

Received Date : 27-Oct-2015

Revised Date : 06-Nov-2015

Accepted Date : 10-Nov-2015

Article type : Breast Images

Author Manuscript

This is the author manuscript accepted for publication and has undergone full peer review but has not been through the copyediting, typesetting, pagination and proofreading process, which may lead to differences between this version and the [Version of Record](#). Please cite this article as [doi: 10.1111/tbj.12690](https://doi.org/10.1111/tbj.12690)

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Extranodal Rosai-Dorfman Disease of the Breast

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A middle-aged woman was noted to have scattered densities and asymmetry on routine mammogram. Diagnostic mammography and ultrasound demonstrated a subcentimeter round mass with indistinct margins and no associated calcifications (Figure 1). CT of the chest, abdomen and pelvis demonstrated multiple subcutaneous nodules but no significant lymphadenopathy.

Breast core biopsy was performed and showed a multinodular lesion comprised of diffuse lymphoplasmacytic and histiocytic inflammation with both loose and well-formed granulomas, some of which were necrotizing. Similar findings were identified in a subcutaneous biopsy from the upper extremity. Immunohistochemical stains showed the histiocytes to express CD68, CD163 and S100 via immunohistochemical staining. Inflammatory cells within the cytoplasm of large histiocytes, or emperipolesis, was also highlighted by these stains (Figure 2). Special stains for fungal and acid fast organisms were negative. These features were diagnostic of extranodal Rosai-Dorfman disease.

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a benign histiocytic proliferation that predominantly involves lymph nodes, most often those of the head and neck region. However, extranodal disease commonly accompanies lymph node involvement and has been identified throughout the body, with skin and soft tissue being the most frequent extranodal sites. Extranodal disease without identifiable lymph node involvement has been estimated to occur in 23% of cases.

Extranodal RDD is a rare diagnosis in the breast. When it does involve the breast, it may be either unilateral or bilateral. Extranodal RDD involving the breast has variable imaging findings but may mimic breast cancer clinically and radiographically, presenting as a mass as in this case.

Diagnostic features on biopsy are characteristic histiocytes with abundant pale cytoplasm exhibiting emperipolesis and co-expression of S100 and macrophage markers such as CD68 and CD163. There is often abundant associated lymphoplasmacytic inflammation. While the latter features were prominent in this case there was also an unusual finding of necrotizing granulomas, which necessitated additional evaluation for fungal and acid fast microorganisms.

RDD has been linked with infectious agents such as Epstein-Barr Virus, human herpesvirus and parvovirus B19 as well as with IgG4-related diseases and with germline mutations in *SLC29A3* in familial cases. RDD may show spontaneous regression in 20% of cases and is typically only treated when there is need for symptomatic relief, with therapies including steroids, immunomodulatory and chemotherapeutic drugs, radiotherapy and surgical excision. At the time of this report this patient had no significant progression of disease and management was observation only.

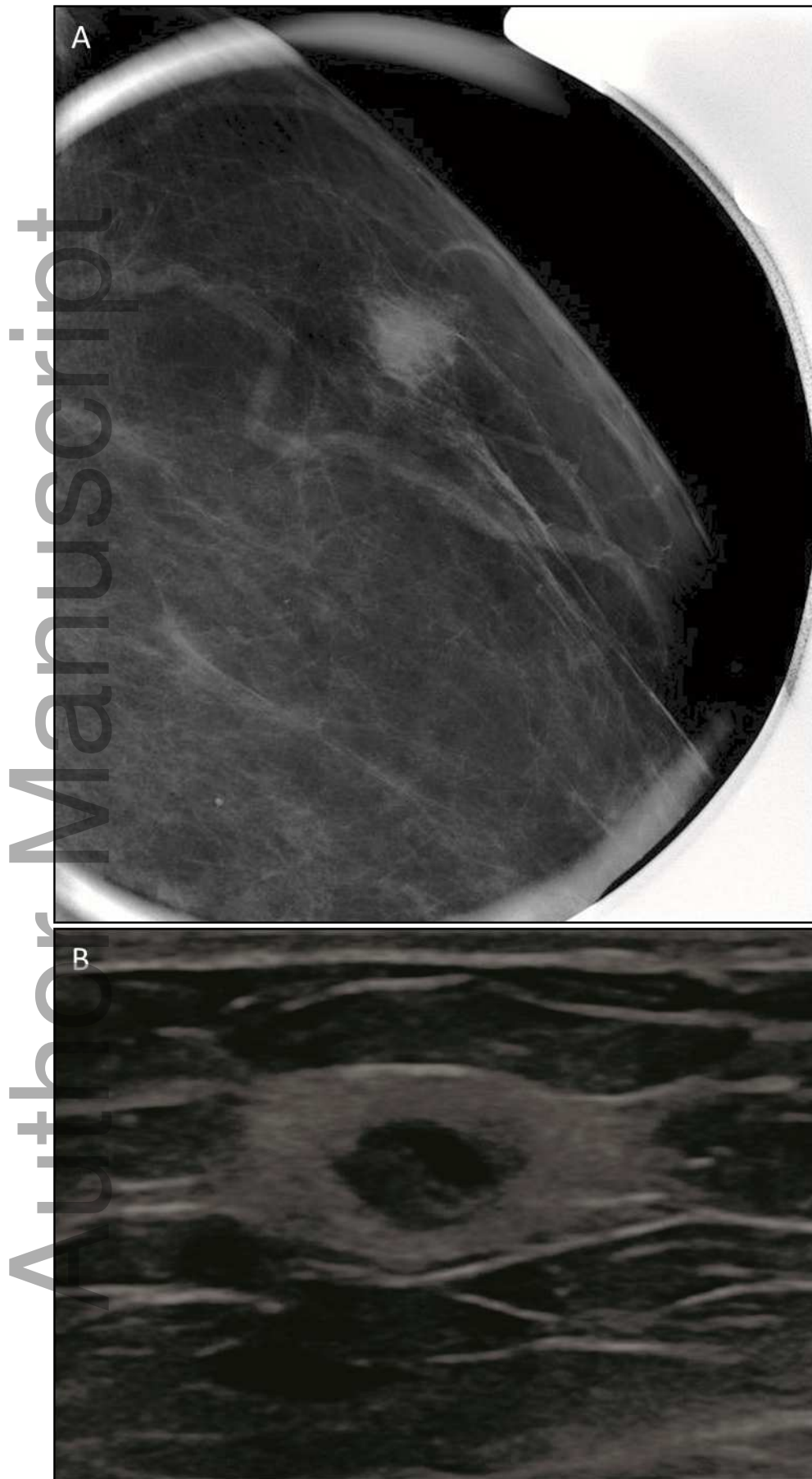
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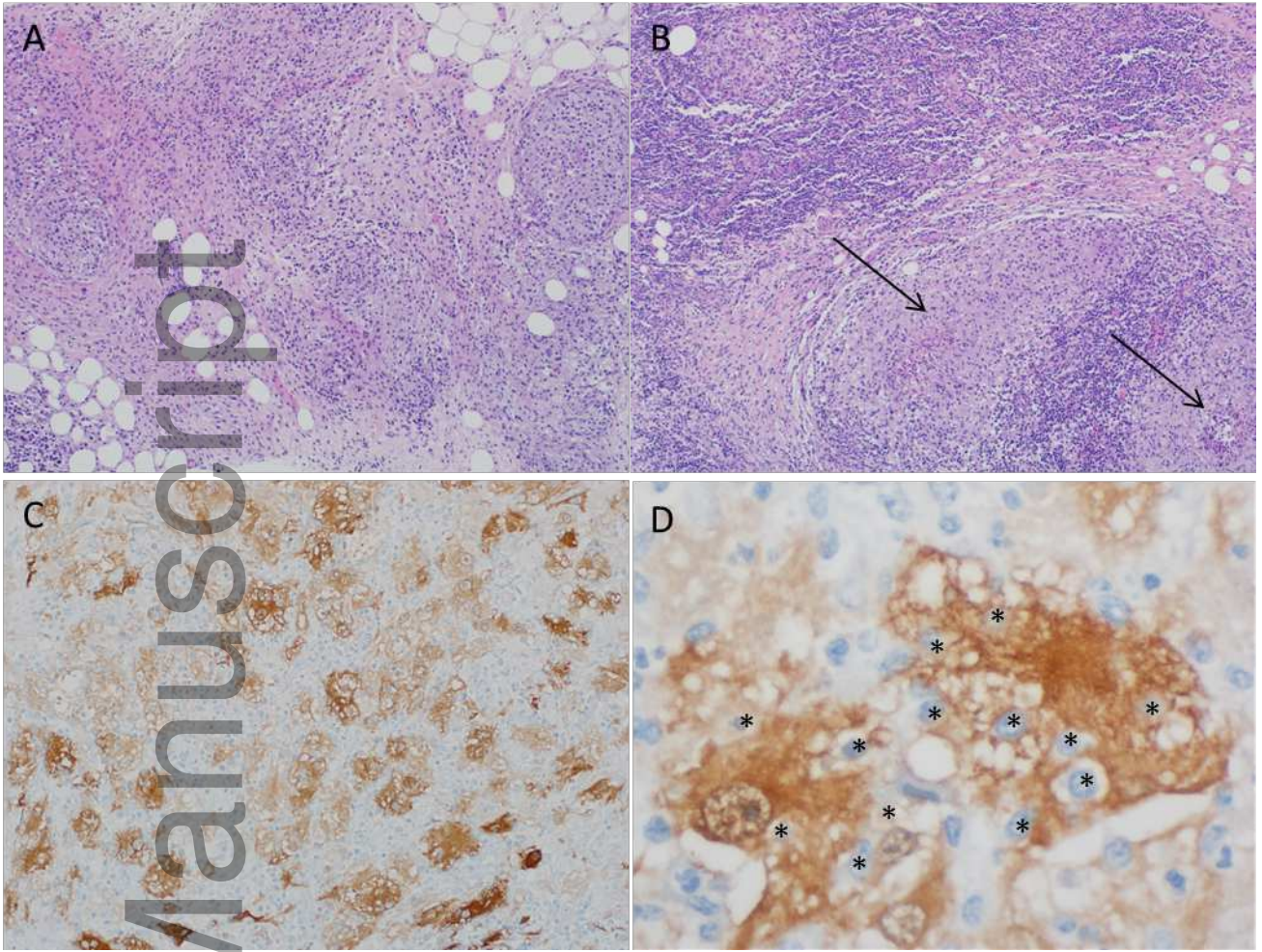
Radiographic images are courtesy of Dr. Leah Carlson, Department of Radiology, University of Michigan.

Figure Captions:

Figure 1. (A) Mammogram spot magnification and (B) ultrasound images of breast mass.

Figure 2. (A-B) Breast biopsy showing lymphoplasmacytic and histiocytic inflammation with granulomas, including necrotizing granulomas (arrows, B) (H&E, 10X). (C) S100-positive histiocytes (IHC, 20X). (D) S100-positive histiocytes with prominent emperipolesis (inflammatory cells within histiocyte cytoplasm are denoted by *s) (IHC, 60X).





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