Clinics in Surgery

Case Report
Published: 09 Feb, 2021

Os Odontoideum as a Cause of Cervical Cord Injury in a Patient with Refractory Epilepsy

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Abstract

Os odontoideum is an anomaly of the second cervical vertebrae in which the odontoid process is separated from the body of the axis. Traumatic injury or congenital fusion failure is thought to be the etiology. The clinical symptoms are variable from cervical pain, torticollis, and myelopathy and vertebrobasilar ischemia. Os odontoideum can cause instability of the neck, and neck injuries can cause life-threatening complications. In this report, we present the case of a 15-year-old girl with refractory epilepsy who developed quadriparesis after a fall and hit to the forehead while traveling. Although the symptoms improved, weakness in her right upper limb persisted at 2 months after the fall. Imaging studies revealed Os odontoideum. Based on her medical history, the recent head trauma due to epileptic seizures accompanied by atlantoaxial instability was considered to result in cervical compression and spinal damage. She was at a high risk of sudden death due to recurrent seizures and cervical injury; therefore, she underwent vertebral fusion surgery.

Keywords: Epilepsy; Os odontoideum; Cervical injury; Vertebral fusion surgery

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Received Date: *28 Dec 2020*
Accepted Date: *22 Jan 2021*
Published Date: *09 Feb 2021*

Citation:
Kusabiraki S, Nakagawa E, Saito T, Takayama Y, Iijima K, Iwasaki M, et al. Os Odontoideum as a Cause of Cervical Cord Injury in a Patient with Refractory Epilepsy. Clin Surg. 2021; 6: 3057.

Abbreviations

CT: Cervical Computed Tomography; MRI: Magnetic Resonance Imaging; SPECT: Single-Photon Emission Computed Tomography; FLAIR: Fluid-Attenuated Inversion Recovery

Case Presentation

A 15-year-old Indonesian girl with refractory epilepsy, who developed seizures and quadriparesis following hitting her forehead in Indonesia. The patient was initially unable to walk, but the symptoms slightly improved in approximately 2 weeks. She visited our department 2 months after the injury owing to persisting difficulty in moving the right upper limb. Her physical examination was normal, with no central nervous system symptoms. Muscle tension was noticeably elevated in her right upper limb, with reduced muscular strength (manual muscle test score, 4). Deep tendon reflexes were increased on both sides, particularly on the right upper limb. She was hospitalized for evaluation and rehabilitation. Cervical Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) (Figure 1) confirmed syringomyelia at C1 level. CT revealed Os odontoideum with a smooth surface that was covered with the bone cortex, indicating this was not a new fracture.

The patient's medical history included 10 episodes of general tonic seizures/day for 10 s, with no consciousness for about one minute, at the age of 3 years and 8 months. At 4 years of age, electroencephalogram revealed frequently occurring diffuse polyspikes and waves. Brain MRI (Figure 2) by Fluid-Attenuated Inversion Recovery (FLAIR) imaging revealed a high-intensity lesion surrounding the left Sylvian fissure. The temporal lobe showed a mushroom-like morphology, which was recognized as ulegyria. Single-Photon Emission Computed Tomography (SPECT) was performed to the construct subtraction ictal SPECT co-registered to MRI image (not shown) showing increased ictal blood flow at the same site. She was diagnosed with combined generalized and focal epilepsy with ulegyria. Despite multiple antiepileptic drugs, her seizures were refractory. At 5 years of age, she had a seizure attack, hit her head on falling and experienced neck pain. She was then diagnosed with a fractured vertebra axis and atlantoaxial subluxation and treated with traction and neck collar. Her epilepsy was intractable; therefore, the ulegyria surrounding the left Sylvian fissure was focally resected (Figure 2).

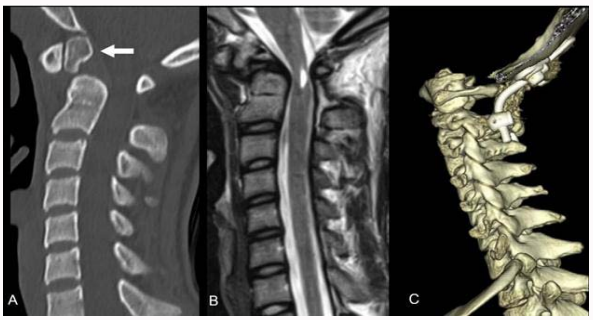

Figure 1: A) Sagittal CT imaging of cervical spine reveals an independent ossicle separated from the odontoid peg (os odontoideum; white arrow). B) T2-weighted sagittal MRI indicating a lesion with high signal intensity at C1 level in the spinal cord. C) Postoperative 3D CT imaging showing posterior occipital cervical reconstruction with screw-rod fixation in occiput-C2 lesion.

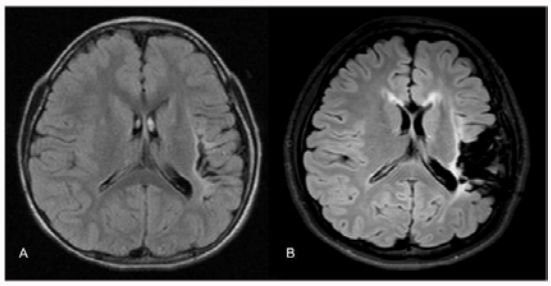

Figure 2: A) At 5 years of age, axial FLAIR MRI showing a high-intensity lesion in the area surrounding the left Sylvian area. B) At 15 years of age, postoperative axial FLAIR MRI revealing dissected anterior corpus callosum and temporal lobe. Glial scars are observed as high-intensity lesions surrounding the dissected area.

She underwent partial corpus callosotomy at 6 years of age when the seizures relapsed, but her epilepsy remained refractory. She was then prescribed zonisamide, phenobarbital, potassium bromide, clobazam and rufinamide, which reduced her seizure frequency to once every 2 to 3 days. Based on the medical history and findings, head trauma due to epileptic seizures with atlantoaxial instability resulting in cervical compression and spinal damage was considered. Her paralysis gradually improved. She was discharged on 11th hospitalized day.

Her seizures were refractory after discharge. The patient was at a high risk of sudden death due to recurrent seizures and cervical injury; therefore, she underwent vertebral posterior fusion surgery 6 months later. Postoperative CT revealed an enlarged spinal canal. Although she experienced refractory spasms, paralysis did not relapse during the 2-year observation period.

Discussion

We believe that this is the first report of a patient with epilepsy and Os odontoideum. Os odontoideum is defined as an independent variably sized ossicle with smooth circumferential cortical margins, separated from the foreshortened odontoid peg [1]. Symptoms vary from minor (pain and torticollis) to serious (myalgia, motor paralysis, and sensory disturbance and cranial nerve defects) [2]. Its etiology is not completely understood. There are two hypotheses: congenital and traumatic. Axis is organized from the proatlas and the C1 and C2 sclerotomes. According to the congenital hypothesis, fusion failure of the odontoid process (C1 sclerotome) with the body (C2 sclerotome) or the odontoid process apex (proatlas) with the odontoid process' main portion causes Os odontoideum. The traumatic hypothesis states that an unnoticed odontoid fracture followed by avascular necrosis and osseous remodeling underlies Os odontoideum [1].

In our patient's case, a fractured cervical vertebra, when she was 5 years of age, may have caused Os odontoideum. However, cervical vertebrae instability was not considered while evaluating refractory epilepsy by MRI. It is necessary to confirm that the cervical vertebrae are present when evaluating sagittal images of patients with refractory epilepsy with cervical injury history. Effective treatment or management approaches are lacking because of unclear Os odontoideum etiology. Arvin and colleagues recommended X-ray examination for forward and posterior bending and head MRI every year for 5 years to evaluate cervical compression risk. The authors recommend not participating in sports with increased contact risk. Asymptomatic patients should be carefully followed, while symptomatic patients with cervical vertebrae instability are advised early surgery [1]. In patients with refractory epilepsy, Kruitbosch et al. [3] reported that the cervical cord injury incidence was 30 to 40 times higher than that in the general population. Furthermore, epilepsy symptoms, impaired consciousness after the epileptic episode and decline in the cognitive ability compared with that before the episode might hinder the cervical cord injury detection [3].

Conclusively, cervical vertebra instability can cause severe injury in patients with refractory epilepsy. Sagittal images during head MRI should be considered to study cervical vertebra. Because falls in epileptic patients are not uncommon, the brain and the spinal cord should be examined. Surgical intervention can prevent spinal cord injury in patients with refractory epilepsy and Os odontoideum.

Statement of Ethics

Informed written consent was obtained from the patient's parents. This study was approved by the ethical committees of the National Center of Neurology and Psychiatry.

Author Contributions

Shohei Kusabiraki, Eiji Nakagawa, Takashi Saito, Yutaro Takayama, Keiya Iijima, Masaki Iwasaki, Ayano Matsui and Tetsuya Abe managed the patient, drafted the article, and revised the final version. All authors read and approved the final manuscript.

Acknowledgement

The authors would like to thank Enago (www.enago.jp) for the English language review.

Funding

We thank the patient and the patient's family whose help and participation made this work possible. This study was supported in part by Intramural Research Grants (1-4, 2-7) for Neurological and Psychiatric Disorders of NCNP, Japan.

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