Rare disease

Idiopathic granulomatous mastitis: rare but important

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Summary

A 61-year-old woman attended the breast clinic with unresolving mastitis and an associated mass, following failed treatment with antibiotics. Triple assessment confirmed idiopathic granulomatous mastitis. Unresponsive to further conservative management and steroid therapy, she underwent surgical excision and made uneventful recovery. No evidence of recurrence was detected at 18 months follow-up.

BACKGROUND

Idiopathic granulomatous mastitis (IGM) is a rare, benign, condition simulating breast cancer. It remains a diagnosis of exclusion, dependent both on a characteristic histological pattern and exclusion of other possible causes. Aetiology and appropriate treatment remain controversial, and the aim is to review current literature on the topic and management recommendations.

CASE PRESENTATION

A 61-year-old woman was urgently referred to our breast clinic with a 3-week history of mastitis of the left breast without systemic symptoms and not responding to antibiotics. Her past medical history included a total hysterectomy and bilateral salpingo-oophorectomy for uterine fibroids and mild asthma. She was a non-smoker and had taken the contraceptive pill and hormone replacement therapy for more than 10 years. There was a positive family history of breast cancer with her daughter diagnosed at age 37 and a cousin at age 50.

Clinical examination revealed a large (7×5cm), erythematous, hard, poorly mobile mass as demonstrated in figures 1 and 2. The mass had all the hallmarks of a malignant tumour, and there was an associated palpable axillary lymph node.

INVESTIGATIONS

On initial assessment both mammogram and ultrasound were normal. Multiple core biopsies were then taken on two separate occasions (1-week interval). The first sample showed only inflammatory changes, and the second sample showed similar changes with the presence of non-caseating granuloma with no evidence of foreign material, fungal or bacterial organisms. There was no evidence of ductal carcinoma in situ or invasive malignant disease. Baseline haematological and biochemical investigations were normal, except for a mildly raised erythrocyte sedimentation rate. Additional tests (serum angiotensin converting enzyme, protein profile, complement profile and rheumatoid factor) were all normal, revealing no immune deficiency.

DIFFERENTIAL DIAGNOSIS

Prior to triple assessment, the differential diagnosis was that of malignant breast carcinoma.

TREATMENT

A diagnosis of granulomatous mastitis was made, and a trial course of prednisolone (10 mg 4 times a day) was advocated. Subsequent follow-up at 2 weeks revealed early signs of fungation with skin necrosis. A simple mastectomy was performed, and histology revealed a mixed inflammatory infiltrate, abscess formation and granulomatous inflammation. There was no evidence of acid fast bacilli or fungi. Diagnosis of IGM was confirmed.

OUTCOME AND FOLLOW-UP

Post-operative recovery was uneventful, and follow-up at 18 months revealed no evidence of recurrence.

DISCUSSION

First described as a separate entity in 1972 by Keller and Wolloch, IGM is, as the name suggests, an inflammatory lesion of unknown aetiology.^{1 2} It is a rare condition, with some institutions seeing only 15–18 cases in a 25-year period. More than 120 cases have been documented and published in the literature worldwide, with little progress made in the past nearly 4 decades in terms of understanding the aetiology and determining the ideal course of treatment.

There has been one case of IGM documented in a male patient; however, most cases of IGM documented are in females with the mean age of 32–36.^{1 3 4} Typically, the patients are of reproductive age presenting commonly during lactation, pregnancy or within 5–6 years from last pregnancy, but the condition can develop in postmenopausal women more than 20 years postpartum as seen in this case.^{1 4–6} It has also been reported in nulliparous women.

Patients are usually referred with lesions suspicious of underlying malignancy. Inflammatory breast changes, palpable (tender or painless) breast mass, nipple discharge, and skin distortion are all common presenting features that mimic breast neoplasm. Clinically, there is little in way of differentiating benign from malignant disease. Diagnosis relies on radiological, but more important, histological findings. It is still a condition diagnosed by exclusion of inflammatory breast malignancy, and of secondary granulomatous mastitis due to infectious causes (eg, *Mycobacterium tuberculosis*, Blastomycosis, Cryptococcosis), autoimmune conditions (eg, Wegner's granulomatosis, giant cell arteritis, foreign body reaction) and Sarcoidosis.

Figure 1

Figure 2

Figures 1 and 2 A large erythematous, hard, poorly mobile mass with all the hallmarks of a malignant tumour.

All of these conditions can present with or be associated with granulomatous mastitis.

Since its recognition, many authors have looked vigorously for distinctive hallmarks that indicate the diagnosis of IGM, and there are some clinical and radiological features that are consistent throughout all the published cases. Mammographic findings characteristically show asymmetrical density in breast tissue more prominent when compared to the contralateral normal breast. Ill-defined

mass effects have been seen, with no other abnormality in a few cases; but, on the whole, there is no evidence of microcalcification reported in all the cases available in the literature.¹⁻⁷

Ultrasonography usually reveals ill-defined hypoechoic lesions that are difficult to measure, showing features of benign disease. However, in the report by Erhan *et al*⁶, 7 of their 15 cases had radiological findings suggestive of malignancy and intraductal papilloma which was histologically

disproven and IGM was diagnosed. MRI has been tried by Schelfout *et al*⁶ showing focal irregular enhancing masses compatible with matitis and malignancy and therefore not valuable in diagnosis. MRI cannot differentiate active inflammatory process from tumorous process but may have a role in determining disease regression over time and may be useful for assessing resolution. The true value of this imaging modality is yet uncertain.

Typical histological patterns are that of lobular distribution of non-caseating granulomas often but not always with the presence of microabscesses.^{1 4 9} In search for plausible aetiology, associations with an autoimmune origin, diabetes, infection, trauma and hyperprolactinaemia have been suggested, but there is no consensus among the numerous authors that have reported on the condition.^{1 4 5} Some authors favour the association with autoimmune disease due to the similarities of IGM with granulomatous thyroiditis.^{6 10} Pourbagher *et al*¹ reported on four cases of IGM with a diagnosis of diabetes mellitus (DM) made 2–4 years prior to presentation of IGM. Whether the connection is a coincidence or DM does predispose to IGM is still uncertain, with no evidence in the literature to support or dispute the theory. Hyperprolactinaemia has been associated with prolonged time from presentation to complete disease resolution, but once again there is insufficient evidence to prove any significant link. Interestingly, Al khaffaf *et al*⁴ noted a higher incidence in the non-Caucasian races and no correlation between IGM and the use of oral contraception as previously suggested. Their study group is small and, therefore, further studies need to be carried out for more reliable and transferable data.

Although self-limiting, the management of IGM remains a challenge. As the condition tends to have a prolonged course and can take up to 109 weeks to fully resolve, it is often poorly tolerated.⁴ It is documented that up to 50% of cases achieve uneventful recovery and the rest are complicated by ipsilateral and/or contralateral recurrence, delayed healing, abscess formation, fistulas and secondary infection.⁸ The importance of the awareness of IGM by surgeons, histopathologists and radiologists is to better manage patients with IGM and avoid unnecessary mastectomies where appropriate. The literature shows that surgical excision alone or in adjunct with steroid therapy is the most commonly adopted approach for the treatment of IGM.^{1 11} Antibiotics have also been used, but Al khaffaf *et al*⁴ demonstrated in their observations that overall outcome was independent of the combination of treatment options.

Some authors argue that with a definitive diagnosis, a non-surgical approach is the ideal treatment option, performing mini incision and drainage of abscesses only when required. The idea is to allow the disease to burn out with time. This has the added advantage of minimal scarring and disfigurement from surgery.^{4 6} Conservative treatment is,

however, not always possible as fine-needle aspiration and core biopsies are not always sufficient to differentiate IGM from malignancy.⁶ Other authors have suggested all patients undergo surgical excision, in adjunct to treatment with steroids, if they present with coexisting conditions such as diabetes and hyperprolactinaemia.

Learning points

- ▶ In conclusion the lesson to be learnt from decades of observation is being aware of the condition and making the correct diagnosis in attempt to provide the most appropriate treatment and avoid unnecessary surgery.
- ▶ It should be considered as part of the differential diagnosis and a definitive diagnosis is essential via the triple assessment.
- ▶ There is no clear clinical consensus regarding the ideal therapeutic management of IGM.
- ▶ Each individual and disease process can vary, and, therefore, with the correct diagnosis conservative versus surgical treatment can be offered to the patient satisfactorily.
- ▶ Further studies need to look into the loose associations made with IGM, in order to understand the condition and thus be able to offer appropriate care.

Competing interests None.

Patient consent Obtained.

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