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# Idiopathic Granulomatous Mastitis, Is It a Surgical Disease? Case Series and Literature Review

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## ABSTRACT

*Idiopathic Granulomatous Mastitis (IGM) is an uncommon benign chronic inflammatory breast disease. It can mimic two frequent breast disorders, breast cancer and breast abscess. Although it is associated with certain risk factors, neither clinical presentation nor radiological findings are diagnostic. Histopathological evaluation plays a crucial role in the diagnosis of IGM and its treatment is controversial. We present two patients with IGM treated at King Fahad Specialist Hospital-Dammam, Saudi Arabia, over a period of 4 years. Both patients had different clinical presentations, and treatment courses. However, they both achieved remission of the disease eventually. We recommend that surgical management should be considered last in the treatment options, to avoid wound related chronic complications and poor cosmetic outcomes. The clinical presentation, diagnosis and management of IGM are discussed in this paper.*

## Keywords

Idiopathic Granulomatous Mastitis, Mastitis, Breast cancer, Inflammatory breast cancer.

## Abbreviations

IGM: Idiopathic Granulomatous Mastitis, BIRADS: Breast Imaging-Reporting and Data system, BMI: Body Mass Index, OCP: Oral Contraceptive Pill, MRI: Magnetic Resonance Imaging.

## Introduction

Idiopathic Granulomatous Mastitis (IGM) was first described by Kessler and Wolloch in 1972 [1]. Its causes remain unknown. However, IGM is frequently found in women in their childbearing age, with history of delivery within 5 years of presentation and who have practiced breast feeding [2-8]. An idea of autoimmune phenomenon has been postulated about IGM [1]. It can imitate a broad spectrum of breast conditions ranging from abscess to inflammatory breast cancer. Hence, it is of diagnostic dilemma as IGM is often difficult to differentiate, either clinically or radiologically, from other infectious conditions or breast carcinoma. Up to date, there is no consensus on the treatment of IGM. Different therapeutic modalities were described in the literature, each with its cons and pros, including conservative management, corticosteroid therapy, immunosuppressant agents, surgical treatment like incision and drainage, lumpectomy or even mastectomy [7,9,10].

## Case 1

A 26-year-old premenopausal lady presented to the emergency room (ER) with a complaint of a right breast painful lump for 1 week duration. The lump first appeared 2 months prior to her presentation. Breast ultrasound (US) was done at that time and revealed a suspicious lesion in the right breast 3 o'clock position with enlarged suspicious ipsilateral axillary lymphadenopathy, reported as BIRADs IVb. The breast mass and the suspicious axillary lymph node were both biopsied and they showed inflammatory changes with granuloma formation, negative for acid fast bacilli (AFB) and negative for atypia or malignancy (Figure 1). The mass gradually increased in size and eventually became painful and was associated with skin redness that developed over the week prior to the patient's ER presentation. The patient is a mother of 2 children, youngest child is 6 years-old, she breast

fed her children for 24 months, and she gave history of oral contraceptive pills use, but denied any history of smoking. Her past history is significant for postpartum thyroiditis and psoriasis for 10 years duration, treated with topical medicine. On physical examination, she was obese, with BMI of 41kg/m $^2$, vital signs were within normal range, including normal body temperature. Local examination of the right breast revealed a tender lump at 3 o'clock position, associated with induration, fluctuation, and local cellulitis. Blood investigation showed no Leukocytosis. A repeated breast ultrasound confirmed the presence of a fluid collection measuring 3.3x3x2.4 cm at 3 o'clock position with internal debris and a posterior acoustic enhancement consistent with an abscess. Aspiration under local anesthesia of 10 cc of pus was successfully performed in the ER and the patient was discharged on antibiotic (oral amoxicillin/clavulanic acid, 1 gram twice daily) with follow up in the breast surgery clinic. On one-week follow up, there was no response to conservative management. There was a persistent mass at 3 o'clock in the right breast, with tenderness, fluctuation and sloughing of the overlying skin. Additionally, a new mass with similar characteristic was palpated at 9 o'clock position in the same breast. Ultrasound examination showed edematous changes in the entire breast with turbid fluid tracking in-between suggestive of progression and an air pocket seen within multiple hypoechoic small collections at the retro-areolar region toward 9 o'clock position (Figure 2). The Patient underwent incision and drainage of the clinically persistent right breast abscess, under general anesthesia. Two elliptical incisions were made at 3 and 9 o'clock to drain both abscesses. However, minimal amount of pus was drained. A swab for culture was taken for aerobic, anaerobic and acid-fast bacilli, as well as tissue biopsy. The two cavities were found to be communicating in a horseshow-like pattern around the superior border of the areola. A Penrose drain was inserted. The patient was discharged and maintained on daily dressing in the clinic. All cultures were negative. Histopathology confirmed the diagnosis of idiopathic granulomatous mastitis, with no evidence of malignancy. The wounds healed in 10 days (Figure 3) and the patient was referred to the rheumatologist who started her on Methotrexate. She received the treatment for 4 months then she stopped it on her own, after which she developed a contralateral breast non painful mass. Local examination of the Left breast revealed a non-tender mass at 3 o'clock peri-areolar position, with no fluctuation, erythema, or discharge. A bilateral breast ultrasound showed a significant interval regression of previously seen right breast collection and small masses. Left breast newly developed 3 o'clock heterogonous hypoechoic lesion with irregular borders and angular margins, measuring 3.1 x 0.8 x 2.6 cm with inflammatory process, (BIRADS II) (Figure 4). These findings were suggestive of another IGM lesion, hence, the patient was reassured and advised to continue on Methotrexate. Biopsy of the new left breast lesion was not performed because it was not suspicious (BIRADS II) on ultrasound, and in order to avoid the possibility of biopsy- induced breast infection. No antibiotics were prescribed, and no surgical intervention was performed. Upon her last clinic visit, the patient had no new breast complaints. Local breasts examination revealed old surgical scars healed surgical wound on the right, with no sinus formation, and no palpable masses in both breasts. The latest US examination showed left breast interval regression in the 3 o'clock heterogonous lesion with irregular borders, currently measuring 1.2 x 0.7 x 1.9. (BIRADS II) benign findings (Figure 5).

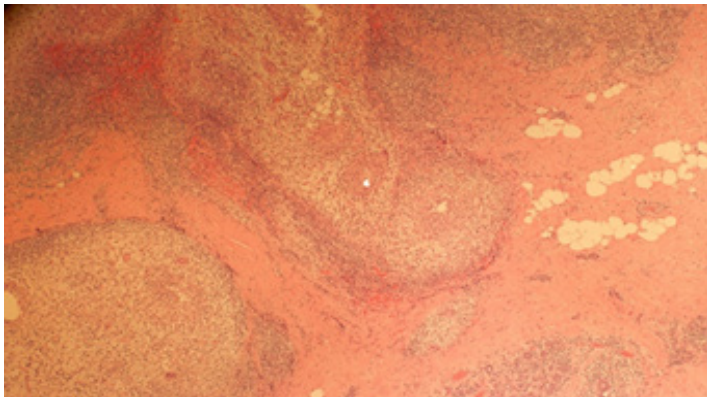

**Figure 1:** A microscopic picture of the breast mass biopsy showing non caseation granulomas with multinucleated giant cells.

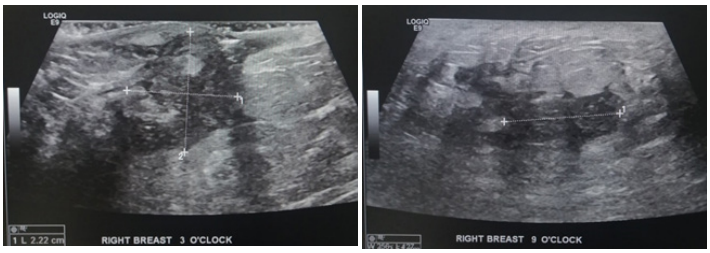


**Figure 2:** Ultrasound of right breast of first case, showing progression of 3 o'clock inflammatory process and an air pocket seen within multiple hypoechoic small collection at 9 o'clock position.

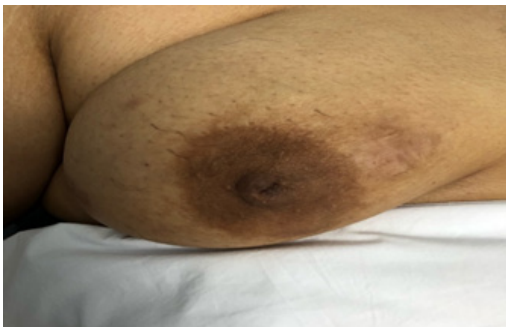

**Figure 3:** Completely healed right breast 3 and 9 o'clock scars.

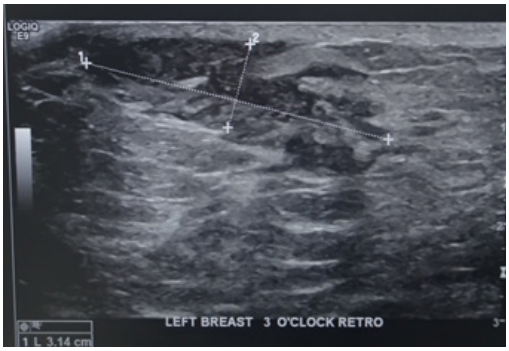


**Figure 4:** Left breast ultrasound showing a newly developed left breast inflammatory mass at 3 o'clock.

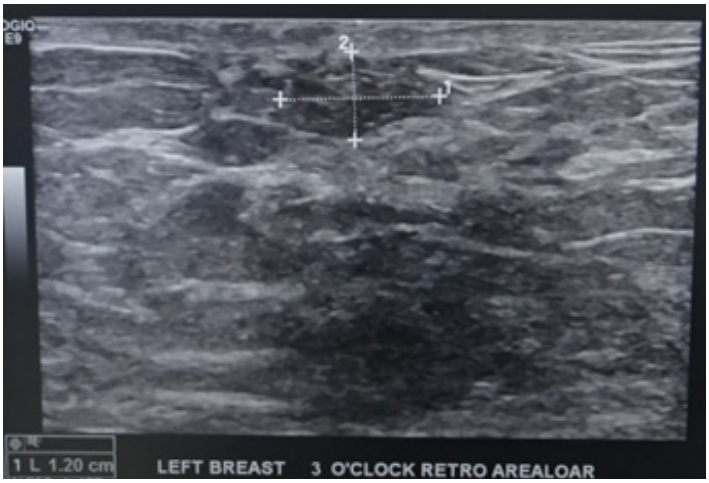


**Figure 5:** Left breast ultrasound showing interval regression of the 3 o'clock heterogonous lesion, currently measuring 1.2 x 0.7 x 1.9cm.

## Case 2

A 49-year-old peri-menopausal lady, presented to the ER with complaint of right breast painful lump for 12 Days duration, which was increasing in size and associated with redness for 3 days prior to presentation, she denied any history of fever, or previous similar problem. She is a mother of 2 children, her youngest child is 7 years-old. She breast fed her children for 2 months only, no history of oral contraceptive pills uses or smoking, and no family history of breast or ovarian cancer. The patient is known to have hypertension treated with Amlodipine and papillary thyroid cancer, post total thyroidectomy 7 years back, currently using L-thyroxin 75 Mcg/day. On physical examination, her vital signs were within normal range including a normal body temperature. Local examination of the right breast revealed tenderness, erythema and hotness involving the peri-areola area at 2 o'clock position, associated with fluctuation and palpable regular non tender lump measuring 2x2 cm, and bilateral congenital nipple inversion. Contralateral breast and both axillae were unremarkable on examination. Laboratory investigation revealed leukocytosis of $12.8x10^9$/L. Breast ultrasound showed two hypo-echoic regular and lobulated lesions at 8-9 o'clock with no acoustic shadowing, both measuring 7x5 mm, and marked diffuse skin thickening likely represent mastitis with no evidence of drainable abscess. No lesion was reported by ultrasound at 2 o'clock. Attempted aspiration of the fluctuating area under sterile measures revealed minimal purulent fluid aspirate. The patient was admitted to the surgical ward for conservative management and was started on intravenous antibiotic. During the hospital course, the pain subsided and the erythema decreased. Culture from the fluid aspirate came back negative for aerobic and anaerobic organisms, fungus and AFB. The patient was discharged home on amoxicillin/clavulanic acid 1 gram twice daily and to be seen in the clinic. However, she traveled to her country on a vacation where she underwent incision and drainage of a right breast abscess and excisional biopsy of a retro-areola lump, and she was informed of a benign histopathology. She returned to our surgical clinic 2 months later complaining of a right breast sinus tract at the surgical scar with on/off minimal purulent discharge. A repeated ultrasound showed right breast anechoic collection at 12 o'clock with some debris which is directly communicating with the skin through a clear sinus tract. Two other small collections were appreciated at 2 and 8 o'clock, (BIRADS II). (Figure 6). Mammogram examination showed a retro-areolar asymmetric increased density at the scar area with no associated suspicious masses or microcalcifications. Multiple enlarged axillary lymph nodes with preserved hilum, likely reactive to her recent surgery (Figure 7). Ultrasound- guidance aspiration of the collections and core biopsy of the cavity were performed. They revealed acute inflammation, giant cells and granuloma formation, consistent with IGM. The patient was treated conservatively and a rheumatology consultation was requested. However, by the time the patient presented to the rheumatologist, her symptoms have already subsided, and their decision was not to place her on any corticosteroid or immunosuppressant therapy. On a six- month follow up, the patient was asymptomatic and her right breast wounds and sinus healed with scaring and mild disfiguration. The 2 o'clock mass was no longer palpable on physical examination (Figure 8). A follow up ultrasound showed interval regression of right breast 2 and 8 o'clock collections, measuring now 1 and 0.8 cm, respectively, and interval regression of the 12 o'clock collection with healed sinus scar, (BIRADS II) (Figure 9).

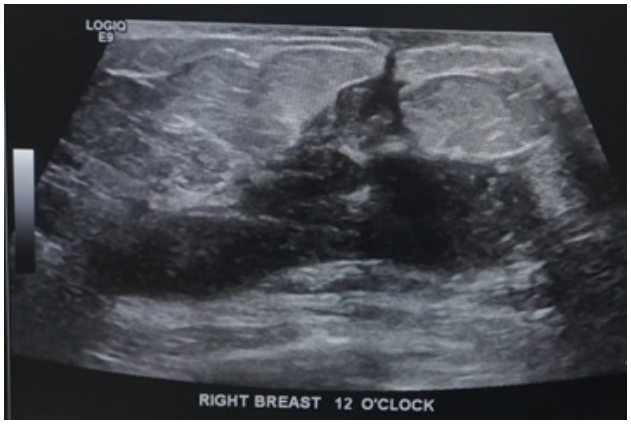


**Figure 6:** Right breast ultrasound showing a fluid collection at 12 o'clock with a sinus tract tracking up through the skin.

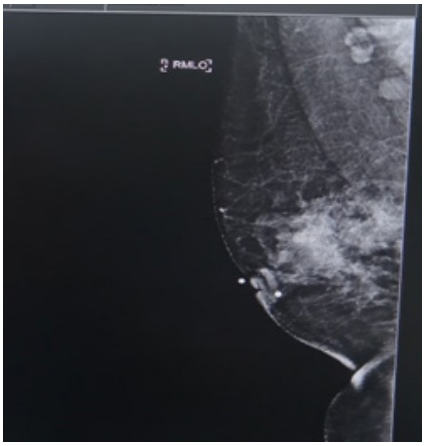

**Figure 7:** Right breast mammogram showing a retro-areolar asymmetric increased density at the scar area and multiple enlarged axillary lymph nodes with preserved hilum, likely reactive.

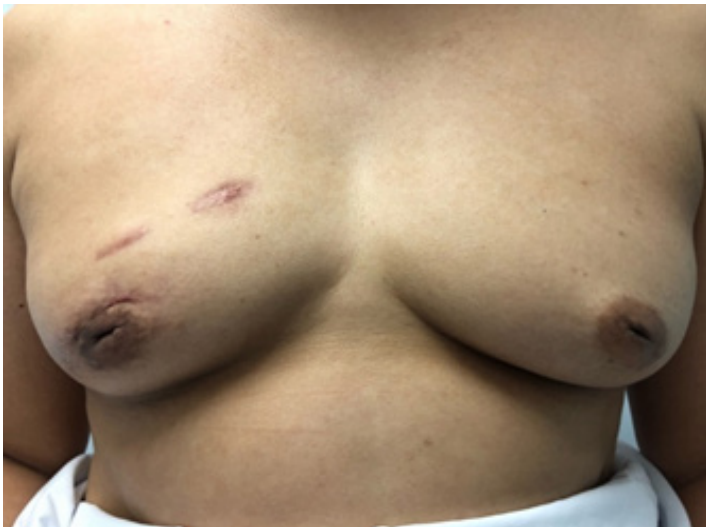

**Figure 8:** Six-month follow up showing healed right breast scars and sinus with mild disfiguration.

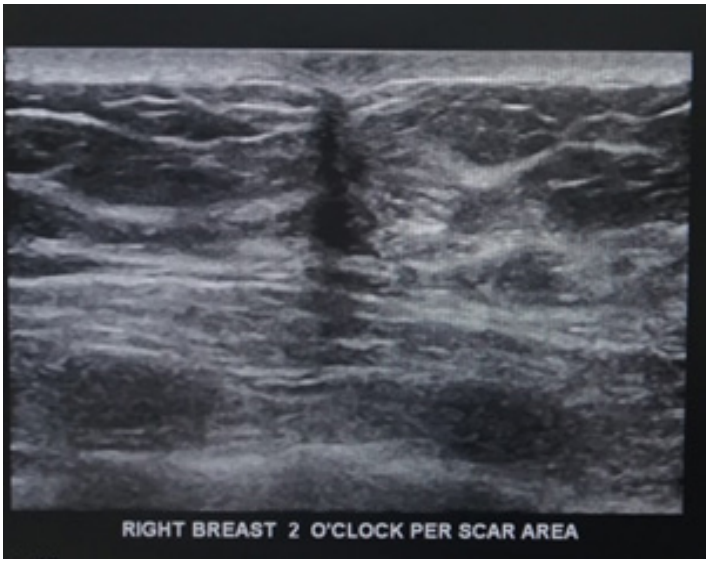


**Figure 9:** Six-month ultrasound follow up of right breast showing interval regression of the 2 o'clock collection with healed sinus track.

## Discussion

IGM usually presents in women around 35 years of age. However, it has been reported in patients as young as 11 years [4] and as old as 80 years of age [4,6,9,11]. The age of presentation of both our patients falls within the common range of age at presentation. The Most common presenting symptom is a palpable mass which can appear at any quadrant of the breast, sparing the retroareolar region. It can be hard and fixed to the surrounding tissue, retraction of the nipple, pain, abscess formation and skin fistula may be present with peau d' orange appearance of the skin or erythema that can mimic breast cancer [12]. Our patients presented with multiple variable symptoms including a painful mass, inflammatory changes, abscess, sinus, and lymphadenopathy suspicious for metastatic breast cancer. The most common presentation is a unilateral breast mass. However, bilateral involvement of the breasts has been reported as well [10]. And here we report bilateral involvement in our first case within few months of her first presentation.

The etiology of IGM remains unclear, but it is most commonly observed in women at premenopausal age, multiparous with history of delivery within 5 years and history of breast feeding [2,3,6-8]. Some of these factors apply to our patients like being premenopausal, multiparas and history of breast feeding. Other potential factors that can contribute to the development of IGM are autoimmune diseases, diabetes mellitus, hyperprolactinemia, Corynebacteria, alpha-1-antitrypsin deficiency, trauma, high BMI, use of oral contraceptive pills, smoking, and foreign body reaction [2-5,7,9,10,13]. Our first patient carried additional risk factors including high BMI and the use of OCP that were not reported by the second patient. Some published cases has reported the development of erythema nodosum and inflammatory arthritis in patients diagnosed with IGM, who responded to steroids and other immunosuppressant therapy. This observation may support the autoimmune hypothesis as a possible etiology for IGM [3]. Moreover, IGM shares some histological features with other lesions like granulomatous orchitis and granulomatous thyroiditis, where an antigen-antibody reaction has been considered as a contributing factor [1,6,7,12]. Our first patient had history of autoimmune disease. She reported having chronic psoriasis and postpartum thyroiditis. Such history should alert the treating physician to consider IGM in the differential diagnosis of breast lesions.

Due to non-specific clinical presentation and lack of pathognomonic findings in the radiological examination, IGM remains a diagnosis of exclusion. And the differential diagnosis includes breast cancer, specifically, inflammatory type, chronic inflammatory or infectious diseases like, duct ectasia, periductal mastitis, bacterial mastitis, tuberculous mastitis, Wagner's granulomatosis, foreign body granuloma, and sarcoidosis [2,4,5,8-10]. Gram stain, cultures, AFB and chest x-ray, are often negative and abscess is mostly sterile as found in both of our presented patients. [1,5,7,9,10,14]. Radiologically, there is no specific pathognomonic feature for IGM. On Ultrasound, it may appear as a hypo-echoic, lobulated, irregular mass with angular margins, increased vascularity of the surrounding breast parenchyma, fluid collection, similar to the ultrasound findings in the first presented case, or communicated sinus tracts as seen in the second case [2,4,7,9,14,15]. Evaluation of the axilla may reveal lymphadenopathy which appears as enlarged lymph node with smooth reactive cortical thickening that can be also present in metastatic lymph nodes from an invasive breast cancer [7,9]. This was one of the differential diagnoses in the first case, and biopsy of the breast mass and the axillary lymph node was necessary to exclude cancer. A mammogram may demonstrate asymmetry either focal or regional, skin thickening, or architectural distortion which can be undistinguishable from cancer [2,3,5,10,15]. Micro-calcification has been reported in the literature as an extremely rare finding in IGM [3]. Our second patient was at the age for mammogram examination, and all the formally described findings were observed in her mammogram. The first patient, however, was 26 years old and therefore

mammogram was not considered in her initial workup due to its low sensitivity in dense breasts. On MRI, the involved area of the breast demonstrates intense mass or non- mass enhancement. Although, MRI cannot differentiate IGM from other breast disorders, it is still considered the best modality to evaluate the extent of the disease, response to treatment, and involvement of the contralateral breast [2-4]. However, due to its cost and inconvenience to the patients, we did not use MRI as a tool to assess response to treatment, especially that the response was easily assessed clinically and by ultrasound in both presented cases. Histological examination is the gold standard in the diagnosis of IGM. Under the microscope, the disease appears as a non-caseation granulomas centered around the breast lobules with multinucleated giant cells, plasma cells, epithelioid histiocytes, lymphocytes, eosinophils, predominantly neutrophilic background and occasionally a micro-abscess may be present [1,4,9-16] These histopathological findings can be identified on incisional/excisional biopsy of an abscess wall, similar to the second case, as well as on a core biopsy of the breast mass, as done for the first patient. Therefore, if the IGM is considered in the differential diagnosis of a breast mass or abscess, we believe that a core biopsy should be performed before any surgical intervention, to avoid unnecessary surgery that may result in chronic wound complications and breast disfiguring.

To date, there is no definitive treatment for IGM, the available therapeutic options include: corticosteroids, immunosuppressive agents, surgical intervention either; abscess drainage or wide local excision, or expectant management with close follow-up [7,10]. The patient's presentation and the clinical course of the disease often indicate on the treating surgeon what treatment option to provide. However, IGM is believed to be a self-limiting disease with a course of 6-12 months, regardless of the type of management that has been taken [6,11,15]. Therefore, surgical intervention should not be considered as the first treatment option and it should be kept for complicated and intractable cases. The treatment plan should be tailored for each patient depending on her presentation. A study that included 20 patients has reported that IGM resolved in 6 weeks with antibiotic therapy alone [9]. Our second patient was treated upon her second presentation with 5 weeks of antibiotics and her symptoms resolved without the need for additional surgery or other treatment modalities. IGM is believed to be an autoimmune disease, therefore, corticosteroid is widely a chosen therapy. No specific dose or duration were established yet, but it is commonly started with a dose of 0.5-1 mg/kg/day of prednisone and tapered slowly according to the disease response. Immunosuppressive therapy like Methotrexate and azathioprine were documented as effective monotherapy [16,17]. This is likely to be true in cases where an autoimmune etiology is suspected. The first patient we presented responded well to methotrexate therapy and relapsed after sensation of treatment. This might not be surprising, knowing that she had history of other autoimmune disorders, indicating that the underlying etiology of her IGM is autoimmune in nature. Immunosuppressive therapy can also be used in cases where steroid therapy is contraindicated or in relapsed cases after discontinuation of steroid, or combined with steroid to facilitate steroid tapering and decrease its side-effect [4,5,8,10,13,17]. Surgical Abscess drainage usually reveals minimal amount of pus and it should include removing all loculi to prevent fistula formation and consequently, non-healing wounds like what happened with the second case who underwent incision and drainage at another hospital then presented for the second time with a sinus. Therefore, we recommend aspiration of small collections if needed to avoid scarring and fistula formation. Local excision of the mass leads to high recurrence rate, ranging between 5% to50% in the literature [3-5,7,10,11,14,15]. If surgery is to be considered, negative surgical margins should be achieved by wide local excision (WLE). WLE can be considered in small masses not involving the sub-areolar area and confined to one quadrant. Surgical interventions can lead to multiple complications such as delayed wound healing, scaring, sinus formation and poor cosmetic result [4,8,10,11,14,15,17]. Mastectomy is reported as a surgical option for sever intractable cases that did not respond to steroid therapy or underwent multiple surgical intervention with multiple sinuses formation and scarring [7,9].

## Conclusion

IGM is an important to recognize benign inflammatory breast condition, with a broad clinical presentation that can resemble mainly breast abscess or carcinoma. It is of diagnostic challenge, histological assessment is crucial for the diagnosis, and non-surgical management is the main stay of treatment. Breast surgeons should have a low index of suspicion in patients with risk factors for IGM, because early diagnosis can prevent the unnecessary surgical intervention that may result in local wound complications and poor cosmetic outcome.

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