Atlanticaoxial Dislocation Associated With a Mass in the Extradural Craniovertebral Junction Unrelated to Rheumatoid Arthritis

—Case Report—

Kiyoshi ITO, Keiichi SAKAI, Takehiro YAKO, Yuichiro TANAKA, and Kazuhiro HONGO

Department of Neurosurgery, Shinshu University School of Medicine, Matsumoto, Nagano

Abstract

A 61-year-old woman without rheumatoid arthritis (RA) was admitted with atlantoaxial dislocation (AAD) and a retroodontoid mass at the craniovertebral junction manifesting as a 1-year history of numbness and mild weakness of the right upper extremity. Computed tomography and magnetic resonance (MR) imaging showed AAD and a mass at the craniovertebral junction. She had no past history of RA or trauma in the head and neck. She underwent surgery to obtain the histological diagnosis of the mass and to improve AAD-induced instability. The lesion was approached through the right transcondylar fossa approach with C-1 laminectomy. Intraoperative pathological examination showed cicatrizing collagen fibers and no obvious tumor cells. After partial removal of the lesion, the AAD was fixed with the posterior approach. The symptoms subsided soon after surgery and the mass decreased on MR images taken 3 months after surgery. If a pseudotumor is suspected based on the preoperative radiological investigation in a non-RA patient with AAD and the symptoms are not progressive, stabilization can be expected to induce spontaneous regression without urgent direct excision of the mass.

Key words: atlantoaxial dislocation, craniovertebral junction, os odontoideum, posterior fixation

Introduction

Atlantoaxial dislocation (AAD) is considered to result from damage to the ligaments around the odontoid process caused by rheumatoid arthritis (RA) and trauma. AAD manifests as various symptoms ranging from posterior neck pain and limitation of neck movement to severe myelopathy and fatal medullary disorders. Patients with RA are likely to harbor a pannus around the odontoid process mimicking a tumorous lesion, so the spinal cord is compressed by both the AAD and the pannus. Pannus associated with RA-induced AAD is quite common, and spinal fusion is considered appropriate for treatment. In contrast, such retroodontoid mass is rare in patients without RA.

We treated a patient without RA who presented with AAD complicated by an extradural mass located in the craniovertebral junction.

Case Report

A 61-year-old woman felt numbness in the fingers of both hands in January 2003, and visited a physician. The patient underwent traction therapy, but the symptom did not improve. The numbness had expanded to the bilateral upper extremities by June 2003. Computed tomography (CT) and magnetic resonance (MR) imaging showed AAD and a mass at the craniovertebral junction. The patient had complaints of occipital pain, numbness of the bilateral upper extremities, and mild weakness of the right upper extremity. These symptoms had not progressed rapidly. She had no past history of RA or trauma in the head and neck. She was referred to our hospital.

Neurological examination on admission found dysesthesia of the bilateral upper extremities, motor weakness of the right upper extremity, increased
deep tendon reflex of the bilateral upper extremities, and no vesicorectal disorder. Serum rheumatoid factors were negative. Cervical radiography and CT showed os odontoideum and irreducible AAD. The atlantodental interval was 8 mm in flexion and 6 mm in extension (Fig. 1). No bone destruction was noted (Fig. 2). Cervical MR imaging showed a mass in the posterior region of the atlantoaxial odontoid process as isointense on T1-weighted imaging and heterogeneously intense on T2-weighted imaging, and the margin of the mass was enhanced with gadolinium (Fig. 3).

Based on these findings, she underwent surgery to obtain the histological diagnosis of the mass and to fix the AAD-induced instability. With the patient in the prone position, the lesion was approached through the right transcondylar fossa approach with C-1 laminectomy. Since the dural capsule was tense, the mass on the ventral side could not be reached.

Therefore, the dura was incised and the cerebrospinal fluid was aspirated. The lesion was white, fibrous, and elastic hard (Fig. 4). The origin of this retroodontoid mass may have been the synovium of the odontoid process. The tumorous tissue was partially excised. Intraoperative pathological examination found cicatrizng collagen fibers and no obvious malignant tumor cells (Fig. 5). These findings were not contradictory with a diagnosis of inflammatory changes. Posterior fixation of AAD was then performed, using the Olerud fixation system (Muranaka Medical Instruments Co., Ltd., Tokyo), from the occipital bone to the C-3 and C-4 lateral masses. Bone chips were scattered in the occipitocervical junction.

Motor weakness of the right upper extremity and sensory disturbance of the bilateral upper extremities improved after surgery. Only headache remained as a subjective symptom. Postoperative cervical radiography showed good condition of the fixation, and MR images taken 3 months after
surgery revealed decreased retroodontoid mass (Fig. 6).

**Discussion**

Tumors are rare in the craniovertebral junction, and the diagnosis may be difficult in some cases, but MR imaging has recently demonstrated several cases of cervical myelopathy associated with non-tumorigenic lesions in the posterior region of the odontoid process. An AAD-associated non-tumorous lesion in the posterior region of the odontoid process was described as a pseudotumor, or a new pathological condition that may cause non-tumorigenic lesions compressing the spinal cord. Four cases of pseudotumor complicating AAD in patients without RA are known, and treated by total excision and partial excision (including biopsy) in two patients each.

Total excision was performed through the posterior transcondylar approach. Posterior fixation was concomitantly performed, and the neurological findings improved after surgery with good outcomes. The histological finding was synovial cyst. Partial excision followed by fixation resulted in lesion shrinkage in 2 and 9 months, and the postoperative neurological condition was good. Both procedures can achieve good postoperative results. Histological examination showed inflammatory tissue, so the AAD-induced instability probably caused the mass lesion as part of the natural course. Therefore, all previous cases of pseudotumor were inflammatory or cicatrizing, suggesting that AAD-induced local inflammation expanded and formed the mass. This pathogenesis is similar to that of pannus, the inflammatory feature of the synovial membrane seen in patients with RA.

MR imaging is useful for the diagnosis of pseudotumors, which have smooth margins, and may exhibit segmented morphology. Growing pseudotumors compress the medulla oblongata or spinal cord from the anterior direction. Pseudotumors may appear as homogeneous hypointense on T1-weighted and hypointense on T2-weighted MR imaging, with no or very subtle
enhancement. In this patient, the lesion appeared as isointense and heterogeneously intense on T1- and T2-weighted imaging, respectively, with slight enhancement of the tumor wall.

The differential diagnosis of tumorous lesions in the craniovertebral junction includes meningioma, chordoma, metastatic tumors, osteoma, osteosarcoma, osteochondroma, and fibrous dysplasia. Differentiation of these tumors from pseudotumor may be possible based on the MR imaging signal intensity and the degree of contrast enhancement. Therefore, if the preoperative radiological investigation indicates a pseudotumor, and the symptoms do not rapidly progress and are not serious, correction of the spinal instability may induce spontaneous regression and urgent direct excision of the tumor may not be needed.

References


Address reprint requests to: Yuichiro Tanaka, M.D., Department of Neurosurgery, Shinshu University School of Medicine, 3–1–1 Asahi, Matsumoto 390–8621, Japan.
E-mail: tanaka@hsp.md.shinshu-u.ac.jp