Case Report

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

Ji Won Choi, M.D., Je Hoon Jeong, M.D., Seung Myung Moon, M.D., Hyung Sik Hwang, M.D.
Department of Neurosurgery, Hangang Sacred Heart Hospital, College of Medicine, Hallym University, Seoul, Korea

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.
Mechanism of Spinal Cord Injury in Congenital Anomalies of C1  |  JW Choi, et al.

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. Currarino et al. 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.

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. Our patient also experienced transient quadriplegia. The anterior arch unites with the lateral centers at 5 to 9 years of age. Clefts or aplasia of the anterior arch are very rare, accounting for only 0.1% in the Geipel series. The anterior arch clefts may occur in the absence of an anterior ossification center, in which lateral masses do not fuse anteriorly. This patient showed a cleft of the anterior arch and partial aplasia of the posterior arch of the atlas.

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. Two centers at the lateral masses extend posteriorly 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.

Defects of the posterior arch are thought to occur due to a failure of local chondrogenesis rather than due to subsequent ossification. This suggestion has been supported by the finding that the connective tissue bridges the bony defect. 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.

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Richardson et al. 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. 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.
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. 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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