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<td>Author(s)</td>
<td>MATSUDA, MASAYUKI; KOYAMA, TSUNEMARO;</td>
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<td>HANDA, JYOJI</td>
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The "Os Odontoideum"
Report of a Case
by
MASAYUKI MATSUDA, TSUNEMARO KOYAMA and JYÖJI HANOA
Department of Neurosurgery, Kyoto University Medical School
(Director: Prof. Dr. HAJIME HANDA)
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The os odontoideum was described as early as in 1863 by BEVAN2l, and in 1886 by GIACOMINI7l. In 1968 GREENBERG9' collected from the literature 46 cases including his own 4 cases. Thereafter several reports12,13,16,17,20 appeared, and we now report an additional case of this infrequent congenital anomaly of the odontoid process.

Case report:
A 15-year-old boy fell head-on during gymnastic hours at school. He was not rendered unconscious, but had neck pain and numbness in the upper extremities. Numbness subsided spontaneously in 10 minutes, but the neck pain persisted. On physical examination 1 hour after the accident, the right sternocleidomastoid muscle was slightly swollen and painful. Movement of the neck was not restricted, and no neurological abnormalities were found. Roentgenograms of the cervical spine revealed an os odontoideum, a moderate anterior dislocation of the atlas on the axis on neck flexion, and a slight posterior displacement on extension (Figs. 1 & 2), but there was no lateral displacement. He was treated conservatively and free from neck pain in a week. Since there were no signs and symptoms referrable to the condition, no specific therapy was recommended. The patient was told to be cautious against a trauma to the head and the neck, and discharged from the hospital. His family members were examined roentgenographically, but no abnormalities were revealed in the cervical spine.

Discussion:
Congenital anomalies of the odontoid process are classified into 5 types9;
Type I : Separate odontoid ........ “os odontoideum”
Type II : Free “apical” segment ........ “ossiculum terminale”
Type III : Agenesis of odontoid base
Type IV : Agenesis of “apical” segment
Type V : Agensis of odontoid process

The odontoid process is formed from 3 separate embryologic anlages18. Two laterally situated centers of ossification appear in the base of the odontoid process at about the 6th month of fetal life, and unite into a single column at the time of
Legends:

Fig. 1.: Open-mouth view.
An ossicle is separated by a gap from the base of the odontoid process, which is projecting as a convexity continuing the upward slope of the superior articular processes of the axis.

Fig. 2.: Lateral views.
Atlanto-axial dislocation is observed. An ossicle is seen posterior to the overdeveloped anterior arch of the atlas; the posterior arch is attenuated.

a) : In flexion
b) : Upright
c) : In extension
At 2 years of age, a separate ossification center appears at the apex of the odontoid process. Fusion of the base and the apex usually occurs by the age of 12 years. A cartilaginous material persists between the apical segment and the base of the dens or between the base of the dens and the body of the axis until late in life. This situation is normal and must not be diagnosed as os odontoideum unless the odontoid is displaced.

Although os odontoideum apparently is a congenital anomaly, we sometimes encounter the difficulty in differentiating this anomaly from the old ununited fracture of the odontoid process, particularly when the separate odontoid process is not associated with other bone anomalies. FIELDING reported the posttraumatic disappearance of the central portion of the odontoid process, which looked like an os odontoideum. According to GREENBERG, a normally developed dens which has failed to gain a bony fusion with the body of the axis should be called an os odontoideum.

On the other hand, an ossiculum terminale is created by a failure of fusion of the apical segment of the dens to its base; the base being normally fused with the body of the axis. This is the most common anomaly of the odontoid. Another type of anomaly which has been often confused with an os odontoideum is agensis of odontoid base. This condition is similar to ossiculum terminale in that the apical segment of the dens is not united. It differs, however, in that the base of the dens has completely failed to develop. Differentiation among these 3 types is practically difficult, and the term “os odontoideum” seems to have been used to describe various types of malformations of the odontoid process in the previous literature.

The diagnosis is usually established by roentgenologic examination. The important finding consists in a degree of atlanto-axial dislocation, which can be demonstrated on the lateral roentgenograms of the cervical spine made with the patient upright and the neck in flexion and extension. WOLLIN stated that the factors which favor the diagnosis of os odontoideum are as follows:

1. Small corticated ossicle separated from the base of the odontoid process by a gap of varying width;
2. The plane of cleavage between the two parts of the odontoid process is above the plane of the superior articular processes of the axis, and the base of the odontoid process may project upward as a “peg” or merely be present as a convexity continuing the upward slope of the superior articular process of the axis;
3. Overdevelopment of the anterior arch of the atlas with attenuation of the posterior arch (which is often bifid) is evidence of an associated bone anomaly.

Our case is in accordance with these criteria (Figs. 1 & 2).

He also described the differentiating criteria for the fracture of the odontoid process:

1. Usually the entire odontoid process can be identified;
2. The gap between the separated fragment and the base of the odontoid process is less;
3. Fracture of the odontoid process extends down into the body of the axis, so that the plane of cleavage is at a more caudal level with a fracture than with an os odontoideum.
The signs and symptoms depend upon the severity of the dislocation. The most common sign is limitation of neck movement due to pain. Cervical cord compression and vertebrobasilar insufficiency caused by the dislocation of the atlas on the axis make up the clinical picture. Meningeal irritation and hypertrophy resulting from the repeated compression, angulation and stretching which the cervical cord and related structures are subjected to by recurrent atlanto-axial dislocations may in turn compromise the blood supply to the spinal cord and eventually lead to degeneration of the affected segments.

Reviewing the available records of 30 cases with os odontoideum, 15 cases showed the spontaneous onset. Among these 15 cases, 7 patients (47%) developed evidence of cervical cord damage before the age of 20. In contrast to this, the patient with odontoid agenesis did not develop myelopathy of spontaneous onset until fairly late in life (28 to 63 years of age). Besides, it is reported that the atlanto-axial dislocation, secondary to an absent odontoid process not associated with other bone anomalies, is not so serious. On the other hand, the presence of an os odontoideum appears to be more hazardous. GREENFIELD reported 2 patients with an os odontoideum who were rendered quadriplegic by relatively simple maneuvers of the head, and one of them died. KLINE also reported the case with atlanto-axial dislocation simulating a serious head injury. The patient apparently had an os odontoideum. Our patient was accidentally found to have an os odontoideum. He had only transient signs and symptoms, and was discharged without any sequelae. It is, however, highly probable that sooner or later he may develop cervical cord damage either spontaneously or traumatically.

Conservative treatment is sometimes effective, but there are cases where surgical intervention is needed. Immobilization, fusion and/or decompression are performed as the surgical treatments, and among them, fusion seems to be the treatment of choice.

Summary:
A case of os odontoideum has been presented and the available literature reviewed. In cases with an os odontoideum, neurological signs and symptoms develop in high percentage and relatively early, while patients with an absent odontoid process remain asymptomatic until fairly late in life.

Acknowledgements:
Dr. KAZUHIKO HAYASHI kindly gave permission to report this case under his care.

References:
2) Bevan. cited by Hohl.
THE "OS ODONTOIDEUM" REPORT OF A CASE


Os odontoideum
症例報告
京都大学医学部脳神経外科学教室

松田昌之 小山素麿 半田譲二

15才男子、学校で体操の時間中転落し頭部を打撲。頭部疼痛及び両上肢のしびれ感をきたしたが、しびれ感は約10分で消失した。意識障害は伴わなかった。1時間後外来時には、頭部運動は正常、右側頸部外反筋の軽度腫張と疼痛のみ認められ、その他神経学的には異常所見はみられなかった。頚椎X線写真ではatlanto-axial dislocation及びOs odontoideumが認められた。保存的治療で頚部の症状は1週間後には消失し、atlanto-axial dislocationによる症状はみられなかった。特別な治療を行うことなく退院した。家族にはX線上頚椎に異常を認める者はなかった。

軸椎脳突起の先天異常は 1) Separate odontoid •...“os odontoideum”, 2) Free “apical” segment...“ossiculum terminale”, 3) Agenesis of odontoid base, 4) Agenesis of “apical” segment, 5) Agenesis of odontoid processの5型に分類される。このうち1, 2, 3, の鑑別は実際上困難であるが、われわれの症例では椎突起が認められず、ossicleはかなりの大きさを保っており、Os odontoideumに相当すると考えられる。又、Os odontoideumと鑑別の困難なものにold ununited fracture of densがある。Os odontoideumのcriteriaとしては、1）椎突起のbaseから離れているcotticated ossicleである。2）椎突起baseとossicleとの境界線が軸椎上関節面より上に存在し、椎突起baseは小さい突起を有するか、又は軸椎上関節面に連続した凸面として存在する。3）軸椎のanterior archが異常に発達し、posterior archが細くなっている。が挙げられている。われわれの症例はこれらのcriteriaに合致しておりOs odontoideumと言える。

Os odontoideumでは神経症状が高率且つ早期に出現し、重篤であることが多いのに対し、Agenesis of odontoid processでは症状の出現が遅く、それほど重篤にはならないと言われている。われわれの症例では偶然にOs odontoideumが発見され、これによる症状は全くみられなかったが、早晩、頭部脊髄障害をきたす可能性が大きいと思われる。