C_1-C_2 cervical myelopathy

KK Lau, SH Yeung, H Cheung

This case report describes a man with C_1-C_2 cervical myelopathy due to an os odontoideum. Over a span of 30 years, the patient developed weakness of his left lower limb, which gradually spread to all four limbs. The diagnosis was not made for three decades despite several medical consultations, a myelogram, and a computed tomography scan of the brain. This case illustrates that neurological disease due to spinal cord compression can have a long history, and demonstrates how advances in magnetic resonance neuroimaging contribute to a definite and accurate diagnosis.

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Introduction

Recognised causes of C_1-C_2 myelopathy include rheumatoid arthritis, osteoarthritis, tuberculosis, cerebral palsy, Down's syndrome, and congenital anomalies of the odontoid. An os odontoideum associated with atlantoaxial instability can lead to narrowing of the spinal canal and cord compression at the C_1-C_2 level. Patients can present with tetraparesis and spasticity of all four limbs.

Case history

This 61-year-old man was confined to a wheelchair and had a long history of progressive weakness of the limbs. Thirty years ago, he had noticed that his left foot was weak, especially on dorsiflexion; his left shoe was worn out more than the right one. There was minimal weakness of his left hand also. Since he was right-handed, however, he could continue his work as a trader. At that time, he had been seen both by an orthopaedic surgeon and a physician. A myelogram did not reveal any abnormality. Ten years later, while he was in Canada, he had a computed tomography (CT) scan of the brain, which was normal. Although he was then fully ambulatory and independent, he noticed a gradual deterioration in his strength. Twenty years later, he had to use a stick for walking.

When the patient presented to us, he was wheelchair bound due to a combination of weakness and spasticity. He employed a strong domestic helper to carry him from his bed to a chair and vice versa. Although his high cortical function was good, there was sphincter dysfunction. There was no history of injury to his neck or head. His family history was unremarkable, and there was no other significant past medical history. He looked smart, was alert, and could give a detailed account of his illness. No skin lesions were present, and his hairline had not receded.

Testing of the cranial nerves and a fundal examination yielded normal results. There was marked spasticity of all four limbs, especially on the left side. The left shoulder abduction and adduction was 2/5 and 4/5, respectively, and the left foot dorsiflexion and plantar flexion was 0/5 and 2/5, respectively. The right counterparts were 3/5 and 4/5 for the right shoulder, and 3/5 and 4/5, for the right foot, respectively. All the jerks were pathologically brisk and there was Babinski sign bilaterally. Abdominal reflex and jaw jerk were not present. Only his right arm was functionally useful as the others were too spastic. The patient had a sensory

Princess Margaret Hospital, Lai King Hill Road, Kowloon, Hong Kong:
Department of Medicine
KK Lau, FHKAM (Medicine), MRCP
Department of Orthopaedics and Traumatology
SH Yeung, FHKAM (Orthopaedic Surgery), FRCS
Department of Diagnostic Radiology and Organ Imaging, Prince of Wales Hospital, Shatin, Hong Kong
H Cheung, FHKAM (Radiology), FRCR

Correspondence to: Dr KK Lau

level below his neck, with diminished response to light touch and pain sensation. Proprioception was intact only in his right arm. There were no clinical signs to suggest rheumatoid arthritis, nor evidence of tuberculosis or osteoarthritis.

Laboratory tests for a complete blood profile, renal function test, liver function test, serum calcium, phosphate, and glucose levels were normal. The test for rheumatoid factor was negative. A lateral cervical radiograph (Fig 1) suggested the presence of an os odontoideum. This was confirmed on sagittal magnetic

Fig 1. A lateral cervical radiograph taken with the neck flexed, suggesting the presence of an os odontoideum (black arrows). The forward displacement of the os and C_1 indicates kinking of the cord at the level of C_{1-2}.

resonance imaging (MRI) [Fig 2], which showed compression of the upper cervical cord at the $C_{1/2}$ level by the os and accompanying soft tissue in the location of the transverse ligament, postero-inferior to the os (Figs 2 and 3). The MRI of the brain was normal.

The patient was referred to the orthopaedic department and an early operation was performed because of his rapid deterioration. The constriction of the upper cervical spine was released by laminectomy of C_1 and C_2, and the cervical spine was then stabilised by an occipito to $C_{3,4,5}$ fusion using a Ransford loop. He was placed in a halo-body vest post-operatively for added external immobilisation.

Fig. 2. Sagittal T_1-weighted magnetic resonance image of the cranio-cervical junction in the supine position confirming the presence of the oval ossicle (white arrows) postero-superior to the anterior arch of the atlas. The location of the os odontoideum is greatly altered compared to that shown in Fig 1, confirming instability. The cord is reduced in diameter and stretched over the os and adjacent soft tissue which is of intermediate- to low-signal intensity on all pulse sequences.

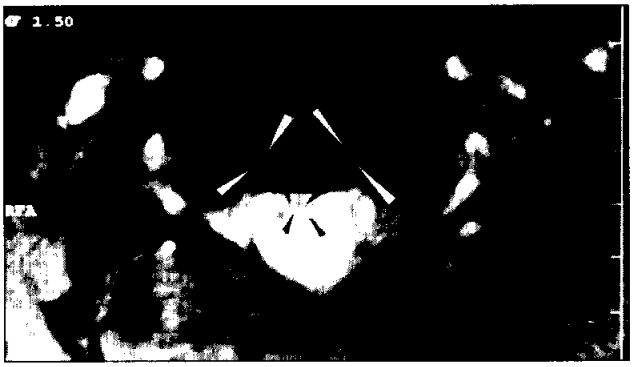

Fig 3. Axial T_2-weighted magnetic resonance image at the C_1 level showing an abnormally high signal within the narrowed cervical cord (black arrows). Tissue with mixed intermediate- to low-signal intensity in the region of the transverse ligament may represent fibrotic hypertrophy and oedema of the ligament (white arrows)

Three months after the operation, there was a major decrease in his spasticity and he could walk with a frame under supervision. Although his left shoulder abduction and adduction were only 3/5 and 4/5, the left foot dorsiflexion and plantar flexion were 0/5 and 4/5, the right counterparts were 4/5 and 5/5 for the right shoulder, and 4/5 and 5/5, for the right foot, respectively. All the jerks remained very brisk, and the bilateral Babinski sign persisted. His right arm and both legs were gaining function with decreasing spasticity. A sensory level was still present, but his response to light touch improved, with positional sense regained on both big toes. He had retention of urine, which he was being treated for.

Discussion

The term "os odontoideum" was first introduced by Giacomini in 1886, and refers to a discrete bony structure that lies cephalad to the axis, in the region of the odontoid process.[1] It is believed to be derived from overgrowth of the "os terminale", an ossification centre at the superior tip of the odontoid process.[2]

The two largest reviews of "os odontoideum" were conducted by Fielding et al[3] and Spierings et al[4] who reported on 35 and 37 patients, respectively. The average age at diagnosis was 18.9 years (range, 3-65 years) and 37.8 years (range, 6-62 years). In Fielding's study, the average duration of symptoms was 1.8 years (range, 1 month - 10 years). Common complaints were pain (64%), numbness (24%), and weakness of all four limbs (16%). Among twelve patients who had cord symptoms in Spierings' study, two had tetraplegia, and both had a history of significant neck injury.

A congenital aetiology of os odontoideum has been proposed.[5] There is, however, more evidence to support an acquired lesion, especially in patients who once had a radiologically-documented normal odontoid process and later presented with an os odontoideum. Schuler documented the natural history of os odontoideum secondary to trauma in a two-year-old girl.[6] Radiographs showed resorption of the dens with subsequent re-ossification and the appearance of a typical os odontoideum 13 months after the injury. Fielding also believes that os odontoideum results from unrecognised fracture through the base of the odontoid in early infancy.[3]

This case is interesting because of the slow progression of symptoms over three decades. The diagnosis was only confidently made by MRI, which was able to show the level and extent of cord compression directly. Magnetic resonance imaging is especially well-suited to the evaluation of the craniovertebral junction by virtue of its direct sagittal imaging capabilities.[1] Occasionally, differentiation between an os odontoideum and a type 2 odontoid fracture is problematic. The presence of associated hypertrophy of the anterior arch of C_1 favours os odontoideum over an acute fracture.[7] This sign was absent in our patient who also denied any previous head or neck trauma. A soft tissue bulge was also seen on the MR images at the level of maximal cord compression. This material exhibited intermediate to low signal on T1 and T2 spin echo sequences and most likely represented fibrous hypertrophy (possibly in association with oedema) in and around the transverse ligament. Fibrous tissue may show variable signal intensity depending on its maturity and tissue composition.[8]

The treatment of patients with os odontoideum is dictated by its likely effect on the cord and the stability of the cervical spine. Prophylactic fusion of the vertebrae is controversial,[4] and probably not recommended,[9] as this procedure severely restricts rotational movements of the head. Surgery was indicated in our patient because he showed progressive cord signs and recent deterioration. The principle of treatment is relatively straightforward—the spinal cord has to be decompressed and the spine stabilised. Post-operative rehabilitation maximises the functional improvement.

The os odontoideum can be excised using an anterior approach through the oral cavity.[10] Incision and reflection of the posterior wall of the oro-pharynx provides access to the anterior arch of the atlas, the removal of which permits excision of the os odontoideum. This approach, however, has inherent theoretical disadvantages. By operating through the oral cavity, strict asepsis is difficult, and may result in higher patient morbidity. Moreover, there is no effective way to secure good stabilisation of the cranio-cervical junction. Hence, the posterior route was chosen in this case. The cord was decompressed by removing the laminae of the atlas and axis from behind. The os odontoideum itself was not touched, allowing the cord to be displaced posteriorly even after decompression. By this means, effective initial stabilisation can be achieved by wiring the Ransford loop to the base of the skull through burr holes, and also sublaminal wires at the C_3, $_4$, and $_5$ levels. Wearing the halo-body vest for three months after the operation helps to protect the bone graft, which will provide a lasting fusion from the occiput to the upper cervical spine.

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