

Case Report

Idiopathic Lobular Granulomatous Mastitis Case Report and Review of Literature

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ABSTRACT

Idiopathic lobular granulomatous mastitis (IGM) is an uncommon inflammatory disease that can mimic breast carcinoma, tuberculosis (TB) of the breast, or a mastitis refractory to antibiotics, with an overall incidence of 2%, only occurring in females. Clinical presentation most commonly consists of a firm unilateral, discrete lump of the breast, and may have inflammation of the skin warranting a biopsy.

This is a case report of granulomatous mastitis in a woman in reproductive age from an endemic area of TB with prior TB exposure. This case report will highlight the clinicopathological findings, treatments, prognosis, and review of literature on granulomatous mastitis.

Due to its rarity, benign pathology, but inflammatory chronicity – it poses both a diagnostic and therapeutic hurdle as it can be confused with mastitis, breast abscess, and breast carcinoma on presentation.

The etiology of IGM is unclear. It is most commonly found in premenopausal women after their last childbirth with oral contraceptive use. The symptoms, diagnosis, and management require a multispecialty approach of several experts to avoid iatrogenic complications and assert the proper diagnosis.

Introduction

Granulomatous Lobular mastitis is divided into two subcategories, Idiopathic granulomatous mastitis (IGM), and granulomatous mastitis.⁴ Granulomatous mastitis is usually secondary to an underlying condition such as TB, sarcoidosis, and rarely, injection of foreign material [1]. Idiopathic granulomatous mastitis is an uncommon inflammatory disease that can mimic breast carcinoma, tuberculosis (TB) of the breast, or a mastitis refractory to antibiotics, with an overall incidence of 2%, only occurring in females [2]. Clinical presentation most commonly consists of a firm unilateral, discrete lump of the breast, and may have inflammation of the skin warranting a biopsy. Other clinical findings may include: nipple retraction, sinus formation, and regional lymphadenopathy. In endemic areas or with travel history to Tuberculosis regions, breast TB should be included with the differential of breast carcinoma. This

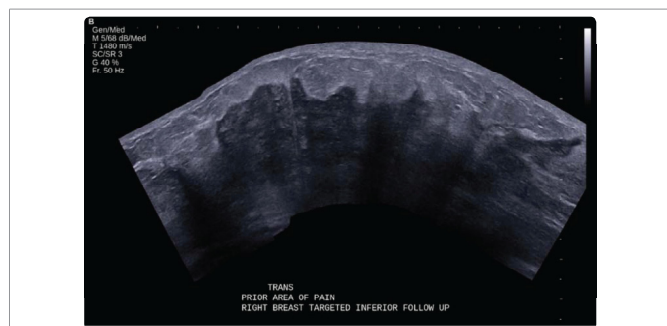
is a case report of granulomatous mastitis in a woman in reproductive age from an endemic area of TB with prior TB exposure. This case report will highlight the clinicopathological findings, treatments, prognosis, and review of literature on granulomatous mastitis.

Idiopathic granulomatous mastitis is a diagnosis of exclusion lending itself to be a challenge to diagnose [3]. Due to its rarity, benign pathology, but inflammatory chronicity – it poses both a diagnostic and therapeutic hurdle as it can be confused with mastitis, breast abscess, and breast carcinoma on presentation [4,5]. The etiology of IGM is unclear [6]. It is most commonly found in premenopausal women after their last childbirth [4] or with oral contraceptive use [7,8]. Idiopathic granulomatous mastitis is more common in pregnant and lactating women and in patients with hyperprolactinemia and may be related to increased ductal secretions which may lead to damaged ductal epithelium with glandular secretions leaking into

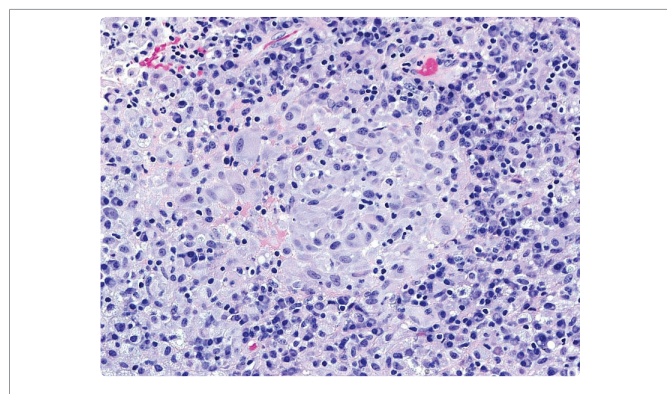
the connective tissue of the breast stroma resulting in a chemical mastitis [9]. Interestingly, Fazio et al. reported that the majority of patients diagnosed with IGM are from developing countries [10]. The symptoms, diagnosis, and management require a multispecialty approach of several experts to avoid iatrogenic complications and assert the proper diagnosis [3,6].

Case Report

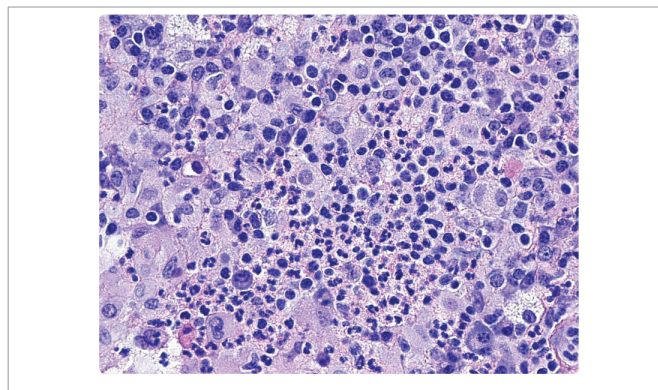
A 30-year-old female, G1P1, with no past medical history presented in 2014 with right breast mastitis and pain. A targeted ultrasound demonstrated non-encapsulating irregular hypoechoic masses and soft tissue edema categorized as a BIRADS 4 (Figure 1). Subsequently, an ultrasound guided right breast core needle biopsy was performed demonstrating chronic inflammatory cells. The patient underwent operative management with excisional biopsy. Pathology demonstrated soft tissue with acute and chronic inflammation, non-necrotizing granuloma with margin involvement (Figure 2 and 3). Patient was treated with oral antibiotics postoperatively and recovered fully. Patient presented four months later with recurrence and discharged home on oral antibiotics with empiric tuberculosis treatment recommended by our infectious disease department. Two months later the patient presented with left mastitis and was treated as an outpatient, failing oral antibiotic therapy. The patient underwent operative incision and drainage with cytology demonstrating inflammatory cells at an outside institution. All of the patient's blood and wound cultures were sterile for each occurrence. The patient was also negative for tuberculosis via quantiferon gold and acid fast bacterial cultures.



Figures 1: Original Ultrasound: non-encapsulating irregular hypoechoic masses and soft tissue edema categorized as a BIRADS 4.



Figures 2: Epithelioid granuloma 200x.



Figures 3: Mixed inflammatory infiltrate including epithelioid histiocytes 400X.

Discussion

Idiopathic granulomatous mastitis (IGM) is a diagnosis of exclusion lending itself to be a challenge to diagnose [3]. Due to its rarity, benign pathology, but inflammatory chronicity –it poses both a diagnostic and therapeutic hurdle as it can be confused with mastitis, breast abscess, and breast carcinoma on presentation [4,5]. The etiology of IGM is unclear [6]. IGM is most commonly found in premenopausal women after their last childbirth [4] or with oral contraceptive use [7,8]. The symptoms, diagnosis, and management require a multispecialty approach of several experts to avoid iatrogenic complications and assert the proper diagnosis [3].

Idiopathic granulomatous mastitis is a rare breast condition with prominent skin findings. It is typically seen in young parous women. Patient's presentations are most commonly local mastalgia accompanied by firm breast mass with possible skin ulcerations, abscesses, or fistulae development [4-6]. Idiopathic granulomatous mastitis may present in any part of the breast but tends to spare the subareolar regions [10]. Radiographic findings on ultrasound (US) are multiple irregular hypoechoic masses and collection with tubular connections with fingerlike aspects [11]. Fistulae formation to the skin in patients with a breast feeding history suggests IGM rather than carcinoma [11]. Mammographic features of IGM are focal symmetric density or mass with definable margin and a dense parenchymal pattern without any abnormalities [12-15]. Magnetic Resonance Imaging can delineate tissue contrast difference and the use of intravenous contrast further improves mass delineation [12]. Kocaoglu et al. [16] suggests that the use of time-signal intensity curves may support US and mammographic results with regards to the ability to distinguish between benign and malignant inflammatory breast disorders.

Histologic examination frequently demonstrates features of lobular non-caseating granulomas but not all cases have the typical non-caseating granulomas with epithelioid histiocytes, multinucleated giant cells, and predominant neutrophilic backgrounds with lymphocytes, plasma cells and eosinophiles of varying numbers without necrosis [17,18]. If caseating granulomas with necrosis and granulomas without a predominant neutrophilic background is present, it would preclude the diagnosis of IGM [18]. However, microabscess formation and fat necrosis with a negative blood and wound culture is supportive of the diagnosis of IGM [17,18]. A highly conservative pathologic feature of IGM is epithelioid histiocytes present within the smears [18]. Previous authors have

advocated for cytopathologist to be aware of the suspicion for IGM when attempting to make a diagnosis [17,18].

The treatment of IGM remains controversial. Traditionally IGM was treated by surgical excision of the mass, but some studies have suggested that the recurrence rate with surgical treatment is higher than with steroid treatment [2,16,19]. Recurrence rates of 5% to 50% are reported after surgical excision of mass [14,20]. An IGM free margin does have a reportedly lower recurrence rate [18,19]. Wide excisions with negative margins are better than limited excision alone, as there is a higher tendency to relapse after limited excision [8,21]. Furthermore, there is a high rate of fistula formation, poor wound healing and disfigurement after surgical intervention in patients with IGM [7]. Surgical excision can result in fistula formation, poor wound healing and cosmetic appearance [5]. A small number of case studies report mastectomies as treatment solutions for patients with complicated IGM [20,22,23].

Providers may opt to allow for close follow-up with or without immunosuppressive agents [5,20,24]. Eric et al. reported spontaneous resolution in 50% of cases of uncomplicated IGM in their study without any treatment with a mean interval of complete resolution of 14.5 months (range 2–24 months) [5]. Immunosuppressive agents, corticosteroids have been proposed as a treatment option but their success rate is variable. The use of corticosteroids was first proposed by Dehetrogh et al. [25] Several case series have also reported successful use of corticosteroids in the treatment of IGM [13,21-23,26,27]. Steroids should be started at a dose of 1 mg/kg per day and tapered slowly according to clinical response [23]. It may be necessary to continue high doses of steroid until the lesions completely resolve. Responses usually occur within weeks of treatment, but patients may need to be treated for several months [7,23]. About half of the cases relapse after stopping or decreasing the dose of steroids [22,23] and the efficacy in the initial and subsequent presentations needs to be further evaluated [25]. The use of steroid sparing agents such as methotrexate or azathioprine may facilitate tapering of steroids [21,22,28]. Due to the clinical presentation, antibiotic therapy is traditionally initiated but have no role in management of true cases of IGM [23].

Conclusion

Idiopathic granulomatous mastitis is a rare disorder of the breast, with an incidence of 2%, that may be encountered in a general practice, and it may mimic breast carcinoma or abscess. We report one case of IGM and describe the clinical presentation, diagnosis, evaluation and management of this disorder. The diagnosis of IGM should be considered in women who develop recurrent sterile breast abscesses and appropriate diagnostic evaluation should be performed on such cases. The available treatment options include close follow-up, immunosuppressive drugs, and surgical excision. Patients with uncomplicated IGM may be observed over time without treatment [7,11,20]. Antibiotics have no role in the management of true cases of IGM [24]. A retrospective review of cases seen over 25 years by Al-Khaffaf et al. showed that regardless of therapeutic intervention, the condition takes about 6 to 12 months to resolve completely. We are aware of only three cases that required complete mastectomy [11,12,22,24].

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