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ORIGINAL ARTICLE

Rib head protrusion into the central canal in type 1 neurofibromatosis

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Abstract
Background Intraspinal rib head dislocation is an important but under-recognized consequence of dystrophic scoliosis in patients with neurofibromatosis 1 (NF1).
Objective To present clinical and imaging findings of intraspinal rib head dislocation in NF1.
Materials and methods We retrospectively reviewed clinical presentation, imaging, operative reports and post-operative courses in four NF1 patients with intraspinal rib head dislocation and dystrophic scoliosis. We also reviewed 17 cases from the English literature.
Results In each of our four cases of intraspinal rib head dislocation, a single rib head was dislocated on the convex apex of the curve, most often in the mid- to lower thoracic region. Cord compression occurred in half of these patients. Analysis of the literature yielded similar findings. Only three cases in the literature demonstrates the MRI appearance of this entity; most employ CT. All of our cases include both MRI and CT; we review the subtle findings on MRI.
Conclusion Although intraspinal rib head dislocation is readily apparent on CT, sometimes MRI is the only cross-sectional imaging performed. It is essential that radiologists become familiar with this entity, as subtle findings have significant implications for surgical management.

Keywords Neurofibromatosis · Dystrophic scoliosis · Rib head · MRI

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Introduction

Neurofibromatosis type 1 (NF1), also known as von Recklinghausen disease, involves the spine in 10% to 69% of patients [1, 2]. Kyphoscoliosis is the most common spinal deformity and presents in dystrophic and nondystrophic forms [2, 3]. Nondystrophic scoliosis resembles idiopathic scoliosis and is usually a long-segment biconvex curve secondary to leg length discrepancy [2]. Dystrophic scoliosis, on the other hand, is characterized by progressive, sharply angulated short-segment curvature with severe wedging, rotation and scalloping of the apical vertebral bodies [3]. Foraminal enlargement, spindling of the transverse processes and penciling of the apical ribs can also be seen with dystrophic scoliosis [2]. A few case reports have described spinal canal penetration by dislocated ribs in patients with NF1 and dystrophic scoliosis. Although most are asymptomatic, spinal cord compression with resultant paraparesis and paraplegia have been documented both before and after spinal instrumentation [1, 4–15].

We report the clinical and radiographic findings of four cases of rib head dislocation into the spinal canal in patients with dystrophic scoliosis secondary to NF1, including

radiographs, CTs and MRIs, as well as clinical manifestations and management strategies in this uncommon but important entity.

Materials and methods

This study was conducted with IRB review and approval. We conducted a retrospective review of imaging studies performed on four children with NF1 referred for surgery at our institutions with rib head protrusion into the spinal canal identified on preoperative imaging and confirmed at surgery. Clinical history, imaging studies, reports and operative reports, and post-operative courses were reviewed. We also present an analysis of children with this entity in the English literature from 1986 to 2009.

Results

Between 2003 and 2009, rib head dislocation into the spinal canal was found at our institutions in four children, ranging in age from 9 to 14 years old. Radiographs, CTs, MRIs, and operative reports were available in all four, along with clinical follow-up from several months to 6 years. Table 1 summarizes clinical and imaging findings. Table 2 summarizes the 17 cases that have been reported in the English literature.

Case 1 is a 14-year-old boy with severe progressive scoliosis who complained of neck and upper back pain. He was mildly tender to palpation at the apex of his curve. He had slight hyperreflexia of the left lower extremity (3+) and a few beats of ankle clonus bilaterally. The preoperative spine radiograph (Fig. 1) demonstrates penciling and displacement of the fourth and fifth ribs at the apex of the curve, although only the fourth was in an abnormal position on CT and MRI. Rib head resection can be seen on the postoperative spine radiograph.

Case 2 is an 11-year-old asymptomatic girl with severe kyphoscoliosis. The preoperative spine CT and MRI demonstrate rib head displacement into the spinal canal at the apex of the curve, with cord impingement on MRI (Fig. 2).

Case 3 is an 11-year-old boy with progressive but asymptomatic scoliosis and kyphosis. Spine CT and MRI demonstrate no evidence of enlarged neural foramina or vertebral scalloping, but displacement of the right ninth rib with spinal canal narrowing (Fig. 3).

Case 4 is a 9-year-old girl with progressive scoliosis who developed back pain and right foot weakness as well as hyperreflexia of both lower extremities and sustained ankle clonus. CT and MRI reveal a neurofibroma destroying portions of the T4 to T6 vertebral bodies and filling adjacent neural foramina, with intraspinal displacement of the right sixth rib (Fig. 4). MRI also shows cord compression by the displaced rib head.

Discussion

Dystrophic scoliosis in children with NF is typically characterized by a short-segment, sharply angulated curve with associated wedging and scalloping of the vertebral bodies. It can be accompanied by vertebral body rotation, widening of the intervertebral foramina and penciling of rib

Table 1 Summary of clinical and imaging findings (4 cases)

Age (yrs)	Sex	Curve (Apex, Cobb angle); displaced rib(s)	Signs & symptoms	Diagnosis made on	Cord impingement/ compression	Presence of neurofibroma	Management
14	M	Levoscoliosis (61°); convex side left 4th rib	Back pain; mild lower limb hyperreflexia and ankle clonus	CT, MRI	No	Yes	T4 laminectomy with rib head resection and posterior fusion/instrumentation
11	F	Hemivertebra T9; Dextroscoliosis T9 (123°); Kyphosis (62°); convex side right 10th rib	Asymptomatic	CT, MRI	Yes	No	Multilevel diskectomies, T9 laminectoy and posterior fusion with resection of 10th rib head
11	M	Dextroscoliosis (90°); Kyphosis (85°); convex side right 9th rib	Asymptomatic	CT, MRI	No	No	9th rib head resection, anterior and posterior fusion and T9 laminectomy
9	F	Dextroscoliosis (60°); convex side right 6th rib	Back pain; right foot weakness, hyper-reflexia and clonus	CT, MRI	Yes	Yes	Resection of neurofibroma and 6th rib head; combined anterior and posterior fusion and instrumentation

Table 2 Summary of 17 cases from literature review

Authors	Age (yrs)	Sex	Curve (Apex, Cobb angle); displaced rib (s)	Signs & Symptoms	Diagnosis Made on	Cord impingement/ compression	Presence of Neuro-fibroma	Management
Flood et al.	15	M	Dextroscoliosis T4–T8 (80°); multiple ribs on convex side	Knee & ankle clonus	CT-M	No	Yes	Two-stage vertebral wedge resection with rib excision and fusion. Traction used perioperatively. Curve decrease to 45 degrees with resolution of clonus
Major et al.								
	13	F	Levokyphoscoliosis (T9, 52°); convex 8, 9 & 10th ribs	Transient loss of sensation below the waist and inability to move lower extremities after fall on rib hump	CT-M	No	No	Two-stage: anterior fusion with resection of rib heads followed by 2nd stage posterior fusion and segmental spinal instrumentation
	5	F	Levoscoliosis (T7, 75); two ribs on convex side	None	CT	No	No	Two-stage: anterior interbody fusion with resection of rib heads followed by 2nd stage posterior fusion and segmental spinal instrumentation
	11	M	Double thoracic scoliosis (29°); convex side 4th rib	None	CT-M	No	No	Posterior fusion with resection of rib head
Deguchi et al.	12	F	Dextrokyphoscoliosis T4–T7 (T5, 90°); convex side 5 & 6th ribs	Weakness of the lower extremities, difficulty walking with eventual paraparesis, hyperesthesia below waist, ankle clonus and knee/ankle hyperreflexia	CT-M	Yes	No	Two-stage combined anterior and posterior spinal fusion and instrumentation. Dislocated rib head was resected. 2 yr f/u with normal neurological status
Dacher et al.	10	F	Dextroscoliosis (T8, 48°); convex side 8th rib	Bilateral ankle clonus and daytime micturition	CT-M	No	No	Two-stage spinal fusion with Cotrel-Dubousset instrumentation. 1 yr f/u with normal neurological status
Kamath et al.	13	M	Dextroscoliosis T7–T11; convex side 10 h rib	None	CT	No	Yes	Intraspinal rib head resection with right T9–10 hemilaminectomy and posterior fusion/ instrumentation
Khoshhal et al.	16	M	Dextroscoliosis T3–T6 (T5, 83°); convex side 5th rib	Postoperative T5 paraparesis	CT	Yes	No	Initially performed in situ posterior fusion without correction or instrumentation. Anterior decompression with rib head resection was performed after T5 paraparesis developed
Mukhtar et al.	10	M	Dextroscoliosis T4–T12 (76°); convex side 9th rib	Back pain; right leg weakness and radiculopathy down to toes when he rolled to his right side	CT and CT-M	Yes	No	Partial rib resection with rib head left in situ. Posterior fusion from T6 to L1 was performed
Gkiokas et al.	13	F	Levokyphoscoliosis T4–T10 (T8, 75°); convex side 8th rib	Gait difficulty, bilateral Bobinski, ankle clonus, right foot drop, decreased sensation/hyperreflexia in lower limbs, and daytime micturition	CT, MRI	Yes	No	Rib head resection with posterior spinal fusion/ instrumentation from T1 to L2. Cobb angle postoperatively was 65 and patient neurological status returned to baseline

Table 2 (continued)

Authors	Age (yrs)	Sex	Curve (Apex, Cobb angle); displaced rib (s)	Signs & Symptoms	Diagnosis Made on	Cord impingement/ compression	Presence of Neuro-fibroma	Management
Crawford et al.								
	5	[a]	Cervicothoracic kyphoscoliosis Protrusion of three rib heads	[a]	CT	[a]	[a]	[a]
	[a]	[a]	Protrusion of one rib head	[a]	CT	[a]	[a]	[a]
	[a]	[a]	Protrusion of one rib head	Asymptomatic	CT	[a]	Yes	[a]
Yalcin et al.								
	14	M	Dextroscoliosis; convex side two ribs	None	CT, MRI	Yes	Yes	Hemilaminectomy followed by rib head resection and posterior fusion with instrumentation
	12	F	Dextroscoliosis T8–L1; convex side rib	None	CT	No	Yes	Hemilaminectomy followed by vertebral translation and pedicle screw fixation. Rib head was not resected
	6	M	Levoscoliosis TL spine; convex side 10 & 11th ribs	None	Radio-graphs & CT	Yes	Yes	5-level annulotomy with resection but not removal of displaced rib heads. Deformity correction with fusionless instrumentation with pedicle screws
Cappella et al.								
	14	M	Double thoracic kyphoscoliosis (kyphosis 120°); convex side 5th rib	Preoperative weakness of lower limbs. Postoperative weakness and paraparesis	CT, MRI	Yes	No	Initially performed two-stage anterior and posterior instrumentation and arthrodesis. Subsequently, rib head resected and posterior cord decompression after postoperative paraparesis developed

[a] Images are demonstrated in a review article on NF1 spine, but additional information is not available

CT-M CT myelography

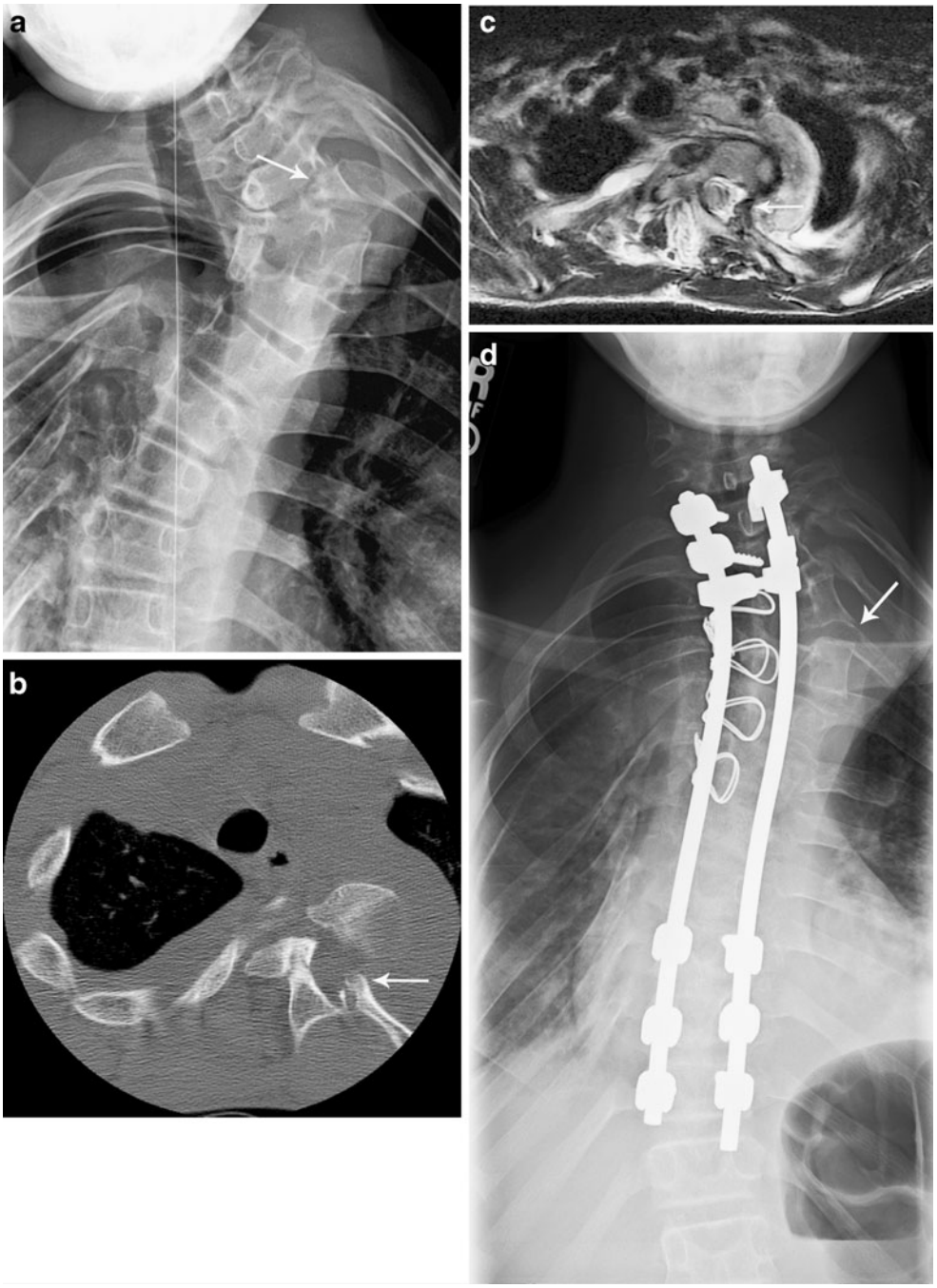

Fig. 1 Case 1. **a** Radiograph shows levoscoliosis of the upper cervical spine with penciling deformity and medial positioning of the left fourth rib head (*arrow*) relative to the pedicle. Note that the fifth rib is also malpositioned, although it was not intraspinal on cross-sectional imaging. **b, c** Axial CT and T2-W MR images demonstrate intraspinal displacement of the left fourth rib head (*arrow*) with narrowing of the spinal canal but without cord impingement. **d** Post-operative spine radiograph shows interval spinal rod and pedicular screw placement with improved levoscoliosis. The displaced left fourth rib head has been resected (*arrow*)

heads. These abnormalities predispose children with dystrophic scoliosis to intraspinal rib head dislocation [1]. To our knowledge, a total of 21 (including our four) cases of intraspinal rib head dislocation in NF1 patients have been reported in the English literature. The majority of documented cases of intraspinal rib head dislocation in NF1 occur during the teenage years (ages range from 5 to 16 years) [1, 4–15], with no gender predisposition. Although generally asymptomatic, the clinical presentation of intraspinal rib displacement varies. Two of our patients had moderate symptoms; the other two were essentially asymptomatic. This parallels the cases we found in the literature, with nine of the 15 for whom this information was available being essentially asymptomatic. The other six had neurological symptoms ranging from mild sensory and motor deficits to paraplegia and paraparesis.

Both Khoshhal and Ellis [10] and Cappella et al. [15] describe the postoperative complication of rib head dislocation in NF patients who developed paraparesis several weeks after posterior spinal fusion without recognition of or attempt at correcting rib head protrusion. In retrospect, rib head protrusion had been present on a preoperative MRI in

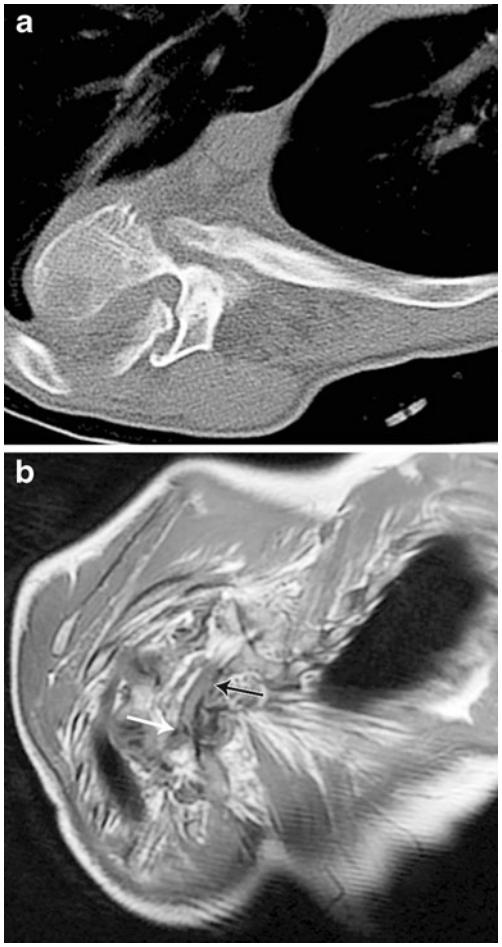

Fig. 2 Case 2. **a** Axial CT shows intraspinal dislocation of the right 10th rib with marked narrowing of the spinal canal. **b** Coronal T1-W MR image demonstrates displacement of the rib head (*white arrow*) intraspinally with narrowing of the canal and impingement of the cord (*black arrow*)

Cappella's patient but had not been recognized. Both cases illustrate the importance of delineating the presence of intraspinal rib head dislocation preoperatively since surgical correction of the scoliosis can bring the displaced spinal cord to its more anatomical location and result in higher risk of cord impingement by the unrecognized dislocated rib head.

Although paraparesis caused by intraspinal rib head dislocation is rare, it remains a diagnostic consideration in NF1 patients who develop acute or progressive neurological symptoms. Of the 17 cases presented in the literature for which clinical information is available, six had evidence of cord compromise or impingement by the displaced rib head. Our case series reveals a similar incidence, with two of our four patients demonstrating such findings.

In our cases, as in the literature, rib head dislocation occurred at the convex side of the apex of the scoliosis, most often involving the mid- to lower ribs. Furthermore, a single rib was involved in the majority of cases (10 out of 17) [5, 8–15], and in all of our patients.

There is no clear consensus regarding the treatment of intraspinal rib head dislocation in dystrophic scoliosis, with most favoring excision of the rib head (under unusual circumstances, the rib head may be left in place) [14]. However, preoperative recognition is essential for surgical planning so that the relationship of rib to cord can be assessed intra-operatively and manipulation performed with appropriate caution.

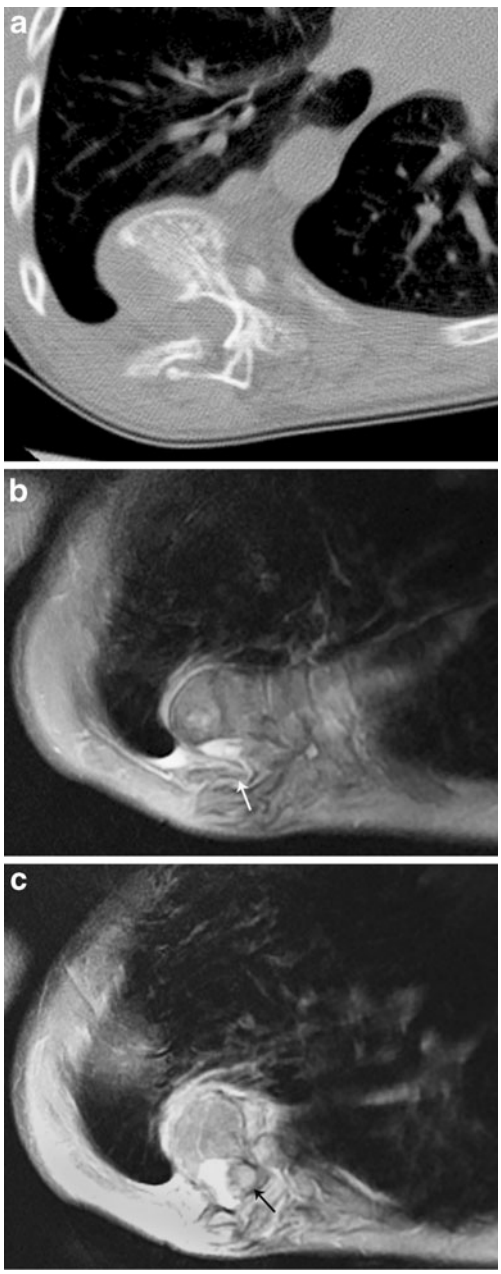

Fig. 3 Case 3. **a** Axial CT demonstrates displacement of the right ninth rib head intraspinally. **b, c** Contiguous post-contrast axial T1-W MR images show the dislocated rib head (*white arrow*) with narrowing of the spinal canal but no cord compromise. The cord (*black arrow*) is located at the concave side of the curvature

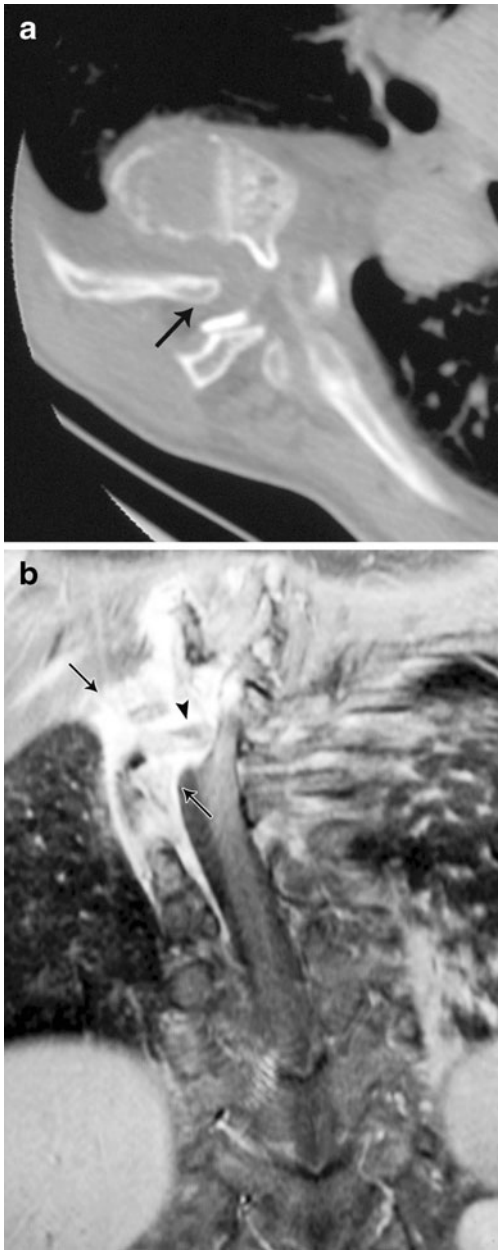

Fig. 4 Case 4. **a** Axial CT shows widening of the neural foramina with intraspinal displacement of the right sixth rib head (*black arrow*). Linear ossific density to the right of the displaced rib head represents the superior articulating facet of the lower vertebrae. **b** Coronal T1-W post-contrast MRI reveals an enhancing right paraspinous neurofibroma (*arrows*) extending into the spinal canal at the apex of the curve. There is displacement of the cord to the left by the dislocated right sixth rib head (*arrowhead*)

Spine radiography is usually the initial imaging for scoliosis, but radiographic diagnosis of intraspinal rib head dislocation is extremely difficult. All reported intraspinal rib head dislocations have been reliably demonstrated by CT, and our cases are no exception. CT myelography accurately depicts the relationship between the spinal cord and the dislocated rib, but MRI demonstrates this noninvasively.

However, MRI – excellent for delineating cord and paraspinous soft-tissue pathology – is sometimes the only cross-sectional preoperative imaging performed. Concern over radiation exposure might limit preoperative CT further in the future, especially if critical findings like rib head dislocation can be diagnosed accurately with MRI. Delineation of bony anatomy is certainly more difficult with MRI than with CT, but MRI can demonstrate intraspinal rib head dislocation, as in our four patients. In our experience, intraspinal displacement of rib heads is best shown on T2-weighted images in the axial and coronal planes.

Conclusion

Intraspinal rib head dislocation in NF1 is an uncommon entity with significant clinical and surgical implications. Although affected patients are generally asymptomatic, presentation ranges from mild back pain to weakness and other myelopathic symptoms. Clinical diagnosis is difficult, and radiological diagnosis requires close scrutiny for subtle findings, such as medial and superior positioning of a penciled rib head – and even then it is extremely challenging. Intraspinal rib head dislocation is reliably diagnosed with CT in all reported cases in the literature and in the cases we present. However, MRI – with its excellent delineation of soft tissues and cord, as well as its lack of radiation – is often performed without CT in the work-up of children with scoliosis, and coronal and axial T2-W sequences can demonstrate intraspinal rib head dislocation. Evaluation of osseous structures with MRI is certainly more difficult than with CT, and it is essential that radiologists become familiar with rib head displacement, as subtle findings have significant implications for surgical management.

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