Idiopathic Granulomatous Mastitis

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Abstract

Idiopathic Granulomatous Mastitis (IGM) is an uncommon benign disorder of the breast that can mimic two frequent breast disorders, breast carcinoma and breast abscess. It is frequently under-diagnosed and poorly managed, with many health professionals unaware of the condition. Timely diagnosis with a clear understanding of the various complications is necessary. IGM usually affects women of childbearing age breastfeeding within the previous 5 years; IGM is a disorder that should be considered in the evaluation of women who present with painful breast disease. Primary health care physicians play a vital role in the identification, diagnosis, referral and long-term management of such patients. This review narrates the care of patients with IGM in Primary care.

Keywords: Mastitis, Granulomatous mastitis, Idiopathic granulomatous mastitis, Granulomatous lobular mastitis, Inflammation

Introduction

Idiopathic granulomatous mastitis (IGM) is a rare benign inflammatory breast entity characterized by holocentric granulomas. IGM has a persistent or recurrent disease course and affects parous premenopausal women with a history of lactation. It has also been associated with hyperprolactinemia and its a etiology has not been fully elucidated. Diagnosis of IGM relied on exclusion of other causes through laboratory workup as well as characteristic clinical examination and histopathologic findings of cystic neutrophilic. IGM is generally divided into two main groups of specific and non-specific. A palpable mass in the breast is the most common complaint, but nipple retraction, hyperemia in breast skin, oedema, ulceration and fistula development during the chronic period are also potential complaints. Systemic symptoms such as fever are generally not treatment, as low-dose prednisolone plus drainage was more effective with a lower recurrence rate than only surgical excision or high-dose prednisolone.

Etiology

GM is generally divided into two main groups of specific and non-specific. The term “specific GM” refers to conditions for which the etiological factors can be identified whether an isolated inflammatory event only applies to the breast, or the breast is involved in a systemic inflammatory event. Nonspecific GM is also known as idiopathic granulomatous mastitis or granulomatous lobular mastitis, which generally refers to conditions that can lead to a granulomatous reaction in the breast or conditions for which the etiological factors cannot be determined.

Causes of granulomatous inflammatory reaction

Infectious	**Mycobacterium tuberculosis** **Blastomycosis** **Cryptococcosis** **Histoplasmosis** **Actinomycosis** **Filarial infection** **Corynebacterium** **Wegener granulomatosis**
Autoimmune process	**Giant cell arteritis** **Foreign body reaction** **Plasma cell mastitis**
Duct ectasis	**Subareolar granuloma** **Periductal mastitis**
Diabetes mellitus **Sarcoidosis** **Fat necrosis** **Idiopathic**	

Presentation

Women presenting to Primary Care physicians show wide variations in their symptomatology Mastitis may present with clinical findings that mimic the two endpoints of breast diseases such as breast abscess and breast cancer. A unilateral palpable mass in the breast is the most common complaint. This condition should be suspected in any woman who presents with unilateral tender palpate mass within a couple of years after giving birth or who has history of at least one live birth and breast-feeding women. However, we have found reports of patients as young as 11 years-old, and as old as 80 years. The true prevalence of IGM is unknown. The disease has been found worldwide and, in all races, but there is a described predilection for Hispanic and Asian women.

Clinical assessment

Women with IGM should be asked about personal, menstrual, medical, and family history in detail. Physical examination should focus on any underlying disorder. The history and examination points to be considered by the Primary care physician are the diagnosis of IGM should be considered in women who develop recurrent sterile breast abscesses and appropriate diagnostic evaluation should be performed on such cases.

Investigation and Diagnosis

Diagnostic work-up of such patients should include confirmation of the diagnosis and the ruling out of other comorbid conditions. Although proper diagnostic accuracy in IGM is lacking, the following are usually used as diagnostic criteria.

The most frequent findings on mammogram and ultrasound are asymmetric diffuse increased density of fibro-glandular tissue and hypoechoic mass lesions or nodular structures, respectively. Breast MRI may be a better imaging modality, but it does not allow a differentiation between a granulomatous process and other disorders. MRI is a promising imaging modality for follow-up of the lesions over time.

Histopathological evaluation plays a crucial role in the diagnosis of IGM. Characteristic histopathology features such as lobular non-caseating granulomas with epithelioid histiocytes, multinucleated giant cells, and a predominantly neutrophilic background with attendant lymphocytes, plasma cells and eosinophils in varying numbers without necrosis and negative microbiological investigation favours diagnosis of IGM. Micro abscess formation and fat necrosis are frequently seen (2). Not all cases have typical non-caseating granulomas on histopathological exam. However, all patients have epithelioid histiocytes present within the smears.

Management

The most important steps after making the diagnosis of IGM are to inform the patient of the diagnosis in a sensitive and caring manner, provide accurate information, and offer referral to appropriate teams. IGM should ideally be managed by a multidisciplinary team comprising of a primary care physician, gynecologist, endocrinologist, rheumatologist, and a psychologist.

The treatment of IGM remains controversial. The treatment options include close follow-up, immunosuppressive drugs, and surgical excision.

Patients with uncomplicated IGM may be observed over time without treatment. Antibiotics have no role in the management of true cases of IGM. The use of corticosteroids have been reported as successful in the treatment of IGM. Steroids should be started at a dose of 1 mg/kg per day and tapered slowly according to clinical response. Responses usually occur within weeks of treatment, but patients may need to be treated for several months. About half of the cases relapse after stopping or decreasing the dose of steroids.

The use of steroid sparing agents such as methotrexate or azathioprine provide options that may facilitate tapering of steroids.

Traditionally IGM was treated by surgical excision of the mass, but some studies have suggested that the recurrence rate with surgical treatment is higher than with steroid treatment. Recurrence rates of 5% to 50% are reported after surgical excision of the mass. Wide excisions with negative margins are better than limited excision alone, as there is a higher tendency to relapse after limited excision. Furthermore, there is a high rate of fistula formation, poor wound healing, and disfigurement after surgical intervention in patients with IGM.

Conclusion

Despite the advances in our understanding of IGM, most cases remain idiopathic, and there is no clear etiology. Regardless of the etiology, (IGM) is a rare, poorly understood condition that presents as inflammatory nodules of the breast. It is often initially misdiagnosed as furunculosis or cellulitis. Despite the painful, scarring, and debilitating nature of the disease, patients often have a delay in accurate diagnosis and treatment. Even when IGM is considered as a diagnosis, it is one of exclusion, with the differential diagnosis including serious conditions such as breast cancer, sarcoidosis, and cutaneous tuberculosis. The management of women with IGM begins in Primary Care with informing the patient of the diagnosis in a sensitive and caring manner and providing accurate information about the diagnosis. A multi-disciplinary approach must be recognized as a cornerstone to effectively manage this complex entity to ensure that physical, psychological, and emotional challenges that result from IGM diagnosis are met and short-term and long-term well-being of these women is optimized.

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Conflict of interest
There are no conflicts of interest to declare by any of the authors of this study

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