

BASILAR INVAGINATION - IMPENDING SUDDEN DEATH - REPORT OF A LIFE SAVING SURGERY

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ABSTRACT

Introduction: Basilar invagination is an uncommon syndrome that occurs when the superior part of odontoid (C2) migrates upward. It occurs both congenitally and in bone/joint diseases. It may lead to static or dynamic stenosis of the foramen magnum, and compression of medulla oblongata, which may manifest clinically as sudden death from brainstem compression. **Case Report:** A 25-year-old man presented with gradually progressive weakness of right arm and left leg for six months, associated with walking difficulty. MRI brain revealed atlanto-axial subluxation with posteriorly subluxed odontoid peg causing severe compression and posterior displacement of brainstem. MRI of spine showed extensive syringo-hydromyelia. He underwent trans-oral odontoidectomy, and posterior fusion, followed by a halo traction. We report a marathon life saving surgery done at our institution that to our knowledge has not been reported from Pakistan.

Key words: Basilar invagination, trans-oral odontoidectomy, life saving

INTRODUCTION

Basilar invagination is an uncommon syndrome that occurs when the superior part of odontoid (C2) migrates upward. It occurs both congenitally and in bone/joint diseases. It may lead to static or dynamic stenosis of the foramen magnum, and compression of medulla oblongata, which may manifest clinically as sudden death from brainstem compression.¹ Obstructive hydrocephalus and/or syringomyelia are common because of direct mechanical blockage of CSF flow.^{2,3} We report a marathon life saving surgery done at our institution that to our knowledge has not been reported from Pakistan.

CASE REPORT

We report the case of a 25-year-old gentleman who presented with gradually progressing weakness of right arm and left leg for past six months. It was associated with difficulty in walking. Power in right upper limb was 2/5

and in left lower limb was 3/5. The power in left upper limb and right lower limb was normal. There was loss of sensations below knee and he had sensory ataxia. He had normal reflexes and planters were down going. MRI brain (figure 1) revealed atlanto-axial subluxation associated with posteriorly subluxed odontoid peg causing compression and posterior displacement of brainstem leading to moderate atrophy of brain stem. MRI of spine showed extensive syringo-hydromyelia of spinal cord extending from C2 up to T10. He was placed in cervical traction pre-operatively, then he underwent a trans-oral odontoidectomy with the inferior clivus and then posterior fusion followed by Halo traction.

Surgical Technique

The patient was anesthetized in supine position with orotracheal intubation. Tracheostomy was performed and orotracheal tube was removed. Dingman retractors were applied, soft palate was split and mucosa on hard palate was retracted. A shelf of bone was removed along with

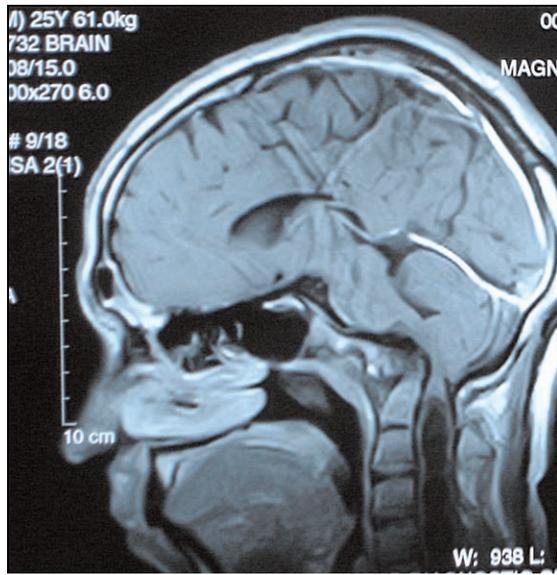


Figure 1: Pre-op MRI brain showing (i) basilar invagination of the odontoid process (ii) platybasia and (iii) cervical syrinx



Figure 2: X ray cervical spine with post fixation and Halo vest

vomer. An incision was made in the pharyngeal wall and pharyngeal muscles were retracted. Arch of atlas and odontoid process were identified and confirmed with the help of C-arm. Resection of odontoid process along with the anterior arch of the atlas was done using bone nibbler and high speed drill. Wound was closed in layers. Patient was turned into prone position carefully and craniocervical fusion was done with a titanium lateral mass plate. This plate was fixed with the help of screws into occipital and C2-3 lateral masses on both sides. Bone graft was also laid down at surgical site for bony fusion. The wound was closed in layers. Patient was then kept in Halo cast for 6 weeks.

Follow Up

He had a good recovery and was sent home on the 11th post-operative day with the Halo cast (figure 2). On three months follow up motor deficit had improved remarkably. Follow up MRI at 3 months after surgery showed partial resolution of the cervical and thoracic syrinx (figure 3). The Halo was removed and the patient was sent home. At five months follow up patient had improved with subtle residual motor deficit and was leading a near normal life.

DISCUSSION

Basilar invagination is a potentially lethal craniocervical anomaly. Its importance lies in its ability to produce atlanto-axial instability which may be complicated by

subluxation following minor trauma. Neurosurgical treatment is quite difficult. Some suggest that ventral brainstem compression should be treated with ventral decompression, followed by occipitocervical fusion.⁴ In Chiari malformation, there may also be occipital decompression surgery followed by occipitocervical fusion with contoured loop instrumentation to prevent further squamooccipital infolding. Ventral decompression means operating through the front of the head, through the mouth. Both the transoral and posterior surgery can be done in one sitting.⁵ Another approach is atlantoaxial joint distraction and direct lateral mass fixation using screws.⁶ A study was performed on 190 patients with basilar invagination by Goel A. et al to assess the appropriate surgical procedure.⁷ Depending on the association with Chiari malformation, the anomaly of basilar invagination was classified into two groups (Group I no Chiari malformation and Group II with Chiari malformation). The principal pathological characteristic was observed to be direct brainstem compression due to odontoid process indentation in Group I and a reduction in posterior cranial fossa volume in Group II. Despite the anterior concavity of the brainstem in both groups, transoral surgery was the most suitable procedure for those patients in Group I and decompression of the foramen magnum was found to be appropriate for patients in Group II. After surgical decompression, a fixation procedure was found to be necessary in most Group I cases, but only in a small minority of Group II cases. When neurological features are absent, a conservative approach may be pursued, such as cervical collar, NSAIDs and simple neck traction. However neurological progression is likely and one year prognosis is poor.⁸ In our patient, anterior decompression with

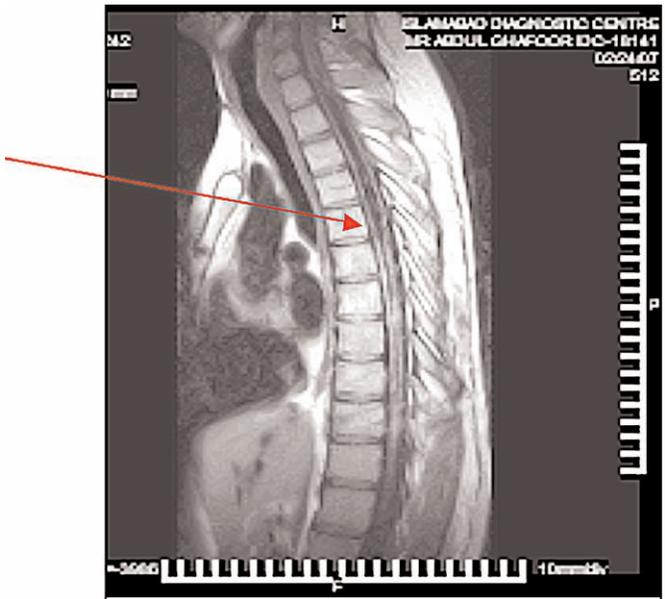
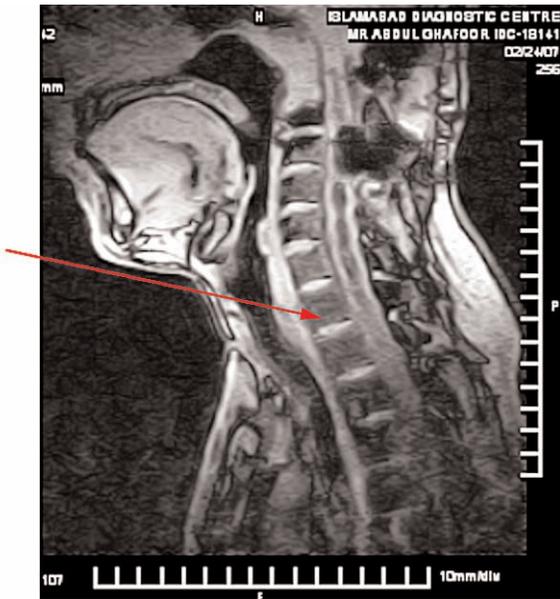


Figure 3: Follow up T1 sagittal image 3 months after surgery showing partial resolution of the (a) cervical syrinx (b) thoracic syrinx (arrows)

posterior fixation was performed with good functional outcome.

CONCLUSION

Basilar invagination is commonly associated with instability of the region and the complex results in a significant degree of neck pain and myelopathy adding considerably to the disability secondary to affection of other joints. For basilar invagination, trans-oral decompression and subsequent posterior fixation have been the most accepted treatment protocol. The surgical treatment of upper cervical spine and skull base remains challenging because of anatomical complexity and limited surgical accessibility. Our case demonstrates an example of successful surgery with avoiding of a potential threat to life.

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