IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 18, Issue 6 Ser. 2 (June. 2019), PP 38-40
www.iosrjournals.org

Idiopathic Granulomatous Mastitis: The Enigma Remains

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Abstract: ***Background:*** *Idiopathic granulomatous mastitis (IGM) is a rare chronic inflammatory breast condition that is characterised by varied local presentations. It is often confused with tuberculosis or malignancy, diagnosis is confirmed only on histopathological examination.*
Methods: *A retrospective study was done on all cases of IGM diagnosed within a span of five years in a tertiary care hospital in West Bengal.*
Results: *During the study period twelve cases of IGM had been diagnosed. All were parous females in the age range of 32-45 years, the mean age being 38.75 years. The disease was on the right side in 66.67% patients. A painless lump was the commonest presentation . Diagnosis was made by either core biopsy or excision biopsy. Ten patients underwent surgery as the primary treatment. Two patients were treated only with corticosteroids. Three patients were treated with corticosteroids after surgery.*
Conclusion : *In conclusion, IBD is a rare disease with uncertain etiology. Further studies are required to come to a consensus regarding management.*
Key Words: *Idiopathic granulomatous mastitis, surgery, corticosteroids*

Date of Submission: 21-05-2019 Date of acceptance: 06-06-2019

I. Introduction

Though Idiopathic Granulomatous Mastitis (IGM) was described more than 45 years ago (1), the enigma surrounding it still remains. The patients present with a wide variety of symptoms which mimic other more common conditions. Clinical diagnosis becomes difficult due to high prevalence of breast tuberculosis in India. We present our experience of twelve cases of IGM which were managed according to the clinical presentations.

II. Methods

A retrospective study was done on all cases of IGM diagnosed within a span of five years in a tertiary care hospital in West Bengal.

III. Results

During the study period twelve cases of IGM had been diagnosed. All were parous females in the age range of 32-45 years, the mean age being 38.75 years. The disease was on the right side in 66.67% patients. There were no bilateral cases in our series. One patient was lactating at the time of presentation. None of the patients had a history of tuberculosis, connective tissue disease or any other infectious disease which could cause granulomatous inflammation. A painless lump was the commonest presentation (**Table 1**). Diagnosis was made by either core biopsy or excision biopsy. Ten patients underwent surgery as the primary treatment. Among them nine patients underwent wide excision and one underwent incision and drainage. Two patients were treated only with corticosteroids. Three patients including the patient who underwent incision and drainage were treated with corticosteroids after surgery.

IV. Discussion

Idiopathic granulomatous mastitis (IGM) is a rare chronic inflammatory breast condition that was first described in 1972 (2). It presents with varied local presentations. Although easily confused with tuberculosis or malignancy, diagnosis is confirmed only on histopathological examination . IGM is an exceedingly rare disease with nonspecific clinical findings, mostly seen in females in their reproductive age (3). In our study all were parous females. IGM should be differentially diagnosed from tuberculosis, sarcoidosis, and mycotic and parasitic infections because its histological features include noncaseating granulomas, small abscesses, and inflammation in the lobules (4).

Many agents, such as local irritants, oral contraceptive pills, viruses, parasitic infections, hyperprolactinemia, diabetes mellitus, smoking, alpha 1 antitrypsin deficiency, and autoimmunity have been proposed to explain the etiology of IGM, however, these have never been proven. It is a diagnosis of exclusion made after malignancy and other known granulomatous diseases such as mycobacterial infections and sarcoidosis have been ruled out (3, 5). A favourable response of the disease to steroids and immunosuppressant drugs has pushed forward the hypothesis of autoimmune nature of the disease. The etio-pathogenesis of the disease is thought to involve the following sequence of events: ductal epithelial damage, transition of luminal secretions to the lobular connective tissue, local inflammation in connective tissue, macrophage and lymphocyte migration to the region, and local granulomatous inflammatory response (1, 6).

IGM usually occurs in parous females of the reproductive age group. Studies have shown that IGM is associated with a history of childbirth and breastfeeding within the previous 5 years (3, 7). The most common clinical presentation is a firm, unilateral, and discrete breast mass that is often associated with an abscess or inflammation of the overlying skin and fistulae (1, 2). The lesions may be located in any quadrant of the breast. In consequence of granulomatous inflammation, IGM can cause skin thickness, sinus and abscess formation, axillary lymphadenopathy, and nipple retraction, which may be clinically mistaken for breast carcinoma. Bilateral involvement is reported very rarely (3, 7).

The information obtained from ultrasound and mammography is nonspecific, and hence the lack of specificity to diagnose IGM or to exclude breast carcinoma. Ultrasound findings include a mass‐like appearance, tubular/nodular hypoechoic structures, and focal decreased parenchymal echogenicity with acoustic shadowing. Some studies have reported parenchymal heterogeneity and areas of mixed echo pattern . The most common mammographic appearance of the lesion is an asymmetrically increased density. An ill‐defined mass, asymmetrically increased density without parenchymal distortion, or microcalcification are the most common findings on mammography (3, 8). Histologically, a granulomatous inflammation is seen, which is closely related to the lobules. Granulomas composed of epithelioid histiocytes, Langhans giant cells accompanied by lymphocytes, plasma cells and occasional eosinophils are found within and around the lobules (9). The same cellular components are present in the FNA smears from these lesions (10). In our study all the patients were diagnosed histopathologically.

An accepted management strategy for IGM is lacking. Surgery and oral corticosteroids have both been used as treatment options. Wide excision/ lumpectomy is considered to be the most effective treatment option, having the lowest rate of recurrence and complications. Most of our patients underwent surgery in the form of wide excision. Surgical management with incision and drainage and expectant management were found to have an unacceptable high rate of complications and recurrence. The role of incision and drainage is controversial because it may not improve the condition and may lead to intractable incision tracks, which subsequently lead to sinus formation. Studies have reported that treatment with oral corticosteroids, alone or in combination with surgery, is effective. Satisfactory results have been reported with high dosages of prednisone (1, 2, 3, 4). In conclusion, IBD is a rare disease with uncertain etiology. Further studies are required to come to a consensus regarding management.

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Table 1: Patient Particulars

Number	Age	Presentation	Side	Treatment
1	43	Lump + sinus	R	Wide Excision + corticosteroid
2	32	Lump	L	Wide Excision

3	45	Abscess	R	Wide Excision + corticosteroid
4	35	Lump	R	Wide Excision
5	35	Lump	L	Wide Excision
6	38	Lump	L	Wide Excision
7	40	Abscess	R	Incision and drainage + corticosteroid
8	40	Lump	R	Wide Excision
9	42	Lump	L	Corticosteroid
10	40	Lump	R	Wide Excision
11	35	Lump + sinus	R	Wide Excision
12	40	Lump	R	Corticosteroid

Dr Madhumita Mukhopadhyay. "Idiopathic Granulomatous Mastitis: The Enigma Remains." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 18, no. 6, 2019, pp 38-40.