Atlantoaxial Subluxation Associated with Os Odontoideum Fused to the Anterior Arch of the Atlas: A Case Report

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There are various anomalies in the craniocervical junction due to the complex nature of the cranial and cervical bone development[1]. Fusion of os odontoideum and the atlas (C1) has been very rarely reported[2]. Since the first case described by Wackenheim in 1971[3], very few cases have been reported in the literature[4-6]. Herein, we report a case of atlantoaxial subluxation associated with the fusion of os odontoideum and the anterior arch of the atlas. A 63-year-old male patient presented with a 2-month history of increasing posterior neck pain, clumsiness, and difficulty in walking. He had hyperactivity of the deep tendon reflex on both sides of the upper extremities but no other neurological abnormalities, such as muscle atrophy and lower cranial nerve dysfunction. He had no history of neck or head trauma. Plain radiographs of the cervical spine revealed a separation of the dens from the axis as well as atlantoaxial subluxation (Fig. 1A), which was reduced in the extended neck position (Fig. 1B). T2-weighted magnetic resonance imaging (MRI) revealed areas of hyperintensity in the spinal cord at the level between the body of the axis and the posterior arch of the atlas (Fig. 1 C). Computed tomography (CT) myelography revealed fusion of the apical segment of the dens and the anterior arch of C1 and narrowing of the spinal canal between the basal dens and the posterior arch (Fig. 2A and D-E). Other anomalous conditions were not identified in the atlantooccipital and atlantoaxial joints (Fig. 2B-C), and there was no additional anomaly in the subaxial cervical spine. In this case with progressive myelopathy, atlantoaxial arthrodesis *via* a posterior approach was planned as it was considered to be reasonable. The operation was performed using the Magerl and Brooks technique (Fig. 3), after which the patient's motor impairment improved over time, with slight numbness in the upper extremities.

In general, the odontoid process separates from the anterior part of the atlas and caudally migrates to fuse with the body of the axis between the 6th and 7th weeks of gestation. After resegmentation of cervical sclerotomes, the odontoid process is composed of the apical dental segment from the caudal proatlas, the basal dental segment from the first cervical sclerotomes, and the body of the axis from the second cervical sclerotomes[7] (Fig. 4). As a result of the complex processes involved in the embryological development of the cervical spine (especially segmentation and resegmentation), various anomalies can occur in the occipitocervical region[1].

Previous reports have demonstrated that os odontoideum, an anomaly of the axis, has an occurrence rate of 0.7%-0.8%[8,9], and 32%-44% patients with os odontoideum have progressive myelopathy[10,11]. However, only 10 cases of os odontoideum fused with the anterior arch of the atlas have been reported thus far, among which only 2 (20%) had progressive myelopathy with atlantoaxial dislocation[2,12]. There are limited data on the differences in the mechanical properties of the atlantoaxial joint between the two anomalous conditions. However, given the occurrence rate of neurological symptoms, there would be no significant differences in the mechanical instability of the atlantoaxial joint between patients with os odontoideum and those with fused anterior arch of the atlas. In our case, the patient developed cervical myelopathy in his 60s without a history of major trauma. We speculated that the cumulative micromechanical stress of daily living with the incomplete bony structures might have caused the atlantoaxial instability.

In this study, we have described a rare case of a 63-year-

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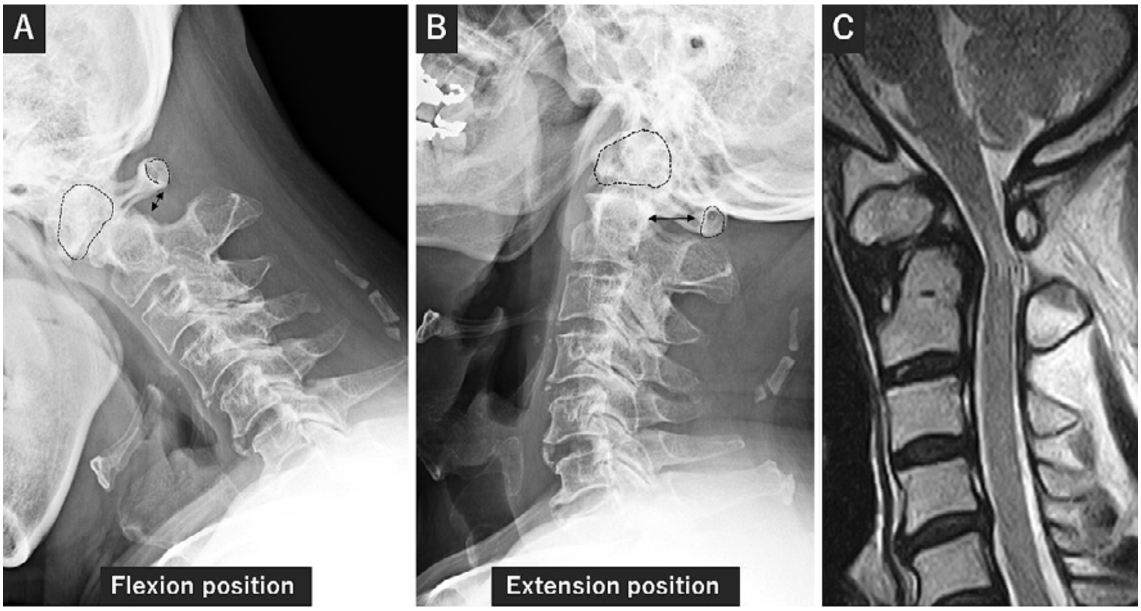

Figure 1. Preoperative plain radiography and magnetic resonance imaging (MRI).
Plain radiographs showing that atlantoaxial subluxation is reduced in the extension position (A–B). MRI showing high-signal intensity within the spinal cord at the level between the basal dental segment and the posterior arch of C1 (C).

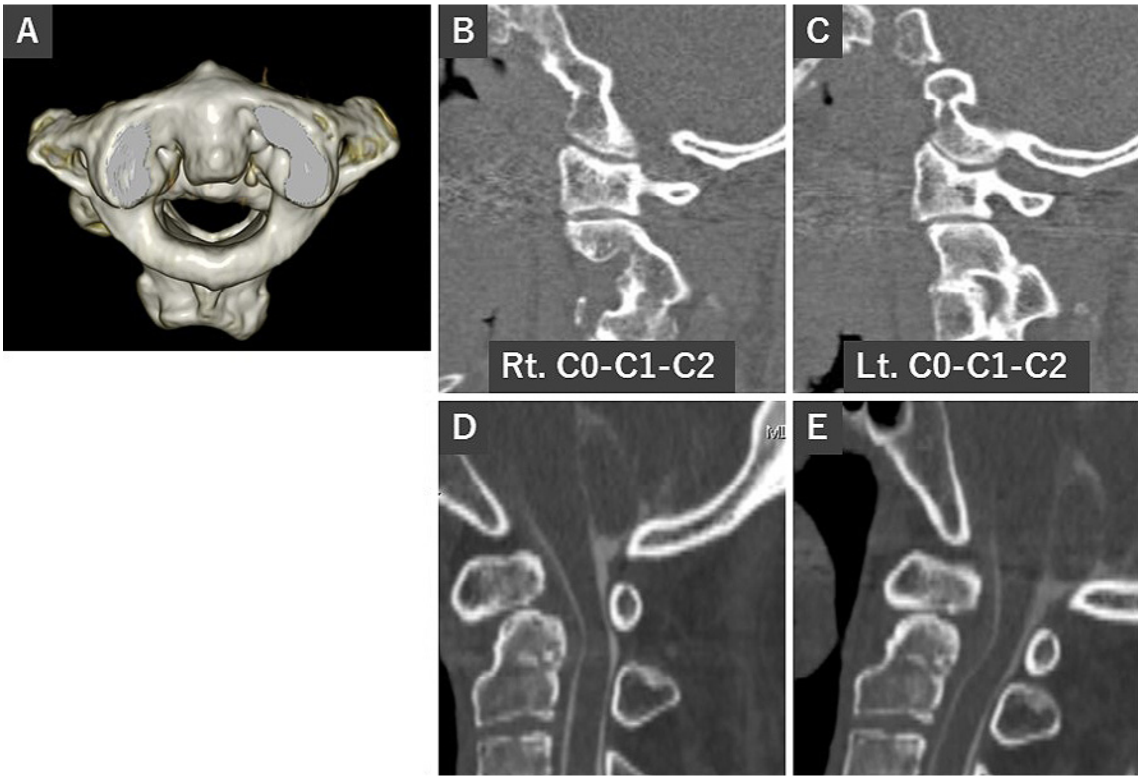

Figure 2. Preoperative computed tomography (CT) myelography.
CT showing the anterior arch of C1 fused with the apical segment of the dens separated from the basal segment of the dens (A). Sagittal reconstruction CT showing the right and left C0–C1–C2 articulations (B–C). Atlantoaxial dislocation in the flexion position (D) is reduced in the neutral position (E).

old male patient with atlantoaxial subluxation and progressive myelopathy associated with the fusion of os odontoideum and the anterior arch of the atlas.

Conflicts of Interest: The authors declare that there are no relevant conflicts of interest.

Figure 3. Atlantoaxial fusion surgery.
Atlantoaxial arthrodesis was performed using the Magerl and Brooks technique.

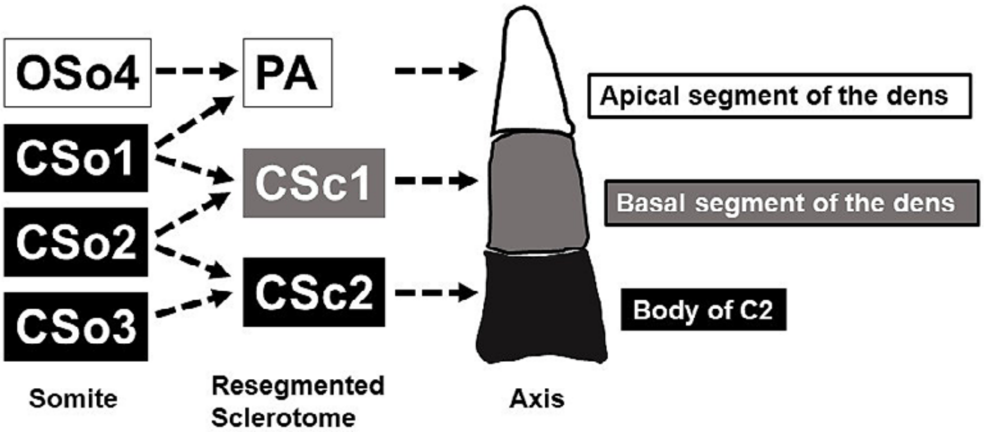

Figure 4. Embryology of the axis.
The odontoid process comprises the apical dental segment from the caudal proatlas, the basal dental segment from the first cervical sclerotome, and the body of the axis from the second cervical sclerotome.
PA: proatlas, CSo: cervical somite, CSc: cervical sclerotome

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Ethical Approval: This article does not contain any studies with human participants performed by any of the authors.

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

References

1. Offiah CE, Day E. The craniocervical junction: embryology, anatomy, biomechanics and imaging in blunt trauma. Insights Imaging. 2017;8(1):29-47.
2. Gil JR, Kim WT, Seo MJ. Congenital anomaly of combined atlas-

odontoid process fusion and bipartite atlas. Jpn J Radiol. 2015;33(12):769-71.

3. Wackenheim A. Densaplasia with atlanto-axial fusion. A special and stable form of densaplasia. Neuroradiology. 1971;2(2):76-9.
4. Olbrantz K, Bohrer SP. Fusion of the anterior arch of the atlas and dens. Skelet Radiol. 1984;12(1):21-2.
5. Pérez-Vallina JR, Riaño-Galán I, Cobo-Ruisánchez A, et al. Congenital anomaly of craniovertebral junction: atlas-dens fusion with C1 anterior arch cleft. J Spinal Disord Tech. 2002;15(1):84-7.
6. Tubbs RS, Oakes WJ, Grabb PA. Nonseparated odontoid process fused to an atlantal hemiarch. Pediatr Neurosurg. 2004;40(3):141-2.
7. Akobo S, Rizk E, Loukas M, et al. The odontoid process: a comprehensive review of its anatomy, embryology, and variations. Childs Nerv Syst. 2015;31(11):2025-34.
8. Ankith NV, Avinash M, Srivijayanand KS, et al. Congenital osseous anomalies of the cervical spine: occurrence, morphological characteristics, embryological basis and clinical significance: a computed tomography based study. Asian Spine J. 2019;13(4):535-43.
9. Perdikakis E, Skoulikaris N. The odontoid process: various configuration types in MR examinations. Eur Spine J. 2014;23(5):1077-83.
10. Klimo P, Kan P, Rao G, et al. Os odontoideum: presentation, diagnosis, and treatment in a series of 78 patients [presentation]. J Neurosurg Spine. 2008;9(4):332-42.
11. Spierings EL, Braakman R. The management of os odontoideum. Analysis of 37 cases. J Bone Joint Surg Br. 1982;64(4):422-8.
12. Wu X, Ling M, Chang Y, et al. Accidental or linked: separated odontoid process fused to the enlarged anterior arch of the atlas associated with atlantoaxial subluxation in a Kashin-Beck disease patient. Eur Spine J. 2017;26(Suppl 1):85-9.