



## Rosai Dorfman Disease: A Rare Benign Disease of Breast

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### ABSTRACT

*Rosai Dorfman Disease also called sinus histiocytosis with massive lymphadenopathy (SHML) is a rare disorder of sinus histiocyte. Mostly involves lymph nodes but can occur in other body parts. This disease is extremely rare in breast. We report a case of breast lump, clinically & radiologically appearing malignant that found out to be Rosai Dorfman Disease on histopathology & IHC.*

### INTRODUCTION

**ROSAI DORFMANS DISEASE** of breast is a benign breast lump which appears clinically and radiologically malignant. Diagnosis is finally based on histology which shows histiocytosis with typical emperipolesis<sup>(1)</sup>. An extremely rare disease of breast as few cases reported so far. This is a rare histiocytic disorder which involves clusters of WBC accumulated in lymph node and other organs. The term RDD given by two pathologists Juan Rosai & Ronald Dorfman in 1969<sup>(2)</sup>. Occurs equally in males & females mostly in 1<sup>st</sup> and 2<sup>nd</sup> decade. Clinically presents as massive bilateral and painless cervical lymphadenopathy with fever, night sweat & weight loss<sup>(3)</sup>. Mediastinal, retroperitoneal & inguinal nodes may be involved. Extranodal site most frequent is skin, soft tissue, upper respiratory tract, bone, eye and rarely in breast.

### PATHOLOGY

Histology shows pericapsular fibrosis and dilated sinuses heavily infiltrated by large histiocytes and plasma cells. EMPERIPOLESIS (engulfment of lymphocytes and erythrocytes by histiocytes) is typical. Stain +ve for S100, CD163, CD68.

### CASE HISTORY

Hamida khatoun 60/f, postmenopausal women presented with lump in lt. breast for 1 month at MAHAVIR CANCER SANSTHAN, PATNA (mcs no.D 63317). Patient had no similar f/h/o breast lump. Patient was non diabetic, non hypertensive & no h/o any significant medical illness. There was no h/o any previous surgery. General examination was normal. On local examination of breast there was 3x2 cm firm, slightly mobile lump in left lower inner quadrant. 1x1 cm skin overlying lump was tethered to it. Mammographic finding was BIRADS cat.3 lesion. Core needle biopsy &

further IHC examination suggested small lymphocytic lymphoma. Pt. was planned for wide local excision with 2 cm margin.HPE of lump suggested ROSAI DORFMANS DISEASE. Pt. recovered well. Follow up examination 3 monthly in May, Aug, Nov 2015 was normal.

## DISCUSSION

ROSAI DORFMANS (RD) disease is a rare histiocytic disorder which involves over production of a type of WBC, called non langerhans sinus histiocyte <sup>(4)</sup>. Generally accumulate in lymph node but can occur in other organs. Etiology may be infectious, environmental or genetic. Sinus histiocytic with massive lymphadenopathy (SHML) synonyms with RD disease may occur in nodal & extranodal site simultaneously. Patient may have fever, pain like systemis complaint or symptoms attributed to organ involved. Majority have spontaneous remission. Some have aggressive disease. Surgical intervention is needed for symptomatic basis or cosmesis. For widespread disease some vinca alkaloid, alkylating agents and steroids have been mentioned.<sup>(5)</sup>

Breast is an extremely infrequent site of occurrence as few cases reported so far. Clinically and radiologically appearing malignant disease needs proper tissue diagnosis to prevent unnecessary overtreatment of this breast disease.

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