Dystrophic Scoliosis with Rib Head Dislocation into Spinal Canal in Neurofibromatosis-1 and its Surgical Management: A Case Report

Tankeswar Boruah[1], Akhilesh Kumar[2], Ramesh Kumar[3]

From [1]Associate Professor, [2]Senior Resident, [3]Head and Director, Spine Unit Central Institute of Orthopaedics, Safdarjung Hospital, New Delhi, India.

Correspondence to: Dr. Akhilesh Kumar, MS orthopaedics, Spine Unit Central Institute of Orthopaedics, Safdarjung Hospital, New Delhi, Delhi - 110029, India. E-mail: akhileshkumar0702@gmail.com.

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ABSTRACT

Neurofibromatosis-1(NF-1) is one of the most common single-gene disorders in humans. Patients with NF-1 may present with a wide variety of clinical manifestations. Spinal deformity is the most common musculoskeletal manifestation of NF-1. Rib head dislocation into the spinal canal is not a common entity but if left untreated can lead to serious neurological complications. The present report describes thoracolumbar dystrophic scoliosis with rib head dislocation into the spinal canal in a 28-year-old male who presented with complaint of spinal deformity with gait disturbance. Cobb angles measured 92 degrees of scoliosis (T9-L2), and 110 degrees of kyphosis and also highlights the importance of preoperative planning and radiological imaging for the successful surgical outcomes and our experience of single-stage posterior deformity correction with pedicle screw construct along with rib head excision.

Keywords: *Dystrophic spinal deformity, Rib head resection, Single-stage deformity correction.*

The neurofibromatosis (NF) is a spectrum of multifaceted genetic disorders. An afflicted patient may present with a wide range of clinical manifestations that include abnormalities of the skin, nervous tissue, bones, and soft tissues [1]. Spinal deformity is the most common osseous defect of NF1 and the prevalence of scoliosis ranges from 10% to 77% of patients [2]. Scoliosis in NF-1 is further classified into non-dystrophic and dystrophic types as per radiographic characteristics. Dystrophic curves tend to progress aggressively. So, dystrophic curves do not respond to brace treatment, and early fusion is advised for these cases [3].

In some dystrophic forms, rib head dislocation into the spinal canal may be encountered due to enlargement of neural foramina by the erosion of adjacent neurofibroma and may cause neurological deterioration by compressing the cord [4,5]. Rib head dislocation into the spinal canal, whether should be excised or not is still a controversy [6,7]. We describe a case in which we treated the deformity by single-stage posterior pedicle screw instrumentation and spinal fusion along with rib head excision.

CASE REPORT

A 28-year-old man, a shopkeeper by profession with a known family history of neurofibromatosis presented to our institute with complaints of spine deformity and gait disturbance. The deformity of spine appeared around 10 years of age and since then it has increased progressively. The patient took the treatment from the charlatan of the local area who fooled him with his quackery and as the patient belonged to the very remote area of our country so he could not manage to reach tertiary medical centre until now for the proper management of his problem.

On systemic examination, the vitals were stable. Physical examination revealed multiple cutaneous neurofibroma, present all over the body along with café-au-lait spots which lead to the diagnosis of NF1 (Fig. 1). On neurological examination, his lower limb deep tendon reflexes were exaggerated with bilateral Babinski and ankle clonus. He had grade 4 motor power in all

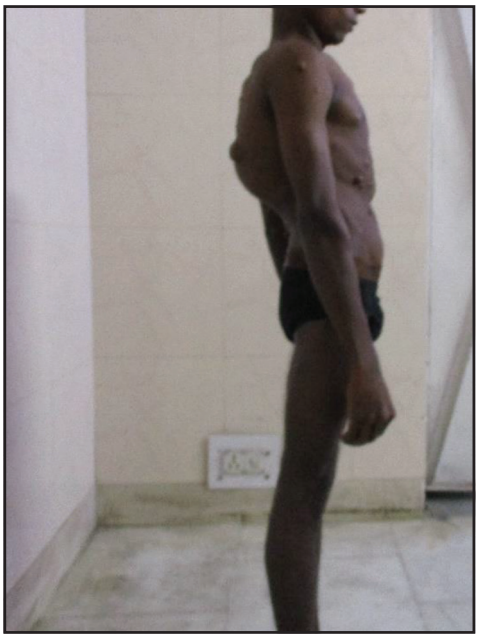

Figure 1: Clinical picture of the patient showing scoliotic deformity along with neurofibromas all over the body

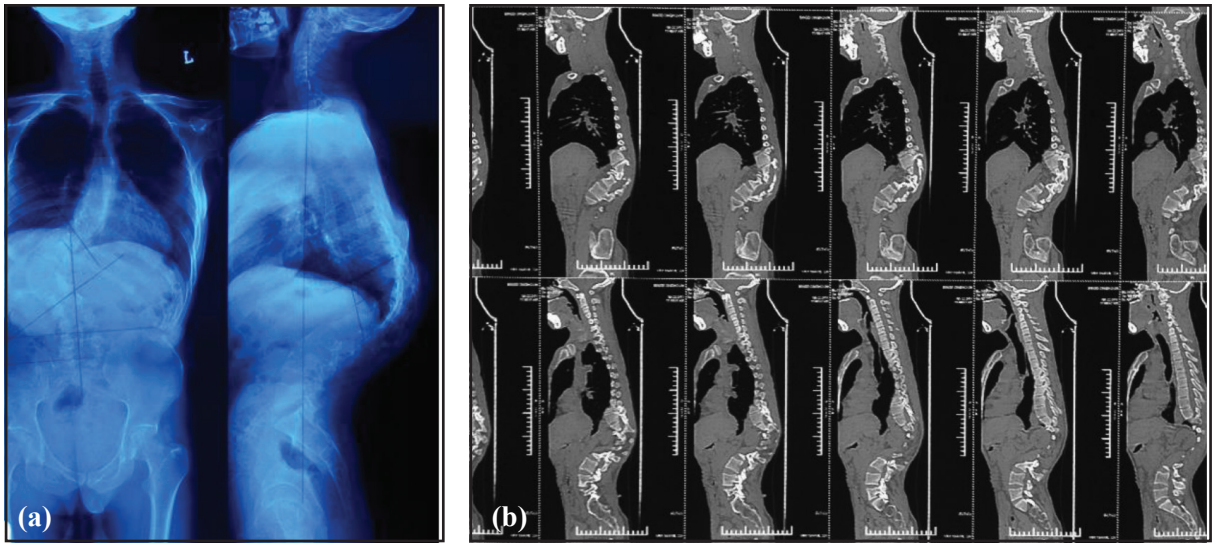

Figure 2: (a). Radiograph showing severe kyhoscoliosis of the lower dorsolumbar spine with convexity towards right side is seen epicenterd at D12 level; (b) CT scan sagittal section showing the kyphotic deformity with wedging of D11 vertebrae.

L2, L3, L4, L5 and S1 key muscles. Sensation and bladder bowel functions were normal. His gait was spastic.

The radiological imaging (X-ray and Computed Tomography (CT) scan) showed thoracolumbar kyphoscoliosis with convexity towards the right side (Fig. 2a and b). Cobb angles measured 92 degrees of scoliosis (T9-L2), and 110 degrees of kyphosis (T10-L2). It was a structural deformity. Magnetic resonance imaging (MRI) showed wedging of D12 vertebrae with acute angulation of cord at D11-D12 level with secondary canal stenosis with subtle altered signal intensity in cord with myelomalacic changes. The CT and MRI showed intraspinal dislocation of rib at the apex of deformity at the D11 level on the convex side (Fig. 3a and b).

After preoperative planning, the patient was operated under neuromonitoring control on January 2018.The patient was operated in a prone position with the posterior midline incision. Posterior instrumentation with pedicle screw fixation was done at D4, D6, D8, D9, D10, L1-5, S1 bilaterally with a bilateral iliac screw. Decompression was achieved by laminectomy and facetectomy from D9-D12 with anterior gibbectomy at D11, D12, L1along with rib head excision at D11level with deformity correction by smith Peterson osteotomy at D9 and D10 level and bone disc bone osteotomy at D11, D12 level followed by application of local bone autograft and allograft (b-ostin calcium phosphate nano-crystalline granules).

The overall surgery was uneventful. The total duration of surgery was eight hours and the estimated blood loss was around 1450 ml. The patient was kept in the intensive care unit for the same night and was extubated the next morning successfully. Post-operatively, the patient had the same power of 4/5 in all L2 to S1 key muscles without any sensory deficit. Bladder bowel function was also preserved, the brisk lower limb reflexes, that were present prior to the surgery, were still present after surgery. Postoperative Cobb's angle was reduced to 38^0 and the kyphotic angle to 17^0 and clinically deformity was not very obvious (Fig. 4).

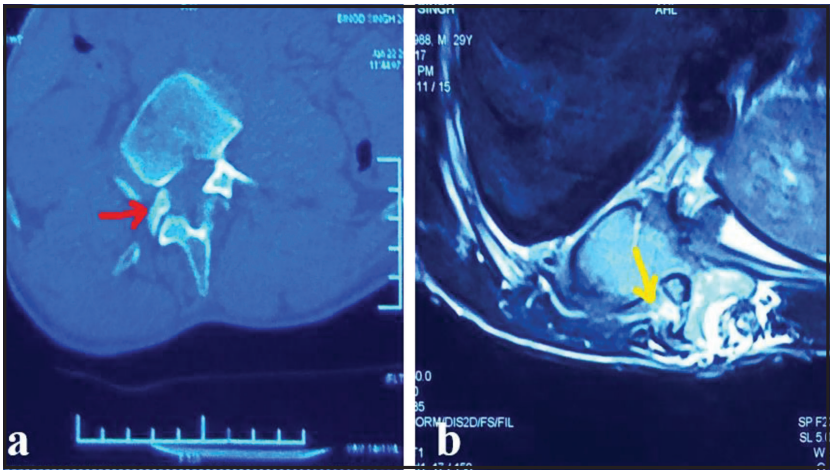

Figure 3: (a) Axial CT image demonstrating intraspinal displacement of the right eleventh rib head (red arrow); (b) T2-W MR images demonstrate intraspinal displacement of the rib head (yellow arrow) with narrowing of the spinal canal but without cord impingement.

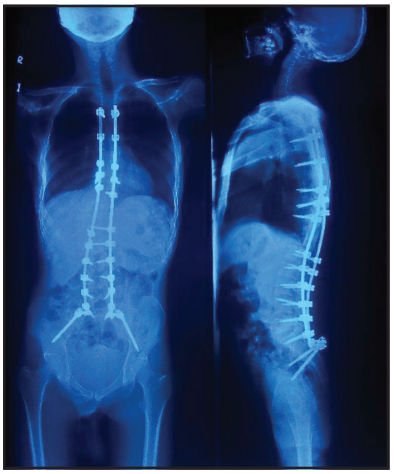

Figure 4: Postoperative spine radiograph showing pedicle screw placement with improved kyphoscoliotic deformity.

Diagnostic Criteria defined by the 1987 Consensus Development Conference of the National Institutes of Health for the diagnosis of neurofibromatosis

Six or more cafe-au-lait macules >5 mm in greatest diameter in pre-pubertal individuals and >15 mm in greatest diameter in post-pubertal individuals
Two or more neurofibromas of any type or more than one plexiform neurofibroma
Freckling in the axillary or inguinal regions
Optic glioma
Two or more Lisch nodules (iris hamartomas) by slit lamp examination
A distinctive osseous lesion, such as sphenoid dysplasia or thinning of a long bone cortex, with or without pseudoarthrosis
A first-degree relative (parent, sibling, or offspring) with NF-1 by the above criteria

For making the diagnosis, at least 2 major criteria are required

Figure 5: Showing Diagnostic Criteria defined by the 1987 Consensus Development Conference of the National Institutes of Health for the diagnosis of neurofibromatosis.

The patient was kept on dorsolumbar sacral orthosis for 3 months. At one year follow-up, the patient was doing his routine activities without any complication and his curvature was still maintained the same as in the postoperative period. His neurological examination was normal with 5/5 muscle power in the lower limb with some residual spasticity.

DISCUSSION

Neurofibromatosis (NF) is an autosomal dominant hereditary disease characterized by the abnormal proliferation of neural crest cells [8]. Two distinct clinical forms of neurofibromatosis are NF-1 or peripheral neurofibromatosis (Von Recklinghausen's disease) and neurofibromatosis-2 (NF-2) or central neurofibromatosis [1]. Bony dysplasia, bony erosion, demineralization, nonossifying fibromas, and scoliosis are all features of NF1 [9].

This patient was diagnosed as NF1 (Fig. 5), he had the dystrophic type of kyphoscoliotic deformity with a severe apical rotation of T9 to L2 vertebrae with an apex at T12. The clinical presentation and radiological imaging which favoured dystrophic features in this patient included a) early occurrence at the young age of <10 years, b) presence of vertebral scalloping of T12 and L2 vertebrae, c) acute angular scoliotic deformity, d) sagittal plane severe kyphosis and e) progression of deformity after skeletal maturity. Rib head dislocation into the spinal canal is also a special feature of dystrophic scoliosis but not a common entity. Although affected patients generally are asymptomatic, some can develop myelopathic features. Few patients present with back pain due to rib hump described as "painful rib hump sign" by Gkiokas *et al.* [10]

As per the literature, dystrophic changes promote faster progression of the curve and cannot be corrected by bracing alone [11]. As the patient had a severe form of kyphoscoliotic deformity with myelopathic features also, surgical correction of deformity with

Figure 6: Postoperative CT scan at 1 year follow-up. Adequate spinal fusion was seen with well-maintained curves as in immediate postoperative.

instrumentation was indicated in this patient. A thorough physical examination and assessment of all radiological images should be done to minimize complications. Features of the vertebral or intraspinal lesion, duralectasia, and spinal cord pseudo meningocele, protrusion of the neurofibroma through neural foramina, spinal cord compression over a sharp angular kyphosis or rib head dislocation into spinal canal need to be identified as they can cause catastrophic neurologic compromise after surgical correction.

In this case, CT-scan with three-dimensional reconstruction was done to visualize the complex bony anatomy, extent of deformity, size of pedicles. MRI of the whole spine is essential for dystrophic scoliosis as it is instrumental in visualizing the internal content of the spinal canal, presence of space-occupying lesion and about the spinal cord itself. In this case, MRI showed lateral wedging of D11 vertebrae with acute angulation of cord at D11-D12 level with secondary canal stenosis with subtle altered signal intensity in cord with myelomalacic changes.

It is difficult to correct intraoperatively such severe dystrophic deformity and to maintain this correction after surgery. Combined anterior release/fusion and posterior fusion using either a pedicle screw construct or a hybrid construct are being done to correct deformity and recommended by authors [12]. After long-term follow-up, it is seen that sagittal correction was maintained with both constructs. However, patients treated with posterior instrumentation using pedicle screw constructs had significantly less loss of coronal correction [13].

Few case series have shown good results with one-stage posterior pedicle screw constructs [14]. We also did single-stage correction

using a posterior pedicle screw construct along with osteotomy, rib head excision and posterior spinal fusion. For achieving fusion, plenty of both autogenous and allograft was used as there are high chances of pseudoarthrosis in NF1 scoliosis. Evaluation of the fusion was assessed on CT scan to identify any evidence pseudoarthrosis because if there is any radiological evidence of pseudoarthrosis, re-exploration and augmentation of the fusion should be undertaken. After one year follow-up, the patient showed good fusion on CT scan (Fig. 6) and his sagittal and coronal curves were well maintained as it was in the postoperative period.

There are still no set guidelines for the surgical management of rib head dislocation into the spinal canal. In the literature, rib head excision has been the preferred method [4,5,8,10] but still, some have shown that spontaneous reduction of rib head following curve correction [6,15]. So,decompression of the spinal cord by resecting the dislocated rib head seems to be safest and advocated by many authors in literature and we also followed the same with a successful outcome.

CONCLUSION

Surgical correction of dystrophic scoliosis is a challenging and demanding surgery. For managing the dystrophic curve with rib head dislocation, a proper preoperative evaluation and study of radiological imaging is of utmost importance in order to decide for its excision or not. Though there are no set guidelines but still rib head excision is a preferred method.

Conflict of interest: The authors declare no conflicts of interest.

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