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Review Article

Idiopathic Granulomatous Mastitis: A Comprehensive Review

Namrata Mahansaria*

1 Consultant Breast oncoplastic surgeon, Centre for Breast disease and Cancer, RINCHI Trust Hospital, Ranchi, Jharkhand

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***Correspondence:**
Dr Namrata Mahansaria

ABSTRACT

Idiopathic granulomatous mastitis (IGM) is a rare chronic inflammatory breast disease. IGM poses diagnostic and therapeutic challenges due to its complex clinical presentation and poorly understood etiology. Although auto immunity has been blamed for its etiology, it has not been fully elucidated. Itshistopathology is characterized by non-caseating granulomas. IGM usually presents as tender mass in the breast, mimicking inflammatory malignancy. This review aims to summarise current knowledge on the epidemiology, pathogenesis, clinical features, diagnostic approach, and management strategies of IGM.

Keywords: Idiopathic Granulomatous Mastitis, Inflammatory Breast Disease, Corticosteroids, Intralesional steroid Injections.

INTRODUCTION

Idiopathic granulomatous mastitis (IGM) is a rare form of non-lactational mastitis, presenting as a chronic inflammatory lesion [1,2]. It was described first by Kessler and Wolloch in 1972 [1]. This inflammatory condition although benign is a challenging disease to treat. It often presenting with non-specific symptoms such as breast pain, swelling, and palpable mass. Despite its benign nature, the differential diagnosis includes malignancy, necessitating a thorough diagnostic workup. This review explores the various facets of IGM to aid in accurate diagnosis and management.

Epidemiology

IGM primarily affects women of childbearing age, typically between the third and fourth decades of life[6]. Although rare, its incidence appears to be rising, possibly due to increased awareness and improved diagnostic techniques. Geographical variations in prevalence have also been noted, suggesting potential genetic or environmental factors. It has high prevalence in the Asian countries, specially South East Asia including India.

Pathogenesis

The pathogenesis of IGM remains unclear, with several proposed mechanisms including autoimmune dysregulation, and hormonal influences. [1,6,7]. Autoimmune response is currently the most accepted theory, supported by histopathological findings of non-caseating granulomas around breast lobules and ducts [2]. Hormonal influence is another very important theory. Most patients are women of childbearing age with a history of pregnancy and lactation

Histopathologically, IGM is characterized by noncaseating granulomas around lobules and ducts in the breast, characterized by necrotizing chronic granulomatous lobulitis and abscess formation.

In Indian scenario, it is extremely important to rule out Tuberculosis as a cause of the granuloma. Other causes of granuloma specially infection, trauma, and foreign body reactions should also be excluded

Clinical Features

Clinical presentation varies widely, ranging from mild breast discomfort to large, ulcerating masses. The most common symptoms include unilateral breast pain, erythema, and the formation of firm, irregular mass[7]. The erythema and pain in presence of breast lump often raise suspicion of inflammatory breast carcinoma[8] The chronic and relapsing nature of IGM can lead to multiple discharging sinus causing significant morbidity and affect quality of life. Definitive diagnosis requires histopathological examination, typically achieved through core-needle biopsy to confirm the presence of granulomatous inflammation. This diagnostic step is crucial to differentiate IGM from malignancy and guide appropriate management.

Management- Diagnosis

Diagnosing idiopathic granulomatous mastitis (IGM) presents a significant challenge due to its rarity and diverse clinical presentation, often mimicking malignancy. The absence of consensus on optimal management underscores the complexity of this condition, characterized by its chjronic nature and propensity for recurrence.

The diagnostic approach begins with a high index of clinical suspicion for IGM. A thorough initial history is crucial to exclude other causes of granulomatous infections, prevalent in the Indian context such as tuberculosis, trauma, and autoimmune conditions like rheumatoid arthritis, including diseases associated with ANA positivity.

Initial imaging typically involves ultrasound (USG) of the breast and/or mammography to evaluate the extent and characteristics of the lesion. USG aids in detecting potential abscess formation, while mammography often categorizes findings under BIRADS 4a due to similarities with breast malignancy[9,10]. Magnetic resonance imaging (MRI) of the breast is generally not recommended as it does not significantly enhance diagnostic clarity in cases of IGM.

Definitive diagnosis relies on histopathological examination via core biopsy of the lesion. Histopathologic features of non-necrotizing

granulomatous inflammation must be confirmed on histopathological examination (HPE). Acid-fast bacilli (AFB) staining should be performed to rule out tuberculosis. In cases of abscess formation, pus should be sent for culture and sensitivity (C/S) testing and Cartridge Based Nucleic Acid Amplification Test (CBNAAT) for tuberculosis.

Treatment

Surgical excision was historically employed in managing idiopathic granulomatous mastitis (IGM) during the 1980s. However, current clinical practice generally avoids surgical interventions due to challenges such as recurrent discharging sinuses, high recurrence rates, delayed wound healing, and suboptimal cosmetic outcomes [1]. Surgical approaches are now reserved for resistant cases, with complete duct excision emerging as a discussed option gaining popularity among patients with refractory IGM.

Conservative Management

Conservative approaches are preferred, starting with antibiotics followed by oral corticosteroids or immune modulators like Methotrexate or azathioprine. Corticosteroids, introduced in the treatment paradigm by DeHertogh et al. in 1980 [3], remain the cornerstone of medical therapy. Typically initiated at high doses and tapered over weeks, corticosteroids are effective but limited by systemic side effects, prompting exploration of alternative therapies such as intralesional steroid injections [12, 13].

Intralesional Steroid Injections

Intralesional steroid injections have emerged as a novel therapeutic modality for IGM in recent years.[11,12] This targeted approach delivers corticosteroids directly to affected breast tissue, potentially reducing systemic side effects while effectively controlling inflammation and minimizing lesion size . Although data in the literature is limited, initial studies indicate promising outcomes in terms of symptom relief and lesion regression, necessitating further investigation to establish their efficacy and safety profile.

Additional Therapeutic Options

Combining low-dose oral steroids with intralesional injections represents another promising treatment strategy. However, larger prospective studies are needed to validate these novel approaches.

Immune Modulators

Immune modulators like methotrexate and azathioprine have been explored in IGM management, yet their success remains limited.[10]

Multidisciplinary Care

Patients with refractory IGM require interdisciplinary management involving Rheumatology consultations to optimize treatment outcomes.[10]

Prognosis and Complications:

Although benign, IGM can result in significant morbidity and complications such as abscess formation, skin ulceration, and recurrent infections. Long-term outcomes are generally favorable with appropriate treatment, although recurrence rates remain high. Close monitoring and prompt intervention are crucial in minimizing complications and optimizing patient outcomes.

Conclusion: Idiopathic granulomatous mastitis remains a challenging diagnosis requiring a multidisciplinary approach for accurate management. Advances in diagnostic techniques and evolving treatment strategies, including the use of intralesional steroid injections, offer promising avenues for improving outcomes and quality of life for affected individuals. Continued research is essential to unravel the complex pathophysiology of IGM and to establish evidence-based guidelines for its optimal management.

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Author Affiliation

Consultant Breast oncoplastic surgeon

Centre for Breast disease and Cancer

RINCHI Trust Hospital, Ranchi, Jharkhand.