Case Report

Cervical Myelopathy Caused by a Split Atlas Anomaly: A Case Report

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Keywords

Case report · Spine · Split atlas · Congenital defect · Myelopathy

Abstract

An 18-year-old male presented with a 6-month history of paresthesia of both arms and legs after a minor neck trauma. CT scan revealed a partial aplasia of the anterior and posterior arches of the C1 vertebra resulting in a split atlas. MRI showed an intramedullary high-signal area. We performed a posterior decompressive laminectomy and occipitocervical fusion. The bony defect into the posterior arch was replaced by a connective tissue cord, resulting in a compression of the dural sheath. The symptoms recovered completely 1 month after surgery. Knowledge of this rare malformation is crucial to the correct management of these cases.

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Introduction

The occipitocervical junction (OCJ) is a complex anatomical area that includes several vital structures and allows mechanical transition between the central nervous system and the axial skeleton. Many types of bone and/or nerve malformations, major or minor, have been described but remain rare [1]. These malformations are diverse, often associated with each other. This highly polymorphism may be responsible for serious neurological disorders. We report the case of a rare “benign” congenital malformation of the OCJ associated with neurological disorders.

Timeline

December 2018	Paresthesia of both arms and legs, cervical pain, and walking difficulty after a minor neck trauma
June 2019	Physical examination revealed a walk with mowing on the right side, pain on palpation of the upper cervical spine, contracture of the paravertebral muscles, and limited cervical range of motion
July 2019	Decompressive laminectomy and occipitocervical fusion
January 2021	Neck pain completely resolved, function was normal, and he remained asymptomatic at the last follow-up

Case Report

A previously healthy, 18-year-old male patient presented with a 6-month history of paresthesia of both arms and legs, cervical pain, and walking difficulty after a minor neck trauma (he fell from his height and landed on the occiput, with hyperflexion of the cervical spine). His symptomatology was improving gradually, but there was no bowel or bladder dysfunction.

At the time of admission, physical examination revealed a walk with mowing on the right side, pain on palpation of the upper cervical spine, contracture of the paravertebral muscles, and limited cervical range of motion. Neurological assessment revealed pyramidal syndrome with bilateral upper and lower limbs hyperreflexia, bilateral Babinski sign, and Hoffman's reflex present. Motor function and sensory examination were normal in the left upper limb and lower limbs. In the right upper extremity, motor function was rated 3–4 out of 5, with hypoesthesia in the C7–C8 territory. Furthermore, examination of the perineal area was unremarkable.

Plain radiographs were interpreted as normal, and dynamic radiological assessment did not show any instability. CT scan revealed partial aplasia of the anterior and posterior arches of the C1 vertebra, resulting in a split atlas with a platybasia and basilar impression (shown in Fig. 1). MRI showed cord compression at the atlas ring and intramedullary high T2 signal intensity area (shown in Fig. 2).

We decided to perform a decompressive laminectomy and occipitocervical fusion using a posterior approach because the patient had neurological symptoms. During the procedure, the bony defect in the posterior arch of C1 was replaced by a connective tissue cord, compressing the dural sheath (shown in Fig. 3).

Our patient underwent laminectomy of C1 (shown in Fig. 4), release of the tissue cord, enlargement of the foramen magnum, occipitocervical fixation, and posterior grafting at the expense of the posterior iliac crest. The intraoperative appearance at the end of surgery is shown in Figure 5. Immediate postoperative radiography is shown in Figure 6. Patient's symptoms recovered completely 1 month after rehabilitation, and the fusion was consolidated at the 4-month visit (shown in Fig. 7). Neck pain completely resolved, function was normal, and he remained asymptomatic at 18-month follow-up.

Discussion/Conclusion

Split atlas (or bipartite atlas) is a rare congenital malformation consisting of the association of partial aplasia (or hypoplasia or defect or dehiscence) of the posterior and the anterior arches of C1 vertebrae [2]. It was considered by several authors as a "benign anatomical variation," without clinical impact, and indeed almost all of the cases reported in the literature were discovered incidentally [3–6]. This malformation has been reported

Fig. 1. CT scan showing partial aplasia of the anterior and posterior arches of the C1 vertebra (anterior arch: red arrow and posterior arch: blue arrow).

in only 0.1% of the population [7, 8]. Isolated hypoplasia of the posterior arch has been reported up to 5% of the population, while isolated hypoplasia of the anterior arch has been reported up to 0.7% [7–10].

Various radiological appearances were reported. Anterior aplasia of the atlas tends to be more narrow and is usually found on midline or paramedial, whereas 97% of posterior arch aplasia tends to be located medially, with only 3% lying laterally [7, 11].

Malformations of the OCJ have embryological origin [2]. Several theories have been proposed to explain the split atlas, but the exact mechanism remains undetermined. The atlas is formed from 3 primary ossification centers: 2 laterals and 1 anterior. The lateral ossification centers extend dorsally, forming 2 hemiarches that should completely fuse and form a union by an average age of 3–5 years. If fusion does not occur, the result would be aplasia of the posterior arch. Aplasia of the anterior arch may occur if the anterior ossification center does not form or if fusion with the lateral masses does not occur properly [2].

Logan and Stuard [12] have suggested that the hypoplasia of the posterior arch is rather the result of lack of local chondrogenesis than an ossification failure. This theory has been adopted by other authors after discovering the presence of connective tissue bridging the bony defect [13, 14].

No classification is reported in the literature of the different variants of the split atlas. However, Currarino et al. [15] have proposed a classification of posterior arch defects into 5 subtypes (shown in Fig. 8), with predominance of type A, found in 97% of all types of posterior arch aplasia [10, 16].

Fig. 2. MRI showing intramedullary high T2 signal intensity at the atlas ring area.

Fig. 3. Intraoperative appearance of the posterior arch aplasia.

Clinically, most cases of split atlas are usually asymptomatic, detected incidentally during radiological exploration [5]. In a few cases, this malformation has been incriminated in various symptoms such as chronic neck pain, headache in the occipital region, torticollis, or neurological disorders. Some publications have reported neurological decompensation following minor head or neck trauma [2].

Fig. 4. Intraoperative appearance after decompression.

Fig. 5. Intraoperative appearance at the end of surgery.

Fig. 6. Immediate postoperative radiography.

Fig. 7. Radiography at 4 months postoperative showing consolidation of the arthrodesis.

Cervical myelopathy was mostly found with anomalies involving types C and D of Currarino's classification. These types contain persistent posterior osseous fragment that can be mobile and possibly causing a mass syndrome on the spinal cord during neck extension, resulting in atypical and varied neurological symptoms [11, 17–19].

For some authors, atlantoaxial instability was another mechanism causing neurological disorders. It has been reported that aplasia of the posterior arch may increase the risk of atlantoaxial subluxation in about 26% of children aged between 2 and 3 years [20, 21].

Fig. 8. Classification of posterior arch defects proposed by Currarino et al. [15]. Type **A** The 2 posterior hemi-arches fail to fuse in the posterior midline. Type **B** Presence of a defect on 1 side of the posterior arch. Type **C** Presence of defects on both sides of the posterior arches. Type **D** Absence of the posterior arch but the posterior tubercle is still present. Type **E** Absence of both the posterior arch and the posterior tubercle.

Aplasia of the anterior arch may be suspected on open-mouth radiographs showing the misalignment of the lateral masses of the atlas, with an anterior arch that appears fat, rounded, and overlapping the outline of the odontoid process [1]. Aplasia of the posterior arch may be seen on a lateral radiography that shows a double shadowing of the split arches revealing the lack of fusion [1, 22].

In the presence of neurological signs, MRI is recommended to view the integrity of the spinal cord. Dynamic radiographs can be performed in the context of cervical spine instability.

This malformation can, in a traumatic context, be confused with a Jefferson fracture [2]. In the case of a split atlas, imaging studies can make the difference by showing a dehiscence of the arches in the midline in the majority of cases, with smooth and corticalized edges and without soft tissue abnormalities (edema and hematoma).

The treatment of the split atlas and its degree of urgency depend on the anatomical and clinical type of the posterior arch malformation. In the case of an asymptomatic incidental finding, management is limited to clinical and radiological follow-up only. Conservative treatment with nonsteroidal anti-inflammatory drugs and usage of cervical orthosis or a halo

Fig. 9. A schematic showing spinal cord compression by the connective tissue cord bridging the bony defect, exerting bowstring effect.

ring may be proposed in case of chronic symptomatology without neurological deficit [3, 7, 23, 24]. In case of instability or neurological decompensation, OCJ fusion should be indicated using either the posterior or anterior approach. Posterior approach [7, 23, 25] allows good fixation of the OCJ but has the drawback of restricting rotatory function of the neck. The anterior transoral approach [23, 26] allows stabilization when preserving rotational neck function, but it is a relatively difficult approach, associated with a high risk of oropharyngeal infection. For types C and D of the Currarino's classification, some authors [18, 24] have proposed surgical excision of the posterior arch at an early stage to prevent cumulative damage to the cord.

This case report shows an atypical discovery mode of a rare malformation, assumed to be benign. The defect of the posterior arch type A, which is in practically all cases asymptomatic, was manifested in this case by neurological disorders following a benign trauma. Myelopathy can be explained by the bowstring effect exerted by the connective tissue cord bridging the bony defect (shown in Fig. 9). Surgical decompression and occipitocervical fusion allowed a total recovery and gave this patient a normal life despite a late diagnosis (after 6 months).

In summary, the occurrence of a split atlas should not be underestimated in this clinical setting. Modern imaging modalities confirm the diagnosis and guide treatment. Patients usually have no symptoms and require no treatment. However, when neurological prognosis is at stake, surgical decompression is the treatment of choice.

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Mehdi Meddeb involved in clinical evaluation of the patient, intervention, and writing and editing of this article. Hassen Makhoulf involved in clinical evaluation of the patient, surgical planning, intervention, and follow-up; conception and design of the work; and acquisition, analysis, and drafting of the work. Sofiene Bouali involved in surgical intervention, analysis and interpretation of the data, and reviewing the manuscript. Khalil Habboubi involved in conception and design of the work and acquisition, analysis, and interpretation of the data. Mondher Mestiri involved in analysis and interpretation of the data and reviewing the manuscript.

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