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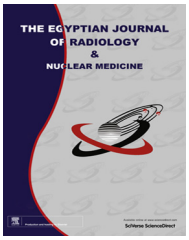

CASE REPORT

Spinal Rosai–Dorfman disease: Case report of a rare disorder

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KEYWORDS

Rosai–Dorfman;
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Abstract *Background:* Rosai–Dorfman disease (sinus histiocytosis with massive lymphadenopathy (SHML)) is a rare, histiocytic, lymphoproliferative disease of unknown etiology affecting young people with male predominance. It is characterized by massive, painless cervical lymphadenopathy with fever and malaise and varying extra-nodal involvement. Isolated spinal canal Rosai–Dorfman disease is extremely rare. We describe a case of isolated Rosai–Dorfman disease with both intradural extramedullary and epidural components.

Clinical presentation: 52 year man presented with 2 week history of progressive lower limb weakness and 2 days of urinary and fecal incontinence. He showed bilateral lower limbs weakness with normal muscle tone, exaggerated deep tendon reflexes and no sensory loss. CT and MRI showed large enhancing soft tissue mass lesion with both epidural and intradural extramedullary components opposite to T2–T5 vertebrae causing spinal cord compression associated with marrow changes of the body of T4 vertebra.

Intervention: A T3–T5 laminectomy and excision of the epidural lesion was performed. We opened the dura and found a large extramedullary well circumscribed mass engulfing the cord. Careful dissection and total resection of the intradural mass was done. The mass was histopathologically proved to be sinus histiocytosis conforming to Rosai–Dorfman disease. Postoperatively the patient showed improvement in the motor power and regained control over urine and stool.

Conclusion: This is a rare case of spinal Rosai–Dorfman Disease with epidural and intradural components causing cord compression. To our knowledge, this represents the first case of combined epidural and intradural extramedullary lesions in the literature.

1. Introduction

Rosai–Dorfman Disease (sinus histiocytosis with massive lymphadenopathy (SHML)) is a rare, histiocytic, lymphoproliferative disease of unknown etiology affecting young people with

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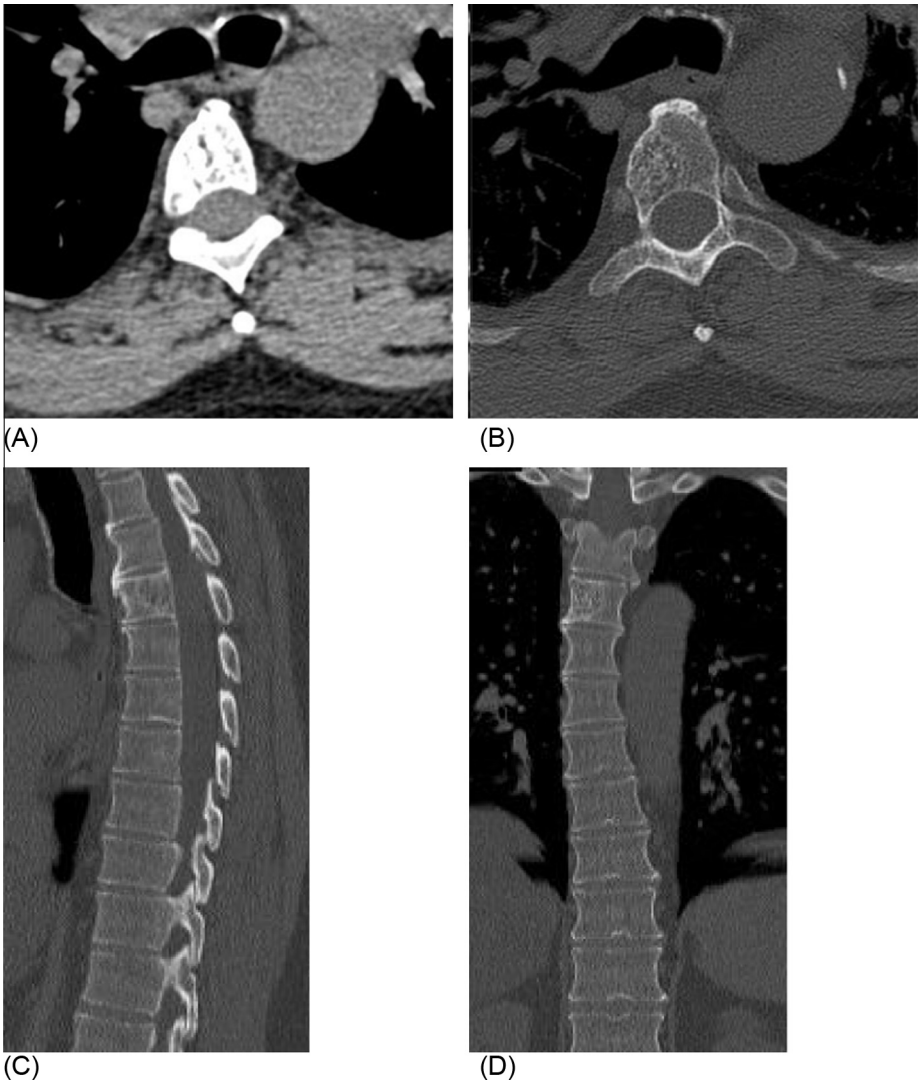

Fig. 1 Axial CT sections of the dorsal spine in (A) soft tissue window setting and (B) bone window setting with reconstructed CT images in (C) sagittal plane and (D) coronal plane showing focal scoliotic deformity with convexity to the right side and altered bony texture of the right aspect of D4 vertebra with no cortical interruption or structural collapse.

male predominance (1). The disease is characterized by massive, painless cervical lymphadenopathy with fever and malaise and varying extra nodal involvement. Isolated spinal cord Rosai–Dorfman Disease is extremely rare with cases described in the literature which are either epidural or intradural (2,3). We describe a case of isolated Rosai–Dorfman disease with intradural extramedullary and epidural components. To our knowledge, this represents the first case of combined epidural and intra-dural (extra-medullary) lesions in the literature.

2. Patient description

A smoker aged 52 year presented with 2 week history of progressive lower limb weakness more on the right side and 2 days of urinary and fecal incontinence. His past medical history is relevant for type 2 DM and schizophrenia since 30 years. Clinical examination showed bilateral Lower limbs weakness 3/5 with normal muscle tone, exaggerated DTR on the right side and sustained clonus in the Right ankle, negative Babinski and no sensory loss. Upper limbs examination was unremarkable.

3. CT findings

CT showed focal scoliotic deformity with convexity to the right side and altered bony texture of the right aspect of D4 vertebra with no cortical interruption or structural collapse (Fig. 1).

4. MR findings

MRI showed a large enhancing dural based soft tissue mass lesion with both epidural and intra-dural extra-medullary components opposite to T2–T5 vertebrae causing spinal cord

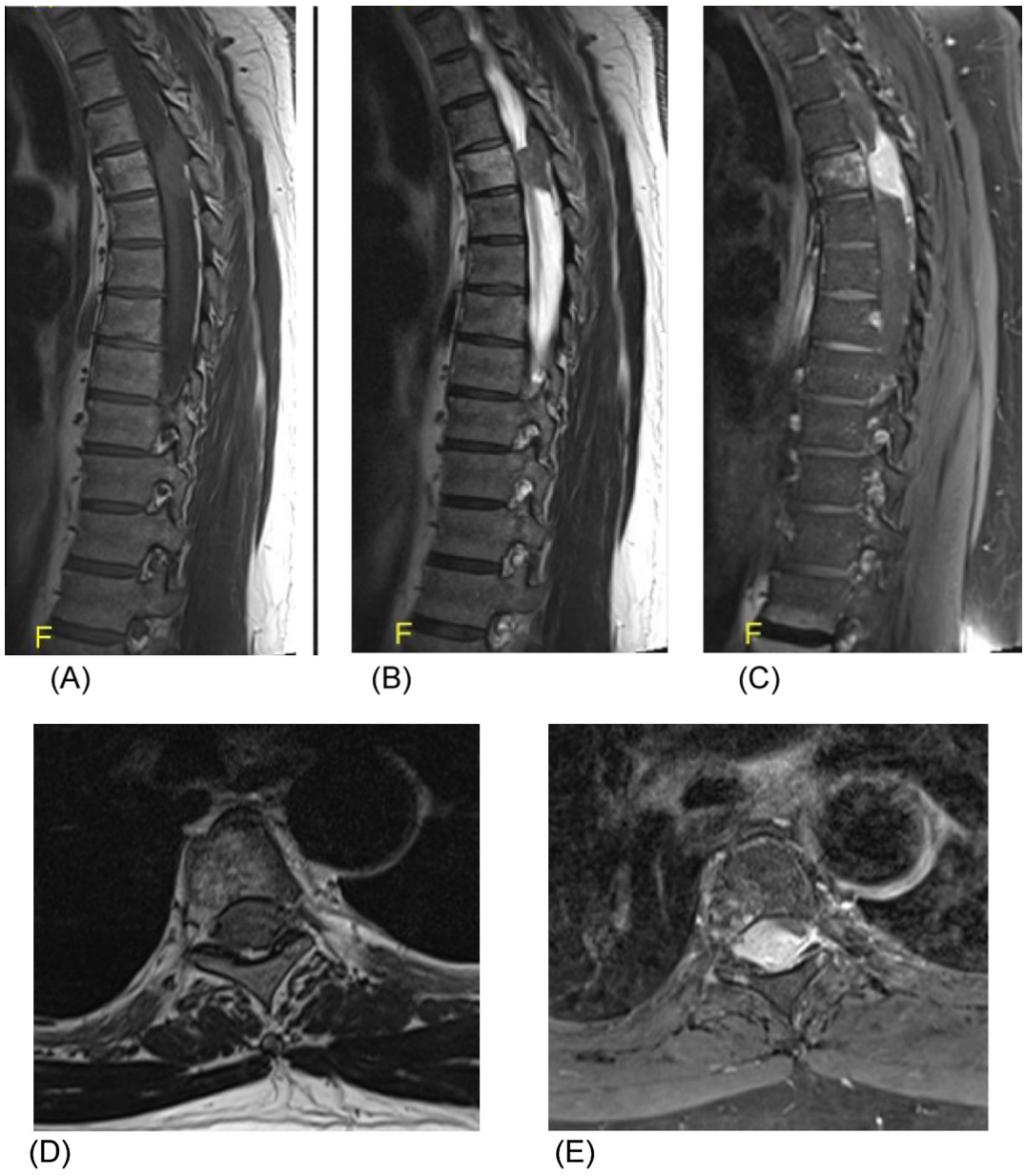

Fig. 2 Sagittal T1 (A), T2 (B) and contrast enhanced T1 (C) weighted images of the dorsal spine, Axial T2 (D) and contrast enhanced T1 (E) weighted images at D4 vertebral level showing an intra-spinal dural based mass lesion with both epidural and intradural components displaying intermediate to low signal intensity in T1 and T2 weighted images and showing intense homogenous enhancement in the post-contrast study extending from D2 to D5 levels and causing marked compression of the spinal cord at D4 level. Associated marrow signal alteration of the right aspect of D4 vertebra is also noted that showed heterogeneous enhancement in the post-contracts study.

compression associated with marrow changes of the body of T4 vertebra (Fig. 2).

5. Intervention

A T3–T5 laminectomy and excision of the epidural lesion was performed. We initially thought it was only epidural component but the Dura appeared bulging with abnormal tissue underneath. We opened the dura and found a large extramedullary well circumscribed mass engulfing the cord more on the right side. The mass was adherent in some areas to underlying arachnoid, and careful dissection and total resection of the intradural mass was done.

6. Histopathology

Two epidural specimens were excised measuring 3×2.5 and 2×1 cm, consisting of multiple soft to firm gray brown fragments in aggregate. One intra-dural specimen was excised and consists of 4 firm to hard gray white fragments in aggregate measuring $3 \times 1.6 \times 0.8$ cm, the largest measure $2 \times 1 \times 1$ cm.

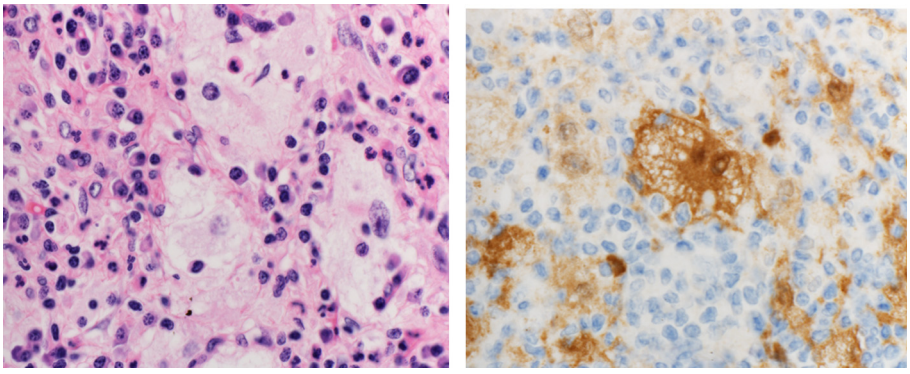

Fig. 3 H&E stain (A) showed aggregate of large histiocytes with plasma cells, lymphocytes and rare neutrophils and the histiocytes strongly expressed the S-100 protein (B).

H&E stain showed Aggregate of large histiocytes with plasma cells, lymphocytes and rare neutrophils. The histiocytes have large round/oval nuclei and abundant pale cytoplasm with some displaying emperipolesis (Lymphocytophagocytosis) with multiple lymphocytes engulfed within histiocyte cells. In addition, the histiocytes strongly expressed the S-100 protein which is not found in the ordinary granulomatous disease (Fig. 3).

7. Response to treatment

The patient tolerated the surgery quite well showed early improvement in the motor power of his lower limbs and regained control over urine and stool.

8. Summary

This is a rare case of spinal Rosai–Dorfman Disease with epidural and intradural components causing cord compression. Neurosurgeons should include it in the differential diagnosis of dural based lesions that resemble meningioma. Management depends on each case and includes observation, corticosteroids, radiotherapy, chemotherapy and surgery; for our case Surgery was the treatment of choice to relieve the cord compression. To our knowledge, this represents the first case of combined epidural and intra-dural extra-medullary lesions in the literature.

Conflict of interest

The authors declare that there are no conflict of interests.

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