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ISOLATED CERVICAL INTRADURAL ROSAI-DORFMAN'S DISEASE – A VERY RARE ENTITY

Neurosurgery

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ABSTRACT

Rosai-Dorfman disease (RDD) is an idiopathic histiocytic proliferative disorder characterized by sinus histiocytosis and massive lymphadenopathy. Extranodal and central nervous system (CNS) involvement is unusual. We describe a rare case of isolated intradural RDD of the spine without cervical lymphadenopathy. RDD should be considered in the rare differential diagnosis of a spinal disease. Resection of the lesions is an effective treatment choice to relieve spinal compression symptoms. The efficacy of the adjuvant therapy to control the disease is unknown.

KEYWORDS

Rosai-Dorfman disease; Sinus histiocytosis with massive lymphadenopathy; Meningiomas; Spinal cord

INTRODUCTION

Rosai-Dorfman's disease (RDD) is a rare form of non-Langerhans cell histiocytosis described by Rosai and Dorfman in 1969 [1]. Clinically, it is an idiopathic histiocytic proliferative disorder characterized by sinus histiocytosis and massive lymphadenopathy. It is characterized histopathologically by the accumulation of CD68-positive, S100-positive, and CD1a-negative histiocytes [2]. It affects predominantly children and young adults with an average age of onset of 20.6 years and occurs more commonly in African patients with a slight male predominance (male to female ratio of 1.4). Inguinal, retroperitoneal, and mediastinal lymph nodes may also be involved.

Extranodal disease is seen in over 40% of cases and may rarely occur in the absence of nodal disease, usually in older patients with different demographics. Common extranodal sites of involvement include the skin (10%), nasal cavity (11%), bone (5%–10%), orbital tissue (11%), and central nervous system (5%, predominantly dural). In less than 5% of the cases, isolated involvement of the central nervous system is seen, with three-fourths in the brain and one-fourth in the spine [1,2]. Isolated involvement of the spine without the involvement of the lymph nodes is very rare with only a few cases reported in literature.

CASE REPORT

A 55-year-old gentleman presented to us with a history of mild neck pain and progressive spastic quadriparesis associated with numbness and tingling sensations on all 4 limbs for 3 years. There was no history of trauma, fever, bladder or bowel disturbances. On examination, the patient had asymmetric weakness in all 4 limbs (Right more than left) - MRC grade 3/5 power in all muscle groups of the right upper and lower limb and MRC grade 4/5 in the left. He had 50% reduced sensory perception to touch and pinprick below C4 dermatome as well as decreased proprioception and vibratory sensations in all 4 limbs with a spastic scissoring gait.

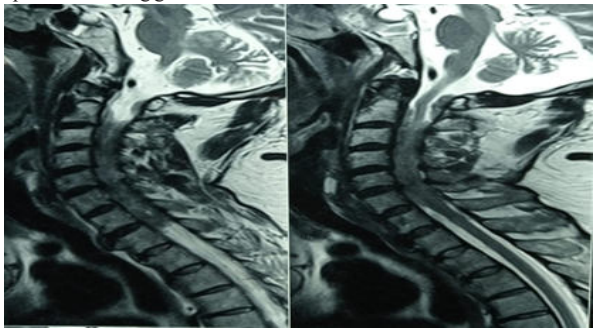

Figure 1 – Pre-operative MRI images showing T2 hypointense cervical intradural lesion with cord compression.

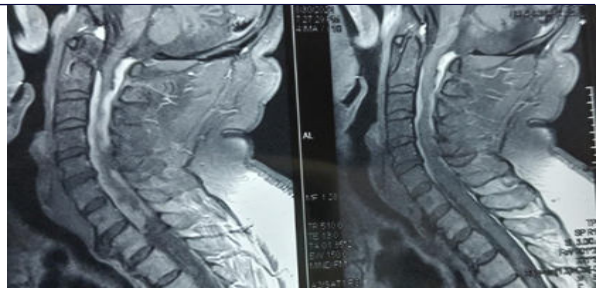

Figure 2 – Pre-operative MRI images showing T1 contrast enhancing cervical intradural lesion with cord compression.

Magnetic resonance imaging (MRI) of the spine showed an irregular C1 to C6 intradural extramedullary lesion which was isointense on T1 images and hypointense on T2 images causing compression of the cord with associated myelomalacia. The lesion was heterogeneously enhanced after gadolinium contrast administration (Figures 1 and 2). No other lesions were visualized on the full-body PET-CT scan including the brain. His blood investigations and chest X-ray were within normal limits.

He underwent C1 posterior arch excision, C2 to C6 laminectomy and sub-total excision of the lesion. Intraoperatively the lesion was pale yellow, was relatively avascular and was completely intradural with minimal adherence to the spinal cord (Figure 3). Considering the anterior extension of the lesion in a setting of an edematous high cervical cord, a sub-total excision was attempted.

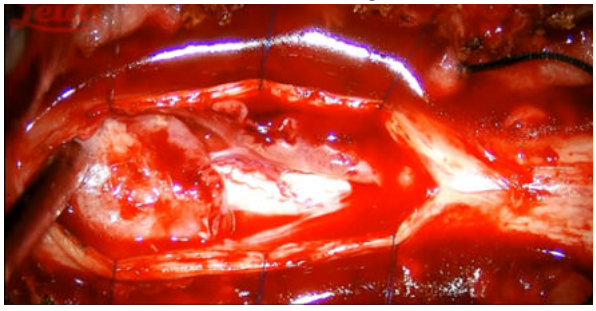

Figure 3 – Intra-operative image showing the lesion with cord compression.

Histopathologic examination of the lesion showed features of a histiocytic neoplasm. The tumor was composed of sheets of histiocytes having pale elongated nuclei, inconspicuous nucleoli, and moderate

pale eosinophilic cytoplasm. There were also dense lymphocytes and plasma cells with eosinophilic cytoplasm and perivascular lymphocytic inflammation. Other changes noted were emperipolesis with engulfed intact lymphocytes and plasma cells in the cytoplasm of histiocytes with inconspicuous mitosis. Stroma was densely collagenized and special stains for acid-fast bacilli and fungal elements were negative. Immunohistochemistry revealed the presence of histiocytes positive for S100 protein, CD68 and CD163 (Figure 4).

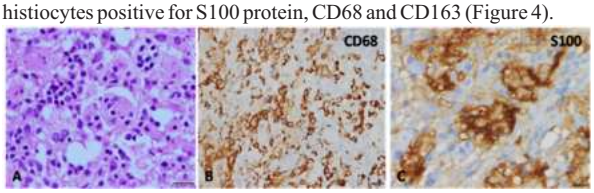

Figure 4 – Histopathological pictures showing histiocytes show emperipolesis of intact lymphocytes and plasma cells (A) Histiocytes are positive for CD68 (B) and S100 (C).

Postoperatively, his power improved to MRC grade 4+/5 in all 4 limbs and he could walk without support. Post-operative MRI spine showed a few residual anterior lesions and cord with no compression (Figure 5). He was continued on oral steroid therapy for 8 weeks and then referred to a medical oncologist for further treatment. A 1-year follow-up MRI showed no increase in the size of residual lesions and clinically patient showed gross improvement in the sensory symptoms as well (Figure 6).

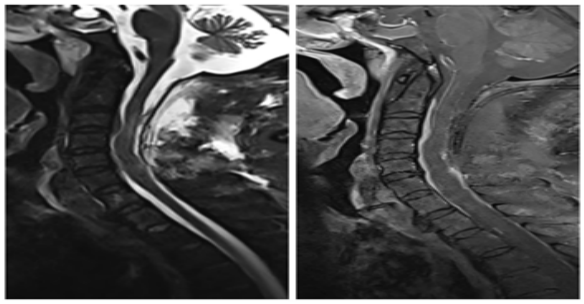

Figure 5 – Post-operative MRI images showing small anterior residual lesion with no cord compression.

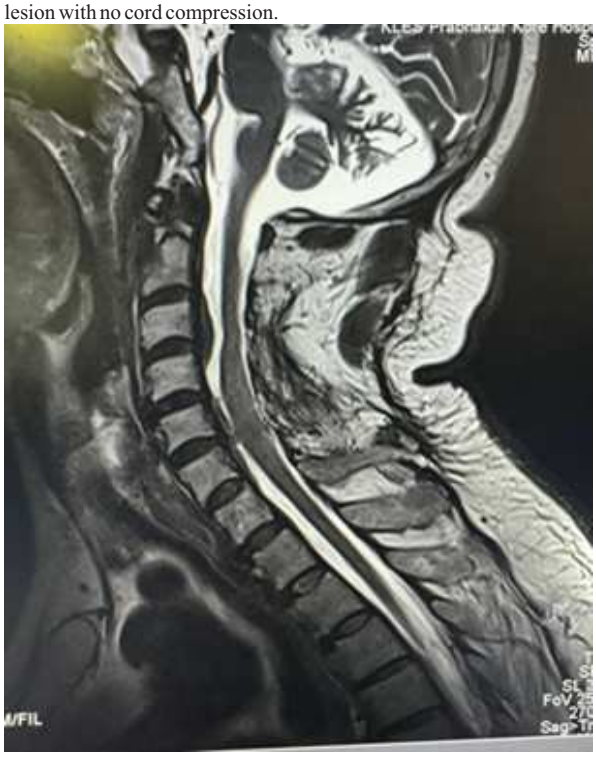

Figure 6 – MRI image after 1 year showing no growth in the residual lesion.

DISCUSSION

RDD is a rare histiocytic illness first characterized by Destombes in 1965 as large lymphadenopathy with sinus histiocytosis [3]. Rosai Dorfman's disease (RDD) was first reported as a distinct clinicopathological entity in 1969 by Rosai and Dorfman in a case series where 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.

RDD's incidence is linked to a clonal origin, even though its etiology remains unknown. Consequently, rather than being viewed as an inflammatory proliferative response, RDD should be treated as a malignancy [5]. The World Health Organization designated RDD as a hematological and lymphoid malignancy in 2016 [6]. Simultaneously, a new classification of histiocytosis was published, which classified several types of RDD, ranging from familial RDD to RDD linked to IgG4. Depending on the place of occurrence, RDD can be further classified into lymph node (the most frequent form), extranodal, and mixed kinds [7]. In summary, more study is necessary to validate the ambiguous pathophysiology of RDD.

Even though the classical presentation of RDD is massive cervical or generalized 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 [8]. Other less common sites include the genitourinary system, gastrointestinal tract, orbit, endocrine gland (particularly thyroid), breast, meninges, and spinal cord [9,10]. RDD is commonly accompanied by fever, elevated ESR, and polyclonal hypergammaglobulinemia.

Isolated RDD of the nervous system has also been reported, and most cases involve the brain [9]. Spinal cord involvement such as intramedullary, intradural, and extradural is very rare in RDD [10]. In neuroimaging it is seen as a dural-based, contrast-enhancing lesion and thus, clinically and radiologically, the disease is thought to represent a meningioma.

The natural history of RDD has been reported to be benign with spontaneous remission. In case of persistent or progressive disease, several adjuvant therapies have been administered with variable success, including radiotherapy and chemotherapy with corticosteroids, 6-mercaptopurine, and methotrexate. Resection of the lesions is an effective treatment choice to relieve spinal compression symptoms [8-10]. There is no established systemic treatment protocol for RDD since it is a rare disease presenting with different clinical features.

CONCLUSION

We hereby present a very interesting and rare case of isolated cervical intradural RDD of the spine. Although rare it should be considered among the differential diagnosis of intradural lesions with cord compression. To date, excision of the lesion is the treatment of choice to relieve spinal compression symptoms. The diagnosis and treatment of CNS-RDD remains a huge challenge due to its rarity and absence of pathognomonic clinical and imaging features. Hence all diagnosticians and pathologists need to be aware of this entity for early and adequate diagnosis and treatment.

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Conflict Of Interest: None

REFERENCES

1. Foucar E, Rosai J, Dorfman R. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): Review of the entity. Semin Diagn Pathol 1990;7:19-73.
2. Duarte-Williamson E, Antony F, Rotarescu R. Sinus histiocytosis (Rosai-Dorfman disease) presenting with solitary cutaneous nodule: a very rare clinical entity. Dermatol

Pract Concept. 2012;2(2):202a04.
3. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy A newly recognized benign clinicopathological entity. Arch Pathol. 1969;87:63–70.
4. Warpe BM, More S V. Rosai-Dorfman disease: A rare clinico-pathological presentation. Australas Med J. 2014;7(2):68–72.
5. Diamond EL, Durham BH, Haroche J, et al.. Diverse and targetable kinase alterations drive histiocytic neoplasms. Cancer Discov. 2016;6:154–65.
6. Swerdlow SH, Campo E, Pileri SA, et al.. The 2016 revision of the World Health Organization classification of lymphoid neoplasms. Blood. 2016;127:2375–90.
7. Abla O, Jacobsen E, Picarsic J, et al.. Consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-Destombes disease. Blood. 2018;131:2877–90.
8. Gaitonde S. Multifocal, extranodal sinus histiocytosis with massive lymphadenopathy: an overview. Arch Pathol Lab Med. 2007;131(7):1117–21.
9. Duarte-Williamson E, Antony F, Rotarescu R. Sinus histiocytosis (Rosai-Dorfman disease) presenting with solitary cutaneous nodule: a very rare clinical entity. Dermatol Pract Concept. 2012;2(2):202a04.
10. Maiti TK, Gangadharan J, Mahadevan A, Arivazhagan A, Chandramouli BA, Shankar SK. Rosai-Dorfman disease presenting as cervical extradural lesion: a case report with review of literature. Neurol India. 2011;59(3):438–42.