

## Bilateral Breast Involvement in Rosai-Dorfman Disease: Report of a Case in a Middle Aged Lady

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### Abstract

A woman aged 40 years sought medical attention for bilateral breast swellings considered to be bilateral fibroadenomata or malignancy. Excised lump revealed to be Rosai-Dorfman disease with characteristic histology. Its incidence in the breast; a rare entity, is discussed in relation to available literature on the subject. One of the perplexing problems; particularly, the origin of the tumor cells is discussed against the background of unusual immunochemical data.

**Keywords:** Breast, Rosai-Dorfman Disease, Clinical and Pathologic Review, Significance of Immunochemical Findings.

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### Introduction

First reported by Rosai and Dorfman in 1969 [1] and later elaborated by them in a detailed report in 1972, Rosai-Dorfman disease [2] (henceforth RDD) is an idiopathic histiocytic disorder; defined by its characteristic histology of lymphohagocytosis and a strong affinity to immunochemical marker of s-100. Initially it was thought to involve lymph nodes massively and subsequently after establishing a registry at Yale University School of Medicine in 1990, [3] it was realized it involves almost all organs except gynecologic organs. In above registry of 423

patients 43% had at least one site of extra nodal involvement as well as lymph node involvement; while 23% cases had extra nodal involvement only. In order of their frequency, the extra nodal sites are skin, nasal cavity and para nasal sinuses, soft tissue, eyelid and orbit and bone [3]. Breast is an uncommon site and because of its rarity, we felt it is worth while to publish this case; lest it would be mistaken for malignancy and overtreated; particularly in a case of this nature, because of its bilaterality, it was likely to be mistaken for invasive lobular carcinoma; which was more common to occur bilaterally.

## Case report

A 40 yr old Muslim lady sought medical help for bilateral lumps in the breast of more than 6 months duration. They were situated in outer quadrant, freely mobile, hard in consistency, and without fixity to deeper structures. Because of bilaterality, lobular carcinoma or fibroadenoma was thought of and operated upon. FNAC didn't reveal any malignancy. No lymphadenopathy was detected clinically.

On operation, both tumors measured 5.5x3.5x2.0cm and 3x3x3 cms respectively and removed without any difficulty. The circumscribed tumors were grayish white and firm. Microscopically the tumors revealed to be of inflammatory origin involving the breast parenchyma as well as surrounding fat. Apart from histiocytes; arranged in sheet like manner, the lesion showed lymphoid nodules; consisting of mature follicles. Fibrosis is marked. The predominant histiocytes showed abundant eosinophilic cytoplasm with irregular nuclei giant cells with lymphophagocytosis were occasional findings (Fig. 1a,b) but they were conspicuous. Immunohistochemically, the histiocytes were strongly positive with s-100 (Fig. 2a). Marker CD1-a was negative throughout, with CD-68, the histiocytes were faint and poorly delineated (Fig. 2b).

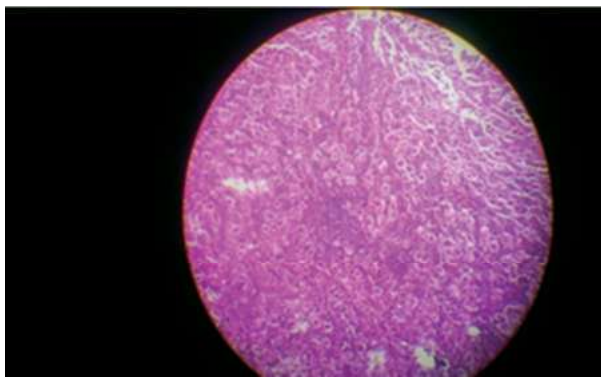


Fig. 1a: Lymphophagocytosis (Medium).

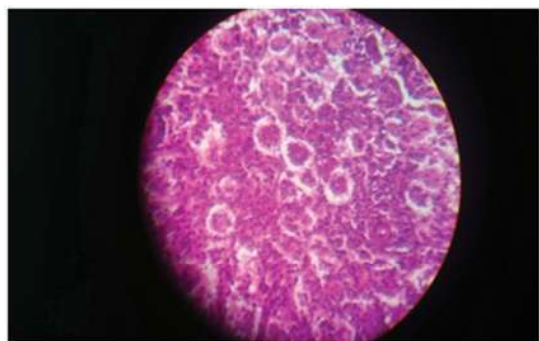


Fig. 1b: Lymphophagocytosis (High power) giant cell

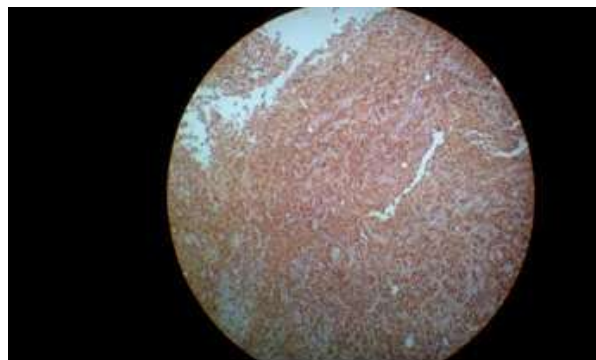


Fig. 2a: S-100+ve histiocytes (Medium).



Fig. 2b: Inset CD.68 faintly positive (Medium)

## Discussion

In the registry mentioned above, only two cases were recorded in the breast; one was considered as a fibroadenoma and the other as a breast mass with axillary lymphadenopathy. Till now, 14 such cases were recorded occurring in breast; most of which were single case reports except in one published by Green et al. [4]; dealing with seven such cases; the maximum recorded to far. Follow up details were available only for three cases in the above series and of them, one proved to be fatal; in a woman 84 years' old, had in addition to breast mass, mediastinal lymph adenopathy, abdominal and hepatic involvement, thereby expanding the biologic behavior of this neoplasm.

Clinical and mammographic findings in all these cases indicated either malignancy or benign cystic disease; where as histologically, it was mistaken for histiocytosis-x, malacoplakia, extraskeletal Erdheim-Chester disease, histiocytic sarcoma, interstitial and follicular dendritic cell tumors, fibrous histiocytoma, idiopathic granulomatous mastitis and infective granulomatous disease. In RDD, two histologic features, seen consistently, are emperipolesis and strong positivity with S100, which are not shared by any one of the above conditions. However in sinus histiocytosis with

massive lymphadenopathy (the other term for RDD and abbreviated as SHML), SHML cells are rare and focal and require effort to identify them.

The diagnosis is facilitated because of its strong positivity with S100 marker; proved in our case admirably and also in all cases reported in the literature. In spite of extensive and detailed description of immunochemical markers of various histiocytes, carried out by Eisen et al. [5] and also discussed elaborately by Ioachim [6], the nature of SHML cells is still conjectural. There are three categories of these cells: those belonging to monocytes and macrophage system and others being dendritic cell system of interstitial and follicular systems; each having distinct site specific presence, morphological, enzymatic, immunochemical features and causing dissimilar diseases. While normal monocytes and macrophages are CD 68 (+), S100 (-), and CD 1a (-) and dendritic cells, CD 68 (-), S-100 (+), and CD.1a (+), SHML cells are CD 68 (+) (in our case, it was weakly positive), S-100(+), and usually CD 1a (+) (in 10% cases only) (Rezketal) [7]. When monoclonal antibody specific for dendritic cells was used, it was negative for SHML cells. Warnke et al. [8] considered these cells express activation antigens and of polymorphic nature as true functionally activated macrophage derived from circulating monocytes. Even though S-100 was positive both in Langerhan histiocytosis as well as in RDD, employing Leu-6 marker, it was found out that Leu-6 has greater specificity for Langerhan cells while it is not expressed in SHML cells. More over, morphology of SHML and Langerhan cells is entirely different and it is not difficult to diagnose these diseases on morphological grounds alone. Cytogenetic and polymerase chain reaction also failed to characterize the origin of these cell (Ioachim) [6] and in spite of easy histologic diagnosis, still the origin of the disease remains an enigma; because of its perplexing immunochemical findings (Jaffe) [9].

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