

# Is idiopathic granulomatous mastitis a surgical disease? The jury is still out

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**Abstract:** Idiopathic granulomatous mastitis (IGM), is a rare entity of chronic inflammatory disorder of the breast of unknown etiology. Very few cases have been described so far, almost exclusively in women. Here we describe a case of IGM in a 53-year-old man presented with a right breast mass, progressively enlarging during the last 6 months. Due to the findings of clinical examination and CT-scan, the suspicion for a potentially malignant lesion was given and the decision for surgical resection was made. Microscopic analysis of the specimen showed non-caseating granulomas around mammary lobules, findings compatible with IGM. The patient is recurrence-free at 18-month follow-up. IGM is a rare benign inflammatory breast disease, usually seen in females of reproductive age. Establishing a diagnosis can be challenging for a surgeon and requires a high index of suspicion as most patients are initially misdiagnosed by their primary care physicians. Steroids and immunosuppressive drugs are considered as fundamental treatment modalities but they are correlated with increased rates of disease response and recurrence. On the contrary, surgical resection demonstrated significantly superior results compared to steroid-alone treatment in terms of recurrence and post-treatment recovery.

**Keywords:** Idiopathic; granulomatous; mastitis; breast; lesion; treatment; surgery

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## Introduction

Idiopathic Granulomatous Mastitis (IGM) is a rare entity of chronic inflammatory disorder of the breast, usually misdiagnosed clinically and radiologically as breast carcinoma or breast abscess (1). Benign lesions of the breast are extremely uncommon in male patients. Only 2 cases of IGM in male patients have been previously described in the literature (1-3). Because of its uncommon etiology and rareness, diagnosis and treatment is still a challenge. The aim of this article is to report a rare case of IGM on a male patient, along with discussion on current debates of the management of this rare entity.

## Case presentation

A 53-year-old male patient presented with a visible mass

occupying the upper half of his right breast (*Figure 1*). The patient was generally in good health without history of autoimmune disease or recent infection. He first noticed the current lesion 6 months prior to his admission at our Department. During this 6-month period, the mass was constantly growing causing progressively increased discomfort and limitation of his right arm movements. Clinical examination revealed a 5 cm, fixed and sensitive to palpation, lump. Computed Tomography (CT) scan with intravenous contrast confirmed the presence of the lesion extending to pectoralis major muscle, measuring 6.3 cm × 3.7 cm × 3.1 cm (*Figure 2A*).

Due to the findings of clinical examination and CT-scan, the suspicion for a potentially malignant lesion was given and the decision for surgical resection was made.



**Figure 1** Macroscopic view of a mass occupying the upper half of right breast.

Under general anesthesia a transverse incision across the chest wall was performed. The lesion found to have a well-defined hard margin and was easily dissected from the surrounding peri-mammary tissues. It was removed intact in a single specimen with macroscopically clear margins. Microscopic analysis showed non-caseating granulomas around mammary lobules, along with a clear space rimmed by neutrophils at the center of the granuloma (*Figure 2B*), findings compatible with IGM. The postoperative course was sound and the patient was referred to rheumatologists for thorough diagnostic work-up for autoimmune diseases that turned out to be negative. The patient is recurrence-free at 18-month follow-up.

## Discussion

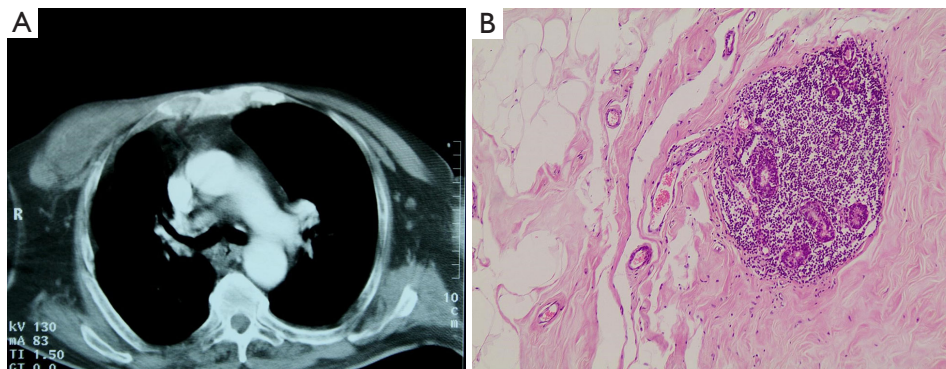
IGM is a rare benign inflammatory breast disease, usually seen in females of reproductive age, with around 200 cases being reported in the literature so far (2,4). Beyond clinical examination and imaging, establishing a diagnosis can be challenging for a surgeon and requires a high index of suspicion with exclusion of infective and autoimmune breast diseases, as most patients are initially misdiagnosed by their primary care physicians, leading to diagnostic confusion and heightened anxiety (1,5). Core biopsy is diagnostic only in around 70% of these patients (1). Although several triggers have been proposed for the development of IGM, the etiologic association of neither of them has been documented. Three main hypotheses about the possible causes of IGM have been suggested, including autoimmune response, infectious disease, and hormonal disruption (1,2,6-9).

There is an ongoing debate in the literature whether IGM is a surgical disease or not (6,7,10,11). The majority of the publications on the subject originate from Mediterranean countries (Turkey, Egypt) as well as countries from the Middle East (1,2,6,10,12,13). Since the disease is more common in women and its pathogenesis seems to be linked to autoimmunity, steroids as well as immunosuppressive drugs are considered as fundamental treatment modalities (14,15). It is commonplace among relevant literature that conservative (medical) treatment is correlated with increased rates of disease response or resolution (around 75%) but also high early disease recurrence (around 30%) (16). Around 40% of the patients under steroids develop side effects, recurrence or persistence of symptoms under steroid treatment and mandate immunosuppressive treatment (6,10,16). On the contrary, studies that included surgical resection as a first-line treatment demonstrated significantly superior results compared to steroid-alone treatment in terms of recurrence and post-treatment recovery (6 months in conservative *vs.* 1 month in surgery;  $P < 0.009$ ) (6,10).

Our case is a very uncommon presentation of IGM since the lesion was extremely bulky (6 cm) and it was developed in a male patient. These findings combined with the imaging results raised the suspicion of a malignant tumor. This was the reason why surgeons decided to proceed to surgical resection with curative intent. Even if the lesion turned out to be benign, the fact that the patient had no recurrence after 18 months, which is the longest follow-up reported in males with IGM, indicates the importance of surgical resection in these patients, no matter whether they will be offered steroid treatment or not.

## Conclusions

All in all, IGM is an extremely rare disease with challenging diagnostic approach and not established treatment in terms of efficacy. It mandates high clinical suspicion, histological confirmation, close monitoring, exclusion of underlying systemic autoimmune conditions and judicious use of steroids, if needed. Even if the jury is still out about the appropriate treatment, lessons learned from our case and from the main body of current literature, indicate that surgical resection should be considered as main treatment modality of curative intent. Despite the difficulty of designing large studies due to its rarity, future comparative studies could shed some light on the debate.



**Figure 2** Evaluation of the lesion. (A) Computed tomography (CT) presenting a lesion measuring 6.3 cm × 3.7 cm × 3.1 cm, extending to the pectoralis major muscle; (B) lobular unit showing dense intralobular and perilobular lymphocytic infiltration, associated with mild lobular atrophy and sclerosis (HE, ×10).

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### Footnote

*Conflicts of Interest:* The authors have no conflicts of interest to declare.

*Informed Consent:* Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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