

OS ODONTOIDEUM

A Cause of Atlanto-Axial Instability

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Os odontoideum is a rare condition in which instability may damage the upper cervical cord. A delay in diagnosis is not uncommon. This paper describes a series of 11 patients with os odontoideum. The presenting symptoms were divided into three groups: posttraumatic neck-pain, gradually appearing signs of medullary compression and an asymptomatic group. Eight patients had atlanto-axial instability. Six had interlaminar fusion between atlas and axis. No serious complications were seen. Stability was obtained in all patients postoperatively. In case of significant instability, fusion is indicated. In patients with cerebral palsy a closer examination of the upper cervical cord is recommended.

Key words: atlanto-axial instability; cervical vertebrae; congenital malformation; os odontoideum

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Os odontoideum is either a congenital malformation or an acquired lesion. A separate ossicle is located in the normal position of the odontoid process (orthotopic os odontoideum) or more cranially (dystopic os odontoideum).

The lesion was first described in 1863 by Bevan (according to LeDouble 1903) and recently by Fielding et al. (1980).

It is possible to diagnose an os odontoideum in a standard X-ray of the skull, but unfortunately it may be overlooked (Figure 1). The condition is rare and constitutes a risk for serious, possibly fatal, damage to the medulla.

PATIENTS AND METHODS

This study comprises 11 patients in whom an os odontoideum was diagnosed at the Odense University Hospital.

The symptoms can be divided into three groups: posttraumatic neck-pain (four patients), gradually appearing signs of medullary compression (four patients)

and an asymptomatic group, where os odontoideum was diagnosed only incidentally in connection with another examination.

Four patients showed neurological symptoms (Table 1): Case 1 had hyperaesthesia and paraesthesia in the upper extremities. Case 3 had a serious tetraplegia, and case 6 had quadriparesis. Case 10 is described below.

Although there was significant instability of the atlas and axis (Figure 2a, b) in some cases it had not caused any symptoms or signs before the diagnosis was established (Table 1).

Instability from 3 to 24 mm was found in eight patients. The lesion was considered stable in one case (Case 10), and in the last two patients the stability was not estimated. One of the latter died of cerebral lesions 5 days after a head trauma. One had os odontoideum diagnosed in 1963. At that time operation had not been considered. Eleven years later, at death, no symptoms of medullary lesion had appeared, and the cause of death had no relation to the os odontoideum.

In the group with gradually onsetting symptoms the lesions were diagnosed after a period of 3–32 years.

In two patients several roentgenographic examinations were carried out before the os odontoideum was diagnosed.

In two patients symptoms (spastic palsy) had already appeared in childhood (Cases 3 and 10).

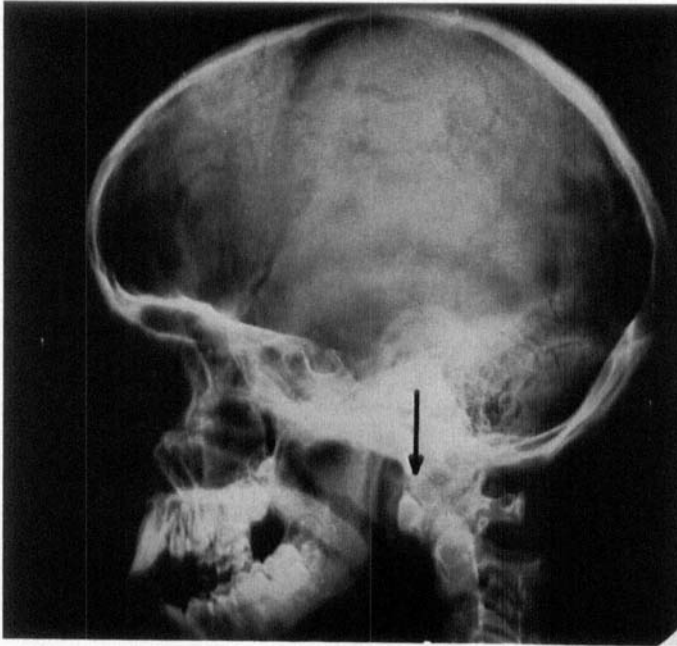


Figure 1. Case 8. Standard plain X-ray of the skull. An os odontoideum is visible (arrow).

Case 10: Three days after an uncomplicated birth a female child showed exanthema and raised cell counts in the spinal fluid. The child was suspected to have an infection and was treated with antibiotics.

At 12 days of age the child developed paresis of the extremities, and electromyography showed signs of polyradiculitis.

The paresis gradually diminished. The child was followed in the Centre for Spastic Children, where an os odontoideum was diagnosed by a screening for congenital cervical anomalies, when she was 6 years old.

Roentgenographically the atlanto-axial region including the dens was normal at the age of 12 days (Figure 3a). At the age of 9 months the X-ray were considered normal, but a later re-appraisal revealed a forward dislocation of the atlas.

At the age of 6 years the dislocation seemed to have an osseous fixation. Supplementary tomograms showed in addition to the os odontoideum an assimilation of the posterior arch of atlas and the occipital condyles. The distance between atlas and the odontoid process was 10 mm.

TREATMENT AND RESULTS

An operation was done on eight patients with instability to gain stability. In six an interlaminar fusion was performed (Sørensen et al. 1978). One had an occipitocervical spondylodesis a.m.

Patte and Rose (Patte & Rose 1969), and one an atlanto-axial osteosynthesis.

In one patient (Case 10) the condition was considered to be stable. In addition to the os odontoideum in this patient there was a forward dislocation of the atlas and assimilation of the atlas and occipital bone, which gave a very narrow spinal channel. A decompression by resection of the posterior arch of the atlas was performed. Osseous formation at the margin of the foramen magnum reappeared (Figure 3b). The patient died at the age of 11.

Except for one case with temporary light paresis of an upper extremity (Case 4) no complications were seen. In all patients with instability the operation achieved roentgenographic verified stability. The course in Case 10 is mentioned above.

The nine patients who were operated upon were followed at least 1 year (1–6 years) after the operation.

The motion of the cervical spine at the last control visit is shown in Table 1 and some reduction in motion was seen. No patient complained of pain and no patient had progression of any neurological symptom.

Table 1. Eleven patients with os odontoideum

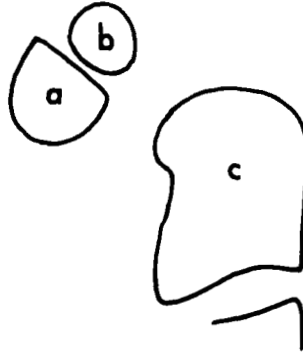
Case no.	Age (years) Sex	Symptoms	Years from onset	Neurological signs	Instability (mm)	Surgical method	Results	
							Movements*	Other
1	46 F	Neck-pain after trauma		+	6	Inter-laminar fusion	CM 3 5,15,20,25,25	
2	49 M	—''—		—	14	—''—	CM 3 30,20,20,30,40	
3	33 M	Cerebral palsy	32	+	12	Occipito-cervical spondylodesis + decompression	Flexion 40 30,30,30,35,60	Headache disappeared. Gait slightly improved
4	55 M	Paraesthesiae in upper limbs	4-5	—	5	Inter-laminar fusion	CM 0 30,20,15,30,35	Continued light paraesthesiae
5	49 M	Neck-pain after trauma		—	3	—''—	CM 5 30,10,10,40,30	
6	67 M	Increasing incoordination in the lower limbs	3	+	8	—''—	CM 0 45,20,20,20,20	The condition stationary
7	43 M	Neck-pain after trauma		—	24	Inter-laminar fusion	CM 0 35,5,5,15,10	
8	9 M	None (unconscious after trauma)		—	13	Atlanto-axial osteosynthesis	CM 2½ 75,0,30,30,45	
9	83 F	—''—		(+)	?	None	Death caused by head trauma	
10	6 F	Cerebral palsy	6	+	4-5 (see text)	Decompression	Progression of symptoms until death 11 years old	
11	44 M	None		—	?	None	Death caused by pulmonary cancer. No symptoms from the os odontoideum	

*Movements in the order: CM (= chin-manubrium distance in cm), extension, lateral flexion right and left, rotation right and left in degrees.

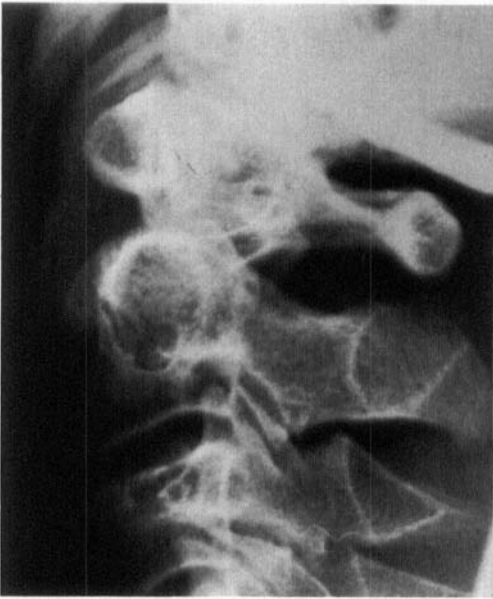
DISCUSSION

At about the fifth prenatal month the base of the cartilaginous dens shows two ossification centres. These centres fuse at the time of birth. The dens

is separated from the body of the axis by the sub-dental synchondrosis which can be seen until the age of 5-11 years. A separate centre near the tip of the dens, the ossiculum terminale, is frequently seen in children, and it is usually completely fused



2a

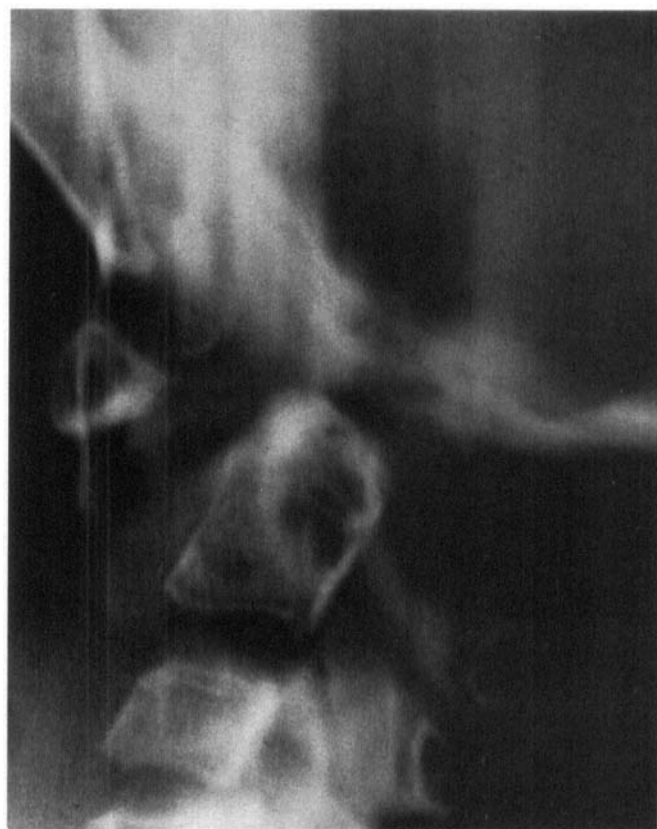


2b

Figure 2. Case 7. a = anterior arch of the atlas. b. = os odontoideum. c = dens. Figure 2a. Lateral view in flexion. The distance between the anterior arch of the atlas and the posterior border of the body axis is 36 mm. In Figure 2b the same distance is 12 mm in extension. A movement of 24 mm.



3a



3b

Figure 3. Case 10. a. AP-projection at the age of 12 days. A normal contour of the dens is seen. b. Lateral tomogram at the age of 8 and 3 years after decompression by resection of the posterior arch of the atlas and the posterior border of the foramen magnum.

with the dens by the age of 10–12 years (Torklus & Gehle 1970).

Fielding et al. (1980) showed roentgenologically that four of their 35 patients and five patients described by other authors on previous occasions had a normal odontoid process. Seventeen of the 35 had had an injury to the neck that required treatment by immobilization. Fielding et al. conclude that most cases of os odontoideum are acquired abnormalities secondary to trauma, perhaps to infection in some cases.

In our study three patients had been X-rayed earlier, and in one case the dens contour was normal at birth, but later on the patient developed an os odontoideum (Case 10, Figure 3a and b). None of these had had an injury to the cervical spine.

Torklus & Gehle (1970) published a case where an os odontoideum was found after tuberculosis of the cervical spine, and Gleason & Urist (1965) described a case of rheumatoid arthritis and odontoid separation.

In one of our patients (case 10) infection in the neonatal period seems a possible cause of the development of an os odontoideum.

Os odontoideum is seen more frequently in patients with diseases like Down's syndrome and, especially, Morquio's syndrome (Lipson 1977). Apart from the lesion in the atlanto-axial region only one congenital defect (bilateral coloboma in Case 11) was observed in our study.

Rezaian (1974) published a case of tetraplegia in a 6-year-old boy with congenital absence of the odontoid process. In this case all symptoms disappeared after occipitocervical fusion. Two of our patients were primarily considered to have a cerebral palsy (Case 3 and 10), but in both cases the symptoms and objective findings may have been caused by the os odontoideum.

When an asymptomatic os odontoideum is diagnosed, the question of whether an operation should be performed arises. Lipson (1977) recommends prophylactic posterior fusion at an

early age to avoid tetraplegia. Fielding et al. (1980) operated on 30 out of 35 cases, with three re-operations because of non-union. Minderhoud et al. (1968) prefer to operate but found a high risk of respiratory failure during or after the operation combined with aggravation of the neurological deficit in four out of nine cases and thus have recommended a more conservative treatment especially for patients with local symptoms, cerebral symptoms and in elderly patients with a transient myelopathy following injury and slight instability.

We recommend an estimation of the stability of the cervical spine through functional analysis and in cases of instability an interlaminar fusion of C₁ and C₂, which in our patients has proved to be almost without risk to them and has given satisfactory results (Sørensen et al. 1978).

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