Balkan Med J 2025;42:565-6

Extranodal Rosai-Dorfman Disease of the Breast Presenting as Inflammatory Breast Cancer

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A 50-year-old woman presented with a left breast (LB) mass that had progressed over 10 months. Her history included a 5 pack-year smoking history. Menarche occurred at age 13, and she delivered her only child by cesarean section at age 27; she did not breastfeed. There was no personal or family history of malignancy.

On examination, a poorly defined, indurated mass measuring approximately 4 cm in greatest diameter was palpated in the upper outer quadrant (UOQ) of the LB. The overlying skin showed inflammatory changes (peau d'orange), and the breast was enlarged compared with the right side (Figure 1a). A firm, mobile, ipsilateral axillary lymph node measuring 1 cm was also detected.

Ultrasonography demonstrated a hypoechoic UOQ mass with edema, fluid collections, and associated inflammatory features, measuring up to 3 cm. Mammography showed a poorly defined, hyperdense lesion in the same region (Figures 1b, c). The Breast Imaging Reporting and Data System (BI-RADS) score was 1 for the right breast and 3 for the LB.

An ultrasound-guided core needle biopsy revealed non-specific inflammatory changes. Because of radioclinical discordance between the imaging and clinical findings, surgical excision of the mass was performed without lymph node sampling.

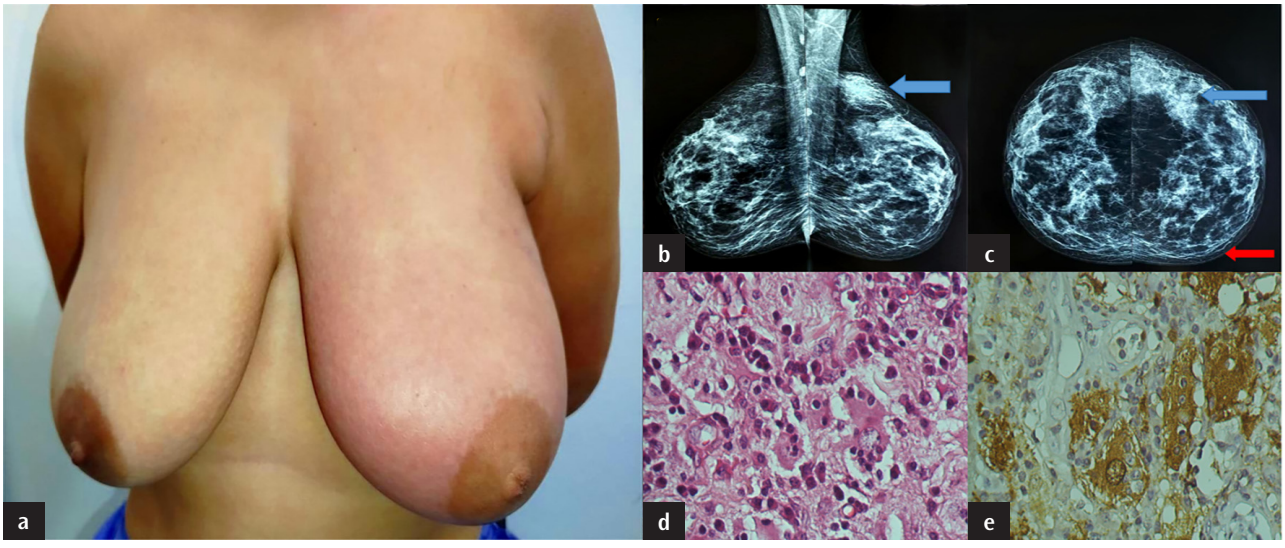

FIG. 1. (a) The left breast demonstrates an inflammatory appearance with an increased size compared to the right breast. (b) Oblique-projection mammography. (c) Craniocaudal-projection mammography: b + c showing a poorly defined, hyperdense mass measuring 3 cm in the greatest dimension, located in the upper outer quadrant of the left breast (blue arrow); the skin is thickened along the entire left breast (red arrow). (d) Hematoxylin and eosin staining: numerous histiocytes with round to oval nuclei, prominent nucleoli, and abundant eosinophilic cytoplasm containing engulfed lymphocytes (emperipolesis). (e) Immunohistochemistry showing positive staining for S100.

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Received: August 04, 2025 **Accepted:** August 16, 2025 **Available Online Date:** 31.10.2025 • **DOI:** 10.4274/balkanmedj.galenos.2025.2025-8-9
Available at www.balkanmedicaljournal.org

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Cite this article as: Bouhani M, Kamoun S. Extranodal Rosai-Dorfman Disease of the Breast Presenting as Inflammatory Breast Cancer. Balkan Med J.; 2025; 42(6):565-6.

Histopathological analysis showed dense inflammatory infiltrates composed mainly of lymphoplasmacytic cells and numerous enlarged macrophages with lymphocytic emperipolesis, a characteristic feature of Rosai-Dorfman disease (RDD) (Figure 1d).

Immunohistochemical staining confirmed that the macrophages were pankeratin-negative and S100-positive, supporting the diagnosis (Figure 1e). Laboratory results were unremarkable except for leukocytosis, with a white blood cell count of 11,400 cells/mm³.

The pathological features were consistent with breast RDD. The patient was monitored for 10 months postoperatively and showed no evidence of recurrence.

Breast RDD is an exceptionally rare benign histiocytic disorder, with fewer than 100 reported cases worldwide. Breast involvement occurs in < 1% of all RDD cases.[1,2] Clinical presentations that mimic inflammatory breast carcinoma or mastitis are extraordinarily uncommon and pose significant diagnostic challenges.

In postmenopausal women over 50 years of age, an inflammatory breast presentation typically raises immediate concern for inflammatory breast carcinoma, which accounts for 1%-4% of all breast cancers and carriers a poor prognosis.[3] Hallmark features include rapid-onset breast enlargement, induration, erythema, and peau d'orange skin thickening.[4,5] However, benign etiologies should also be considered. Non-puerperal mastitis, particularly in smokers, remains an important differential diagnosis even in the absence of fever.[6] Leukocytosis, as observed in this case, further complicates interpretation because of its non-specific nature.

The discordance between clinical and radiological findings necessitates tissue sampling for definitive diagnosis. Breast RDD usually appears as an ill-defined hypoechoic mass, with BI-RADS scores commonly falling into suspicious categories, making radiological distinction from malignancy difficult.[5,7,8] Core needle biopsy establishes the diagnosis in approximately 60%-70% of cases, while the remainder require surgical excision, as in our patient.[9]

Definitive diagnosis depends on characteristic histopathological features and immunohistochemical patterns. Pathognomonic findings include dense lymphoplasmacytic infiltrates and large histiocytes containing intact inflammatory cells (emperipolesis), although this feature may be less prominent in extranodal disease.[1,10] Immunohistochemistry is pivotal: RDD histiocytes typically express S100 protein and CD68 but are negative for pankeratin, distinguishing RDD from epithelial malignancies and other histiocytic disorders.[2]

Surgical excision is the preferred treatment for isolated breast RDD, providing both diagnostic certainty and curative potential. Prognosis is excellent, with mortality exceedingly rare and recurrence uncommon.[1,7] Long-term follow-up has shown favorable outcomes in most patients.

Informed Consent: Informed consent was obtained from the patient.

Authorship Contributions: Concept- M.B.; Design- M.B.; Materials- M.B., S.K.; Data Collection or Processing- M.B., S.K.; Analysis or Interpretation- M.B.; Literature Review- M.B.; Writing- M.B.; Critical Review- M.B.

Conflict of Interest: No conflict of interest was declared by the authors.

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