Clinical History:

An 8-year-old boy presented with torticollis. There was a history of head injury a few years ago. (**Figure 1**).

Figure 1: Lateral radiograph of the cervical spine

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What is the diagnosis?

a) neurocentral synchondrosis
b) ossiculum terminale
c) acute fracture of odontoid process
d) os odontoideum

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Answer:

d) os odontoideum

Radiological findings

The lateral cervical spine shows an ovoid, corticated ossicle at the upper end of the odontoid process. The superior margin of the odontoid process is smooth and rounded. The os odontoideum, together with the arch of the atlas, is anteriorly subluxed in relationship to the rest of the cervical spine. No prevertebral soft tissue swelling is noted. (**Figures 2 & 3**).

Figure 2: Same radiograph as Figure 1. The os odontoideum (arrowheads) is displaced posteriorly in relationship to the rest of the C2 vertebra (C2 vertebral body = 2). The upper part of the remnant C2 vertebra is smooth and well-corticated. The anterior (a) and posterior (p) arches of the atlas are subluxed posteriorly together with the os odontoideum

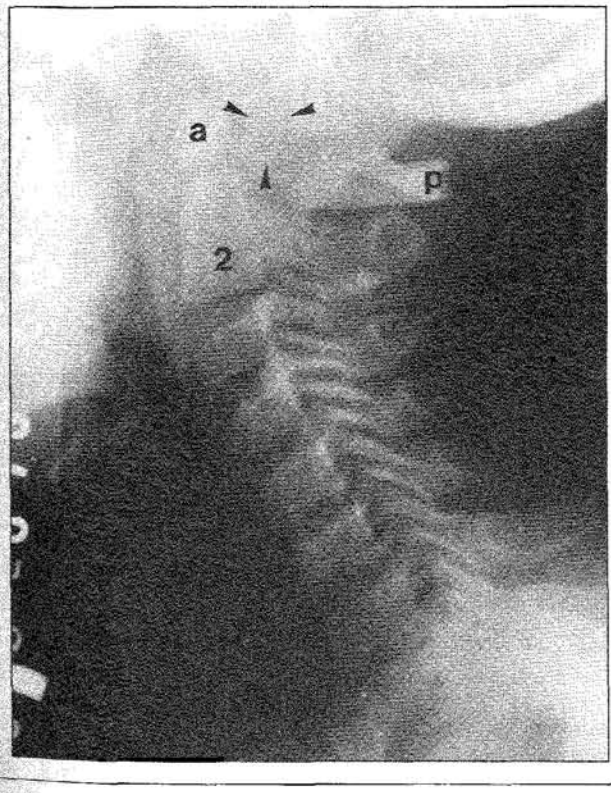
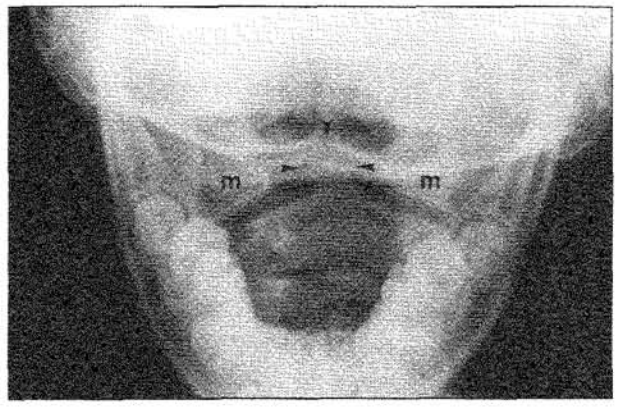


Figure 3: Open mouth view of the upper cervical spine showing the os odontoideum (arrowheads) clearly detached from the rest of the C2 vertebra. The gap is indicated by arrows. [Lateral mass of the atlas = m]



Discussion

Interpretation of abnormalities of odontoid process relies on understanding of the embryology and the postnatal development of craniocervical junction.

Neurocentral synchondrosis

Embryologically, the body of the odontoid process is derived from the mesenchyme of the first cervical sclerotome. Early in fetal life, the odontoid body is continuous with the centrum of the first cervical vertebra, from which it separates and fuses with the axis to become the odontoid process.[1]

At birth, the odontoid process is separated from the body of the axis by a wide cartilaginous plate. This epiphyseal plate does not lie at the anatomical base of the odontoid process at the level of the superior articular facets of the axis where most fractures are anticipated. Instead, it is well below the anatomical base and lies within the body of the axis. This normal epiphyseal plate (neurocentral synchondrosis) is present in nearly all children younger than four years and in 50% of children between four and five years old. It rarely persists after the age of six years.[1] In this case, the older age, as well as the higher position of the gap

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between the ossicle and the odontoid base, excludes this diagnosis.

Ossiculum terminale

The apex of the odontoid process is derived from the mesenchyme of the fourth occipital sclerotome or proatlas and is not ossified at birth. Radiologically, it is seen as a V-shaped depression in the odontoid process.[1] There is a secondary ossification center (ossiculum terminale) at the tip of odontoid process, which usually appears in most children between ages of seven and nine years.[2] It gradually enlarges and fuses with the body of the odontoid process by the age of twelve years. Failure of fusion may occur, leading to a small persistent ossicle at the tip of odontoid process. As the persistent ossiculum terminale is firmly bound to the rest of the odontoid process by cartilage, it is rarely of clinical significance.

In this case, the size of the ossicle in this film is too large for ossiculum terminale. The odontoid process is also abnormally short as compared with the well-formed odontoid expected in association with ossiculum terminale.

Acute fracture through the base of odontoid process

In this case, the well corticated margin of the ossicle and superior surface of the odontoid base are radiological features against those of an acute fracture. In addition, no prevertebral soft tissue swelling is evident. Moreover, the patient did not present with a history of recent neck trauma.

Os odontoideum

The radiological features of os odontoideum include a round or ovoid corticated ossicle situated superior to the body of the axis. The odontoid process is short with a smooth and rounded superior margin. The os odontoideum may be situated directly above the odontoid (orthotopic) or may be located near the base of occipital bone where it may fuse with the clivus (dystopic).[1]

The ossicle is fixed firmly to the anterior ring of the atlas due to an intact transverse ligament and moves with it in flexion and extension. The anterior portion of the atlas is usually hypertrophied.[3] The resultant atlantoaxial subluxation is in the anterior direction in the majority of patients. Instability of the atlantoaxial articulation is best assessed by flexion and extension views.

There remains considerable controversy concerning the aetiology of this condition, as to whether it is congenital or acquired. A number of reported cases in which individuals with a previously normal odontoid process subsequently developed an os odontoideum supports the concept of an acquired entity.[1,4] Moreover, the formation of os odontoideum is also difficult to explain embryologically. It is now generally accepted that os odontoideum is the result of unrecognised trauma in the infantile period.

The clinical presentation is variable. It may be an incidental finding in asymptomatic patients or may present after neck trauma with symptoms such as neck pain or torticollis. Os odontoideum must be suspected in a child with persistent neck discomfort and irritability.[4] Neurological deficit after the initial trauma is infrequent as instability develops gradually with os odontoideum. The prognosis is worst if there is insidious onset of progressive neurological impairment due to spinal cord compression.[5] Computed tomography and magnetic resonance imaging are very useful for delineation of the bony abnormality and the degree of spinal cord compression, respectively. ■

References

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