Eur J Med Res (2008) 13: 136-138

A Delayed Diagnosis of an Unstable Os odontoideum

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Abstract
We report the case of a 2-year old child from Kuwait with known history of tetraparesis from birth. Radiological evaluation including computed tomographic scan and magnetic resonance imaging confirmed an unstable orthotopic os odontoideum which was complicated by cervical myelopathy.

Introduction

Os odontoideum is a separate ossicle of variable size over a dens with smooth corticated margins. Its etiology is debated controversially in the literature [1]. A congenital or a traumatic origin have been taken into consideration [1, 2]. Clinically, it may be either asymptomatic or it presents with signs of atlantoaxial instability and myelopathy [2, 3, 4]. Here, we present a case of an os odontoideum complicated by a congenital tetraparesis.

Case Report

A 2-year old girl from Kuwait was referred to the pediatric clinic for neurological evaluation. The child had a tetraparesis from birth. There was no history of trauma or surgical procedures. In Kuwait the diagnosis of periventricular leucomalatia was made. Routine laboratory test results were within normal limits.

A magnetic resonance imaging (MRI) of the head showed no abnormality. Sagittal T1 and T2-weighted MR images of the cervical spine demonstrated an ossicle (os odontoideum) separated from the body of the dens with a retrodental soft tissue mass restricting the spinal canal. The spinal cord signal was hyperintense on T2-weighted images at C2 (Fig. 1 A). A gadolinium-enhanced T1-weighted MR image did no show any abnormal enhancement (Fig. 1 B). A sagittal computed tomographic (CT) scan demonstrated a well corticated os odontoideum, located over a dens basis (Fig. 2A, B). There was no abnormality of the atlas. During a CT-scan the child turned the head by chance, leading to lateral dislocation of the os odontoideum (Fig. 3A). In the coronal T1-weighted image the os odontoideum was positioned centrally (Fig. 3B). Five days after admission the child was discharged from our clinic on request of the parents, and the family left Germany.

Discussion

Most authors indicate, that os odontoideum was first identified by Giacomini in 1886 [5]. However, according to LeDouble, this entity was first described by Bevan in 1863 [6]

The exact incidence of this event is unknown. In the study of Sankar et al. os odontoideum was diag-

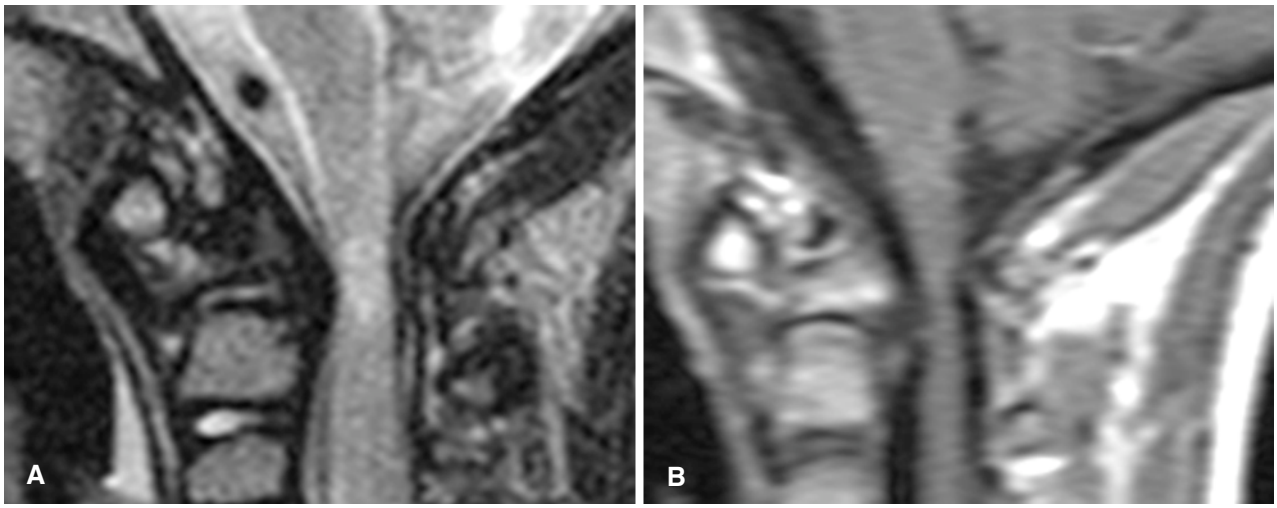

Fig. 1. MR images: A T 2-weighted image of the cervical spine shows os odontoideum, retrodental soft tissue mass narrowing down the spinal canal at C1-2, and a hyperintense spinal cord signal. B A gadolinium-enhanced T1-weighted MR image did no show any abnormal enhancement.

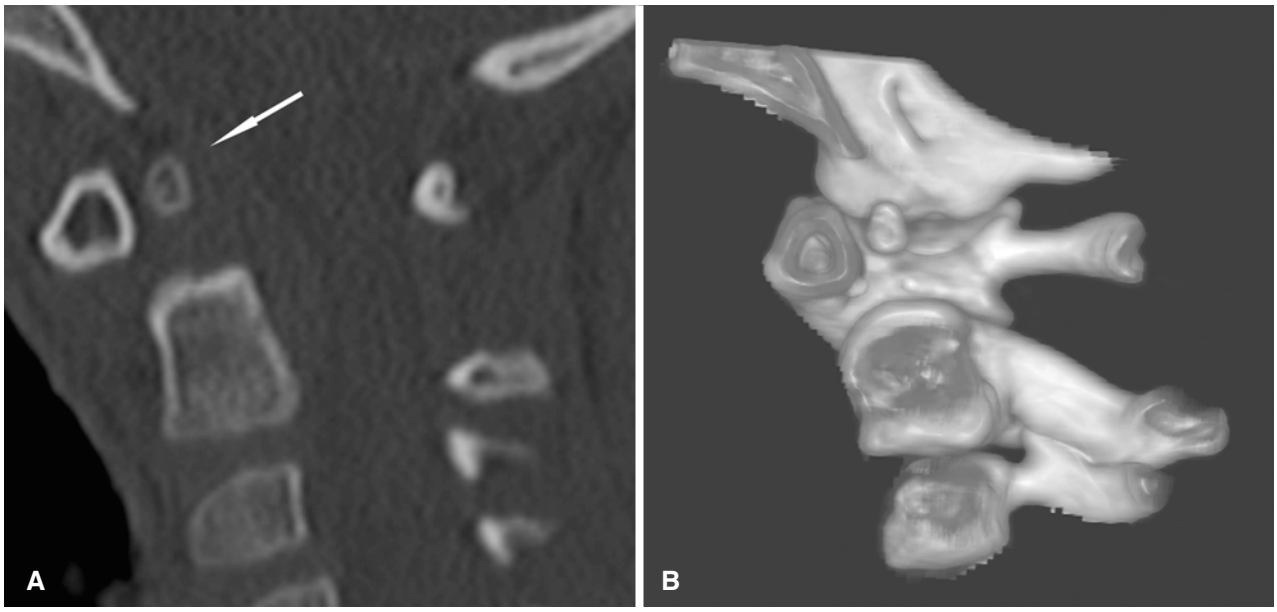

Fig. 2. CT scan: A CT-scan showing an oval well corticated os odontoideum (arrow), B Lateral view of a CT three dimensional reconstruction.

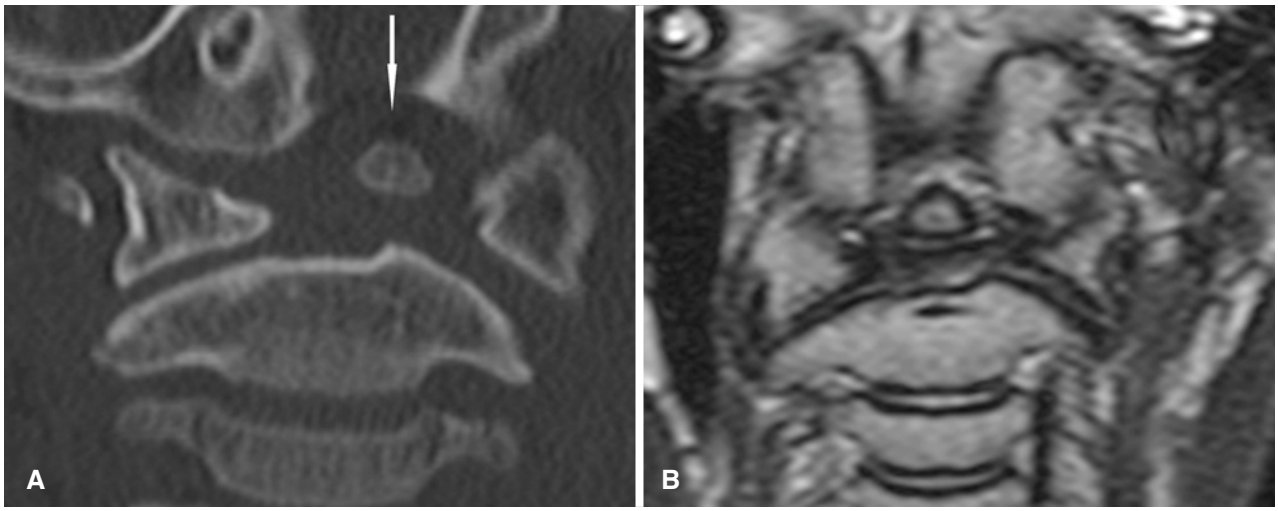

Fig. 3. Instability of the os odontoideum: A Dislocation of the os odontoideum to the left side during a rotation of the head (arrow), B Coronal T1-weighted image demonstrated a central position of the os odontoideum.

nosed in 3.1% of patients with abnormal cervical spine radiographs [1]. However, Lowry et al. identified os odontoideum in 11 of 25 children, in whom an upper cervical spine fusion should be performed [7]. It may be associated with other anomaly disorders such as Down syndrome, Klippel-Feil malformation, or Laron syndrome [2, 8, 9].

Os odontoideum may be due to an unrecognized fracture at the basis of vertebra 2 [2, 10], or it could represent a congenital abnormality, resulting from an incomplete fusion of the ossification center within the odontoid process [4, 11]. Our patient had no previous history of trauma.

Os odontoideum may be located orthotopic, in the normal position of the odontoid process, or dystopic, fused or articulated with the clivus [2].

Clinically, os odontoideum may be present with a broad spectrum of symptoms, ranging from neck pain to profound neurologic deficit [1, 12]. It may also be asymptomatic [1, 2, 12]. Clinical symptoms are related to atlantoaxial instability, resulting from os odontoideum, and spinal cord compression. In our patient instability of the orthotopic os odontoideum resulted in cervical myelopathy and tetraparesis. To our knowledge, this is the first report of a tetraparesis from birth due to os odontoideum.

Cervical Instability can be diagnosed by flexion and extension views with measuring of the anterior atlantodental interval [4, 13, 14]. Choit et al. suggested that MRI is not needed to visualize the instability [13]. However, the evaluation of the instability in dynamic cervical radiographic studies may be associated with hazardous complications. We did not perform this ex-

amination. In our case the patient rotated the head during CT-scan, and the os odontoideum dislocated here.

According to the literature, an operative C1-C2 fusion is recommended in patients with neurological symptoms [2, 3, 12]. Patients managed nonoperatively should be regulary monitored for signs of upper cervical instability or of myelopathy [4, 12].

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Received: August 10, 2008 / Accepted: January 9, 2008

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