Idiopathic granulomatous mastitis

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Abstract
The rare condition of idiopathic granulomatous mastitis (IGM) is presented here, unusually, in a 54-year-old woman. IGM mimics breast carcinoma and further differentials include tuberculosis and fungal infections of the breast together with other chronic granulomatous conditions. Of note is its characteristic ultrasound features that can suggest the diagnosis. Diagnosis is made by core biopsy and histology. Patients have been shown to respond to steroid and other immunosuppressive therapy, with surgical excision reserved for those whose condition is recurrent and unresponsive to medical treatment.

Case report
A 54-year-old woman presented with a 3-week history of a painful right retro-areolar mass which was unresponsive to a course of antibiotics. She had no significant past medical history, including no previous history of tuberculosis. She was not on any chronic medication, and in particular had not used the oral contraceptive pill. She had no history of breast trauma, and there was no family history of breast cancer. She had breastfed her two children for 3 months each. Her first child was born when she was 24 years old and her second child when she was 36. She was currently peri-menopausal.

Clinically, there was a unilateral firm right retro-areolar breast mass which was tender to palpation. The overlying skin was thickened and slightly warm. There was no associated nipple discharge or skin sinus.
Mammography revealed bilateral moderately dense fibro glandular breast parenchyma. In the right retro-aireolar region was a poorly defined area of increased density (Fig. 1). Ultrasound revealed an irregularly outlined, hypoechoic mass measuring 19 mm x 17 mm x 20 mm. Hypoechoic tubular extensions into the breast parenchyma were noted. There were no posterior acoustic features. Colour Doppler demonstrated increased vascularity in the surrounding tissue but not in the mass (Fig. 2). Associated with these findings was a right axillary lymph node measuring 14 mm x 10 mm. It was round, with a thickened cortex and minimal visible fatty hilum.

The imaging features raised suspicion for a carcinoma, and ultrasound-guided core biopsy of the mass and fine-needle aspiration of the right axillary lymph node were performed.

The core biopsy demonstrated features of granulomatous mastitis. Stains for demonstrating acid-fast bacilli (Ziehl-Neelsen) and fungal elements (Pas, Grocott’s) were negative (Fig. 3). The fine-needle aspirate of the right axillary lymph node was consistent with a reactive lymph node.

The patient was treated conservatively without steroids and showed good resolution of her symptoms with expectant management.

**Discussion**

Idiopathic granulomatous mastitis (IGM) is a granulomatous inflammation of the breast lobules of unknown aetiology. It was first described by Kessler and Wolloch in 1972. Clinically, it most commonly affects parous women, aged 22 - 42, who have had their children between 6 and 15 years before diagnosis. Unusually, our patient presented at 54 years of age, having delivered her last child 18 years prior to her breast condition.

The pathological findings of IGM are unrelated to specific infection, trauma or foreign body reaction. No consistent history of breast feeding or oral contraceptives can be associated with the condition. The response of IGM to steroids implies an autoimmune process. However, histological features of an immune-mediated inflammation, such as vasculitis and predominantly plasma cell and lymphoid aggregates, are not seen in IGM. In a large series of 54 women reviewed between January 2000 and April 2008, the histological result of IGM represented less than 1% of all breast biopsies done at that institution, confirming as in other reports that IGM is a rare disease.

Most commonly, IGM presents as a unilateral breast mass which can, as in our patient, be tender. While in some series no skin and nipple changes were reported; others report erythema and draining sinus tracts to the skin. Since most patients present with a fixed, immobile non-tender mass, the main differential diagnosis of concern is that of carcinoma of the breast. This proves to be of further concern as the mammographic appearance of IGM is commonly an asymmetric increase in density, in either outer or medial quadrants and peripherally, or less commonly (as in this patient) retro-aireolar in position. IGM may also present as an irregular lobulated mass on mammogram. Diffuse increase in breast density of the affected side may be the only mammographic indicator of pathology, but this is the least common presentation; in dense breasts, pathology may be masked.

Ultrasoundography is helpful in characterising IGM. In one case series, the most common ultrasound feature is that of a hypoechoic or heterogeneous hypoechoic mass. The distinguishing characteristic of IGM, as is demonstrated in this case, is that of tubular hypoechoic extensions from the dominant mass that may connect to other nearby masses if there are any. Parenchymal distortion with acoustic shadowing and no discreet mass may be seen on ultrasound instead of the findings of a dominant mass. Benign regional axillary adenopathy may be present.

Pathologically, inflammation affecting breast lobules or lobulitis and the formation of non-caseating granulomas composed of clustered epitheloid histiocytes is present. In addition, lymphocytes, plasma cells, neutrophils and giant cells comprise the inflammatory infiltrate. The granulomas can become confluent and liquefy. These positive findings need to be associated with the negative findings of tuberculosis or any other possible infectious cause of inflammation in the specimen.

The differential diagnosis of IGM remains carcinoma of the breast, tuberculosis and fungal infections, especially when the mass is associated with sinus tracts to the skin. Other considerations include sarcoidosis, fat necrosis, Wegener's granulomatosis, plasma cell mastitis and a ruptured cyst. Sarcoidosis of the breast can give a similar histological picture to IGM, and it may be difficult in the absence of systemic disease to differentiate between the two.

Treatment of IGM has been conservative, allowing mild disease to recover. This was the management and outcome to date of the patient presented here. Surgical excision has proved more successful than incision and drainage in the past, with cases of poor wound healing with incision and drainage being reported. Post surgery, a course of steroids may be prescribed. Antibiotics are unhelpful, and a persistent mass unresponsive to antibiotics prompts patients to seek further care. Current therapy favours a course of steroids, repeated if necessary, and followed by immunosuppressive therapy with methotrexate. Surgical excision should be reserved for recurrent disease that does not respond to medical therapy.

In conclusion: IGM is a rare benign breast condition. It mimics breast carcinoma both clinically and radiographically. Infective mastitis and inflammatory breast disease of known aetiology must also be considered in the differential diagnosis. Recognising the condition and obtaining histology is essential in making the diagnosis. The value of making this diagnosis is that the prognosis and treatment differ significantly from that of breast carcinoma, and the radiologist can be pivotal in the diagnosis of this rare condition.