



Rosai Dorfman Disease - A Rare Presentation of Cervical Lymphadenopathy - A Case Report

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Article History	Abstract
Received: 06 June 2023 Revised: 05 Sept 2023 Accepted: 25 Nov 2023	<p><i>Rosai Dorfman disease is an uncommon histiocytic disorder presenting as bilateral cervical lymphadenopathy in children and young adults. Under the revised classification of histiocytic disorders, it is classified in the 'R' group of histiocytosis. Cutaneous Rosai -Dorfman disease is regarded as a separate entity which falls under the 'C' group of histiocytosis according to this classification system. We herein report a case of cervical lymphadenopathy who initially developed swelling in the right side of the neck and later developed in the left side of the neck. Patient underwent mantoux test which showed induration of 22mm which was significant. No other symptoms like cough with expectoration, loss of appetite was present. chest x ray was normal. Tuberculous Lymphadenitis was suspected. FNAC was done but confirmatory diagnosis was not obtained, so Excision biopsy was done. Finally, microscopic examination revealed marked emperipolesis with sheets of mature lymphocytes and plasma cells which is pathognomic cytoarchitecture of Rosai Dorfman disease. No evidence of granuloma/ RS cells/ atypia seen. Immunohistochemistry showed S -100 – strong positivity.</i></p>
CC License CC-BY-NC-SA 4.0	Keywords: Disease, Lymphadenopathy

1. Introduction

Rosai Dorfman disease, a rare benign histiocytic disorder characterised by massive, bilateral, and predominantly cervical lymphadenopathy. Although the clinical manifestations and histologic appearance are suggestive of an infectious process, no microorganism has yet been identified.

Case Details: A 65 yr old male, came with the complaints of swelling in the neck for 2mths, apparently normal 2mths back. Initially, he developed swelling in the right side of neck and later he developed on his left side of neck. Ultrasonogram of neck revealed multiple discrete lymph nodes noted in the cervical level 3 and 4 on both sides. largest measuring 2x1.5 cm in the C4 level on left side. clinically, there was a 2x2 cm swelling left side of neck, smooth surface, firm in consistency. Mantoux test, chest xray was done. Mantoux was positive. Patient had nil significant past /present history. FNAC of the swelling was done, suspected colloid cyst of thyroid. To obtain confirmatory diagnosis, Excision biopsy was done.

HISTOPATHOLOGY: GROSS: we received a single globular mass measuring 2.5x2x1.5cm with nodular surface. On cut surface: - capsule was thickened, firm, grey tan and grey white.

MICROSCOPIC EXAMINATION: Microscopy showed a thickened capsule (Fig .1) with mild architectural effacement. Several enlarged sinuses (Fig.2) showing large clusters of foamy macrophages with evidence of marked emperipolesis (Fig 3 & 4), paracortical effacement with sheets of mature lymphocytes, plasmacells with Russell bodies (Fig 5 & 6) seen. Also, evidence of fibroblastic proliferation (Fig .7) seen. No evidence of any granuloma /RS cells /atypia seen. IHCmarker S 100 showed strong positivity (Fig.8).

We arrived at the diagnosis of ROSAI DORFMAN DISEASE /SINUS HISTIOCYTOSIS WITH MASSIVE LYMPHADENOPATHY (SHML). Following excision, patient condition improved and later discharged and on follow up.

Fig. 1 Thickened Capsule

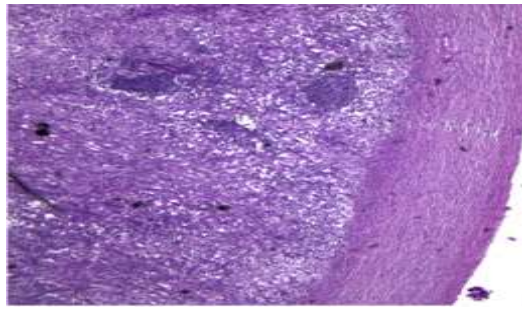


Fig. 2 Enlarged Sinuses

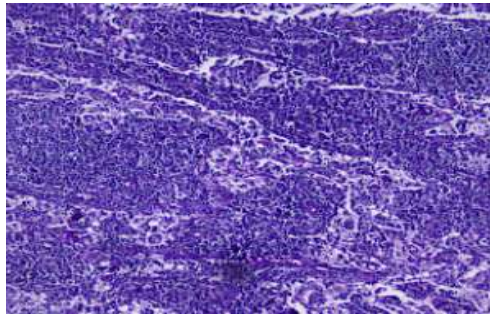


Fig. 3 Low Power -Emperipolesis

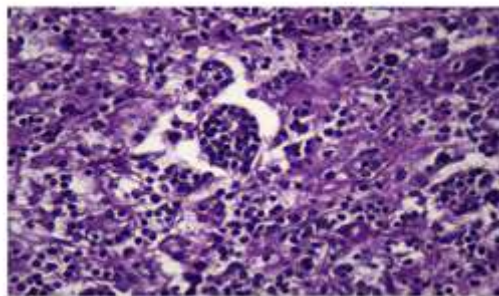


Fig. 4 High power - EMPERIPOLESIS

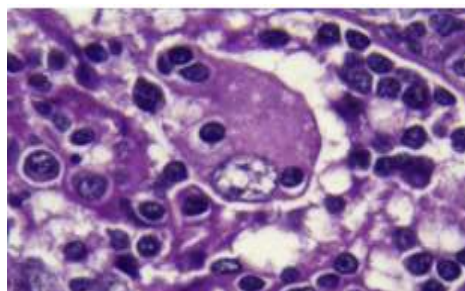


Fig.5 Plasma Cells

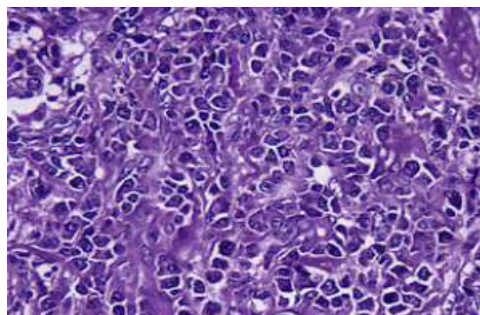


Fig 6. Russel Body

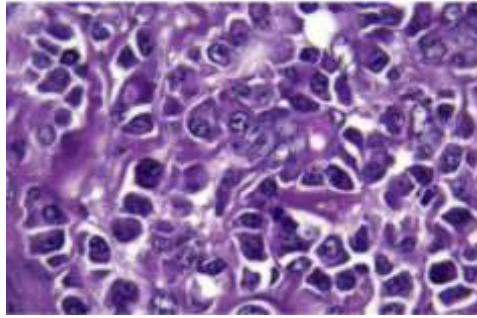


Fig.7 Fibroblastic Proliferation

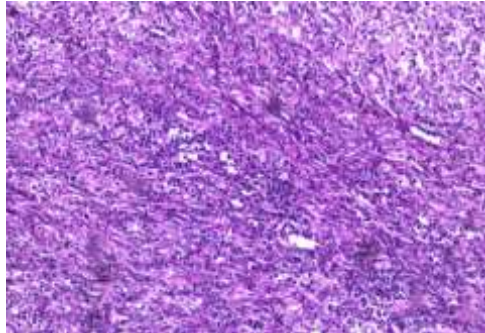
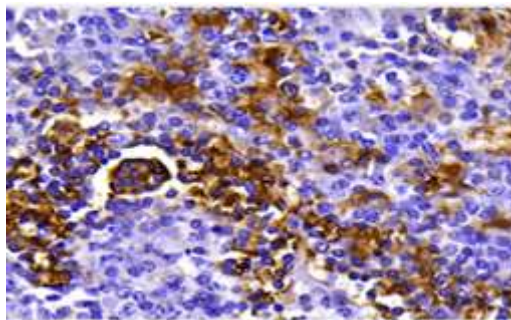


Fig. 8 IHC – S 100



Discussion:

According to the histiocyte society, Rosai Dorfman disease classified in to Sporadic (non-cutaneous), Familial RDD and Cutaneous RDD. Sporadic RDD, being the most common form includes the classic nodal form, extranodal RDD, neoplasia-associated RDD and immune – associated RDD.

Classical (Nodal and cutaneous Rosai Dorfman disease) is self-limiting and benign. Patients have one or more immune disorders, such as polyarthralgia, asthma, or hemolytic anaemia, preceding or associated with the onset of disease. Familial RDD includes H syndrome, an autosomal recessive genetic syndrome caused by mutation in the SCL29A3 gene. syndrome comprises of hyperpigmentation, hypertrichosis, hepatomegaly, hearing loss, heart anomalies, hypogonadism, short stature, hyperglycemia, and hallus valgus.

Extra nodal sites were later recognized and they were shown to represent over 40% of cases. Common extranodal sites of involvement include skin, nasal cavity, bone, orbital tissue and central nervous system. Rosai Dorfman disease remains a disorder of unknown etiology, it has been previously perceived to be a reactive, non -neoplastic that lacks clonality.

Recently evidences in support of a clonal nature of a few subsets of RDD has been found. kinase mutations have been recently noted in nodal RDD, including MAP2K1, NRAS, ARAF, KRAS. Targeted DNA /RNA sequencing and whole exome sequencing performed showed kinase driver mutations involving KRAS, MAP2K1, NRAS and CSF1R.

4. Conclusion

Rosai Dorfman disease share some morphological features with Ig G4 related disease, such as storiform fibrosis, abundant plasma cells, thereby we may consider this condition to be differential diagnosis. Other differentials to be kept in mind are Langerhans cell histiocytosis in which eosinophils and necrosis are more frequent. Langerhans cells positive for CD1A, Langherin. Hodgkins's lymphoma replaces the

architecture, but not usually situated within sinuses. Reed- Sternberg cells are absent in Rosai Dorfman disease.

Sporadic Rosai Dorfman disease is usually self-limiting and has a good outcome. Although, Rosai Dorfman has unique, histopathological cytoarchitecture it may be mistaken for other neoplastic and inflammatory histoproliferative diseases.

Surgical resection is indicated in symptomatic disease. Multifocal, irresectable extra nodal disease may require systemic therapy which include corticosteroids, sirolimus, immunomodulatory therapy etc.

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