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Case report on 10 year child with Rosai Dorfman disease

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Abstract
Background: RDD is a rare proliferative histiocytic disease with a benign course. In children it is a rare cause of rapidly progressive lymphadenopathy, which sometimes mimics malignancy.
Case presentation: This report describes a case of Rosai Dorfman disease of 10 year old female child. The clinical, radiological, histological evaluation and management are presented in case report.
Conclusion: The course of RDD is unpredictable, episodes of remission and exacerbation may occur for several years. There may be a spontaneous and permanent remission or suffer progressive and generalized disease.

Keywords: Rosai Dorfman disease, histiocytosis, child, multiple bony lesions

Introduction

RDD is a rare proliferative histiocytic disease with a benign course. The condition was first described in 1965 but only recognized as a clinical and pathological entity in 1969 through a publication by Rosai and Dorfman. Found worldwide and affecting individuals with an average age of 20.6 years, RDD is slightly more common among men (1.4:1) and is significantly more common among Caucasians and blacks than among Asians. In children it is a rare cause of rapidly progressive lymphadenopathy, which sometimes mimics malignancy.

The disease typically presents with extensive cervical lymphadenomegaly, most often bilateral and painless (87%). At first lymph nodes are mobile and discrete, but over time they become adherent and tend to develop into a large multinodular mass. The axillary (23.7%), inguinal (25.7%) and mediastinal (14.5%) regions may be affected, though not as severely as the cervical region. This case report discusses the presentation of Rosai–Dorfman disease in a 10 year old female patient, who presented with bilateral knee pain and subcentric sized lymph nodes in both jugular chain. The histopathology from femur epiphysis and tibia metaphysis demonstrated numerous histiocytic infiltrate with fibrosis with emperipolesis.

Case report

A 10 years old female child was admitted in our institution with complain of bilateral knee pain and difficulty while walking. Episodes of pain and remission occur since 6 months. Pain and stiffness and weakness gradually progressed with time and episodes of sudden fall down while walking occurred.

The patient has history of taking treatment for it at initial event but did not get relief. Patient had no History of trauma, fever, weight loss, loss of appetite

On physical examination both lower limb have no swelling, redness, crepitus, abnormal movement no history of local rise of temperature seen. All deep reflexes are normal no complain of numbness, tingling, or decreased sensation over both lower limbs.

On plain radiograph of right knee joint with tibia and fibula anteroposterior and lateral view:

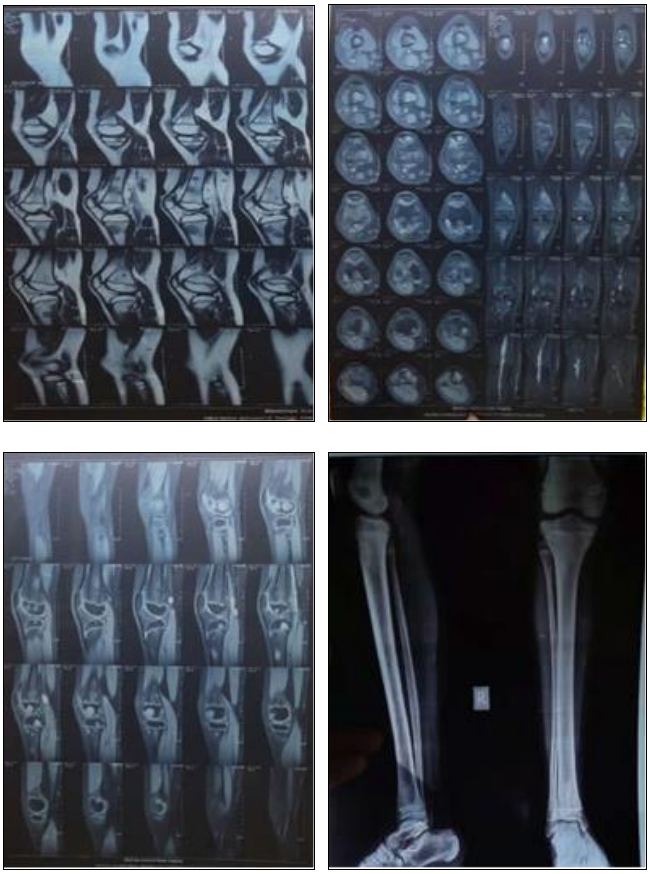
1. Both tibia and fibula under vision appear normal
2. No e/o lytic or sclerotic lesion
3. No e/o fracture or dislocation

On USG neck-multiple subcentimetric sized lymph nodes are noted in both jugular chain. Major vessels of neck appear normal, both lobes and isthmus of thyroid gland appear normal in size shape and echotexture.

On USG hip and knee-few varying size lymph nodes are noted, some of them are rounded hypoechoic, largest measures approx 10×6 mm in left popliteal fossa.

MRI of both knee with screening PBH and whole spine

- Multiple well defined altered signal intensity lesion Are noted involving bilateral femur, tibia, patella, fibula
- screening of PBH shows 2 similar intensity well defined Lesions in left sacral ala and iliac bone
- Few rounded lymph nodes are noted in popliteal and posterior Lower thigh on either side. The largest in right popliteal region Measures 12×11mm.largest in left popliteal region 18×15mm



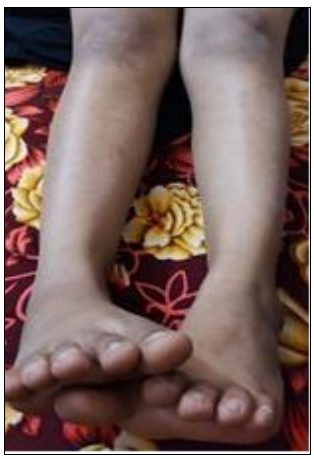


Fig 1: Histological evaluation

Histological evaluation

Specimens taken from left inguinal lymph node, right upper tibia and distal femur, slide and block of bone of knee region, bone/bone marrow biopsy.

Macroscopic description

1) Elongated bony tissue measuring 1 cm in height
2) Elongated bony tissue measuring 1.9 cm in height
3) Few tiny greyish soft tissue measuring 0.1×0.1 cm

Microscopic description

Histological examination shows bone with intervening marrow and foci with dense infiltrate of large.
Histiocytes with emperipolesis. There are admixed plasma cells with few eosinophils. no RS cells,

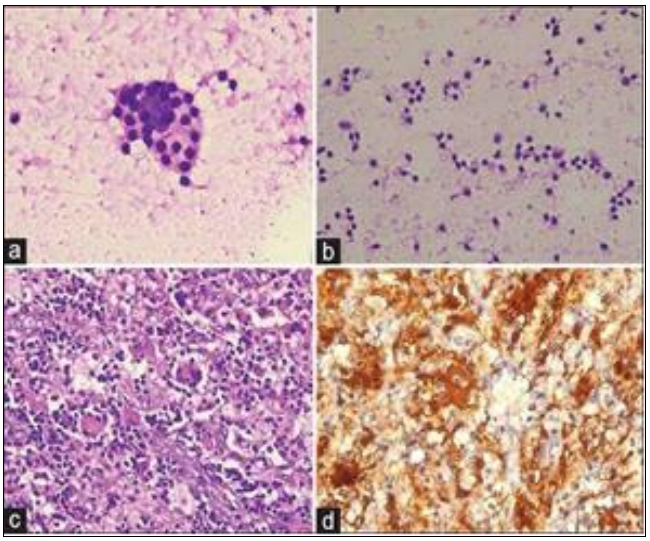


Fig 2: Granuloma or langerhans cells

IHC
PAX-5 – some cells positive CD79a, CD20-many cells positive
Synaptophysin, CD34, NKX2.2, CD1a- negative CD3-moderately positive
Tdt-few cells positive Ki67-low
Histiocytes positive for PGM-1 and s-100 & negative for CD1a

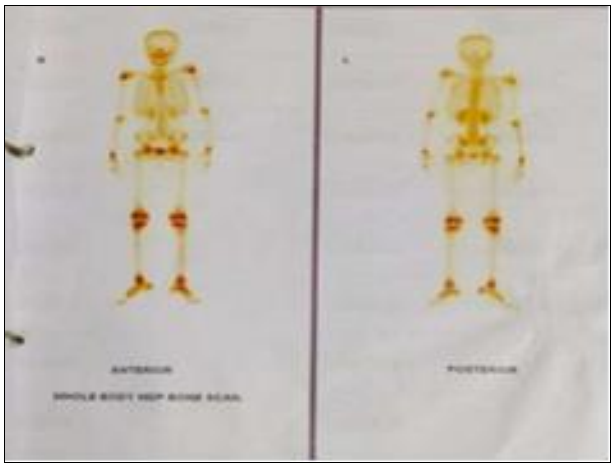


Fig 3: Bone scintigraphy- reveals no evidence of osteoblastic skeletal metastasis

Diagnosis and treatment

Biopsy from bone around knee with histiocytic lesion with emperipolesis Suggestive of Rosai dorfman disease.
Dexamethasone given for 15 days with tapering and started prednisolone10mg for 10 days

Discussion

RDD is a rare proliferative histiocytic disease with a benign course. The condition was first described in 1965 but only recognized as a clinical and pathological entity in 1969 through a publication by Rosai and Dorfman [1]. Found worldwide and affecting individuals with an average age of 20.6 years, RDD is slightly more common among men (1.4:1) and is significantly more common among Caucasians and blacks than among Asians [2]. In children it is a rare cause of rapidly progressive lymphadenopathy, which sometimes mimics malignancy [3].

The disease typically presents with extensive cervical lymphadenomegaly, most often bilateral and painless (87%). At first lymph nodes are mobile and discrete, but over time they become adherent and tend to develop into a large multinodular mass. The axillary (23.7%), inguinal (25.7%) and mediastinal (14.5%) regions may be affected, though not as severely as the cervical region [4]. Our patient presented painless unilateral cervical lymphadenomegaly with compromised lymph nodes in the mediastinum (pretracheal, prevascular, aortopulmonary window, right hilar and subcarinal), retroperitoneum and mesentery.

Extranodal tissue involvement is documented in 43% of RDD patients, especially of the skin, soft tissues, upper airway, bones, urogenital system, lower airway and oral cavity [4-7]. In addition, RDD may be associated with other conditions, including other forms of histiocytosis [8].

Up to 30% of RDD patients report fever [9], frequently associated with a high BSR and polyclonal hypergammaglobulinemia (up to 90% of cases), anemia and neutrophilic leukocytosis [5]. Our patient presented with fever (preceding two weeks), a high BSR, neutrophilic leukocytosis (28 x109/L) and anemia (Hb = 8.6 mg/dL).

The etiology, pathogenesis and natural history of RDD remain obscure. Some authors have suggested a role for human herpesvirus six (HHV-6) supported by reports of peculiar patterns of expression of HHV-6 antigens in abnormal histiocytes from RDD patients [10]. Other authors believe RDD may be the consequence of an exacerbated response of the immune system to infection by the Epstein-Barr virus, cytomegalovirus, *Brucella* or *Klebsiella* [11, 12].

The diagnosis of RDD is based on the clinical history and confirmed by histopathological examination. Specimens may be obtained by open surgical biopsy or fine needle aspiration. The latter is by many considered a sensitive and reliable diagnostic method and has the advantage of being possible in the outpatient setting [13]. Excised lymph nodes are often grayish with capsular fibrosis or pericapsular fibroadipose tissue. In general, the architecture of the lymph node is subverted and changes are observed in patients with long-standing lymphadenopathy. Usually the sinusoids of the lymph nodes are markedly expanded due to lymphatic stasis and display a mixed cell population, including lymphocytes, plasmocytes and histiocytes. The most characteristic cells in the sinuses are histiocytes of accentuated phagocytic appearance. These cells are large and irregular with abundant eosinophilic and sometimes vacuolated cytoplasm, usually displaying a round or oval nucleus with well-defined and delicate membranes and a single prominent nucleolus. Mitosis is rarely observed. However, histiocytes with foamy cytoplasm may be predominant in the cellular environment. The most important histological finding in RDD is histiocytes with a variable number of phagocytosed cells, usually lymphocytes, plasmocytes or erythrocytes.

Some cells, especially lymphocytes, remain viable inside the vacuoles, giving rise to a phenomenon known as lymphophagocytosis or emperipolesis, defined as the presence of intact lymphocytes inside other cells, in this case, histiocytes. The most useful marker of histiocytes in RDD is the expression of protein S100. Histiocytes may also express pan-macrophage antigens (CD68, HAM 56, CD14, CD64 and CD15), phagocytosis-related antigens (CD64 or the Fc receptor for IgG) and lysosomal activity (lysozyme and alpha-1-antitrypsin). In addition, histiocytes are negative for CD1a and contain no Birbeck granules [5].

The anatomopathological evaluation of our patient revealed, among other things, emperipolesis, positivity for PS100 and CD68 and negativity for CD1a. The differential diagnosis of RDD includes histiocytosis of Langerhans cells, histiocytic sarcoma, lysosomal storage diseases (such as Gaucher's disease), classic Hodgkin's lymphoma, melanoma and metastatic carcinomas and infections caused by Histoplasma and mycobacteria involving the lymph node [5].

Due to its low incidence, no standard treatment has yet been defined for RDD. However, since the condition is self-limiting, it is often unnecessary to intervene, except when the airways are obstructed or vital organs are compressed. Several forms of therapy have been described involving corticosteroids, chemotherapy combined with periwinkle alkaloids, anthracyclines, antimetabolics and alkylating agents, interferon, antibiotics, radiotherapy and partial or total surgical resection [12]. A review of the literature revealed that 50% of patients with RDD require no treatment and that 82% of untreated patients experience spontaneous and complete disease regression [12]. Some reports describe disease control in children without therapy [14, 15]. In this case, after careful analysis of the biopsied specimen, a conservative approach was adopted with progressive reduction of the lymphadenomegaly.

The course of RDD is unpredictable. Episodes of remission and exacerbation may occur for several years. In approximately 70% of cases the disease is permanent but stable, 20% experience spontaneous and permanent remission and 10% suffer from progressive and generalized disease.

References

1. Rosai J, Dorfman RF. Sinushistiocytosis with massive lymphadenopathy: a newy recognized benign clinicopathological entity. Arch Pathol 1969;87(1):63-70.
2. Brenn T, Calonje E, Granter SR, Leonard N, Grayson W, Fletcher CD *et al.* Cutaneous Rosai-Dorfman disease is a distinct clinical entity. Am J Dermatopathol 2002;24(5):385-91.
3. Duval M, Nguyen VH, Daniel SJ. Rosai-Dorfman disease: An uncommon cause of massive cervical adenopathy in a two-year-old female. Otolaryngol Head Neck Surg 2009;140(2):274-5.
4. Foucar E, Rosai J, Dorfman RF. Sinushistiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): review of entity. Semin Diagn Pathol 1990;7(1):19-73.
5. McClain KL, Natkunam Y, Swerdlow SH. Atypical cellular disorders. Hematology Am Soc Hematol Educ Program 2004, 283-66.

6. Landim FM, Rios HO, Costa CO, Feitosa RG, Rocha Filho FD, Costa AA.[Cutaneous Rosai-Dorfman disease]. An Bras Dermatol. Portuguese 2009;84(3):275-8.
7. Zhang JT, Tian HJ, Lang SY, Wang XQ. Primary intracerebral Rosai-Dorfman disease. J Clin Neurosci 2010;17:1286-88.
8. Sachdev R, Shyama J. Co-existent Langerhans cell histiocytosis and Rosai- Dorfman disease: a diagnostic rarity. Cytopathology 2008;19(1):55-8.
9. Bist SS, Varshney S, Bisht M, Pathak VP, Kusum A, Gupta N. RosaiDorfman syndrome - a rare clinical entity. Indian J Otolaryngol. Head Neck Surg 2007;59:184-6.
10. Luppi M, Barozzi P, Garber R, Maiorana A, Bonacorsi G, Artusi T et al. Expression of human herpesvirus-6 antigens in benign and malignant lymphoproliferative diseases. Am J Pathol 1998;153(3):815-23.
11. Lu D, Estalilla OC, Manning JT Jr, Medeiros LJ. Sinushistiocytosis with massive lymphadnopathy and malignant lymphoma involving the same lymph node: a report of four cases and review of the literature. Mod Pathol 2000;13(4):414-9
12. Pulsoni A, Anghel G, Falcucci P, Matera R, Pescarmona E, Ribersani M et al. Treatment of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman Disease): report of a case and literature review. Am J Hematol 2000;69(1):61-71.
13. Kumar B, Karki S, Paudyal P. Diagnosis of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman Disease) by fine needle aspiration cytology. Diagn. Cytopathol 2008;36(10):691-5.
14. Stones DK, Havenga C. Sinushistiocytosis with massive lymphadenopathy. Arch Dis Child 1992;67(4):521-3.
15. Dearth J, Hunter D, Kelly D, Crist W. Sinushistiocytosis with massive lymphadenopathy. CA Cancer J Clin 1980;30(1):55-8.