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Case report

Idiopathic granulomatous mastitis mimicking inflammatory breast carcinoma: What are the odds?

B. Zulfiqar, Residents, U. Appalaneni, Medical student, B. Ahmed, Residents*, A. Hassan, Residents, P. Boddu, Residents, A. Carey, Pathologist

Advocate Illinois Masonic Medical Centre, United States

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Background

While inflammatory breast cancer is rare, idiopathic granulomatous mastitis is more uncommon. We hereby present a very rare case of granulomatous mastitis which thought to be inflammatory breast cancer.

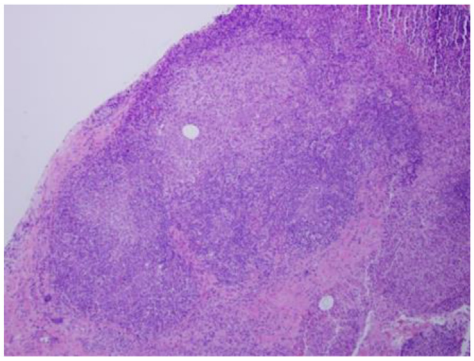
Case

A 27-year-old Hispanic woman presented with a breast lump, fever, joint pain, and a rash over her anterior shins. Since 1 one month of presentation she had noticed a breast lump which was slowly increasing in size. She was a mother of 2 children and had last breastfed 3 years ago. She denied any trauma to the breast or any nipple discharge. In the week prior to admission, the patient also began to experience fever and joint pain, most severely in her left knee. During this time, she also developed multiple tender lesions over her lower extremities.

On physical examination, patient was noted to have a 7×5 cm indurated mass over the right upper quadrant of her right breast, with overlying fluid and ecchymosis, no nipple discharge or retraction and no axillary lymphadenopathy. The patient had multiple tender erythematous nodules measuring 1–2 cm in diameter over her anterior shins in addition to one larger lesion measuring 5 cm on her right lateral thigh. The patient had mild effusion of the left knee with tenderness to palpation of the popliteal fossa and significant pain with both passive and active range of motion of her left knee.

On admission, she was febrile, tachycardiac and distressed. Laboratory work revealed leukocytosis of 12.8 (87% neutrophils, 7% lymphocytes). Furthermore, inflammatory and autoimmune work up showed elevated CRP 12.1 and ESR 77, a procalcitonin level <0.05. Investigations for HIV, RPR, Histoplasma urine antigen, Group A Streptococcus, antinuclear antibody, rheumatoid factor, and anti CCP antibody were negative. Quantiferon Gold testing was indeterminate. In regards to her left knee swelling, a lower venous duplex study showed no evidence of DVT but did find a likely Baker's cyst measuring 3–4 cm in the left popliteal fossa.

Moreover, breast ultrasound and an ultrasound guided core needle biopsy confirmed findings of granulomatous mastitis with some neutrophilic infiltrate. Patient had been treated empirically with IV vancomycin until the diagnosis was confirmed. Following diagnosis, the patient was started on corticosteroids after which she improved clinically and was discharged on 6 weeks of steroid taper.



(100 x) magnification showing granulomatous inflammation

* Corresponding author.
E-mail address: beenish_zulfiqar_ali@yahoo.com (B. Zulfiqar).

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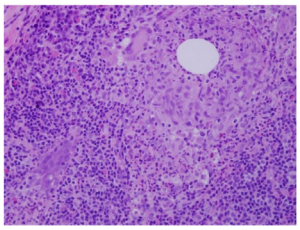

(400x) magnification showing the granuloma with giant cells, acute and chronic inflammation, and damaged lobule.

Discussion

Granulomatous mastitis (GM) is a rare chronic inflammatory disease of the breast of unknown etiology [1,2]. It was first described by Kessler and Wolloch in 1972 [12]. Thought to be an autoimmune condition, it typically occurs in young parous women as a painful extra-areolar breast lump [1]. Studies have shown its association with pregnancy and lactation [4]. Clinically and mammographically, it may mimic a carcinoma of the breast [1,3].

The most common presentation is of a hard lump in one breast without any sign of a systemic disease. Other possible symptoms include nipple retraction, pain, inflammation of the overlying skin, nipple discharge, fistula, enlarged lymph nodes, in rare cases peau d'orange-like changes [13]. Presentation is generally unilateral although a significant share of cases is bilateral. Several cases occurring together with fever, polyarthralgia and erythema nodosum have been documented, though fever is a rare presentation [14].

Histopathological of IGM is characterized by the presence of multinucleated giant cells and epithelioid histiocytes forming non-caseating granulomas around lobules. Minor ductal and periductal inflammation is usually present. The lesion, in some cases, can be difficult to distinguish from carcinoma, infectious causes (tuberculosis, syphilis, corynebacterium, mycosis), autoimmune conditions (sarcoidosis, granulomatosis with polyangiitis), and foreign body reaction [5,6].

Pathophysiological of the disease follows a sequence initiated by ductal epithelial damage followed by transition of luminal secretions into the surrounding lobular connective tissue. Extravasation of luminal contents incites a local inflammatory response and promotes macrophage and lymphocyte migration into the region, resulting in a granulomatous inflammatory respons [7].

IGM is believed to be of autoimmune etiology. However classical serological tests used in autoimmune disorders (such as antinuclear antibody, ANA, and rheumatoid factor, RF) provide conflicting results in IGM patients. A case series identified negative ANA and RF titers in all of their 18 study patients [8], while another 8-patient case series reported RF and ANA positivity in six and two patients, respectively [9].

Some sources report that approximately half of the patients may recover fully after a lengthy period of expectant management (mean time 14.5 months, range 2–24 months) [10]. Others suggest treatment with steroids, which should be prolonged with a gradual taper over 6 months. Our patient was found to have clinical improvement on a prednisone taper. While some sources report very good success with steroids, most report a considerable risk of recurrence after a treatment with steroids alone. Some studies suggest that surgical excision may be the best treatment. Wide local excision serves the dual purpose of diagnosis and treatment. A study by Gluten et al. reports recurrence in one of 15 patients treated with wide local excision [11]. However, there is no established treatment protocol for IGM.

Conclusion

Granulomatous mastitis is a rare chronic inflammatory disease of the breast of unknown etiology. Thought to be an autoimmune condition, it may involve multiple organ systems. Our patient presented with fever, painful breast mass and was in the reproductive age group. This presentation would put cancer higher on differential diagnosis. Hence it is of paramount importance for physicians to be aware of such rare presentations.

Conflicts of interest

No conflicts of interest.

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Author contribution

Medical Student: case report narration.
Residents: literature review, discussion.
Pathogist: pathology images.

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