

Unilateral Fusion of the Odontoid Process with the Atlas in Klippel-Feil syndrome: A Case Report¹

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Klippel-Feil syndrome (KFS) displays congenital fusion of the cervical vertebrae; it is a relatively common condition and has many associated malformations such as Sprengel's deformity, scoliosis, rib anomalies, congenital defects of the brain or spinal cord, renal anomalies, congenital heart disease, deafness, cleft palate, cranial and facial asymmetry, and enteric cysts. There are various types of cervical fusion observed in KFS. However, fusion of the odontoid process with the atlas is a very rare finding. We report here on a 4-year-old boy with unilateral fusion of a separated odontoid process with the lateral mass of the atlas, and this was associated with a spontaneously closed ventricular septal defect, a small patent ductus arteriosus and a horseshoe kidney.

Index words : Atlas and axis

Spine, abnormalities

Computed tomography (CT)

Klippel-Feil syndrome (KFS) indicates congenital fusion of two or more cervical vertebrae and this may be associated with various anomalies, including cardiovascular abnormalities, genitourinary tract abnormalities, scoliosis, Sprengel's deformity, rib abnormalities, abnormalities of the spinal cord, otolaryngological anomalies and enteric cyst. The classic triad (a short neck, low posterior hairline and a limited range of motion of the neck) is apparent in fewer than 50% of these patients. KFS is relatively common, yet fusion of the odontoid process with the atlas is a very rare finding (1, 2). In all the reported cases of fusion between the odontoid process and the atlas, the odontoid process was shown to be fused to the anterior atlantal arch (1 - 7). In a few cases, the odontoid process was fused to the lateral mass

as well as the anterior arch of the atlas (3 - 5). We present here a case of fusion between a separated odontoid process and the lateral mass of the atlas. To the best of our knowledge, this type of case has not been previously reported in the English medical literature.

Case Report

A 4-year-old boy presented with torticollis at birth. His chin pointed to his left side and his neck range of motion appeared normal. He had no weakness in any extremity and no history of trauma or illness of the head and neck. A heart murmur had been found at birth and his mother knew he had congenital heart disease. Computed tomography (CT) of the cervical spine (Fig. 1) revealed pseudoarticulation with partial bony fusion between the odontoid process and the right lateral mass of the atlas. The gaps between the anterior arch and both lateral masses of the atlas were asymmetrically open, with the left gap abnormally wide when compared with that seen on normal postnatal development. There was also

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an abnormally wide gap between the posterior arches of the atlas. The odontoid process was enlarged and deformed, like the body of the atlas. There was no bony fusion between the odontoid process and the body of the axis. This gap was located above the level of the superior articular facets of the axis and it resembled the intervertebral discs between the other cervical vertebrae. On rotation to both sides, the separated odontoid process was fixed to the right lateral mass of the atlas and it rotated with the atlas, but differently from the axis. The other cervical vertebrae were normal.

The patient underwent echocardiography and abdominal ultrasonography to evaluate any other possible asso-

ciated anomalies. The echocardiography revealed a spontaneously closed ventricular septal defect with pseudoaneurysm formation and a small patent ductus arteriosus (Fig. 2A). The abdominal ultrasonography revealed a horseshoe kidney (Fig. 2B).

Discussion

In all the reported cases of fusion between the odontoid process and the atlas, the odontoid process was either separated or not separated from the body of the axis. In the all cases of the nonseparated type, the odontoid process was fused with the anterior arch of the atlas (1,

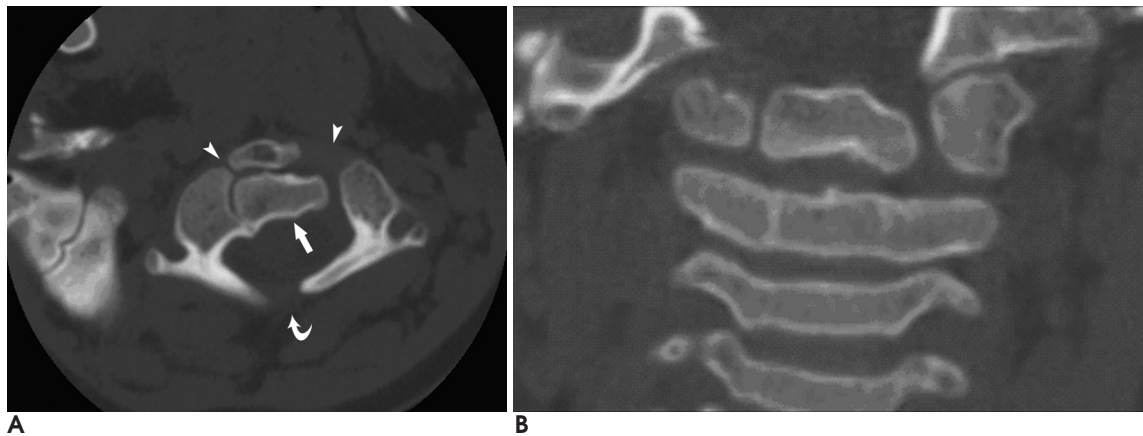


Fig. 1. Nonenhanced computed tomography (CT) scans (bone window) of the cervical spine.

A. Axial CT scan through the atlas shows the enlarged odontoid process (arrow) fused to the right lateral mass of the atlas. The neurocentral synchondroses (arrowheads are the cartilaginous gaps between anterior arch and both lateral masses) and posterior synchondrosis (curved arrow are the cartilaginous gap between the neural arches) are open.

B. Coronal MPR image of the upper cervical vertebrae shows no bony fusion between the odontoid process and the body of the axis, and this resembles the intervertebral discs of the other cervical vertebrae.

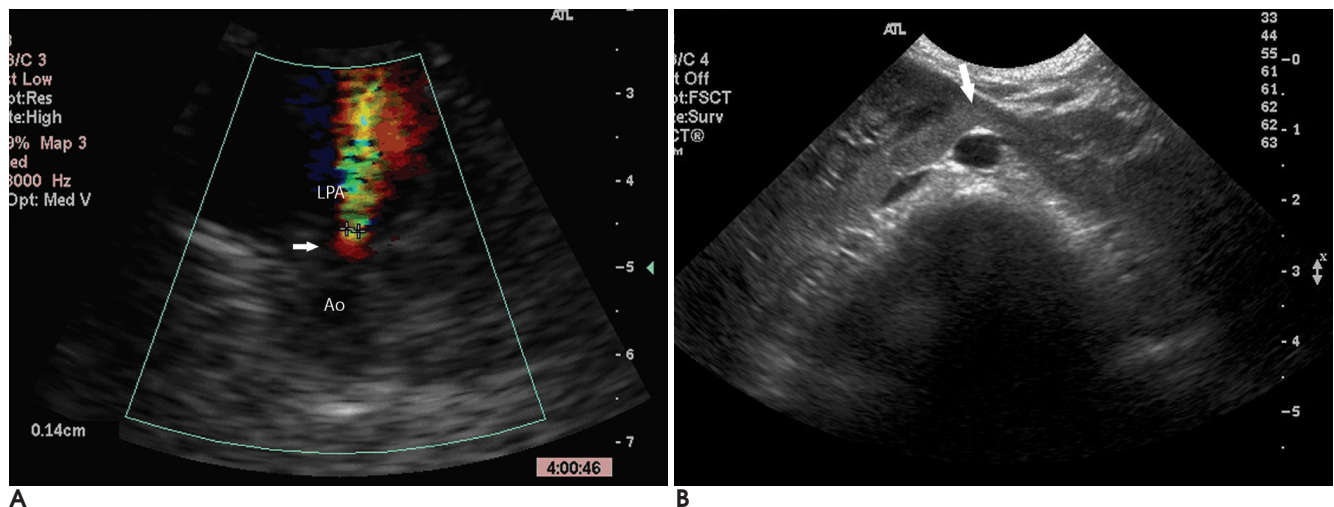


Fig. 2. A. The echocardiography reveals a small shunting (small arrow) from the aorta (Ao) to the left pulmonary artery (LPA) and spontaneously closed ventricular septal defect with pseudoaneurysm formation (not shown). RV: right ventricle; LV: left ventricle.

B. The abdominal ultrasonography demonstrates the isthmus (arrow) of a horseshoe kidney anterior to the aorta.

2, 5 - 7). Only one case of the nonseparated type was fused to the lateral mass as well as the anterior arch of the atlas, and this was associated with atlanto-occipital instability (5); the other cases had no evidence of instability. Two cases of the separated type have been reported in the English literature late in the twentieth century (3, 4). In both cases, the anteroposterior and lateral radiographs demonstrated fusion of the separated odontoid process with both lateral masses as well as fusion with the anterior arch of the atlas. Both cases were associated with atlantoaxial instability and one of them was treated with posterior atlantoaxial fusion using a bone graft from the iliac crest (4).

Most of the reported cases displayed associated anomalies of the atlas, axis or other cervical vertebrae such as fusion between the lateral masses of the atlas and axis (1, 2), complete fusion of the lateral masses and the posterior arches of the atlas and axis (5), a anterior arch cleft of the atlas (2), a posterior arch midline fusion defect or a hypoplastic posterior arch of the atlas (6, 7), and other cervical or craniovertebral junctional anomalies (2 - 4).

During postnatal development, the atlas develops from 2 posterior ossification centers and usually a single anterior ossification center. The two posterior centers form the lateral mass and the posterior arch on each side, and they form the posterior synchondrosis at a rudimentary spinous process. The anterior and posterior centers meet to form the neurocentral synchondrosis anteromedially to each lateral mass. The neurocentral and posterior synchondroses of the atlas are fused by the age of four to six years (8). The body and posterior elements of the axis develop from a central ossification center and two posterior ossification centers, and the odontoid process develops from two primary centers. The two primary centers of the odontoid process extend inferiorly into the body to form the dentocentral synchondrosis below the level of the superior articular facets and just within the eventual body of the axis. The dentocentral synchondrosis closes by five to six years of age (9).

In our case, the gap between the odontoid process and the body of axis was abnormally wide and located above the level of the superior articular facets of the axis; this was different from the normal dentocentral synchondrosis. That is, the odontoid process was separated from

the body of the axis. The separated odontoid process was not fused to anterior arch, but to the unilateral lateral mass of the atlas. There were also abnormally wide gaps between the ossified anterior arch and the left lateral mass of the atlas and between both ossified posterior arches of the atlas. In the future, these have the possibilities of developing into a cleft in the anterior atlantal arch and spina bifida occulta, respectively.

KFS may result from failure of both segmentation and resegmentation of the sclerotomes. The dense cranial part of the first cervical somite forms the atlas, and the loose caudal part of the first cervical somite forms the dens. The dense part of the second cervical somite forms the rudimentary disc between the dens and the body of the axis (10). The presence of congenital fusion between the lateral mass of the atlas and the odontoid process of the axis could be explained by a segmental defect of the first cervical somite (2); the separation between the odontoid process and the body of the axis could be explained by the failed involution of the dense part of the second cervical somite.

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