ROSAI DORFMAN DISEASE IN A PREGNANT FEMALE: A CASE REPORT

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ABSTRACT:
Rosai Dorfman disease (RDD) or sinus histiocytosis is a rare, idiopathic, non neoplastic histiocytic proliferation of unknown etiology typically presenting with massive cervical lymphadenopathy with or without systemic involvement. It may occur in any age group, but is most commonly seen in children and young adults. We report a case of RDD with bilateral massive cervical swelling in a 21 years old pregnant female. The swelling progressively increases in size after the onset of pregnancy. FNAC was advised. The cytomorphology revealed lymphocytes, plasma cells and uni to multinucleated histiocytes with lymphophagocytosis and the diagnosis of RDD was rendered. It is concluded that FNAC is a reliable and useful tool for the diagnosis of sinus histiocytosis with massive lymphadenopathy (RDD) and biopsy can be avoided, thus reducing the inconvenience to the patients especially in pregnancy.

Key words: Rosai Dorfman disease, Lymphophagocytosis, Pregnant
INTRODUCTION:

Sinus histiocytosis with massive lymphadenopathy, commonly known as Rosai Dorfman Disease; a rare and benign entity of unknown etiology was first described by Rosai and Dorfman in 1969.² It mainly involve the lymphnodes and characterized by painless, bilateral cervical lymphadenopathy accompanied by fever, leukocytosis, elevated ESR and hypergammaglobulinemia. It is very important to distinguish it from lymphoma and other causes of histiocytosis, for proper treatment of the patient. The disease is usually self limiting. Here, we report a case of progressive enlargement of bilateral cervical swellings in a 21 years old pregnant woman.

CASE REPORT:

A 21 yrs old female in third trimester of pregnancy presented in FNAC clinic with chief complaint of progressively increasing bilateral swellings for last one year. Initially the swelling was 2x1 cm as in OPD records but it progressively increased after she conceived and now she presented with bilateral multinodular swellings of size 4x3 cm approximately. There was no history of fever or weight loss. General physical examination was within normal limits with absence of jaundice, cyanosis and oedema. On local examination revealed swellings were firm, non tender and varying in diameter from 6 to 7 cm. Skin overlying the swellings was normal. There was no history of night sweats, rashes and bleeding manifestations. Her past and personal history was not significant. There was no family history of similar illness. Other systems were within normal limits. Hematological examination revealed Hb 8.4 gm%, PCV 40.6%, MCV 78 fl, MCH 28 pg, MCHC 30 %, Retic 1.2% . The peripheral blood count revealed leukocytosis (11,800 / cu mm) with neutrophilia (82%), platelet count 3.49 lakhs/cu mm. ESR was raised
外围血液涂片显示微细的低色素红细胞，伴有轻度的异形性。胸部X光片正常。腹部超声检查正常，未发现任何器官增大或淋巴结肿大。常规生化和尿液检查均在正常范围内。对HIV I & II、HBV和Toxoplasma的血清学检查均为阴性。

FNAC的颈部肿块被进行。用Romanowsky染色的涂片，被发现是细胞性的，并显示了大量的巨噬细胞，呈小群或大群，有丰富的浅粉红色的嗜酸性细胞质和多叶或单叶的核。这些巨噬细胞显示了吞噬淋巴细胞、中性粒细胞和浆细胞的能力。背景是淋巴细胞和红细胞。细胞病理学上，考虑为Rosai Dorfman Disease的双侧颈部淋巴结受累的可能性。

**DISCUSSION:**

RDD是一种罕见的、非肿瘤性巨噬细胞增生症，具有原发性特征。1大多数病例发生在第一或第二十年龄，然而任何年龄都可能发生。男性比女性更常见。2,3

大部分患者（95%）出现巨大的颈部淋巴结肿大。淋巴结肿大的程度有时是非常显著的，有报道称淋巴结肿大达到6厘米，导致呼吸功能障碍。在43%的病例中，同时累及外周病变，而仅仅23%的病例为孤立的外周病变。3临床患者可能被误诊为淋巴增生性疾病或其他感染性疾病，如结核病，后者在发展中国家如印度更为常见。非特异性全身症状包括发热、体重减轻和乏力。患者也可能出现咽炎、鼻腔分泌物和阻塞。1实验室异常包括贫血、白细胞增多、血沉升高和高γ球蛋白血症。2

确切的病因、病理生理和自然病史仍然是未知的，尽管在过去20年中进行了大量的研究。8免疫介导的起源被少数人提出，而其他人则认为是一种反应性过程，由于某种感染，如HHV-6或未定义的免疫缺陷，由某些其他生物如EBV或CMV引起的，可能引起疾病。1,2少数研究者认为它可能是骨髓干细胞的起源。2目前，它被认为是反应性增生，表现出自发性消退。3 RDD是骨髓单核细胞从外周血中招募到
lymph nodal sinuses or extranodal sites and their transformation into the immunophenotypically distinct RDD histiocytes which demonstrate emperipolesis and functional uniqueness in terms of cytokine expression profile. Release of cytokines like TNF-α from these cells is responsible for the genesis of fever and other systemic symptoms.⁷ There is no ideal therapeutic regimen, treatment option ranges from surgery, radiotherapy and steroids to chemotherapy.³

Although RDD is a histopathological diagnosis, yet FNAC has proved to be a reliable tool in diagnosing it. Only a few cases of RDD diagnosed by fine needle aspiration cytology have been reported in the literature. Cytology reveals numerous large histiocytes with abundant pale cytoplasm with phagocytosed lymphocytes, neutrophils and plasma cells (emperipolesis). In emperipolesis the lymphocytes are not attacked by enzymes and appear intact within the histiocytes in contrast of phagocytosis. The phenomenon of emperipolesis is highly useful for the diagnosis of RDD using FNAC.¹² All these features were also present in our case. None of the case has been reported in pregnant female as in our case.

Histologically, the lymphatic sinuses are dilated and are reveal presence of lymphocytes, plasma cells and numerous large histiocytes with emperipolesis resulting in partial or complete architectural effacement.⁵,⁸

Differential diagnosis includes nonspecific sinus hyperplasia, Langerhans cell histiocytosis, hemophagocytic syndrome, tuberculosis, lymphoma and metastatic malignant melanoma.⁹ Reactive sinus hyperplasia shows large clusters of histiocytes, accompanied by reactive lymphocytes, germinal center cells, immunoblast and tingible macrophages, emperipolesis is absent. In LCH, langerhans cells have grooved and twisted nuclei and the background has eosinophilic microabscesses. LCH is positive for both S-100 protein and CD 1a. Hemophagocytic syndromes should be differentiated from RDD on the basis of the presence of hemophagocytosis, absence of emperipolesis and the presence of pancytopenia and hepatosplenomegaly. Tuberculous lymphadenitis shows epithelioid cell granuloma with or without caseous necrosis which are absent in RDD. Smears from patients with Hodgkins disease show lymphocytes, plasma cells, histiocytes, eosinophils and Reed- Sternberg cells.¹² They are S100 protein negative, CD15 positive, and CD30 positive. Negative staining for HMB-45 and pankeratin differentiate RDD from melanoma and metastatic carcinoma.⁹
Clinical photograph of the patient

Figure showing FNAC and Histopathology from the specimen
References:


Legends:

Figure 1: Photograph of patient showing massive enlargement of cervical lymphnodes.

Figure 2a&amp;b: Cellular smears showing large number of histiocytes in groups or scattered singly and showing phagocytosis of lymphocytes.