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Case report

**Idiopathic granulomatous mastitis masquerading as carcinoma of the breast: a case report and review of the literature**

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**Abstract**

**Background:** Idiopathic granulomatous mastitis is an uncommon, benign entity with a diagnosis of exclusion. The typical clinical presentation of idiopathic granulomatous mastitis often mimics infection or malignancy. As a result, histopathological confirmation of idiopathic granulomatous mastitis combined with exclusion of infection, malignancy and other causes of granulomatous disease is absolutely necessary.

**Case Presentation:** We present a case of a young woman with idiopathic granulomatous mastitis, initially mistaken for mastitis as well as breast carcinoma, and successfully treated with a course of corticosteroids.

**Conclusion:** There is no clear clinical consensus regarding the ideal therapeutic management of idiopathic granulomatous mastitis. Treatment options include expectant management with spontaneous remission, corticosteroid therapy, immunosuppressive agents and extensive surgery for refractory cases.

**Background**

Originally described by Kessler and Wolloch in 1972 [1], idiopathic granulomatous mastitis (IGM) is a rare disease of unknown etiology, which often mimics infection or malignancy and remains a diagnosis of exclusion. As a granulomatous form of lobular mastitis, IGM may be differentiated from granulomatous forms of periductal mastitis, as well as from granulomatous mastitis caused by sarcoidosis, Wegener's granulomatosis, giant cell arteritis, polyarteritis nodosum, foreign body reaction, and tuberculous, syphilitic, parasitic and mycotic infections [2,3]. Herein, we present a case of a young woman with IGM, initially mistaken for mastitis as well as breast carcinoma, and successfully treated with a course of corticosteroids.

**Case presentation**

A 36-year-old pre-menopausal Asian woman previously in excellent health, presented with a three-month history of a progressively worsening tender right breast lump, with associated ulceration, induration, and erythema, draining serosanguinous fluid. She denied systemic symptoms, as well as any known recent fungal or tuberculosis exposure. The patient is a G2 P2002 who nursed her children without difficulty. She has had no history of breast disease, and her past medical and surgical histories are otherwise...
unremarkable. She denied tobacco use, any family history of breast diseases, use of any medications including oral contraceptives, or any known drug allergies. Physical examination was otherwise unremarkable with no palpable lymphadenopathy.

During initial presentation to an outside hospital, mammography and breast ultrasound revealed an increased density and ductal dilatation of the right breast with associated edematous changes. All laboratory and other radiological studies were otherwise normal. Although malignancy had not been excluded, the patient received a presumptive diagnosis of mastitis and was treated with a 7-day course of amoxicillin and cefadroxil without improvement. This was followed by a course of cefdinir and gatifloxacin, which was complicated by an antibiotic-induced erythema nodosum. As a result, antibiotics were discontinued and the patient was scheduled for an open breast biopsy. Interestingly, the patient’s symptoms spontaneously resolved before tissue sampling was performed.

Two months after her initial presentation, a second similar lump appeared in the right breast (Figure 1). A repeat ultrasound of this area again demonstrated dilated ducts and debris. Bilateral breast magnetic resonance imaging (MRI) revealed regionally dilated ducts with prominent regional ductal and parenchymal plateau enhancement. Given these findings were not typical for mastitis, and clinical and radiological data could not exclude breast carcinoma, tissue sampling was performed. Microscopic examination revealed chronic inflammation and macrophage, giant histiocyte and epithelioid-like cellular infiltration, with cytologic features suggestive of a granulomatous process (Figure 2). Further histopathological analysis showed no evidence of carcinoma or abscess formation, and all cultures and stains for infectious organisms remained negative. Secondary to exclusion of malignancy, infection, and other causes, the patient was given the presumptive diagnosis of idiopathic granulomatous mastitis (IGM). She was treated with a 6-month tapered course of prednisone with an excellent response, and has since remained free from recurrence.

**Discussion**

Although the exact etiology of IGM remains unclear, associations with autoimmune disorders, oral contraceptive use, pregnancy, hyperprolactinemia and alpha-1-antitrypsin deficiency have been suggested [2,4-6]. Most studies report an average age of presentation in the third decade of life (range 11 to 83 years) with symptoms often developing within a few years of pregnancy [3]. Moreover, conflicting data exists regarding the significance of oral contraceptive use in patients diagnosed with IGM, with percentages of patients using contraception ranging from 0% to 33% [2,7,8]. However, no true associations with pregnancy, breast-feeding, prolactin levels, or oral contraceptive use have been established to date.

Interestingly, the majority of published reports of IGM have come from outside the U.S [9-13]. Whether this disproportional lack of published reports from the U.S. represents a lower prevalence of IGM, under diagnosis in the U.S., over diagnosis in developing countries, or a combination of these factors is uncertain.

**Figure 1**

A patient with IGM of the right breast status-post incisional breast biopsy. Clinical presentation of IGM may mimic common entities such as breast mastitis, as well as more involved diagnoses such as malignancy.

**Figure 2**

Photomicrograph of hematoxylin and eosin specimen from (1). Arrows indicate granulomatous inflammation centered on breast lobules, while arrowhead indicates the presence of multinucleated giant cells within non-caseating granulomatous inflammation (magnification 100×; bar = 150 µm).
nation of the above remains to be addressed, yet under-
scores the necessity of appropriate diagnosis and treat-
ment of IGM. Most frequently, the primary clinical
finding is a unilateral firm breast mass affecting any quad-
rant of the breast, which may be tender in 25% of cases
and present bilaterally in 25% of cases [2,3,14]. Patients
with a more chronic presentation may go on to develop
fistulae, abscesses, nipple inversion, and skin inflamma-
tion and ulceration over the course of several years [15].
In a recent study, Lai et al. reported that 100% of women
with a histopathologically confirmed diagnosis of IGM
initially presented with palpable breast masses [12].
Moreover, approximately 56% of women within this
study were initially suspected to have breast carcinoma
[12]. Given this initial presentation, combined with mam-
mography, ultrasound and fine needle aspiration biopsy
results that often mimic malignancy, several studies have
documented the use of unnecessary mastectomies in
patients with IGM [2,4,5]. Clearly, this underscores the
importance of thorough histopathological analyses in
patients with suspected breast carcinoma. A recent study
assessed the potential utility of dynamic contrast-
enhanced MRI in diagnosing IGM. Using patients with a
histopathologically confirmed diagnosis of IGM, Kocaoglu
et al. found varied appearances on MR and were unable to
identify any imaging features characteristic of the
disease, thereby suggesting its limited diagnostic util-
ity [16].

The predominant and characteristic histopathological fea-
ture of IGM, granuloma formation, is also commonly
found in other entities, thereby rendering it a diagnosis of
exclusion. The lobular distribution of IGM represents a
mixed chronic inflammatory process composed of lym-
phocytes, plasma cells, epithelioid histiocytes, multinu-
 cleated giant cells and less frequently, neutrophils [13,17].
However, confirmatory diagnosis is obtained only
through identification of granulomatous inflammation
centered on lobules (granulomatous lobulitis) with an
absence of caseating necrosis. In more severe cases, con-
fluence of the granulomatous inflammation may obliterate
its typical lobulocentric distribution, thereby further
complicating the diagnosis. Additionally, microabscess
formation may occasionally involve the entire lobule, and
squamous metaplasia of lobular and ductal epithelium
may occur [3,18,19]. Although IGM patients rarely
present with systemic signs of infection, culture speci-
 mens must nevertheless be analyzed for subtle microor-
 ganisms.

Likely secondary to the lack of published reports, particu-
larly from the U.S., there is no clear clinical consensus
regarding the ideal therapeutic management of IGM.
Although several studies have reported varying
approaches to the treatment of IGM, many of these treat-
ment algorithms were formulated without a definitive ini-
tial diagnosis. Histopathological confirmation of IGM
combined with exclusion of malignancy and other causes
of granulomatous disease is of utmost importance in
guiding clinical decision making and preventing inapprop-
riate and unnecessary treatments. Therefore, following
careful confirmation of diagnosis, the initial treatment for
IGM clearly should be non-operative. Indeed, in a recent
study, 50% of patients receiving expectant management
had spontaneous and complete resolution of disease fol-
lowing a mean of 14.5 months [12]. An initial treatment
option for patients with new-onset IGM with mild to
moderate symptoms may be expectant management with
close regular surveillance. For patients with clinically
advanced disease or more severe symptoms, in whom
infectious etiologies have been excluded, oral daily pred-
nisone, starting with 0.8 mg/kg/day and tapering with
clinical improvement, is a common regimen. Unfortu-
nately, aside from the well-established side-effects of cor-
ticosteroid therapy, patients often relapse with cessation
of therapy with one study reporting recurrence rates as
high as 50% [11]. In cases of recurrent disease or those
refractory to the above therapies, immunosuppressive
agents, like methotrexate or azothioprine, have been uti-
 lized with variable responses [9,20], but minimal clinical
evidence exists. Finally, surgical options should be
explored in refractory cases or those with persistent collec-
tions. A retrospective study by Erhan et al. reviewed 18
women with clinicopathologically confirmed IGM treated
with excisional biopsy [14]. According to the authors,
recurrence was seen in only 3 women, 2 of whom were
found to have hyperprolactinemia that was successfully
treated with repeat excision and anti-prolactinemic ther-
apy without subsequent recurrence. In cases with persist-
ent abscess and fistula, wide surgical excision and even
mastectomy may be indicated [2,10].

Conclusion
IGM is a rare and benign inflammatory process com-
monly mistaken for malignancy and other disease enti-
ties. As a result, it is often incorrectly treated. It typically
occurs in women during the third decade of life, present-
ing as an inflammatory palpable mass without systemic
symptoms. Correct diagnosis requires the exclusion of
infectious etiologies, other causes of granulomatous mas-
titis, and malignancy, combined with definitive his-
topathological confirmation. Treatment should initially
be non-operative and depending upon the severity of ini-
tial presentation, may range from expectant management
to corticosteroid therapy. In refractory cases, immuno-
 suppressive agents and surgical excision may be indicated.

Competing interests
The author(s) declare that they have no competing inter-
ests.
Authors’ contributions
RT performed chart review, literature search, and wrote and prepared the manuscript. BIO carried out histopathological analyses and review of the manuscript. JH treated the patient and reviewed the manuscript. ALR was the patient’s surgeon and helped write and edit the manuscript. All authors have read and approved the final manuscript.

Acknowledgements
AR has personally spoken to the patient and obtained consent to present the case and included photographs.

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