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Case Report

Spinal Rosai Dorfman disease: A rare case scenario

Roshni P. S[1*], Sudha Haridas[1], Chothisha Poovan[1]

[1]Dept. of Pathology, Meitra Hospital, Calicut, Kerala, India

Abstract

Rosai Dorfman disease (RDD) is a histiocytic disease, which usually involves the cervical lymph nodes in majority of the cases, though it can occur less commonly in extra-nodal sites like skin, orbit, upper respiratory tract and bones. As far as bone involvement is concerned, it usually involves the long bones and contribute to around 10% of cases. Rosai Dorfman disease presenting with primary involvement of spine is a very rare scenario, with less than 1% of cases according to literature. Here is the case of a 45-year-old female who presented with complaints of mid back pain. Radiological examination showed well defined expansile lytic lesion involving the right transverse process, lamina and pedicle of T7 vertebra causing mild narrowing of T7-8 neural foramina. Excision biopsy from spinal lesion showed features of Rosai Dorfman disease and this was confirmed on immunohistochemistry.

Keywords: Rosai Dorfman disease, Histiocytes, T7 vertebra.

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1. Introduction

Rosai Dorfman disease (RDD) was initially reported in 1965[1] and later in 1969, Rosai and Dorfman recognized it as a specific entity.[1] RDD commonly presents as a nodal disease. It can have extra-nodal presentations as well and the sites of involvement include skin, orbit, upper respiratory tract and bones.[3] Bone involvement is seen in less than 10% of patients.[4-6] Involvement of spine is very rare and is seen in only less than 1% of cases.[7]

2. Case Presentation

A 45-year-old female presented with complaints of mid back pain since 2.5 months. Pain was radiating to the right anterior chest along the 7th rib. There was no history of trauma or fever. Patient had no neurological deficits or night pain. General examination and systemic examination was within normal limits. Musculoskeletal examination showed normal gait and there were no tenderness points in the spine. Routine laboratory investigations were within normal limits. Computed tomography (CT) of Thorax revealed a well-defined expansile lytic lesion involving the right transverse process, lamina and pedicle of T7 vertebra causing mild narrowing of T7-8 neural foramina. Radiological differential diagnosis included plasmacytoma, metastasis and lymphoma. Excision of T7 lesion with T5-9 posterior instrumented stabilization was done and the specimen was sent for histopathological evaluation. Gross examination showed multiple bits of bony tissue, which aggregate measured 3.0x 3.0x 2.0 cm. Microscopy of hematoxylin and eosin(H&E) stained sections showed an infiltrating lesion within the marrow cavity, composed of sheets of histiocytes with abundant pale eosinophilic cytoplasm and round nucleus with prominent nucleoli (**Figure 1**). Histiocytes showed emperipolesis of lymphocytes, plasma cells and neutrophils (**Figure 2**). Background showed prominent mixed inflammatory infiltrate composed of lymphocytes, plasma cells and neutrophils. Focal areas showed neutrophilic microabscess. Morphology favored Rosai Dorfman disease and immunohistochemistry (IHC) was done for confirmation. Histiocytes were positive for s100 and CD 68 (**Figure 3 and Figure 4**). Histiocytes were negative for CD1a and Langerin (**Figure 5 and Figure 6**).

*Corresponding author: Roshni P. S
Email: roshni.ps@meitra.com

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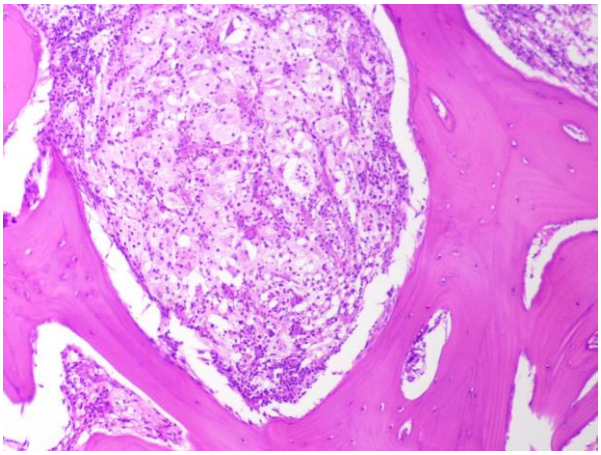

Figure 1: H&E - Intertrabecular spaces showing sheets of histiocytes, surrounded by mixed inflammatory infiltrate

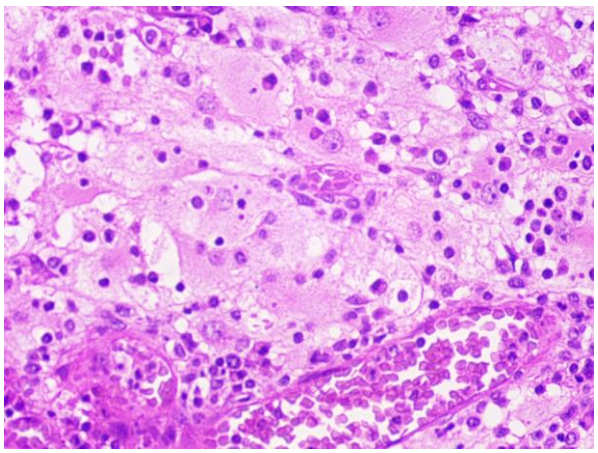

Figure 2: H&E- Histiocytes show emperipolesis and are surrounded by lymphoplasmacytic infiltrate

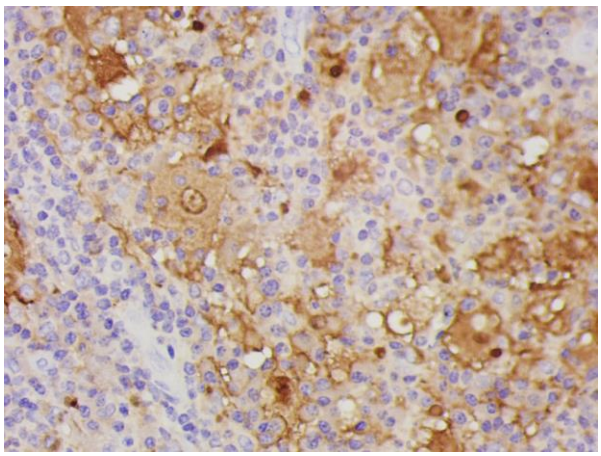

Figure 3: IHC-Histiocytes are positive for s100

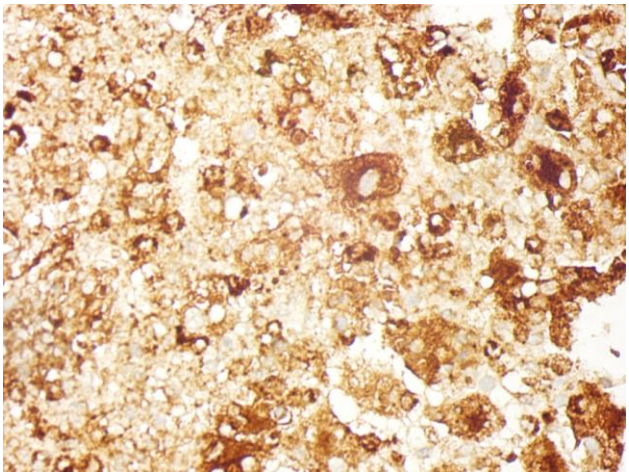

Figure 4: IHC-Histiocytes are positive for CD68

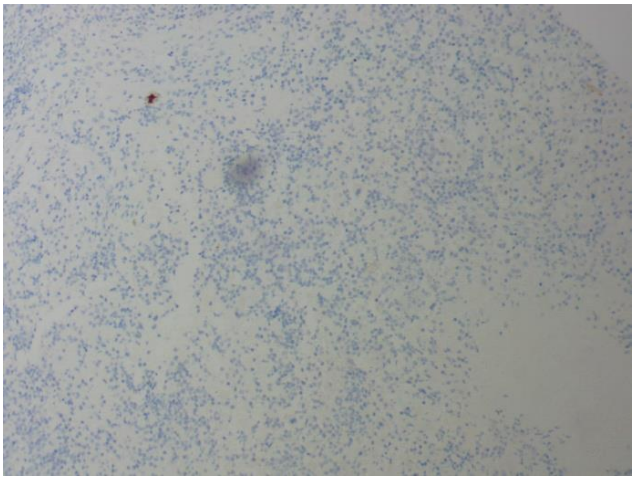

Figure 5: IHC-Histiocytes negative for CD1a

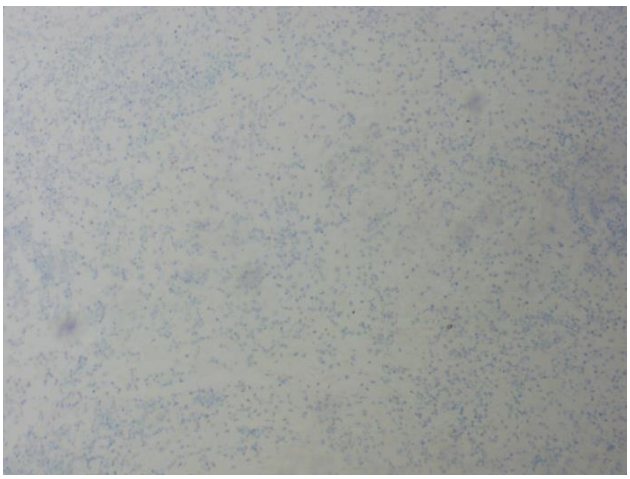

Figure 6: IHC-Histiocytes negative for Langerin

Later, PET CT (positron emission computed tomography)was done to assess the disease extent and it showed flurodeoxyglucose (FDG) avid lytic lesions in the manubrium sternum & bilateral iliac bones, intense FDG avid soft tissue lesion involving the posterior wall of right side of sphenoid sinus and faint FDG avid subcentimetric lymph nodes (likely infectious/ inflammatory pathology) in level II.

Patient was started on Lenalidomide - Dexamethasone therapy. Lenalidomide 10 mg was given once daily for 2 weeks, followed by rest for 1 week. Dexamethasone 20 mg was given once weekly for 4 weeks. Patient improved symptomatically with treatment.

3. Discussion

Rosai Dorfman disease (RDD) was first reported in 1965[1] and later in 1969, Rosai and Dorfman recognized it as a distinct disease entity. In a case series, they referred it as "sinus histiocytosis with massive lymphadenopathy".[2] RDD is a lympho-proliferative disorder of uncertain aetiology which generally presents as bilateral, massive, painless peripheral cervical lymphadenopathy. These patients often have associated constitutional features, such as fever, increased serum erythrocyte sedimentation rate, leucocytosis, polyclonal hyperglobulinemia and anaemia. Apart from nodal disease, RDD can present as extra-nodal disease in around 43% of cases. Sites of extra nodal involvement include skin, orbit, upper respiratory tract and bones.[3] Skeletal involvement is seen in around less than 10% of patients and generally involves long bones like tibia, femur, humerus, clavicle and bones of hand.[4-6] Only few cases of spinal RDD are reported and they contribute less than 1% of extra nodal disease.[7]

Clinically, most of the patients belong to the age group of 20-30 years of age.[8] Cervical and thoracic spine are the common sites of spinal Rosai Dorfman disease (RDD).[9] Lin etal has conducted a study on 51 patients with isolated spinal RDD and they observed that 72.5% of isolated spinal RDD were centered in thorasic spine.[10]

No conclusive or specific findings are obtained on radiological evaluation.[11] Histopathology remains the cornerstone in accurately making a diagnosis of RDD. The characteristic feature of RDD is the accumulation of histiocytes with enlarged, round to oval hypochromatic nuclei and abundant eosinophilic cytoplasm, often containing engulfed intact inflammatory cells known as emperipolesis. It is accompanied by marked lymphoplasmacytic inflammatory infiltrate in the background.[3] Occasional neutrophilic infiltrate also noted. On immunohistochemistry, the histiocytes are typically positive for S100 and negative for CD1a. Negativity for CD1a and Langerin rules out Langerhans cell histiocytosis, which is a close differential diagnosis.[4]

RDD is reported to show spontaneous resolution.[3] Pulsoni et al have observed 82% remission in their review of 40 cases.[12] Surgery is the preferred treatment modality in symptomatic lesions.[13] Other treatment options include steroids, radiotherapy, chemotherapy, supportive therapy, and α-interferons.[14] Number of nodal groups involved and the presence of extra-nodal involvement play an important role in prognostication. Nodal RDD tends to have better outcome when compared to extra-nodal disease.[4]

4. Conclusion

Rosai Dorfman disease presenting primarily as a spinal lytic lesion, without obvious nodal involvement is very rare. It has no definite clinical or radiological features. Diagnosis is typically made on histopathological evaluation, which shows the presence of histiocytic cells with emperipolesis and marked lymphoplasmacytic inflammatory cell infiltrate. On immunophenotyping, histiocytic cells show positivity for s100 and negativity for CD1a & Langerin. Surgical excision forms the mainstay of treatment in symptomatic lesions. So the take home message is that, RDD can present at uncommon sites including spine. It can clinically mimic other pathological conditions. However, possibility of RDD should be considered in the differential diagnosis of spinal pathologies, even in the absence of nodal involvement.

5. Source of Funding

None.

6. Conflict of Interest

None.

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