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# High cervical C3-4 'disc' compression associated with basilar invagination

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A 20-year-old male had torticollis and short neck since birth. He presented with symptom of progressive quadriparesis over a two-year period. Investigations revealed basilar invagination with marked rotation in the craniovertebral region and relatively large C3-4 region osteophytes. Serial MRI over two years showed persistent signal opposite C3-4 disc space suggestive of cord compression. Although the cord was humped over the odontoid process, there was no clear radiological evidence that the cord was compromised at this level. During surgery, instability was identified only at the craniovertebral region and not at the level of C3-4. Distraction of the lateral masses of atlas and axis and fixation using interarticular spacers and bone graft and direct screw implantation in the lateral mass of the atlas and pars of the axis resulted in reduction of the basilar invagination and of atlantoaxial dislocation. The patient had marked clinical recovery, despite the fact that no direct procedure was done for C3-4 disc decompression. The case suggests that C3-4 disc changes could be secondary to primary instability at the craniovertebral junction.

**Key words:** Assimilation of atlas, atlantoaxial dislocation, basilar invagination, high-cervical disc prolapse

## Introduction

Basilar invagination is frequently associated with a complex of bony and neural anomalies in the craniovertebral region and spine including platybasia, 'fixed' atlantoaxial dislocation, assimilation of the atlas, malformation of the occipital bone and a range of fusion defects of cervical and other spinal vertebral bodies. High cervical disc prolapse is rare in general and particularly its presence in association with basilar invagination is rarely reported.<sup>[1-6]</sup> This may probably be due to the limited range of movements in the region and the strength of the ligaments. We had earlier reported two cases where high cervical disc-related cord compression was associated with basilar

invagination.<sup>[2]</sup> In both cases, the C3-4 disc compression was treated by the transoral surgical route. In one of these two cases the disc was resected and inter-body plate and screw fixation was done and in the other C3-4 disc was resected in addition to odontoidectomy. No fixation surgery was done in the latter case. Apart from these two cases, we could not locate any other report where the management issues and association of high-cervical disc and basilar invagination is discussed. We report another case of basilar invagination with a fixed rotatory atlantoaxial dislocation, wherein investigations showed that the main site of neural compression was at the level of the C3-4 disc. The clinical issues raised by the successful treatment by craniovertebral fixation alone are discussed.

## Case Report

A 20-year-old male patient had short neck and torticollis since birth. For about two years, he had neck pain and progressive quadriparesis. Investigations done one year ago had shown basilar invagination, 'fixed' atlantoaxial rotatory dislocation, assimilation of atlas and fusion of C2 and C3 vertebrae. There was clear evidence of cord compression due to large osteophytes opposite the C3-4 disc space [Figure 1]. Although the cord was humped over by the odontoid process, no cord changes were evident at this level. Repeat investigations at the time of admission showed similar radiological features. Considering the presence of clinical and radiological features of instability of the craniovertebral region, a stabilization procedure was planned that would have had incorporated the occipital bone and the C3-4 vertebra. However, during operation, a marked instability was identified at the craniovertebral- atlantoaxial joint, whilst the C3-4 vertebrae appeared remarkably stable. As per our earlier published protocol, we performed a distraction, reduction and fixation of the craniovertebral instability that included basilar invagination and 'fixed' atlantoaxial dislocation.<sup>[7,8]</sup> Specialized spacers and

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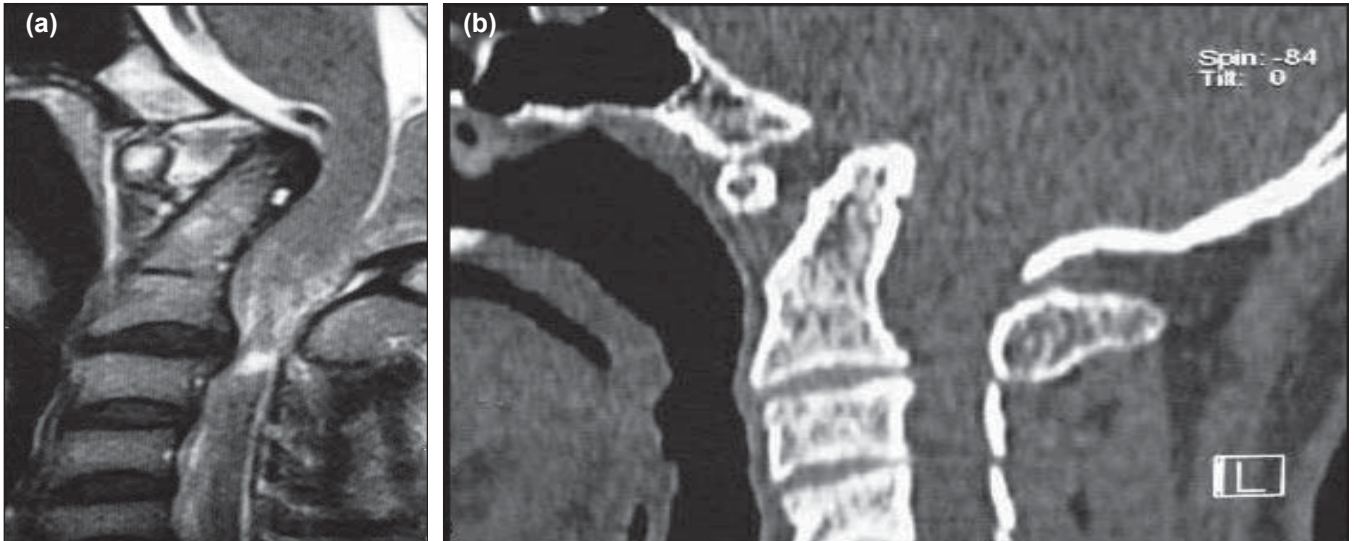


Figure 1: (a) T2-weighted MRI showing basilar invagination, atlantoaxial dislocation, assimilation of atlas, C2-3 fusion and hyperintensity in the cord at the level of C3-4 disc, (b) CT scan showing basilar invagination, atlantoaxial dislocation, assimilation of the atlas, C2-3 fusion and relatively large posterior osteophytes at the C3-4 disc level

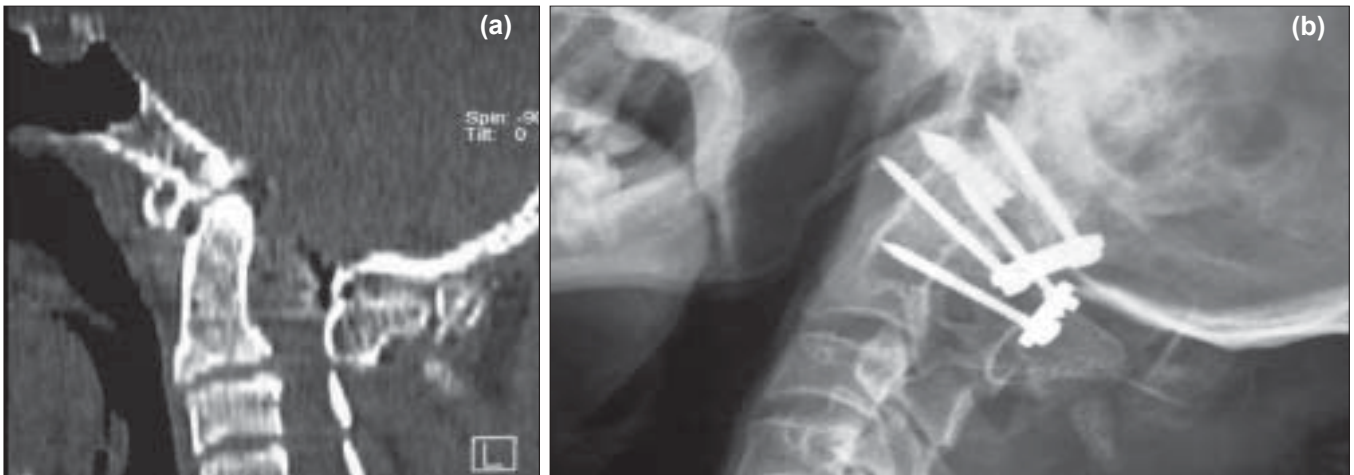


Figure 2: (a) Postoperative CT showing reduction of the basilar invagination and atlantoaxial dislocation, (b) Postoperative X-ray, showing fixation of the region using plate and screws and intra-articular spacer

bone graft pieces harvested from the iliac crest were impacted in the distracted atlantoaxial joint and direct plate and screw fixation incorporating C1 facet and C2 pars was carried out. The procedure was technically remarkably difficult due to severe invagination and joint rotation. The C2-3 and C4 spinous processes revealed no evidence of instability and no fixation of the region was attempted. He was placed on hard-cervical collar for three months. Postoperatively, the patient had remarkable clinical improvement. He was able to walk unaided swiftly, within few days of surgery. As stainless steel plates and screws were used for fixation, the status of C3-4 region cord compression could not be analyzed [Figure 2].

### Discussion

High cervical disc-related compressive lesions

are relatively rare and the surgical strategy and philosophy for such lesions is still under evaluation.<sup>[1,3,6]</sup> Radiological presence of high cervical disc-related cord compression in the presence of basilar invagination and other craniovertebral region anomalies has been rarely identified earlier. We had earlier reported two cases, wherein, we had done transoral decompression and fixation of the C3-4 vertebra using inter-body plates and screws in one case and decompression of both the odontoid process and the disc lesion in the other.<sup>[4]</sup> The MRI picture in our present case suggested cord compression opposite the C3-4 disc space. In the clinical situation, treatment of the C3-4 disc compression appeared to be the most rational form of treatment. Considering the additional presence of basilar invagination and 'fixed' atlantoaxial dislocation it was decided to fix the craniovertebral region incorporating the C3-4 segments and consider

a direct surgical decompression of the region by an anterior approach at a subsequent stage. C1-2 facet distraction, realignment of the region and fixation as discussed resulted in remarkable clinical improvement. Such a treatment was essentially a level above the actually demonstrated site of cord compression. The remarkable clinical recovery following the treatment raises a philosophical issue regarding the pathogenesis and treatment of such a complex combination of anomalies.

As has been discussed in the literature earlier, high cervical disc protrusion can be a result of excessive strain on this joint due to the abnormally 'fixed' craniovertebral junction.<sup>[5,6]</sup> Exact cause of high cervical disc compression and the effect of craniovertebral fixation are difficult to comprehend and evaluate. However, it does appear that the craniovertebral anomaly was the primary pathology and the C3-4 disc region compression a secondary phenomenon. Treatment of the primary pathology probably resulted in the relief from a factor that caused secondary effect on the cord.

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Accepted on 15-06-2007

**Source of Support:** Nil, **Conflict of Interest:** None declared.