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Original Article

MANAGEMENT OF IDIOPATHIC GRANULOMATOUS MASTITIS: DILEMA IN DIAGNOSIS AND MANAGEMENT

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Abstract

Background: Idiopathic granulomatous mastitis is a rare disease of the breast. Clinically and radiologically it may mimic breast carcinoma. To date, there is no agreed gold standard treatment for IGM. The clinical management often starts with tests to eliminate microbial infection and when microbial infection can be ruled out, long-term oral steroid treatment or surgery is administered.

Objective: To see the response of different treatment options of idiopathic granulomatous mastitis.

Methods: This is a retrospective study involved 50 patients who were diagnosed with IGM and were treated in our institution between January 2009 to December 2016.

Results: The mean age of patients was 34.3 years. Breast lump was the most common presentation. The right breast was affected in 30 cases & left breast 20 cases. 4 patients were pregnant at the time of presentation. Lactation within 6 months of presentation was documented in 4patients.45 patients used contraceptives pills. A clinical suspicion of malignancy was present in 17 cases. Mammography was performed in 20 patients and showed focal asymmetrical dense lesions in 9, well-circumscribed opacity in 8, spiculated lesion in 1, and was normal in 2. Fine-needle aspiration was performed in 40 patients. Wide local excision was the mainstay of treatment. One patient underwent mastectomy due to recurrence and the final histology confirmed idiopathic granulomatous mastitis with no evidence of malignancy. 12 patients developed recurrence after a mean follow-up of 31.2 months.

Conclusion: IGM is not so rare disease in our country. Clinical, radiologic, and even cytologic findings are sometimes confused with malignancy. Adequate Surgical treatment as well as adjuvant therapeutic treatment are necessary. Knowledge, awareness and co-operation among surgeons and pathologist should also be strengthen to avoid unnecessary misdiagnosis and treatment.

Key Words: breast cancer, granulomatous mastitis, idiopathic granulomatous mastitis.

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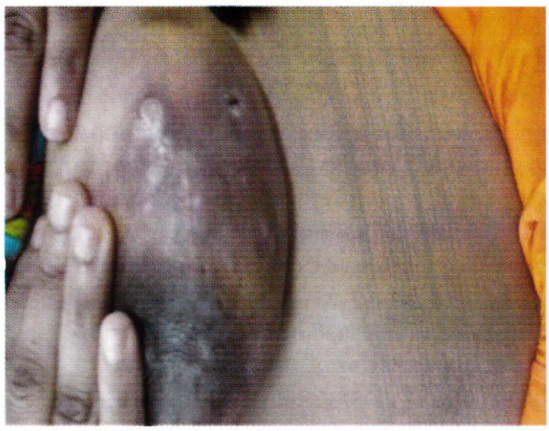
Introductn

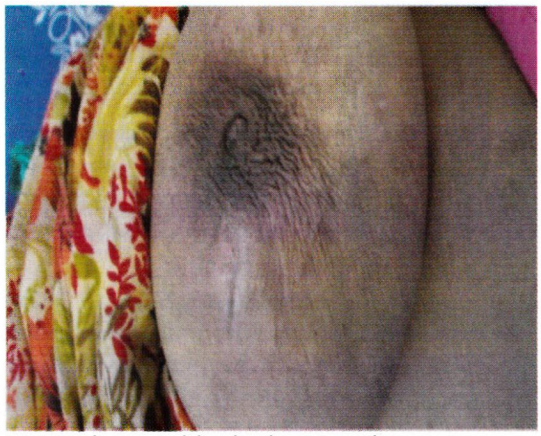
Granulomatous mastitis (GM) is a rare benign inflammatory disease of the breast with multiple etiologies such as tuberculosis (TB), sarcoidosis, foreign body reaction, and mycotic and parasitic infections[1]. In contrast, idiopathic granulomatous mastitis (IGM), first describedas a specific entity by Kessler and Wolloch in 1972 [2] and further elaborated by Cohen in 1977[3], is characterized by the presence of chronic granulomatous lobulitis in the absence of an obvious etiology. Clinically patients present with a hard lump that mimics carcinoma, which may lead to nipple retraction and sinus formation. Even mammographic and fine-needle aspiration cytology (FNAC) findings are sometimes interpreted as malignant. It is linked to lactation and was reported to occur during pregnancy and postpartum period, with a good proportion of the patients developing the lesion within 3 years of delivery.Associations with alpha1-antitrypsindeficiency and hyperprolactinemia have been reported4. Surgical excision with or without steroid therapy is the mainstay of treatment, with more than half of patients requiring more than one operation 1.

Materials and methods

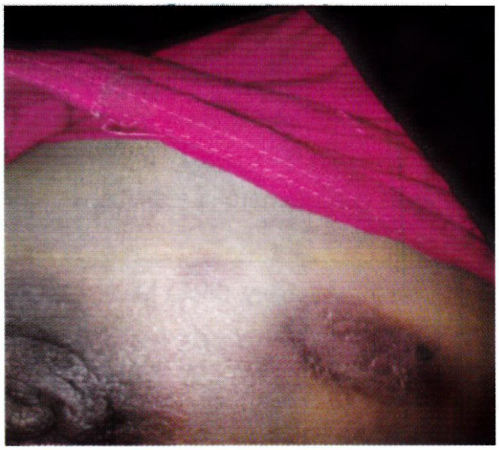
This study involved 50 patients who were diagnosed with GLM and were treated between January 2009 to December 2016. Clinical presentation, treatment processes, and results were analyzed. In all these patients, the lesions were confirmed with a breast examination and imaging evaluation using ultrasonography (US), mammography. A definitive diagnosis was made pathologically after excision biopsy was performed. In addition, tuberculosis was ruled out in each patient from the patient's medical history, chest X-ray and polymerase chain reaction test results. The criteria for selecting the method of initial treatment were as follows: patients with mild symptoms or small lesions were observed; patients who were initially thought to have bacterial mastitis underwent antibiotic treatment; patients with abscess at the time of their diagnosis underwent drainage; and surgical excision was performed on patients who showed mass forming lesions or localized lesions at the time of their diagnosis. The recovery period was defined as the time between the initial visit and the disappearance of the symptoms or the completion of wound healing in the case of surgical treatment. Recurrence was defined as the reappearance of the same symptoms after the surgical removal of the lesion. The patients were followed up in outpatient department.



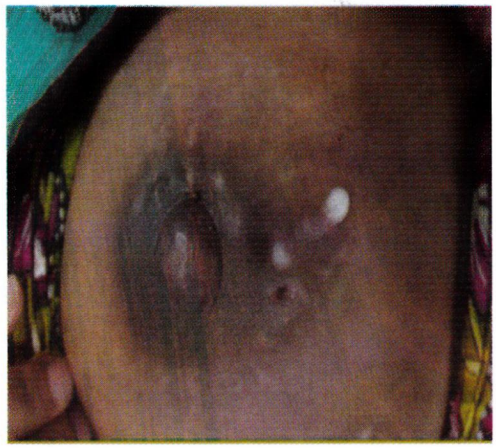
Sinus with skin thickening



Lump with nipple retraction



Abscess



Nipple discharge

Fig1 Presentation of IGLM.

Results

The mean age of patients was 38 years (range 21–55years).

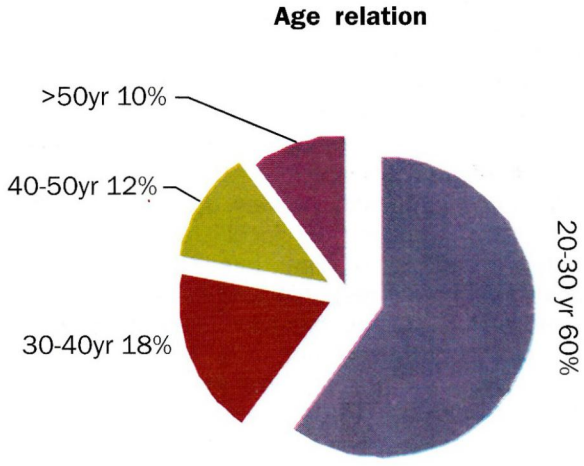


Fig-2: Age relation of subject (n=50)

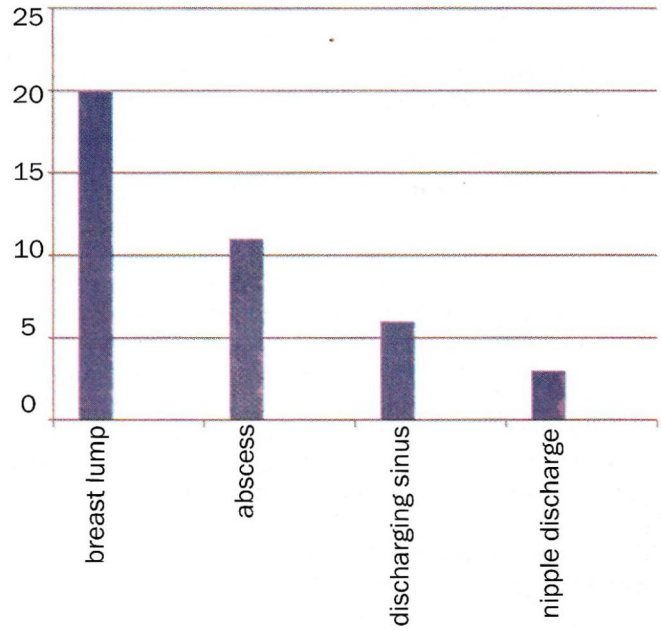


Fig-3: Clinical presentation of IGM.

Out of 20 patients (40%) the main presentation was a breast lump measuring 2.5 to 8 cm in diameter (mean 4.2 cm), 11(22%) patients presented with breast abscess, discharging sinus 6(12%), the remaining 3(6%) patients presented with nipple discharge.

The mass was unilateral in all patients. The right breast was affected in 30(60%) cases. The upper outer quadrant was affected in 29 (58%) cases. All patients were parous. 4(8%) patients were pregnant at the time of presentation. Lactation within 6 months of presentation was documented in 4(8%) patients. No cases of puerperal mastitis were seen. Serum prolactin level was measured for 8(16%) patients and hyperprolactinemia was documented in 2(4%) of them. 45(90%) patients used contraceptive pills. There was no previous history of TB, sarcoidosis, or other infectious or granulomatous diseases in any of the patients. None of the patients had a family history of breast cancer or evidence of autoimmune diseases. A clinical suspicion of malignancy was present in 17cases (34%). Palpable axillary lymph nodes were found in only 4(8%) patients, and the nodes were biopsied and the histology showed nonspecific hyperplasia. Ultrasonography was performed in 45(90%) patients and the predominant findings were the presence of heterogeneous hypoechoic pattern 30(60%) patients or hypoechoic tubular structures 15(30%) patients. Mammography was performed in 20 (40%) patients and showed focal asymmetrical dense lesionsin 9(18%) patients, well-circumscribed opacity in 8(16%), spiculated lesion in 1(2%), and was normal in 2(4%). Fine-needle aspiration cytology was performed in 40 (80%) patients. In 19 (38%) patients the aspirated material was highly cellular and showed evidence of granulomatousinflammation. In 7(14%) patients the diagnosis of fibrocystic disease was suggested, while in the remaining 14(28%) nonspecific inflammatory lesions were found.

Table -1: Clinical, imaging & pathological finding of IGLM in our series

Clinical presentation	Mammographic finding (20 cases)	Ultrasonography (45 cases)	FNAC(40 cases)
Breast lump 20(40%)	Focal asymmetrical dense lesions-9(18%)	Heterogeneous hypoechoic pattern 30(60%)	Highly cellular and showed evidence of granulomatous inflammation 19(38%).
Abscess 11(22%)	Well -circumscribed opacity in 8(16%)	hypoechoic tubular structures 15(30%)	Fibrocystic disease7 (14%).
Dischargin sinus 6(12%)	spiculated lesion in 1(2%)		Nonspecific inflammatory lesions 14(28%).
Nipple discharge 3(6%)	Normal in 2(4%)		

Wide local excision was performed in all patients. None of the patients had an associated breast carcinoma. Mastectomy was performed in one patient due to persistent suspicious lesion & multiple discharging sinus in different quadrants. Clinical, radiologic, histopathologic, and laboratory testing, including periodic acid-Schiff (PAS) and Ziehl-Neelsen (ZN) acid fast stains, failed to identify any specific causative organisms in all the cases. However, in one case who developed recurrence 9 months after excision, which flared up with steroid treatment, mycobacterial DNA sequences were detected using PCR on paraffin-embedded tissues taken from the recurrent mass. The ZN stain of thebreast biopsy was negative. Chest radiograph revealed no remarkable findings. This patient was treated with antituberculous therapy and she showed a complete remission within 12 months.

The follow-up period extended to December 2015. All patients had initial satisfactory results, with no wound morbidity. However, 12 patients (24%) developed recurrence after a mean follow-up of 31.2 months (range12-94 months) and underwent further reexcision.

Discussion

Idiopathic granulomatous mastitis is a rare benign breast disease that mimics breast cancer both clinically and mammographically. It may raise important diagnostic and therapeutic dilemmas, as more than 50% of the reported cases of IGM were initially mistaken for breast carcinoma[5]. The practice of performing mastectomy on the basis of triple assessment (clinical, mammographic, and FNAC findings consistent with malignancy), especially in young patients, does not seem to be quite justified. We believe that frozen-section or true-cut biopsies should be an integral partof the assessment of patients suspected of having breast cancer before resorting to mastectomy or breast-conservingsurgery.The diagnosis of IGM is one of exclusion; all known infectious and non-infectious causes of granulomatous inflammation have to be excluded. In addition to carcinoma and carcinomatous mastitis, IGM should be differentiated from other chronic inflammatory breast diseases such as mammary duct ectasia, Wegener granuloma, sarcoidosis, TB, and histoplasmosis[6]. IGM is characterized by chronic lobular inflammatory process and noncaseating granulomatous inflammation. All our patients had unilateral involvement. IGM is usually unilateral and can affect any quadrant of the breast[6], although bilateral involvement has been reported[7]. We could not explain the predominance of right breast affection among our patients. As shown in this series, and consistent with the reported literature, most patients with IGM are relatively young parous women less than 50 years of age 1.It has been reported that IGM is associated with pregnancy and lactation [1]. Four (8%) of our patients were lactating and another four (8%) were pregnant at the time of diagnosis. The etiology of IGM is still unknown. However, inreviewing the literature, we identified four main possible mechanisms [2,6].

(a) IGM may be caused by a chemical reaction associatedwith oral contraceptive pills.
(b) It may have an underlying autoimmune process.
(c) It may have an underlying infective etiology that cannot be detected by the current means.
(d) It may be an immune response to extravasated secretions from lobules.

In the above-proposed theories, damage to the ductal epithelium produced by local trauma, a local or systemic irritant, or viral infection may cause a localized immune response and subsequent lymphocyte and macrophage migration[6]. Immunohistochemical staining shows that, the lesion contains T lymphocytes[8]. A multifactorial etiology seems to be more likely. In this study 11 patients presented with breast abscesses. The complications of IGM include abscess formation, fistulization, and chronic suppuration[9]. We noticed that the largest reported series of IGM are coming from the developing countries[10]. The incidence of TB mastitis among surgically treated lesions of the breast is less than 0.025%[11]. ZN stain, purified protein derivative (PPD), and routine histology studies are sometimes not sufficient to rule out the diagnosis of TB mastitis. So we suggest that in cases of recurrent IGM, PCR testing be used as a safeguard. Mammography and ultrasonography are used mainly to rule out malignancy rather than to confirm the diagnosis of IGM. However, the presence of a focal asymmetrical density on mammography coupled with ultrasonic findings of a large inhomogeneous hypoechogenicity with internal hypoechoic tubular lesions might suggest the possibility of IGM[12]. Nonetheless, IGM might mimic breast carcinoma radiologically, and the final diagnosis should be reached histopathologically due to the high false-positive and false-negative mammographic appearances. Actually the possibility of IGM was not raised by our radiologists preoperatively, although in retrospect we could identify some of the features mentioned above. This reflects the importance of awareness of this entity by radiologists. There is no ideal treatment for IGM, but wide local excision, with or without steroid therapy, is the most common method of treatment[13]. Limited excision aloneis of little benefit, as there is a strong tendency for persistence or recurrence[5]. Twelve of our patients relapsed after surgery, and underwent additional surgery, after which no sign of recurrence was observed.

Conclusion

The diagnosis of IGM can be challenging. Awareness of this condition is of great significance for surgeons, radiologists, and pathologists because it can be mistaken for invasive carcinoma.

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