Case Report

Atlantoaxial dislocation adjacent to kyphotic deformity in a case of adult Larsen syndrome

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Abstract

Kyphotic deformity is often seen in Larsen syndrome. However, its progress in adults is not clear. The adjacent level compression in these patients adds to the difficulty regarding the level that needs to be operated. A 56-year-old male presented with neck pain and spastic quadriplegia. Radiology showed kyphotic deformity (sequelae of Larsen syndrome) with atlantoaxial dislocation. Cord compression was apparent at both levels but careful evaluation showed C1-2 level compression and some compression below the kyphotic deformity. The kyphotic spine was already fused and the canal diameter was adequate. The adjacent level C1-2 was fused and he improved dramatically. Correction of long-standing kyphotic deformity may not be necessary, as it unlikely to progress because of its tendency to fuse naturally. Rather, the adjacent levels are likely to compress the cord due to excessive stress. A proper clinical history and a thorough radiological examination help the surgeon to make an appropriate decision.

Key words: Atlantoaxial dislocation, cervical kyphotic deformity, Larsen syndrome, two level compression

INTRODUCTION

Progressive cervical kyphosis is often seen in Larsen syndrome.1,2 However, little is known about its progress or presentation in adulthood. Occasionally, compression is seen at other levels apart from the kyphotic deformity. The clinical features are often overlapping and it is difficult to decide the level to be operated. We describe a case of Larsen syndrome in an adult with cervical subaxial kyphotic deformity and adjacent atlantoaxial dislocation. A proper informed consent was obtained from the patient.

CASE REPORT

A 56-year-old male presented with complaints of neck pain radiating to occiput and progressive spastic quadriparesis (JOAS-12 at presentation) for 12 months. There were no clinical features of posterior column or spinothalamic tract involvement. There was no history of trauma or tuberculosis. X-ray of the cervical spine revealed a gross kyphotic deformity at C2-C4 and increased atlantoaxial interval, suggestive of atlantoaxial dislocation. Anterior osteophytes were seen at C4-5 level [Figure 1a and b]. Careful evaluation of computed tomography (CT) of the cervical spine revealed the C1-2 level compression.

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spine showed kyphotic deformity due to anterior wedging of C2-4 vertebrae. The vertebral bodies were fused to each other anteriorly. The disc space was narrow at C4-5 with anterior osteophytes but no posterior osteophyte. Atlantoaxial dislocation was apparent in addition to cervical kyphosis. The atlantoaxial joints showed degenerative changes [Figure 1c]. Magnetic resonance imaging (MRI) demonstrated significant compression of the cord with signal changes at the craniovertebral junction and moderate compression at the subaxial spine [Figure 1d].

The patient underwent direct posterior reduction with fusion of C1-2. The C1-2 facets were drilled flat and spacers with bone graft were placed. C1 lateral mass and C2 pedicle screw were put and fixed with a rod. Intraoperative manipulation was done to achieve reduction.

Spasticity improved significantly in the postoperative period. Radiology showed good anteroposterior and vertical reduction [Figure 1e]. At a 12-month-follow-up, he is able to carry out his daily activities independently (JOAS-17). Currently, the patient is being followed up for a lower level pathology at C5-6.

**Diagnosis**

The diagnosis of Larsen syndrome is usually established in childhood. They often present with progressive cervical kyphotic deformity. Both surgical and conservative treatment options have been described with variable success in children.[1-3]

However, the natural history of kyphotic deformity in Larsen syndrome is not clear. The presentation in adults is rare.[4] The kyphotic deformity in adults is likely to be long-standing and is unlikely to progress. It is possible that the kyphotic vertebrae fuse with each other, as seen in our case. The compression seen due to kyphotic deformity is quite deceptive. In fact, the adjacent levels (both cranial and caudal) are likely to get affected due to such kyphotic fusion. It is the adjacent level that causes cord compression, giving rise to symptoms due to increased stress.

The decision-making in managing these cases is difficult due to its rarity and the multi-level compression. Treating kyphotic deformity is not necessary, as it is nonprogressive. In addition, the radical resection and reconstruction of such kyphotic spine is not without danger. Usually, it is one adjacent level that compresses more than the other and requires surgical intervention. In our case, the kyphotic deformity deceptively appeared to compress the cord. However, careful evaluation showed compression at C1-2 level and mild compression just caudal to kyphotic deformity. In addition, the kyphotic deformity showed fusion. MRI showed signal change at cervicomedullary junction and cervical syrinx. This helped us take a decision to operate at this level. The patient improved remarkably. The lower adjacent level can be tackled at a later date in case the patient becomes symptomatic.

The adjacent levels should be carefully evaluated in cases with such kyphotic deformity. The genetic defect in the filamin part of cytoskeleton results in ligamentous laxity. These patients usually have multiple joint dislocations with kyphoscoliosis.[1]

**CONCLUSION**

In conclusion, the subaxial kyphotic deformity itself may not be the cause of cord compression in all cases of adult Larsen syndrome. Adjacent level degenerative changes may be responsible for cord compression. Careful evaluation of clinical features and radiology, especially the level adjacent to the deformity is of paramount importance.

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**Conflicts of interest**

The authors have no conflict of interest.

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