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Case Report

Cervical Klippel-Feil syndrome progressing to myelopathy following minor trauma

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ABSTRACT

Klippel-Feil syndrome (KFS) is a rare disease with a clinical triad of low posterior hairline, short neck, and limited neck motion. Frequent fusion of two or more cervical vertebrae resulting from a congenital segmentation defect can lead to adjacent level hypermobility, instability, and even neurologic symptoms that require surgical intervention. However, surgical results in adults with KFS with concomitant atlantoaxial subluxation and cervical spinal stenosis have not been reported. We report a 58-year-old man with complaints of an unsteady gait, general weakness, and clumsiness in both hands for 6 months. Deep tendon reflexes in both knee joints were increased, with a positive Babinski sign. Bladder and sphincter function were intact. Radiographic findings included C2–C7 congenital fusion with atlantoaxial subluxation and spinal cord compression. He was treated with posterior occipitocervicothoracic fusion, instrumentation, and posterior decompression with a partial craniectomy under the diagnosis of cervical myelopathy. Postoperatively, the neurologic deficits improved without any complications, although bilateral rod breakage was noted at consecutive outpatient department (OPD) follow-ups. He recovered well with residual left hand numbness.

1. Introduction

Klippel-Feil syndrome (KFS), an uncommon disease, is now recognized in some clinical and radiographic retrospective studies [1–3]. Most studies reported no patients with cervical spine-related symptoms. Some symptomatic patients had neck pain, radiculopathy, or myelopathy since childhood and adolescence and the risk of cervical stenosis in these patients was high [4]. However, there are no reports of symptomatic adult patients with KFS presenting with concomitant atlantoaxial subluxation and cervical spinal stenosis. We report a 58-year-old man who sustained cervical myelopathy because of KFS with concomitant atlantoaxial subluxation and cervical stenosis.

2. Case report

A 58-year-old man complained of unsteady gait with clumsiness in both hands for 6 months. He had no other systemic diseases. The symptoms exacerbated in the previous 2 weeks after he fell against a bathroom washstand, resulting in a head contusion with a hyperflexion injury to the cervical spine. He reported bruising over his forehead. He had limited neck extension and flexion movement. On physical examination, he was noted to have a short neck and low posterior hairline. He also complained of general weakness with a sensation of tightness over his body. The deep tendon reflexes in both knee joints were increased, with a positive Babinski sign. Sphincter function was intact. Plain radiography (Fig. 1) showed C2–C7 congenital fusion with atlantoaxial subluxation. The anterior atlantodens interval (AADI) was 8.7 mm and 4.6 mm in the flexion and extension views, respectively. Magnetic resonance imaging (MRI) of the cervical spine showed C1–C5 spinal stenosis with a posterior ossifying enlargement at the C3 and C4 levels (Fig. 2). The space available for the cord was 6 mm at the

Conflicts of interest: none.

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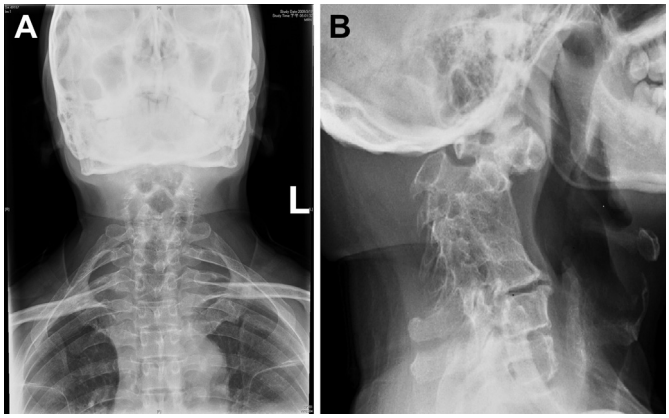

Fig. 1. (A) Anteroposterior and (B) lateral radiographs of the cervical spine of a 58-year-old man with type III Klippel-Feil syndrome with atlantoaxial subluxation.

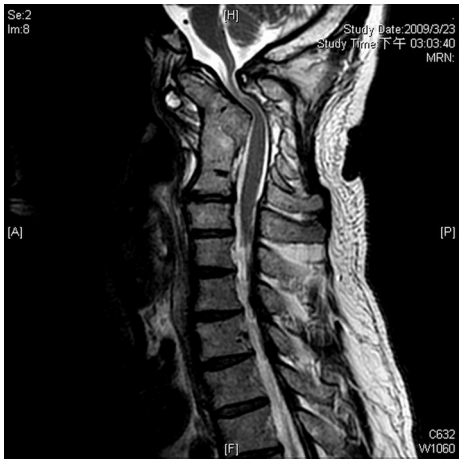

Fig. 2. Sagittal T2-weighted MRI scan shows extensive C2–C7 fusion with ossifying enlargement and C1–C5 spinal stenosis. There is no increased signal intensity within the spinal cord.

C1 level. Computed tomography (CT) showed occipitocervical junction instability (Fig. 3).

After the diagnosis of KFS with cervical myelopathy was made, the patient underwent a two-stage operation. First, he received halo vest external fixation under local anesthesia, assisted by fluoroscopy. The aim was to maintain correct craniocervical alignment and help the patient adapt to postoperative conditions without further neurologic deterioration (Fig. 4). After being observed for 2 days, he was treated with posterior decompression with a laminectomy from C1 to C5 and a partial craniectomy of the occiput. Good pulsation of the dura was observed immediately after decompression. The altered anatomy of the congenitally fused segments made it too difficult to apply lateral mass screws. Occipitocervicothoracic fusion was extended to the T3 level using a titanium screw-rod fixation system (Summit occipitocervicothoracic spinal fixation system, DePuySpine, Rayham, MA, USA). Occipital screws were bicortically purchased and secured with the plates on the midline. Then the occipital screws and plate were connected to the thoracic pedicle screws with rods, followed by assembly of the crosslinks. The exposed dura was covered with absorbable gelatin sponge. A humerus strut allograft plate was placed posteriorly over the decompressed segments between the cranium and C6 for occipitocervicothoracic fusion (Fig. 5). There was no worse change of the somatosensory-evoked potential intraoperatively. After the surgery, the patient reported subjective improvement. The Nurick score [5] improved from 3 preoperatively to 0 at 6 months postoperatively. The halo vest was removed 1 month postoperatively under stable conditions.

The patient tolerated the rehabilitation program well. The muscle power in his four limbs recovered well except for residual

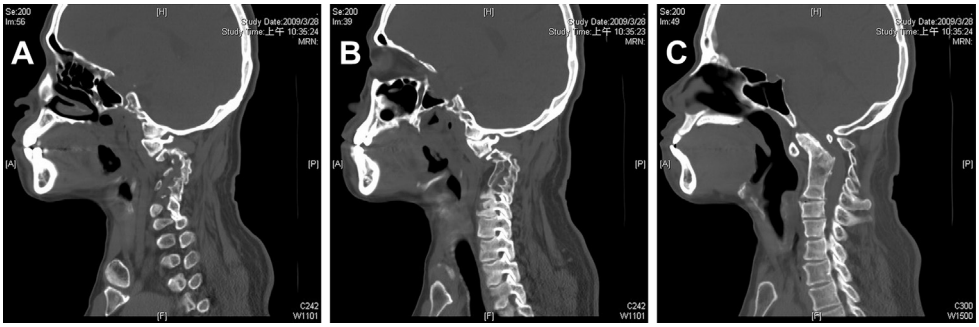

Fig. 3. (A, B, and C) Sagittal CT scans showing superior odontoid migration without occipitalization. Angular motion has caused an increase in the atlantodens interval, indicating instability.

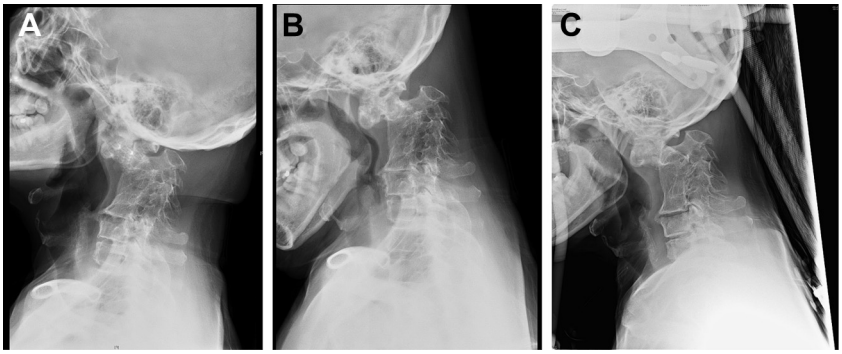

Fig. 4. (A and B) Instability of the occipitocervical junction preoperatively. (C) After application of a halo vest. Atlantodens interval = 4 mm, upper hard palate-upper edge of T1 angle: 0°.