is generally a coincidental finding and requires no specific treatment.

Until now, various classification systems of AAD have been published [10 11]. First proposed in 1968, Greenberg's classification in two groups – reducible and non reducible – each connected to different treatment modality, has been generally adopted even though treatment strategies still remain quite controversial [12]. Of course, due to the limited number of cases, preparation of universally accepted guidelines is a demanding objective. Currently, management of AAD and associated neural compression – with or without OO – includes three key-stages: closed reduction attempt (cervical traction), orthotic immobilization and surgery in case of persistent instability or recurrence of deformity after a period of immobilization [4 10 12]. Nonoperative management with cervical traction and/or orthotic immobilization is considered a viable option only for asymptomatic patients provided that atlanto-dental interval is less than 5 mm in adults and less than 4 mm in children [12]; in these cases longitudinal radiological surveillance is also recommended. Surgical options include anterior approaches (odontoidectomy, transoral reduction plate, anterior transarticular C1-C2 screw fixation) posterior approaches (trans-articular and pedicle C1-C2 screw fixation, occipitocervical stabilization, bone decompression) and combined approaches. The best surgical technique is still debated; however, posterior fusion is regarded as the treatment of choice for reducible atlantoaxial deformity.

CONCLUSIONS

This case should serve as reminder that subtle, frequently age-related and slowly progressive signs and symptoms such as head-neck movement restriction, neck pain and headache, unsteady gate, numbness, weakness, lightheadedness, lower cranial nerves dysfunction, sphyncter dysfunction, presyncopal events need to be thoroughly considered and properly investigated. In patients harboring congenital o acquired unstable deformities of the craniocervical junction consequences of neglect may be extremely serious.

Since even a minor head-neck trauma can precipitate the onset of a catastrophic neurologic syndrome with quadriplegia or death [12 13], early detection, although challenging, is critical to ensure timely treatment and to prevent major complications. Plain radiographs, CT scans and MRI of the cervical spine provide all the information needed to plan appropriate treatment (upper cervical spine anatomy, presence of bony fusion, myelopathy, instability); if foramen magnum stenosis is diagnosed, Transcranial Doppler Ultrasound allows for noninvasive evaluation of vertebral arteries hemodynamics. On the other hand, it should be kept in mind that congenital anomalies of the craniovertebral joint as well as fragility/traumatic cervical fractures may be silent [12 13]. According to the Canadian C-Spine Rule, individuals over 65 years of age should always undergo a good quality 3 view X-ray study of the cervical spine (AP, lateral, odontoid) in the case of cervical trauma no matter the absence of specific symptoms [14].

As upper and lower cervical segments are the most difficult to visualize on radiographs, CT scan (or MRI) which is essential in the event of abnormal or doubtful X-ray findings, is strongly advisable also in the presence of incomplete radiological examination [13 15].

Conflict of interest

The Authors declare to have no conflict of interest.

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