

Idiopathic granulomatous mastitis: a case report of breast abscess and review of literature

Mastite granulomatosa idiopática: caso clínico e revisão da literatura

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Abstract

Idiopathic granulomatous mastitis is a rare chronic breast inflammatory disease of unknown etiology, often mimetizing two other common diseases, breast abscess and breast carcinoma. We report a case of a 53-year-old woman with granulomatous mastitis, initially mistaken for infectious mastitis with an abscess. Surgery and systemic steroids were not enough and immunosuppression with methotrexate resulted in an improvement of symptoms.

Keywords: Breast abscess; Idiopathic granulomatous mastitis.

INTRODUCTION

Idiopathic granulomatous mastitis (IGM) is a rare idiopathic benign breast disorder, self-limiting, but with slow resolution: 14.5 months in average¹. It is more prevalent in developing countries, particularly in Asian and Hispanic women^{2,3}.

Clinical and imaging studies are nonspecific, often mimetizing breast abscess⁴ or carcinoma⁵. The definitive diagnosis is based on a histological result that reveals lobocentric non-caseating granulomas in which no microorganisms or features of other pathologic entities are identified².

No consensus exists on the best treatment modality. Surgical excision and steroid therapy are the preferred modalities.

We report a case of IGM, diagnosed after prolonged ineffective treatment of presumed infectious mastitis with abscess. After surgical excision and steroid therapy proved ineffective due to relapse, methotrexate resulted in improvement of symptoms. Recognising this condition is essential to prevent the morbidity caused by delayed diagnosis and misguided therapy.

CASE REPORT

A 53-year-old woman presented with two painless masses in the right breast. Physical examination revealed two unilateral, freely mobile, well-defined and non-tender right breast masses: one with 60 mm at the inner upper quadrant (IUQ) - lump 1; and other with 20 mm at the transition of the lower quadrants (LQ) - lump 2. Lump 1 appeared seven months before and corresponded to a hyperechoic nodular mass that had increased from 10 to 60 mm in this period. There was no discharge, nipple inversion, overlying breast skin changes or palpable axillary lymphadenopathy. Remainder of her physical examination was normal.

The patient denied systemic symptoms and any known fungus exposure. She had pleuropulmonary tuberculosis at age 16, gestational history of G2P2 and previous hysterectomy with bilateral adnexectomy for fibroids six years before. Her medical and family history was otherwise unremarkable except taking oral contraceptive (OC) for six years (from 24 to 30 years old).

Breast ultrasound images revealed a nodular, hypoechoic and predominantly cystic, 45 mm diameter image, not vascularized (lump 1); and a second nodular image, solid with cystic images inside, 25 mm diameter, regular borders and slight posterior acoustic shadowing (lump 2). No axilar adenomegaly were visualized.

All laboratory exams including blood count, bio-

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chemistry, erythrocyte sedimentation rate, CEA and viral markers were normal, except the CA15.3 level that was 39.91 U/ml (normal: < 35 U/ml). Thorax and skeletal radiography were normal.

The patient received a presumptive diagnosis of mammary abscess and was oriented to surgical aspiration, with resolution after drainage of 60 cc of purulent liquid. Amoxicilin was empirically prescribed (500 mg three times/day) and discontinued when the culture revealed no evidence of bacterial or mycobacterial growth. The pathology showed the presence of infected cystic areas with necrotic material, histiocytes and neutrophils.

In the following month, a relapse occurred with appearance in the right breast of a tender mass, swelling in the IUQ and erythematous changes around the lesion and, in spite of two surgical drainages and two separated courses of antibiotics (flucloxacillin and amoxicillin) and anti-inflammatory (ibuprofen), the mass relapsed two other times. Breast ultrasound identified two heterogeneously hypoechoic collections suggestive of abscesses in the IUQ and inner LQ (57 mm and 37 mm, respectively – Figure 1) corresponding to two medium density nodular formations with well-defined contours on mammography (Figure 2). No microcalcification or adenomegaly in right axilla were evident.

An excisional biopsy was performed and the histopathological analysis showed features of granulomatous mastitis: chronic lobulocentric noncaseous granulomas composed of clustered epithelioid histiocytes. Culture of tissue failed to grow any pathogens or

mycobacteria. No features of other pathologic entities (like vasculitis) were identified. Further evaluation for autoimmune, connective tissue and infectious granulomatous disease was negative, including stains and cultures for fungi and acid-fast bacillus. There was no evidence of carcinoma and serum tumour markers were normal (CA125, CA 17.5) or normalized after six months (CA15.3). Initially elevated angiotensin-converting enzyme levels (98 U/L; N: 20-70) normalized after two months to 28 U/L. Thorax-abdominal-pelvic computed tomography and cephalic MRI revealed no evidence of other granulomas, but scintigraphy revealed mild to severe hyperactivity in two areas of the right breast.

Considering the patient's clinical history and all the laboratory findings, the diagnosis was interpreted as IGM.

After the last surgery there was a persistent drainage of a serous exudate at the scar and, two months later, a nodule was detected in IUQ of the right breast measuring 23x13 mm, accompanied by a long hypoechoic tubular extension into the skin and reactive ipsilateral lymphadenopathy (Figure 3).

The patient was started on 30 mg of deflazacort daily with good initial clinical response: discharge decreased and no masses were palpable after eight weeks. At the eleventh month of therapy, an asymptomatic recurrence in her right breast was detected by MRI (gra-



FIGURE 1. Breast ultrasound: two heterogeneously hypoechoic collections suggestive of abscesses

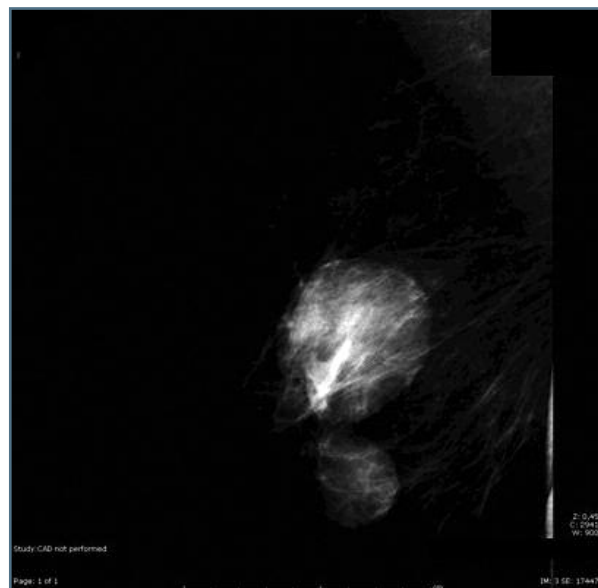


FIGURE 2. Mammography: two medium density and well-defined nodular formations

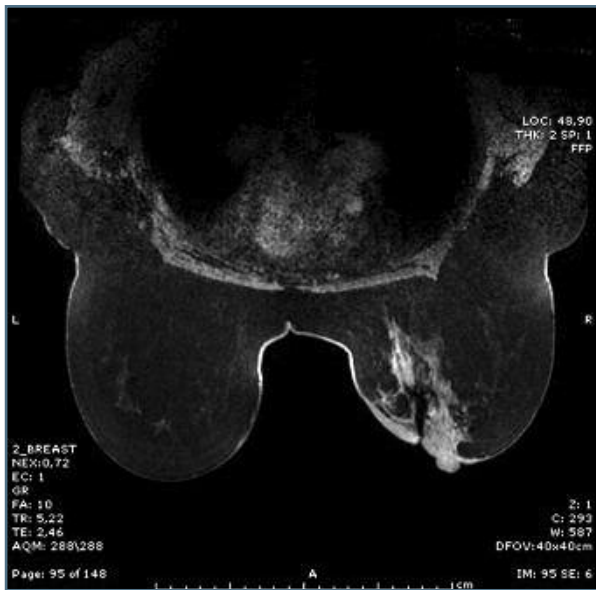


FIGURE 3. Breast magnetic resonance image: heterogeneous hiperdensification with extension to the skin, compatible with mastitis

nulomatous change), followed by sero-hematic discharge at the fourteenth month and appearance of a 50 mm tender mass with superficial erythema at the seventeenth month, despite steroids dose increase (45mg at thirteenth month and 60mg at seventeenth month). Breast ultrasound at eighteenth month identified two heterogeneously hypoechoic fluid collections with 50 and 30 mm extending up to the nipple in the IUQ and LQ, respectively. At this time, methotrexate (10mg/week) was added, deflazacort tapered to 30 mg/day over the next month and tapered off completely after five months.

Methotrexate therapy was continued for seven months with progressive shrinking of the breast mass. At the end of therapy, no mass was palpable and the breast tissue was soft, without erythema or fistulae.

DISCUSSION

IGM is an exceedingly rare diagnosis with approximately 200 cases reported in the literature during the past three decades. There is no clear etiology; possible etiologic factors include autoimmune response to extravasated secretions from lobules due to trauma, infections or chemical irritation^{6,7}; hyperprolactinemia⁵; diabetes mellitus; smoking and lack of alpha-1 antitrypsin^{7,8}.

This disease most commonly affects women of child-bearing age^{6,9}, with a history of pregnancy and lactation^{6,10} six to fifteen years before^{5,6}. In the present case, this was not found: the patient was 53-year-old, having delivered her last child 26 years before, and with no previous breast-feeding.

Conflicting data exists regarding the significance of OC use in IGM, with percentages of use ranging from 0% to 33%^{7,11}. Although our patient had used OC, that occurred for a limited period of time and more than twenty years ago. Besides that, the response to immunosuppressive therapy implies an autoimmune process.

Five months elapsed between the first consultation and diagnosis, with three therapeutic fluid aspirations and three courses of antibiotics cycles performed. These findings are in keeping with reports in the literature that IGM can mimic breast abscesses⁴, since the physical exam and radiological findings are similar. This should alert us to consider this disease when a presumed abscess is being unresponsive to usual therapy.

Our diagnosis was performed through a histological finding and exclusion of other known causes of granulomatous diseases.

Treatment of IGM depends on the severity of the disease but treatment protocol is not well established^{5,12}. In this case, wide surgical excision and immunosuppression were the primary treatment modalities.

Although surgical wide excision was useful in providing an exact diagnosis, it was complicated by cutaneous fistula and recurrence. In fact, the literature points to a high rate of fistula formation⁵ and a recurrence rate of 5.5-50%³. In complicated and resistant cases, steroids should be administered after excision: prednisolone 30-60 mg daily is recommended for at least 6 weeks, with improvement in 77% of the cases⁹ but with 50% of recurrences after dose reduction or discontinuation¹³. In case of resistance to corticosteroids, a second cycle or replacement by immunosuppression can be tried (methotrexate: 10-15 mg/week in the first six months and 20 mg/week after)¹⁴. Our patient required deflazacort administration (30 mg daily) during the first ten months for stabilization of the disease. Afterwards, there was worsening of the disease, resistant to increasing the dose of steroids, and methotrexate therapy was required for complete resolution of the disease.

Methotrexate has the advantage of controlling the inflammatory process and thus preventing further complications without the complications of steroid

therapy.

Other therapeutic approaches have more limited indications: antibiotic should be prescribed only in case of cellulitis, abscess or open draining sinuses¹⁰; and total mastectomy is the latest approach in resistant cases⁵. Cultures of all aspirate and biopsy samples should be obtained. In this case, cultures proved negative except one culture that grew a small number of colonies of *Staphylococcus epidermidis* (one month after wide surgical excision), treated with cotrimoxazole.

A retrospective review of cases seen over 25 years by Al-Khaffaf et al. showed that regardless of therapeutic intervention, the condition takes about six to twelve months to completely resolve⁸.

Because of the high frequency of recurrence a close surveillance is recommended.

CONCLUSION

To prevent morbidity, IGM should be considered in the differential diagnosis when presumed infectious mastitis with breast abscess does not respond to usual treatment. Immunosuppression with methotrexate is a therapeutic weapon in complicated and persistent cases after failure of surgery and steroids treatment.

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