Spinal Rosai-Dorfman Disease: A Rare Case Report

1S Syed Ali, 2Shailendra Markad Uttamrao, 3Balamurugan Mangaleswaran

ABSTRACT
A 28-year-old male presented with recurrent spinal space occupying lesion who was treated with antitubercular medications in suspicion of granulomatous lesion before. He underwent surgical excision and histopathology with immunohistochemistry was confirmative of extranodal-spinal Rosai-Dorfman disease.

Keywords: Spinal Rosai-Dorfman disease, Extranodal histiocytosis, Lymphoproliferative disease.


Source of support: Nil

Conflict of interest: None

INTRODUCTION
Rosai-Dorfman disease (RDD) or sinus histiocytosis with massive lymphadenopathy, is a rare, non-neoplastic lymphoproliferative disease exhibiting typical histopathological features, but has an unknown origin and pathogenesis.5

CASE REPORT
A 28-year-old male with no comorbidities presented with history of low backache with bilateral sciatica for 3 months. Magnetic resonance imaging (MRI) lumbosacral spine with contrast showed contrast enhancing L3-4 to L5 extradural space occupying lesion for which he underwent L4 and L5 laminectomy and near total excision elsewhere 6 months back. Histopathology was reported as lymphocytosis with plasmacytes, no evidence of malignancy. He was treated with antitubercular medications. His backache and leg pain improved for few days. Later, he again started having similar illness with numbness of both lower limbs, which was gradually increasing in intensity. There was no history of weakness of limbs, bowel/bladder disturbances. He did not have any constitutional symptoms of infection. On examination, there were no neurological deficits. He was evaluated with MRI lumbosacral spine with contrast and screening of whole spine. It showed T1 isointense and T2 hyperintense, homogenously enhancing extradural lesion from L3 to L4 and L4-5 to S4 levels causing secondary canal stenosis (Figs 1A to E). He underwent re-exploration, L3 and S1 laminectomy and near total excision of tumor. After surgery, his pain subsided. Histopathology showed multiple histiocytes with mixed inflammatory cells composed of lymphocytes and plasma cells. Immunohistochemistry showed CD-68, S-100 and LCA positivity which was suggestive of RDD (extranodal sinus histiocytosis) (Figs 2A to E). He underwent postoperative radiotherapy. There was no pain/recurrence at 6 months follow-up.

DISCUSSION
Rosai-Dorfman disease is a rare lymphoproliferative disease with variable manifestations. In the classic form, the patient presents with a massive cervical lymphadenopathy and fever. Neurological symptoms rarely occur1 and are usually the result of cerebral2 or spinal4 involvement. Other manifestations also include nasal obstruction, tonsillar enlargement or hearing abnormalities. This also often accompanies with leukocytosis, an elevated erythrocyte sedimentation rate, weight loss and hypergamma-globulinemia.

In literature, only six cases have been reported so far (Table 1).

EXTRANODAL SITES
Most commonly the skin, upper respiratory tract, orbit, and testes.2 Spine involvement is rare which accounts for less than 5%.

PATHOGENESIS AND IMAGING
Rosai-Dorfman disease may represent either an autoimmune disease or a reaction to some infectious agent, such as a virus. On MRI, most of the cases of spinal RDD present with dura-based hypointensity or isointensity on T1- and T2-weighted images, with marked homogeneous enhancement after gadolinium administration. Clinically and radiologically, this disease is often believed to be a meningioma,6 but meningiomas in the spine usually present as a solitary mass.

PATHOLOGY
The definitive diagnosis of RDD relies on pathological examination, and the immunohistochemical characteristics of the lesions contribute to determining the final diagnosis.
Spinal Rosai-Dorfman Disease: A Rare Case Report

Table 1: Literature review of spinal Rosai-Dorfman disease

<table>
<thead>
<tr>
<th>Year</th>
<th>Journal</th>
<th>Author</th>
<th>Place</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>1985</td>
<td>J Bone Joint Surg Am Dec 1,67(9):1427-1431</td>
<td>Chan KW et al</td>
<td>Hong Kong</td>
<td>C5-6</td>
</tr>
</tbody>
</table>

The characteristic histopathological pattern of RDD is emperipolesis, a process of cellular ingestion by the histiocytes. Large pale histiocytes of RDD are immunoreactive for S100 protein and CD68, but negative for CD1a, a feature that helps to distinguish this tumor from Langerhans cell histiocytosis (which is
positive for CD1a). The discovery of classic Reed-Sternberg cells and a lack of histiocytes and emperipolesis help to distinguish Hodgkin disease from RDD.

Natural history of RDD is relatively benign and self-contained, but cases of rapid progression with multisystem involvement resulting in death have been reported.¹

**TREATMENT**

Surgical removal is generally regarded as the treatment of choice. Adjuvant therapies, including radiotherapy and small-dose chemotherapy with corticosteroids, cyclophosphamide, and vincristine have been administered with variable success.⁷

Although uncommon, vertebral canal involvement of RDD should be considered in the differential diagnosis of vertebral canal tumors. Resection of the lesions is an acceptable treatment of choice.

**REFERENCES**