DISLOCATION OF ATLAS CAUSED BY CONGENITAL ABSENCE OF ODONTOID PROCESS
REPORT OF A CASE

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Received for publication July 2, 1960

Congenital anomaly of the odontoid process of the axis, first described by Bevan in 1863, is very rare, and prior to this report not over thirty cases could be found in literature and as far as we could review, this is the first case ever reported in Japan. This anomaly is caused by the failure of "physiological fusion" between the body of axis and the odontoid process, which embryologically equivalent to the body of atlas. In most cases with such anomalies, abnormal motion in the atlantoaxial joint exists, and sooner or later the dislocation of atlas would be followed by a mere violence or even spontaneously.

Case report

The patient, 23 years old, laborer of lumber-mill, was first seen in January 1960. Since February 1959 he began to notice pain in the nuchal and lower back region, and the motion of the neck was limited. In November a lumber, though not so heavy, dropped on his head. Immediately after the injury he felt weakness of the right arm, and motion of the neck was found fully obstructed, but he did not consult with any doctor and continued his labour until January 1960.

At the examination the patient's cervical spine was backward flexed and the nuchal muscle spasm were noticed. The motion of the neck towards any direction was impossible. The alignment of cervical spine was almost normal but the space between the occiput and the spinous process of axis was abnormally wide. There was tenderness over the suboccipital-triangle and the right brachial plexus. All muscle reflexes of the upper extremities were markedly exaggerated, especially on the right side. Knee and ankle jerk also increased. On the right upper extremity and right side of the neck, hypaesthesia was found (Fig. 1).

Fig. 1 Range of hypaesthesia (dots).
Motion of the extremities, however, remained undisturbed.

Roentgenograms of the cervical spine showed forward dislocation of the atlas and absence of the odontoid process of the axis (Fig. 2, 3). Myeologram revealed a constricted passage of the iodized oil at the level of the atlanto-occipit joint (Fig. 4).

Fig. 2 (a)

(a) Lateral roentgenogram of cervical spine.
(b) Antero-posterior roentgenogram of axis.
(c) Axial roentgenogram of axis.

Fig. 2 (b)

Fig. 2 (c)

Fig. 3 Tomogram of upper cervical spine.

Fig. 4 Myelogram of atlanto-occital region.
Besides these cervical anomalies, eunuchoidism and delayed development of the bones were also found in this patient. That is, genital organs were found very small and pubic hair and beards were not seen (Fig. 5). Extremities were very slender, and epiphyseal lines were still remained in all long bones.

But the form and size of sella turcica normal.

Laboratory findings were as follows:

- Calcium in serum 7.0 mg/dl.
- Phosphate in serum 4.0 mg/dl.
- Alkaline phosphatase in serum 6.1 Bodansky unit
- 17-Ketosteroid in urin 4.0 mg.p.d.

Though the spinal fusion was recommended, this was refused by the patient, and traction was tried for 2 months without improvement of the neurological symptoms and signs.

Comment

The odontoid process of the axis belongs to the atlas embriologically, and in the normal person its fusion with the axis is to be completed before 6 years of age. Though it is very rare, when this “physiological fusion” is not completely performed, the states that is called “dens aplasia” or “os odontoideum” result. According to the grade of the insufficient fusion, this anomaly is classified radiographically into following three types, that is, total absence of the dens (aplasia), partial absence of the dens (hypoplasia), and failure of fusion between dens and body of axis (os odontoideum). However, even in dens aplasia the odontoid process of axis is not completely lacking but cartilaginous rudiment is found at the position. Also in the case of os odontoideum cartilaginous connection with axis is found (Bevan et al.).

In such anomaly, the atlanto-axial joint is unstable, because of the lack of odontoid process which has a important role for the stabilisation of the joint, dislocation of the atlas may sometimes result by a mere violence or spontaneously. And the dislocation more likely occurs forewards rather than lateral or backward.

As the associated abnormalities, Klippel-Feil syndrome and platybasia were reported. In our case remained epiphsiccal lines were found in almost all long bones. This may probably have something to do with the hormonal disturbance which was actually shown in the presence of eunuchoidism, though the laboratory data on this particular case were not conclusive.

Diagnosis of absence of the odontoid process is not always easy, and tomogram is a helpful tool in diagnosing such case. Fracture of the odontoid process and
inflammatory changes in the atlanto-axial joint must be differentiated from this disease.

Various treatments have been reported, such as traction, immobilization in cast and fusion operation.

Summary

A case of atlas dislocation caused by congenital absence of the odontoid process with delayed skeletal development and eunuchoidism, found in 23 years old male, was reported.

Reference