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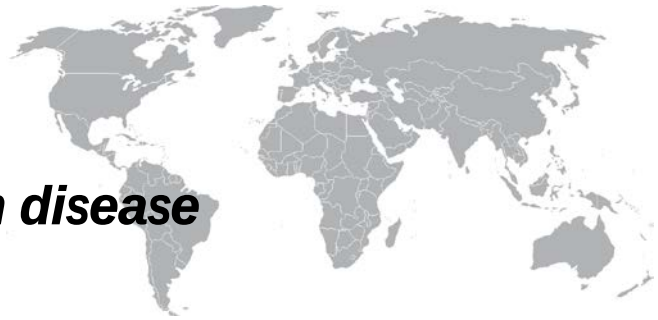
*Case Report*



# *Spinal extradural Rosai Dorfman disease*

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***ABSTRACT***
Rosai Dorfman disease is a rare disorder of histiocytes, which present as massive cervical lymphadenopathy. Here we describe a case of extranodal Rosai Dorfman disease involving the spinal cord. The patient presented with subacute progressive spastic paraparesis and numbness of both lower limbs. MRI of the spine showed an extradural mass extending from C3 to L1 level causing compressive myelopathy. Biopsy of the lesion was done through a D1 D2 laminectomy and the histopathology was consistent with Rosai Dorfman disease. We treated our case with one month steroids which resulted in clinical improvement and radiological resolution of the lesion.

**Keywords:** histiocytosis, sinus, magnetic resonance imaging, epidural neoplasms, spinal cord compression

## INTRODUCTION

Rosai Dorfman disease also known as sinus histiocytosis with massive lymphadenopathy is a rare, benign, disease of young adults characterised by painless massive cervical lymphadenopathy, fever, elevated ESR and polyclonal hypergammaglobulinemia (1). It is characterised by neoplastic proliferation of histiocytes within the sinuses of lymph nodes. Isolated involvement of extranodal sites occurs in 23% cases (2). Here we describe a case of spinal extradural Rosai Dorfman Disease which responded to steroid treatment.

## CASE

A 35y old male presented with progressive weakness, stiffness and numbness of both lower limbs of two months duration. He had no symptoms pertaining to upper limb, bladder or cranial nerves. There were no systemic symptoms. Examination revealed a moderately built and nourished person. There was no pallor, icterus or lymphadenopathy. Temperature was 36.8ºC, PR-76/min and BP-130/80mmHg. Neurological examination showed normal higher functions, cranial nerves and upper limb. Lower limbs were spastic with exaggerated knee and ankle jerks. Power was grade II in both lower limbs. Abdominal reflex was absent in all quadrants and plantar response were extensor bilaterally. All modalities of sensation were impaired below D6. There was no spinal tenderness or signs of meningeal irritation. Examination of respiratory system, cardiovascular system and gastrointestinal system were also normal.

Routine blood investigations showed a Hb – 14g/dl, TC – 9,800/mm$^3$, DC-$N_{60}L_{25}B_1E_5M_9$, ESR – 20mm/hr, Platelet-3.5 lakh/mm$^3$, Blood Urea – 35mg/dl, S.Creatinine – 1.2mg/dl, Random Blood Sugar – 180mg/dl. Liver function tests were within normal limits. USG abdomen and Chest X ray were normal. MRI of the spine showed a large diffusely enhancing soft tissue mass lesion in the posterior and lateral epidural space in the cervical and dorsal spinal canal extending from C3 to L1 level with spinal cord compression. (Figure 1, 2, 3) Biopsy was taken from the lesion through a D1 D2 laminectomy which showed a firm, soft lesion with low vascularity extending along the posterior aspect of spinal canal adherent to the dura. Histopathology showed sheets of histiocytes admixed with lymphocytes, numerous plasma cells, neutrophils and proliferating blood vessels with many histiocytes showing emperipolesis. Immunohistochemistry was positive for both S-100 and CD 68 and negative for CD1a, consistent with Rosai Dorfman disease. He was treated with a course of steroid. He made remarkable improvement and the power of both lower limbs became Grade IV. Repeat MRI of the spine showed good resolution of the lesion.

## DISCUSSION

Rosai Dorfman disease (RDD) was first reported in 1965 (3), but it was described as a distinct clinicopathological entity from histiocytosis X in 1969 by Rosai and Dorfman in a case series were they called it “sinus histiocytosis with massive lymphadenopathy” (4). Histologically the involved lymph nodes show dilated sinuses infiltrated with large histiocytes, lymphocytes and plasma cells. The presence of emperipolesis or engulfment of lymphocytes by histiocytes is characteristic of RDD. Immunohistochemically RDD cells are positive for S-100, CD68 and CD163 while CD1a is typically negative.

Even though the classical presentation of RDD is massive cervical lymphadenopathy with systemic symptoms, isolated extranodal involvement can occur in 23% of cases. The most common extranodal sites involved are skin, upper respiratory tract and bone (5). Other less common sites include genitourinary system, gastrointestinal tract, orbit, endocrine gland (particularly thyroid), breast, meninges and spinal cord (6-8). RDD is commonly accompanied by fever, elevated ESR and polyclonal hypergammaglobulinemia.

Neurological manifestations have been reported in about 4% of cases (9). The most common is brain lesions presenting sub acutely with headache, seizure or focal deficits due to mass effect. The typical lesion appears as a dural based, extraaxial lesion with contrast enhancement on MRI, mimicking a meningioma (10). Spinal cord involvement is rare. Most of them present as focal extradural mass even though intradural and intramedullary involvement has also been reported (11,12). Longitudinally extensive spinal lesions are extremely rare. Our case presented as an extradural lesion extending from C3 to L1. Such extensive involvement of the spinal cord is uncommon with only one case reported previously (13).

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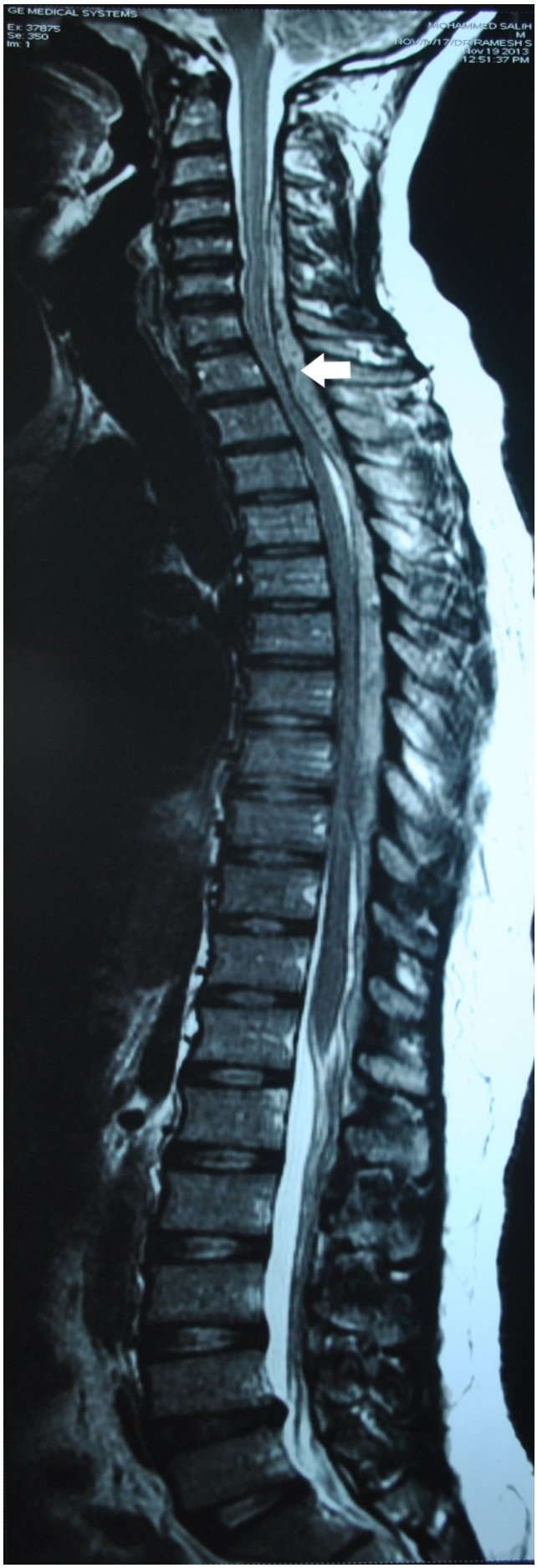


**Figure 1:** *T2 weighted sagittal MRI showing extradural lesion extending from C3 to L1 level*

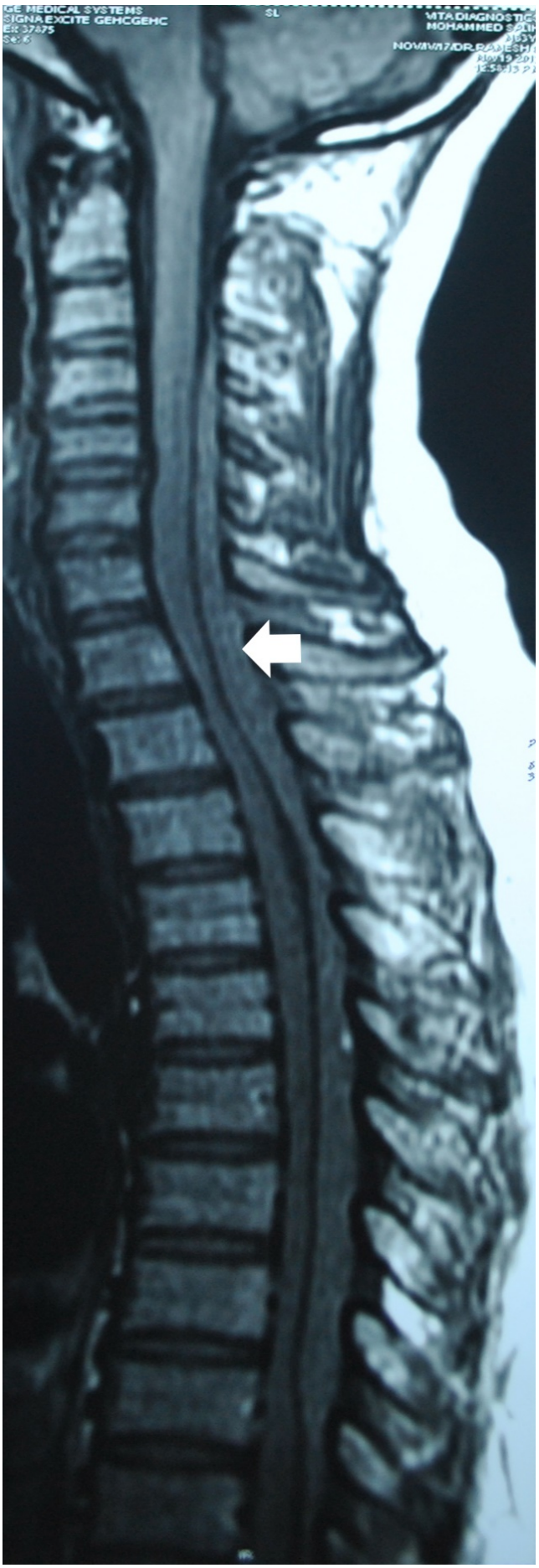


**Figure 2:** *T1 weighted sagittal MRI showing the lesion causing compressive myelopathy*

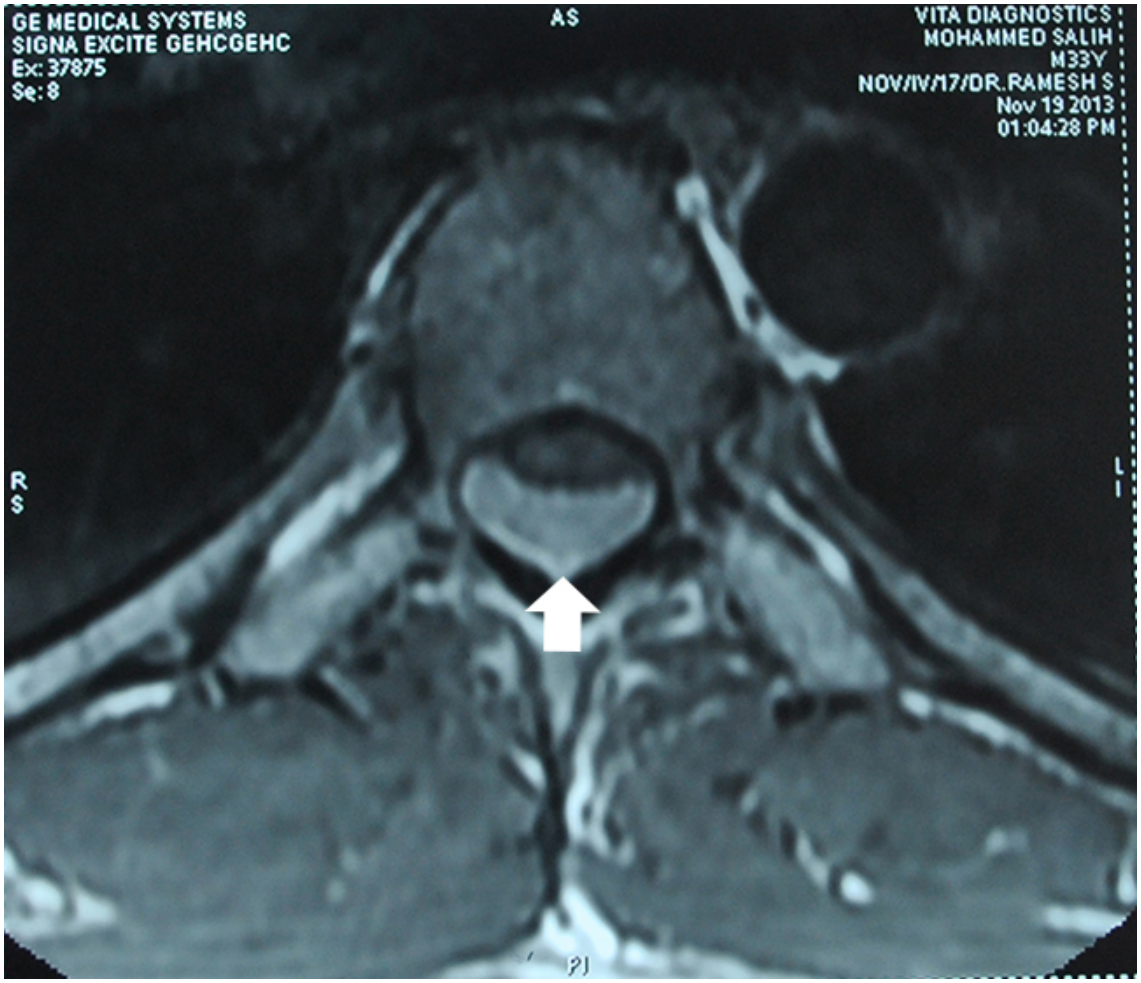


**Figure 3:** *T1 weighted axial MRI of the same lesion*

Focal lesions in brain and spinal cord are usually treated surgically. Other modalities of treatment are radiotherapy and steroids. Since the lesion was extensive, we treated the case with steroids, which resulted in clinical improvement with radiological resolution of the spinal cord lesion. Chemotherapy has no role in the treatment of RDD (14).

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