Os odontoideum associated with hypertrophic ossiculum terminale

Case report

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The authors report on the case of a 20-year-old man who presented with a transient tetraparesis. Neuroimaging studies demonstrated atlantoaxial dislocation and ventral compression of the rostral spinal cord caused by a quite rare association of os odontoideum and hypertrophic ossiculum terminale. The patient underwent removal of two free ossicula via a transoral approach and posterior fusion in which an autogenous bone graft was placed.

The majority of cases of os odontoideum are believed to be an acquired form; however, controversy with regard to the congenital causes of os odontoideum remains. One hypothesis is that os odontoideum results from the failure of fusion and the hypertrophy of the proatlas, although considerable confusion surrounds this hypothesis because definitive classification of os odontoideum—to differentiate between similar anomalies—has not been established. This rare coincidence in the current case supports the belief that os odontoideum has a different embryological origin from ossiculum terminale, which is thought to be a proatlantal remnant.

Key Words • atlantoaxial dislocation • craniovertebral junction • odontoid fracture • os odontoideum • ossiculum terminale • occipital condyle

A wide variety or association of congenital and/or acquired anomalies can affect the CVJ. The persistence of the terminal ossicle and os odontoideum are well known and much less common atlantoaxial abnormalities; however, the association of these two anomalies in one axis is quite rare.

In this report, we describe a case of os odontoideum associated with hypertrophic ossiculum terminale, and we discuss the causes of os odontoideum, focusing on its embryological origin.

Case Report

Presentation. This 20-year-old man suffered from neck pain and a transient tetraparesis after he performed a “header” during a soccer match. He had no significant history of head or neck injury.

Examination. Neurological examination on admission was unremarkable. Plain x-ray films and tomograms revealed two separate ossicula with intact cortical margin above the remainder of the odontoid process and behind the hypertrophic anterior arch of the atlas (Fig. 1). Flexion–extension tomograms demonstrated that the atlas and two ossicula moved relative to the remainder of the odontoid process by 7 mm anteriorly on flexion and 2 mm posteriorly on extension. The spinal canal had a minimum diameter of 11 mm between the apical ossicle and the posterior arch of the atlas. A remnant of the neurocentral synchondrosis was seen between the odontoid base and the axial body. Coronal reconstructed CT scans revealed three-part separation of the odontoid process and the scoliotic axial malalignment of the three parts, as well as asymmetry of the lateral mass of the atlas (Fig. 2). Magnetic resonance imaging demonstrated ventral compression of the cervicomedullary junction caused by two anomalous ossicula (Fig. 3). A remnant of the synchondrosis was also seen at the anatomical base. An MR angiogram revealed no vascular anomaly. No attachment of the anomalous ossicula to the surrounding bone structures including skull base was detected on neuroimaging.

Operation. After a diagnosis of atlantoaxial dislocation due to os odontoideum associated with hypertrophic ossiculum terminale was made, we performed a transoral decompressive procedure and posterior fusion in which an autogenous iliac crest bone graft was applied. During the
transoral resection, two ossicula were shown to have intact smooth cortical margins and no attachment to surrounding bone structures. A fibrocartilaginous layer was found between the apical ossicle and the posterior arch of the atlas (PA). A persisting remnant of the neurocentral synchondrosis (arrow and dashed lines) is seen between the odontoid base (OB) and the body of the axis (BA).

Postoperative Course. The patient was placed in a halo brace for 3 months and discharged without neurological deficits.

Discussion

A wide variety of congenital and/or acquired anomalies can affect the CVJ. Although most lesions in this region are thought to be congenital forms, the cause of os odontoideum has remained controversial since it was first described by Giacomini in 1886. In the past, proponents of the hypothesis that os odontoideum is congenital in origin considered the failure of the fusion of the ossification center within the odontoid process. Recently, considerable published evidence on cases of acquired os odontoideum indicates that an unrecognized occult fracture and subsequent avascular necrosis might cause the lesion. The fact that definitive classification of os odontoideum—to differentiate among similar anomalies such as third occipital condyle, ossiculum terminale, and old odontoid fracture—has not been established has led to considerable confusion with regard to the causes of os odontoideum.

On an embryological basis, Greenberg classified os odontoideum as Type I congenital incompetence of the odontoid process in which the bone separated below the level of the superior articular facet of the axis, and he classified ossiculum terminale as Type II congenital incompetence of odontoid process in which the bone separates above the superior articular facet of the axis. He, therefore, thought that Giacomini’s original illustration should not be termed os odontoideum but ossiculum terminale. In the majority of cases reported as os odontoideum, the
dislocation line was above the plane of the superior articu-
lar facet of the atlas; and 3) the tip of the odontoid process.

Embryologically, the “proatlas,” which is derived from
the cranial portion of the fourth occipital sclerotome,
mainly divides into three parts and forms the following
bone structures: 1) the lower portion of the clivus and the
occipital condyles; 2) the dorsal cranial articular facets of
the atlas; and 3) the tip of the odontoid process. 

Because of such a complicated developmental process,
the proatlantal remnant is thought to have
various manifestations. 

Because the failure to integrate with the clivus of the cranial part of proatlas results in the
third occipital condyle (condylus tertius), the basioccpital
fissure, the basilar process, and some other malformations
summarized as the manifestation of the occiput. Ossi-
fication of the caudal part of proatlas begins in humans at
age 3 years and fuses with the odontoid process between
ages 10 and 13 years. The failure of this portion to fuse
with the remainder of the odontoid process results in per-
sisting ossiculum terminale. This anomaly is usually sta-
ble and of little clinical significance because the separa-
tion lies above the anterior atlantoaxial joint, where the
odontoid process is fixed to the atlas by the transverse lig-
ament. Cases of atlantoaxial dislocation due to ossiculum terminale have been reported in association
with a specific condition such as mongolism or traumatic
avulsion.

The embryological origin of the major part of the odon-
toid process (odontoid base) is the “primitive atlas,”
which derived from the caudal portion of the fourth occi-
pital and the cranial portion of the first cervical sclerotomes. The odontoid base fuses with the body of axis,
which is derived from the “primitive axis” when a child is
4 years of age. A rudimentary cartilaginous disc may
remain until advanced age in this junction, and the pres-
ence of this fragile cartilaginous plate is thought to be
the reason why the Type II odontoid fracture is much more
common in children. Proponents of the congenital ori-
gin initially considered that the os odontoideum was the
main part of the odontoid process that failed to fuse at this
anatomical base, although they could hardly explain the
inconsistency of the line of cleavage.

The radiological features by which an os odontoideum
and a fracture of the odontoid process are distinguished
have been emphasized; however, radiologically it may be difficult to distinguish os odon-
toid from a long-standing odontoid fracture.

In the last four decades there have been numerous
reports in which there was radiological evidence of an
intact or normal-appearing odontoid process prior to
the development of the os odontoideum.

In these cases, the initial traumatic event was not al-
ways associated with an obvious odontoid fracture, and
vascular necrosis of the odontoid process, not non-
union, has been suggested to be the cause of os odon-
toid. The vascular anatomy of the dens has been described in
detail elsewhere. 

When the ascending arteries, mainly from the vertebral arteries, are compro-
mised at the odontoid base by displacement, overstretching,
or compression due to fracture, hemorrhage, or dislo-
cation, the vascular supply of the odontoid becomes
dependent on the supply via the apical arcade. In this case
ischemic necrosis may occur in the watershed area of odon-
toid process, where the line of cleavage is usually found
in os odontoideum. This condition is often associated with the
hypertrophy of the separated ossicle or anterior arch of the
atlas because of the preservation of blood supply from the
apical arcade via the apical or alar ligament. The corticated
appearance of the ossicle and the presence of the joint carti-
lage between the ossicle and the remainder of the odon-
toid process, which have been thought to be the evidence in
favor of a congenital origin, may be explained by a repara-
tive process after the avascular necrosis occurs in the grow-
ing individual. It is likely that there is a critical age
when the vascular supply and development of the odontoid
process are such that traumatic injury may compromise
the blood supply and result in resorption of the odontoid
base because most documented cases of acquired os odontoideum follow trauma in patients before the age of
3 years. With or without radiological evidence of odontoid fracture, the specific feature such as a
retropharyngeal swelling observed after the initial trauma is thought to be consistent with an injury to the vascular supply of the developing odontoid process. Infectious and/or inflammatory disease, such as tonsilitis, rheumatoid arthritis, or other deep cervical infections, can result in the same condition. Moreover, if the blood supplied not only by the ascending artery but also the apical arcade is compromised simultaneously, or if severe infection or inflammation is inflicted on the vascular damage, the bone absorption of the odontoid process may be so excessive as to result in a total absence of the odontoid process.

The relatively infrequent association of other anomalies is thought to support the belief that os odontium is an acquired lesion. Furthermore, reports that it is commonly associated with Down syndrome, Klippel–Feil malformation, Morquio syndrome, or other anomalies can be explained by the occurrence of avascular necrosis due to increased laxity of ligaments and atlantoaxial instability.

Based on this evidence, we and many authors believe that in the majority of cases os odontoideum is an acquired lesion. There remain, however, a few unresolved issues supporting the congenital origin of os odontoideum. One hypothesis is that the os odontoideum results from the failure of fusion and the hypertrophy of the proatlas associated with hypoplasia of the main portion of the odontoid process, a variant of ossiculum terminale.

In the patient in the present report a unique association of two anomalous ossicula of the odontoid process was demonstrated. Little information was available about such a rare condition. In 1974, Wackenheim reported a similar illustration of this anomaly with radiological evidence and proposed the term “dens tripartitus” for such a coincidence of anomalies in the same adult. Atlantoaxial dislocation observed in the present case was a consequence of the bone separation between the caudal ossicle and the remainder of the odontoid process. Given the radiological and operative findings, we thought that the apical ossicle was a hypertrophic ossiculum terminale and the caudal one was an os odontoideum. Because the apical ossicle was not related to the atlantoaxial instability, it was less likely that the caudal ossicle was precipitated by the avascular necrosis due to the presence of the apical ossicle. Of note in our case was an additional neuroradiological finding: the line of cleavage of os odontoideum could be distinguished from a persisting remnant of the neurocentral synchondrosis. This finding suggested that os odontoideum in this case was neither the consequence of the failure of fusion of the ossification center at the anatomical base nor the nonunion Type II odontoid fracture. The significance of the present case lies in the fact that os odontoideum can be associated with ossiculum terminale. In other words, these two anomalies have different embryological origins, and os odontoideum is less likely to be a remnant of the proatlas. Therefore, some possibility of a congenital origin was thought to exist, because this

Fig. 4. Postoperative neuroimaging studies. A: Sagittal T1-weighted MR image. B: Axial CT scan obtained at the level of atlas after total removal of two free ossicula via the transoral approach. C: Axial CT scan obtained at the level of axis after posterior fusion in which iliac crest bone graft and sublaminar wiring were used to achieve fixation.
coincidence of anomalies could not be explained sufficiently by the theory of acquired os odontoideum, including the unrecognized neck trauma in his early childhood, although the evidence for such a conclusion is weak.

Several questions remain to be answered: 1) the propriety of such a hypothesis that os odontoideum may result from the failure of migration of the ossification center,19 2) the reason for the increased prevalence of os odontoideum associated with third occipital condyle that may lead to limitation in the range of motion of the CVJ;26 3) the reason a preponderance of males are affected;11 4) the reason odontoid dysplasia is more frequently associated with Morquio syndrome, although the patients with this disease often appear normal until the age of 2 or 3 years and the associated atlantoaxial instability is usually mild;29 and 5) the cause of the anomaly of the odontoid process demonstrated in few-month-old children with floppy infant syndrome.14,21 In addition, we would like to emphasize that the definition of third occipital condyle, ossiculum terminale, and os odontoideum, including the orthotopic and dystopic variations,16,26 should be again standardized to discuss the remaining controversies.

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References


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