

Idiopathic Granulomatous Mastitis: A Condition With no Definitive Treatment

Mastitis granulomatosa idiopática. Una entidad sin tratamiento definitivo

Idiopathic granulomatous mastitis (IGM) is a rare chronic inflammatory disease of the breast that simulates other diseases, such as cancer, abscess and periductal mastitis.^{1,2} It does not present defining characteristics on imaging diagnostic tests³ and treatment continues to be controversial.⁴ We present 3 cases of IGM that were diagnosed in our hospital over the course of a year.

The patients were 41, 34 and 36 years old. They all presented with painful breast lumps: the first 2 patients had solitary lumps and the third had bilateral lumps (2-5 cm in size). They reported no history of trauma or contraceptive use. In the first patient, the lesion had inverted the nipple, although no inflammatory signs were observed in any of the patients. All patients had been pregnant followed by prolonged breastfeeding in the last 4 years.

In all 3 patients, ultrasound studies revealed poorly defined hypoechoic lesions (Fig. 1) with no axillary involvement. Mammography demonstrated a nodule in the left lower quadrant of the right breast in only one patient, while in the other 2 the test was negative. Magnetic resonance imaging (MRI) showed alterations consistent with malignancy and breast abscesses in the second and third patients, while in the first patient the test was negative.

In one patient, fine needle aspiration (FNA) was performed, which provided no diagnostic data. Meanwhile, in all three cases core needle biopsy (CNB) established the diagnosis of necrotizing granulomatous mastitis (Fig. 2). Ziehl, PAS and

Giemsa stains as well as bacteriological cultures for tuberculosis and fungi were negative.

The lesion was excised in one patient and, after 18 months, no relapse has been detected. The other 2 patients developed fistulas (in one at the site of the puncture and in the other the fistulas were bilateral and periareolar); cultures were negative in both cases. The fistulas received no treatment other than local therapies. Two patients, one 5 months and the other 3 months after diagnosis, are asymptomatic and have had no lesions either on examination or on ultrasound.

IGM was first described in 1972 by Kessler and Wolloch.¹ Average age at diagnosis is 36 (18-67 years) years.^{2,5} Most women have a history of pregnancy or breastfeeding² in the previous 5 years.

The prevalence of this disease is unknown, although one study reported IGM in 1.8% of 1106 women with benign breast disease.⁶ This condition has been detected more frequently among Asian or Hispanic women.^{2,6} In our series, 2 patients

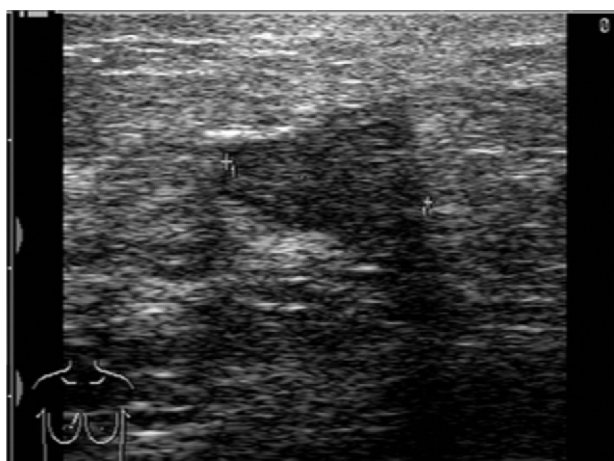


Fig. 1 – Poorly-defined hypoechoic lesion.

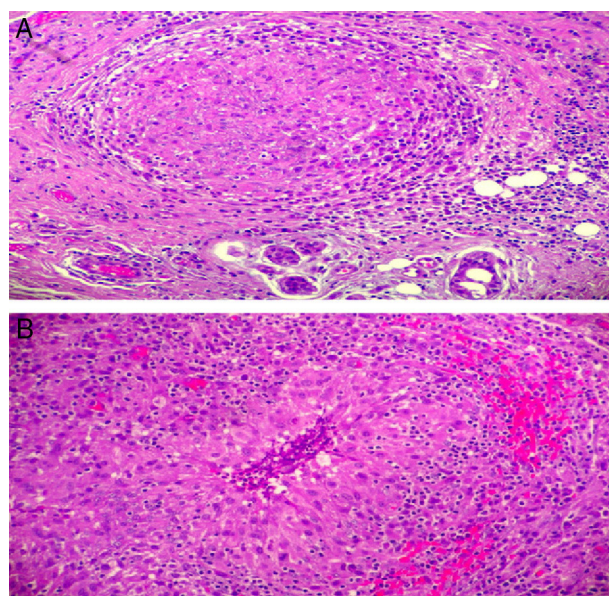


Fig. 2 – (A) Non-caseating granuloma consisting of epithelioid histiocytes that are surrounded by lymphocytes and plasma cells (HE, 100×); (B) necrotizing granuloma: central necrosis surrounded by palisading epithelioid histiocytes, lymphocytes and plasma cells (HE, 100×).

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are Hispanic (Latin American) and the other is Spanish (Mediterranean).

Several mechanisms have been described that could explain this disease: a reaction associated with contraceptives, trauma, autoimmune phenomena, infection (although the pathogen has not yet been identified) and localized immune response to the extravasation of the secretion of the breast lobules.^{1,2}

IGM is manifested clinically as a breast lump (78%)⁴ that varies in size^{4,5} and is usually unilateral, although occasionally it can occur in both breasts and in any quadrant.^{4,5,7} Some 22% of cases may present a fistula⁴ and 7% have associated nipple secretion,⁵ or there may be acute inflammatory signs with nipple retraction and "orange peel" skin.^{1,2,6,8} Enlarged lymph nodes are not usually palpable.⁶

Although lesions consistent with this disease can be detected by mammography, ultrasound and MRI,^{3,4,7} the spectrum of possible images is so broad that no specific features of IGM have been demonstrated.³

The diagnosis can only be established by histologic study.^{6,8} In our cases, CNB was sufficient for diagnosis, as in other series.^{4,5,7,8} Characteristic findings are non-caseating granulomas made up of epithelioid histiocytes, Langhans giant cells, neutrophils and a variable number of lymphocytes and plasma cells, all of which are associated with a negative exhaustive microbiological study.⁵⁻⁷ The absence of casein and the predominance of neutrophils are important data that favor the diagnosis of IGM.⁷ With FNA, it is difficult to establish a diagnosis of IGM, since it does not provide enough material, especially for the exclusion of other etiologies.⁴

Treatment of this disease is controversial.^{2,4} Patients presenting uncomplicated IGM may be observed without treatment because spontaneous resolutions have been reported.⁸ Antibiotics do not play a role in the treatment of true IGM.⁹ Corticosteroids can be used as a single treatment for the initial reduction of the tumor to achieve excision with good esthetic results.¹⁰ Prolonged corticosteroid therapy may be necessary,⁸ and relapse occurs in fifty percent of cases.⁹ The use of agents such as methotrexate and azathioprine has been described to improve the response to corticosteroids.⁹

Surgical removal, which may be the definitive cure, is associated with the appearance of fistulas, wound retraction and recurrences (5%–50%), which would decrease if negative margins were achieved.^{4,7,8} These patients need long-term monitoring because of the possibility of recurrence.¹⁰

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