

The treatment conundrum that is idiopathic granulomatous mastitis

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Mastitis—inflammation of breast tissue—is a benign, yet potentially debilitating condition that affects women of childbearing age, and its aetiology is usually infectious or autoimmune.

Judging from multiple publications in recent years, there has been increased interest in mastitis of autoimmune origin, of which idiopathic granulomatous mastitis (IGM) is a subset. However, evidence regarding the best treatment approach is still lacking as most reports are retrospective and hence skewed towards treatment bias of the service where patients are seen and managed. Despite this, no hard data regarding the incidence and prevalence of IGM as a medical condition are available from existing literature.

It is important to remember that IGM is a diagnosis of exclusion, and all other causes—such as tuberculosis, sarcoidosis, autoimmune conditions (Behçet's disease, Sjögren's syndrome and systemic lupus erythematosus), granulomatosis with polyangiitis (polyglandular autoimmune syndrome [PGA], formerly Wegener's granulomatosis), eosinophilic PGA (formerly Churg-Strauss syndrome), Crohn's disease, diabetic mastopathy and mammary duct ectasia—should be considered before starting treatment.¹ Needless to say, breast cancer should always be considered as one of the differentials.

IGM by definition is of unknown aetiology. However, based on its response to treatment (corticosteroids and immunosuppressants), and considering the postulated “known” triggers for the granulomatous response (contraceptive pills, hyperprolactinaemia, *Corynebacterium* infection, recent trauma and smoking) that gives rise to its symptoms, an autoimmune origin is the favoured underlying cause for now.¹

The disease is benign, but can be locally aggressive, potentially causing widespread destruction of the breast, leaving patients with permanent scars. Scarring could be the result of severe inflammation that leads to sinus formation, skin ulceration, and cosmetic deformities from tissue loss. The condition is eventually self-limiting, burning out in the majority of patients

within 1–2 years of onset.^{2,3} However, a significant minority of patients do have a protracted course and suffer multiple relapses, which significantly impair quality of life⁴ and leave a scarred, deformed breast that serves as a permanent reminder of the traumatic period in their lives.

Coupled with its symptoms and signs at presentation (hard, irregular masses, lymphadenopathy and irregular hypoechoic densities on ultrasound scans),⁴ IGM may cause patients and their physicians continuous anxiety over the possibility of an underlying carcinoma, until there is complete resolution of the disease with no visible lesions on imaging. IGM commonly presents as a mass in the breast in mostly pre-menopausal women, and occasionally may be associated with pain, erythema and swelling.⁵ Pus and abscess formation may be part of the acute presentation, whereas cases that present late often have mammary fistulae and ulceration of the skin on physical examination.⁶

The wide spectrum of therapeutic modalities recommended for this condition reflects the confusion concerning its aetiology, and can range from observation to prolonged use of antibiotics or immunosuppressants, and surgery (with certain patients opting for mastectomy to rid themselves of this disease).^{4,6}

A recent study by Tang et al. in this issue of the *Annals* provides additional Singapore data regarding IGM in women.⁷ Consistent with international evidence, the condition affects women with a history of breastfeeding in their pre-menopausal years. A positive *Corynebacterium* culture was found in a subset of patients and if present, symptoms were successfully abrogated with antibiotic therapy. However, while no modality of treatment offered superior outcomes, smoking was shown to increase relapse rates.

Thus, in light of the current literature, the following algorithm can be considered. Heightened awareness of IGM is necessary, and early core biopsy (fine needle aspiration is not diagnostic for this condition) of all masses should be performed. This not only excludes malignancy as a cause, but also provides histological

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proof of granulomatosis, which would then trigger the consideration of IGM as a diagnosis. Upon histological confirmation of the condition, patients can be offered observation (mild cases that present as a mass only, without any hallmarks of active inflammation, and patients are educated to detect when these become significant); a short course of steroids (high dose initially for control, and then tapering down as the disease resolves), unless there are active signs of infection (pus formation) that may necessitate antibiotic therapy; and/or surgical intervention. Patients should be reviewed regularly early on, and cases that are refractory to steroids can be referred to rheumatologists for consideration of immunosuppressant therapy (methotrexate or azathioprine) to avoid complications associated with long-term steroid use. The difficulty lies in discerning which patients require what treatment. Over-treatment could result in unwanted and unnecessary prolonged exposure to steroids, while late or inadequate treatment would fail to control the condition sufficiently and result in progressive scarring and deformity of the breast.

In any case, the principles of treatment would be to remove the triggering condition (such as oral contraceptive pill or infection) where possible, and to

suppress the over-exuberant immune response (steroids or immunosuppressive therapy) once the initiating trigger has been controlled.

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