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Case Report

Congenital Cleft of Anterior Arch and Partial Aplasia of the Posterior Arch of the C1

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Congenital anomalies in arches of the atlas are rare, and are usually discovered incidentally. However, a very rare subgroup of patients with unique radiographic features is predisposed to transient quadriparesis after minor cervical or head trauma. A 46-year-old male presented with a 2-month history of tremor and hyperesthesia of the lower extremities after experiencing a minor head trauma. He said that he had been quadriplegic for about 2 weeks after that trauma. Radiographs of his cervical spine revealed bilateral bony defects of the lateral aspects of the posterior arch of C1 and a midline cleft within the anterior arch of the atlas. A magnetic resonance imaging revealed an increased cord signal at the C2 level on the T2-weighted sagittal image. A posterior, suboccipital midline approach for excision of the remnant posterior tubercle was performed. The patient showed significant improvement of his motor and sensory functions. Since major neurologic deficits can be produced by a minor trauma, it is crucial to recognize this anomaly.

Key Words : Congenital anomalies · Cervical atlas · Spinal cord injury · Head trauma.

INTRODUCTION

Congenital anomalies in arches of the atlas are rare, but well described[1,2,4,10,14]. These defects are usually discovered incidentally[5,12]. However, in some instances, patients who experienced transient quadriparesis after a minor head trauma have unique anatomical features[11]. In this report we describe such a case and with review of pertinent literature.

CASE REPORT

A 46-year-old male presented with a 2-month history of tremor and hyperesthesia of the lower extremities after a minor head trauma. He said that he had been quadriplegic for about 2 weeks after the trauma and had been under conservative management in another hospital. He also said that his motor weakness was improving gradually. He did not complain of any neck pain or motor weakness but reported tremor of the right leg and hyperesthesia below the T12 dermatome. He also complained of having difficulty with voiding and defecation. On neurological examination, his motor function was normal and his sensory examination revealed a decreased sense of light touch in his lower extremities and right hand. He had a positive Lhermitte's sign and saddle anesthesia and decreased anal sphincter tone. A plain radiograph of the cervical spine revealed that the posterior arch of the atlas was partially absent.

Radiographs taken in flexion and extension showed no instability of the occipitoatlantoaxial area and the definite movement of a bony remnant of the posterior arch. A computed tomography scan showed the midline cleft within the anterior arch of the atlas and the bilateral bony defects of the lateral aspects of the C1 posterior arch with the most dorsal part of the posterior arch preserved (Fig. 1). A magnetic resonance imaging (MRI) revealed that there was an increased cord signal at C2 in the dorsal aspect of the spinal cord on the T2-weighted images without compression of the cord (Fig. 2). For further evaluation of the pathogenesis of the spinal cord injury, fluoroscopy was used to evaluate the patient's dynamic cervical spine movements. Fluoroscopy showed that the anterior movement of the bony remnant of the posterior arch was independent of the C1 anterior arch and was compressing the spinal cord during extension (Fig. 3).

A posterior, suboccipital midline approach with a pinned headrest was performed. At surgery, we found dense fibrous bands connecting the bony fragment to both sides of the lateral masses. We removed only the bony remnant of the posterior arch and did not carry out occipitocervical fusion because we confirmed no instability of the occipitocervical area pre- and intraoperatively.

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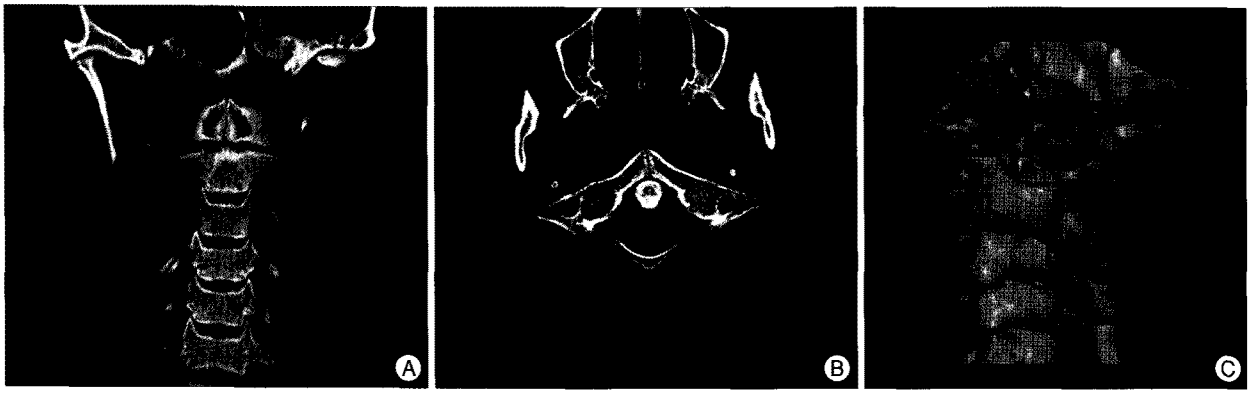

Fig. 1. A CT scan reveals a midline cleft in the anterior arch of the atlas (A) and partial aplasia of the posterior arch (B and C).

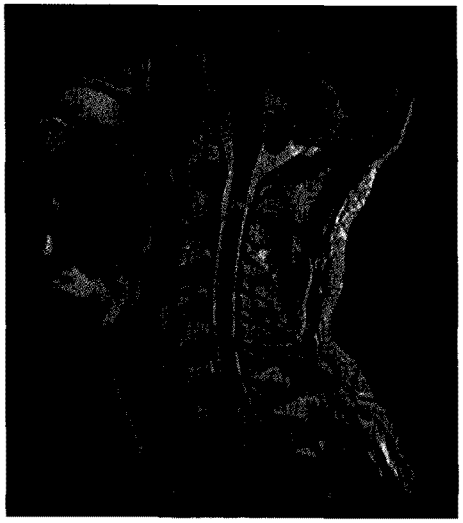

Fig. 2. T2-weighted sagittal image of the cervical spine shows an intramedullary signal abnormality at the level of C2 suggestive of cord contusion.

The 12 month follow-up radiograph showed no instability of the occipitocervical complex (Fig. 4). By 6 weeks after the operation, the patient showed significant improvement of his neurological symptoms. Furthermore, the patient did not show neck pain, tremor of the legs, Lhermitte's sign and hyperesthesia below the T12 dermatome. However, the sphincter tone of the anus remained decreased.

DISCUSSION

The atlas is formed from three primary ossification centers: an anterior ossification center that forms the anterior tubercle and two lateral centers that form the lateral masses and the posterior arch[1,6,11,12]. Two centers at the lateral masses extend posteromedially to form the posterior arch usually in the fourth year. In about 2% of the population, the fourth ossification center forms a posterior tubercle between the two neural arches around the second year of life[6].

Defects of the posterior arch are thought to occur due to a failure of local chondrogenesis rather than due to subsequent ossification[11]. This suggestion has been supported by the finding that the connective tissue bridges the bony defect[11,13]. We also identified the remnant connective tissue band, in our case intraoperatively. Therefore, this patient seems to have both an error of chondrification as well as the rare fourth ossification center in the posterior arch of atlas.

Malformations of the atlas include both clefts and aplasias[1-4,10,14]. Currarino et al.[4] have divided the posterior arch anomalies into five types depending on the extent of absence of the posterior arch and the presence or absence of the posterior tubercle. These five types include of median clefts of the posterior arch of C1 (Type A), varying degree of unilateral defects (type B), bilateral defects (type C), absence of the posterior arch with a persistent posterior tubercle (type D), and total agenesis of the posterior arch including the tubercle (type E). Type A occurs in 5.4% of the population and 97% of all posterior arch defects. Types B through E have been reported to occur in 0.69% of the population[4,12]. Among the patients with the five types of posterior arch anomalies, those with type C anomaly developed sudden neurological symptoms after a head or neck trauma. The patients with this type experienced episodes of transient quadriparesis after a minor trauma[4,8,9,13,14]. Our patient also experienced transient quadriplegia. The anterior arch unites with the lateral centers at 5 to 9 years of age[11]. Clefts or aplasia of the anterior arch are very rare[7], accounting for only 0.1% in the Geipel series[2,7]. The anterior arch clefts may occur in the absence of an anterior ossification center, in which lateral masses do not fuse anteriorly[2]. This patient showed a cleft of the anterior arch and partial aplasia of the posterior arch of the atlas.

Richardson et al.[13] were the first to propose a mechanism by which neurological deficits occur. They emphasized that the isolated posterior fragment moved anteriorly and that when the neck was extended, the traumatized dorsal spinal cord caused inward buckling of the ligaments. However, they could not demonstrate this on flexion-extension films. Sharma et al.[14] were the first to reveal the movement of the bony tubercle with neck extension. In the two cases in their literature, they could validate inward movement of the tubercle in dynamic cervical spine radiographs and the signal abnormality of the spinal cord in an MRI. In this case, we were unable to confirm the movement of the bony tubercle by a dynamic simple x-ray workup. The patient's MRI, however, showed a signal change of the spinal cord.

Fig. 3. Dynamic fluoroscopic findings. This figure shows the anterior movement of the bony remnant of the posterior arch independent of the C1 anterior arch and compressing the spinal cord with extension.

Fig. 4. Follow-up plain lateral radiograph of the cervical spine in flexion (A), neutral (B) and extension (C) position. There is no significant instability of the occipitoatlantoaxial complex.

For further evaluation of pathogenesis of the spinal cord injury, the patient was evaluated on his dynamic cervical spine movements by fluoroscopy and we confirmed the movement of the bony tubercle (Fig. 3).

Treatment strategy is arguable. However, none of the patients in Sharma's studies underwent resection and only one case of cervical instability associated with these congenital anomalies was reported[14]. We think that in the absence of definite instability, a sufficient treatment is resection of the tubercle, which results in resolution of the symptoms.

CONCLUSION

We have a rare congenital anomaly of an anterior arch cleft and a posterior arch partial aplasia. Since major neurologic deficits can be produced by a minor trauma, it is crucial to recognize this anomaly.

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