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Review Article

Lupus mastitis as a first manifestation of systemic disease: About two cases with a review of the literature

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ABSTRACT

Lupus mastitis is an uncommon manifestation of systemic lupus erythematosus (SLE) that affects the subcutaneous fat in the breast, much like lupus panniculitis, but additionally involves the mammary gland. We report on two women for whom lupus mastitis was the initial manifestation of SLE and provide a literature review of 34 additional cases reported in the Anglo-Saxon and French literature since 1971, making this the largest review to date.

Lupus mastitis (LM) can manifest clinically as subcutaneous masses that may be painful, or may present cutaneous involvement such as thickening and discolouration. The radiologic manifestations of LM are broad and include calcifications, masses and asymmetries. Most often, excluding malignancy requires percutaneous biopsy, with histologic findings that are virtually pathognomonic for SLE. Thus, surgery is avoided and medical management can begin, antimalarial drugs and corticosteroids in most cases.

1. Introduction

Lupus erythematosus is an autoimmune disease that demonstrates systemic, cutaneous or both systemic and variable cutaneous manifestations. Cutaneous lesions are classified as specific (ex: discoid lesion) or non specific (ex: alopecia, vasculitis, nail changes, lupus panniculitis) [1,2]. Lupus panniculitis is a non specific inflammatory condition involving the subcutaneous fat and is most often seen in patients with a known diagnosis of lupus. It is referred as lupus erythematosus profondus (LEP) and it occurs most often in patients with a discoid variant lupus [1,2].

Lupus mastitis (LM) is an uncommon form of manifestation of systemic lupus erythematosus (SLE). It is estimated that 2–3% of patients with SLE will develop lupus panniculitis [1,2], characterized by distinctive subcutaneous nodules as described by Kaposi in 1883. [1,3–6]. LM is simply defined as extension of lupus panniculitis to the mammary gland [7]. LM is a very rare pathology, with 44 cases reported in the Anglo-Saxon and French literature since 1971. Of these, only 34 well-documented pathologically confirmed cases have been found. LM often presents both clinically and mammographically as a mass. Suspicious calcifications can also be detected on mammography. Acutely, LM is usually painful and the skin can appear erythematous or violaceous. The course of the disease is chronic. Some lesions will ulcerate or become atrophic but others will resolve [7].

This article presents two new diagnoses of LM in women not known for SLE and compares and contrasts these two cases with published reports.

2. Case 1

A 64 year-old postmenopausal white woman presented with a slightly painful palpable mass in the upper central right breast that had developed over 3 months. On physical examination, the mass was associated with skin thickening and an erythematous/bluish discoloration. She had no previous history of breast disease or SLE and was otherwise healthy. One sister had been diagnosed with breast cancer at age 60.

A mammogram showed trabecular thickening and a right upper quadrant regional asymmetry (Fig. 1A) that had progressed since the year prior (not shown), without distortion or associated microcalcifica-

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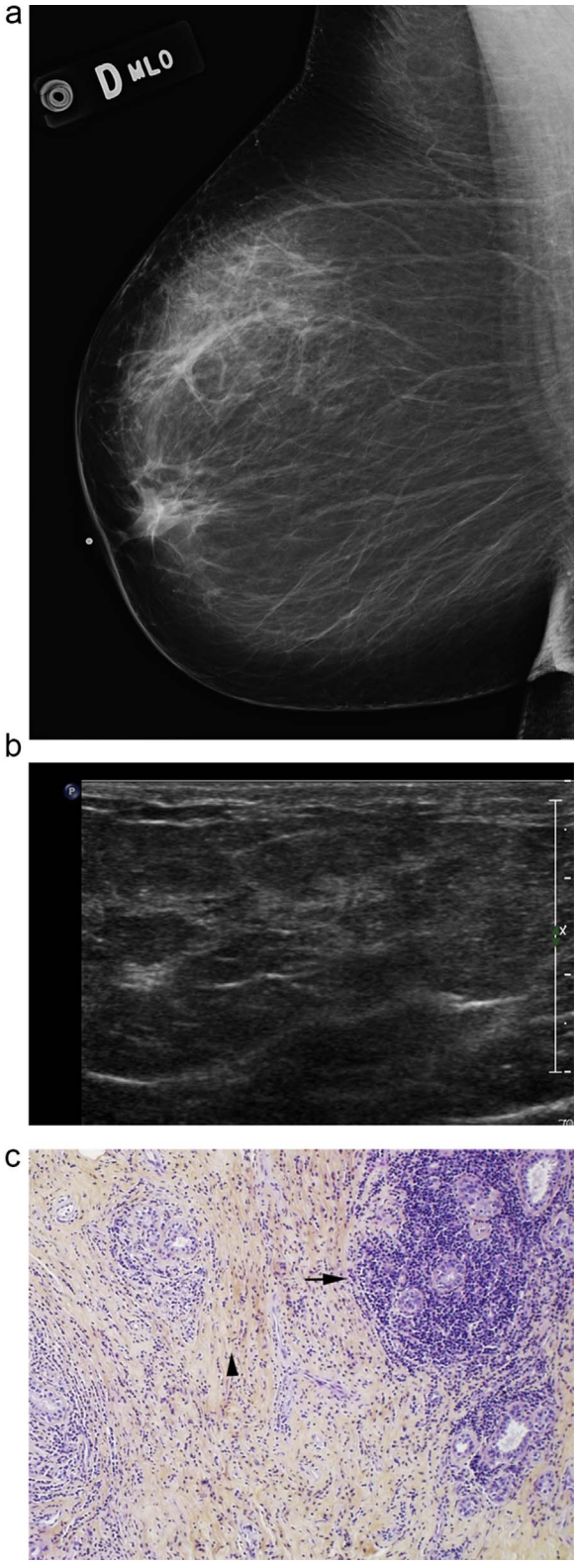

Fig. 1. A 64 year-old postmenopausal white woman presented with a slightly painful palpable mass in the upper central right breast that had developed over 3 months.A. Right mammographic mediolateral oblique (MLO) view shows trabecular thickening and regional asymmetry of the superior quadrant.B. Breast ultrasound reveals diffuse hyperechogenecity of the subcutaneous fat and cutaneous thickening, without any mass.C. Open surgical biopsy was performed and pathology revealed lymphocytic infiltrates involving both lobules (arrow) and stroma (arrowhead) on hematoxylin and eosin stain.

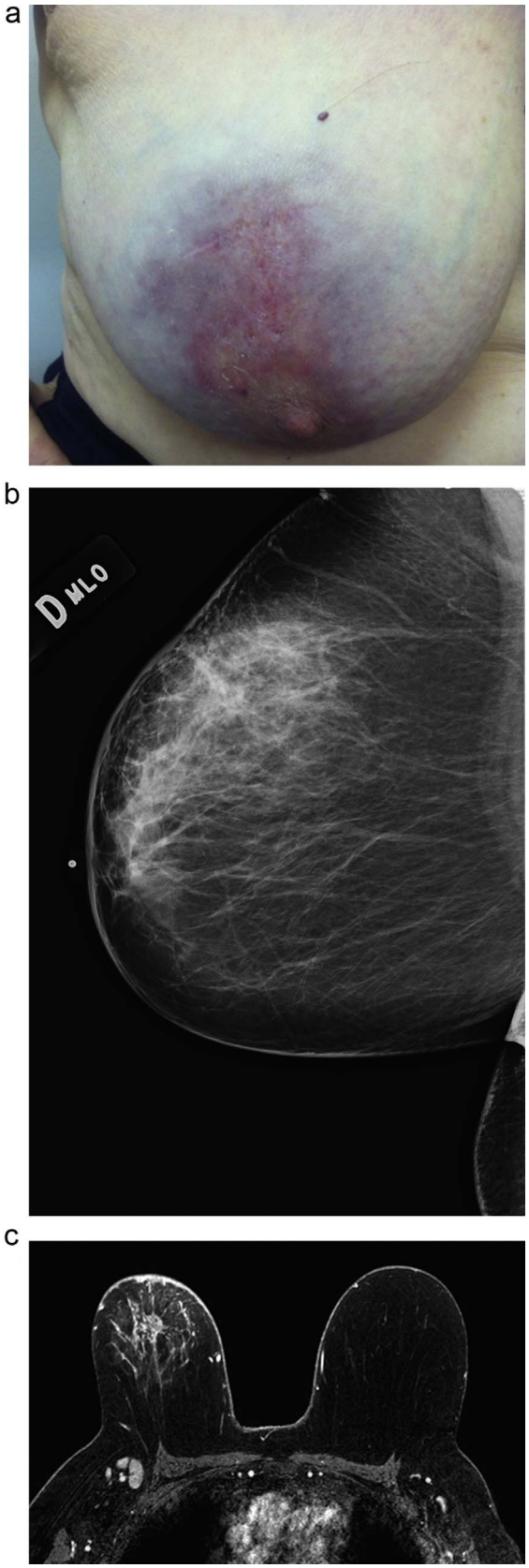

Fig. 2. Same patient as Figure 1, six months later.A. The skin is erythematous with a bluish discoloration and desquamation. Note scar from recent open biopsy in the upper quadrant.B. Right MLO view confirms progression of both skin and trabecular thickening as well as progressive asymmetry in the superior breast.C. Axial delayed post-contrast T1-weighted MRI shows skin thickening of the upper right breast, along with mass enhancement in the central breast and an enlarged ipsilateral lymph node.