Case Report

Atlantoaxial arthrodesis using C1-C2 transarticular screw fixation in a case of Morquio syndrome

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Abstract

Prophylactic or therapeutic arthrodesis is recommended for atlantoaxial instability in Morquio syndrome. Occipitocervical fusion, the common approach for upper cervical fusion in Morquio syndrome sacrifices the movements at the occipitoatlantal joints. The use of C1-C2 transarticular screws for achieving C1-C2 arthrodesis, without compromising mobility at the occipitoatlantal joint in Morquio syndrome has not been reported. We report a case of Morquio syndrome with atlantoaxial instability and odontoid hypoplasia, where we successfully achieved C1-C2 arthrodesis using transarticular screws and bone graft. The advantages of this method over other methods of atlantoaxial arthrodesis in Morquio syndrome have also been discussed.

Key words: Atlantoaxial arthrodesis, Morquio syndrome, occipitocervical fusion, transarticular screw

Introduction

Atlantoaxial instability in Morquio syndrome can be due to ligamentous laxity, hypoplastic/dysplastic odontoid, or failure of fusion of the C1 ring anteriorly or posteriorly.[1–4] It is notorious not only for producing neurologic symptoms but also because of the associated mortality. Prophylactic fusion of the upper cervical spine has been recommended in patients even before the appearance of neurologic symptoms.[1,4,5] Occipitocervical fusion is the recommended method of upper cervical fusion in these patients.[4,5] We report a case of Morquio syndrome where atlantoaxial instability was managed successfully with C1-C2 arthrodesis only, using transarticular screws and bone grafts.

Case Report

A 6-year-old girl, a known case of Morquio syndrome, presented with neck pain of insidious onset, along with progressive weakness of all the four limbs since 3 months. She had an unstable gait, with history of repeated falls. Neurological examination revealed grade 3 power and spasticity in all four limbs, with extensor plantar responses bilaterally. All sensations were intact.

Radiographs and computed tomography (CT) images of the cervical spine revealed a hypoplastic odontoid [Figure 1a], bifid posterior arch of C1, atlantoaxial subluxation, and increased atlantodens interval. MRI confirmed atlantoaxial subluxation with spinal cord compression at C1-C2 level [Figure 1b].

The child was operated on an emergency basis in view of the progressive neurological deficit. CT scan images of the patient were assessed for suitability for placement of 3.5-mm transarticular screws across the C1-C2 facet joints. Use of an operating microscope assisted in the dissection. This was coupled with intraoperative fluoroscopy to enhance safety in screw placement. The patient, in cervical traction, was placed prone, with the head end of the table elevated to 30°. Satisfactory reduction of the atlantoaxial joint was confirmed fluoroscopically. A posterior midline incision was made extending from the occiput to the C2 spinous process and was extended deep into the relatively avascular midline structures. The posterior arch of the atlas was exposed by subperiosteal dissection. It was confirmed to be bifid as had been revealed by preoperative radiographs. It was completely excised and utilized as bone graft for C1-C2 lateral mass fusion. An adequate decompression was performed and the C1-C2 facet joints on both sides were

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Access this article online	
Quick Response Code:	Website: www.ijoonline.com
	DOI: 10.4103/0019-5413.83766

exposed. After widely excising the capsule of the joint, it was packed with bone graft. Under image guidance, a guide wire was passed across the C1-C2 facet joint bilaterally in the trajectory previously described.[6-7] A 3.5-mm screw with a length of 34 mm was placed along the predetermined screw trajectory bilaterally. A midline stabilization could not be performed due to the presence of the bifid C1 posterior arch.

Postoperatively, the neck was immobilized in a Philadelphia collar for 3 months. The patient was ambulated with support after 2 days. The power in the limbs improved gradually and was restored to MRC grade 5/5 at the end of 3 months, with no residual spasticity. The patient was then ambulating normally without any aid. Postoperative MRI revealed adequate decompression of the spinal cord [Figure 1c]. There was no evidence of instability on the flexion–extension lateral radiograph of the cervical spine at 24 months and the fusion did not extend beyond the desired level [Figures 2a and b].

Discussion

Disability and life expectancy in patients with Morquio syndrome depends upon skeletal complications, amongst which cervical myelopathy is the most important.[1,4] Atlantoaxial instability in these patients can result in life-threatening dislocation. Prophylactic fusion of the upper cervical spine can prevent the development of myelopathy and can be lifesaving.[1,4] Occipitocervical fusion is the commonly recommended method in these patients.[4,5] Ransford *et al*.[4] performed occipitocervical fusion in 17 patients with Morquio syndrome. They used onlay femoral or tibial autografts placed posteriorly, secured to the laminae of C1 and C2 to obtain occipitocervical fusion. Gluf and Brockmeyer[8] reported a case of Morquio syndrome where C1- C2 transarticular screws were utilized within the occipitocervical construct. However, occipitocervical fusion for atlantoaxial instability sacrifices the movements at the occipitoatlantal joint [50% flexion-extension (30°) and 10° lateral bending]. In this case, only atlantoaxial

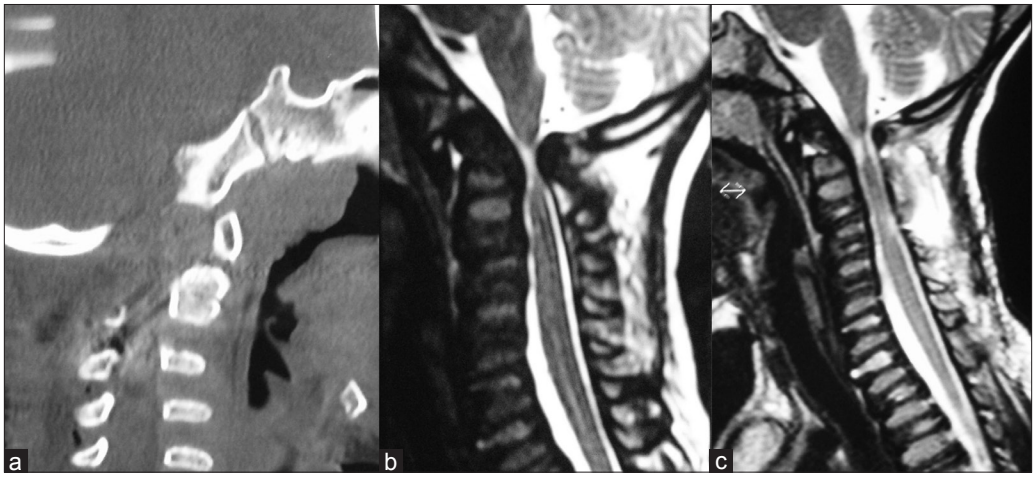

Figure 1: (a) Mid-sagittal CT image showing odontoid hypoplasia (b) Preoperative MR image showing the wafer-thin spinal cord at the craniovertebral junction due to the atlantoaxial subluxation (c) Postoperative MR image showing adequate decompression of the spinal cord

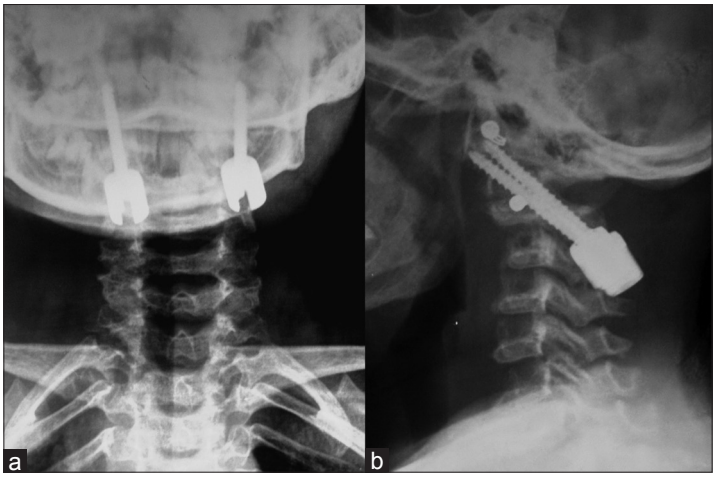

Figure 2: Anteroposterior radiograph of upper cervical spine (a) and lateral radiograph (b) at 24 months follow-up showing C1-C2 transarticular fixation

arthrodesis was performed, thus preserving occipitoatlantal motion. Atlantoaxial arthrodesis was accomplished using C1-C2 transarticular screw fixation as this method has certain advantages. It provides excellent mechanical immobilization, thus maximizing the chances of fusion.[7] Moreover, the bone graft placed in the C1-C2 facet joints is in compression mode, which is physiologically optimum for successful fusion; on the other hand, the onlay graft in occipitocervical construct is in tension mode.[8] Fusion rates of up to 100% have been reported in pediatric patients with C1-C2 transarticular screws.[8] Solid osseous union between C1-C2 facet joints was noted radiographically at 3 months follow-up in our patient. The power in the limbs was restored and there was no residual spasticity. The fusion did not extend beyond the desired level and the occipitoatlantal joint was mobile. This method may be associated with complications like screw malpositioning or catastrophic vascular or neural injury. However, meticulous preoperative planning to determine the screw trajectory and to assess the position of the foramen transversarium, along with the use of intraoperative fluoroscopy, can help avoid complications.[7–8] C1-C2 arthrodesis using wires and grafts has been extensively used. However, biomechanical studies have shown that they provide only semirigid fixation and therefore require a halo body jacket to increase the chances of fusion.[9] Studies comparing C1-C2 fusion rates using screws with that by wires plus bone grafts and halo body jacket report significantly higher fusion rates with the former (98% *vs* 86 %; P=0.003).[8]

To the best of our knowledge there is no mention in the literature of a case where atlantoaxial instability in Morquio syndrome was managed exclusively with C1-C2 transarticular screws and bone graft. In contrast to occipitocervical fusion, there was no arthrodesis of the mobile occipitoatlantal joint and thus motion at that level could be preserved. We chose C1-C2 screws as this provides rigid internal fixation and achieves fusion rates significantly higher than those with cable fixations. This technique can be used for both prophylactic fusions as well for treatment of symptomatic patients, where it can be performed after surgical decompression.

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How to cite this article: Kulkarni AG, Shah SM. Atlantoaxial arthrodesis using C1-C2 transarticular screw fixation in a case of Morquio syndrome. Indian J Orthop 2011;45:470-2.

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