CLINICAL VIGNETTE

Breast Pain and Fevers – A Case of Idiopathic Granulomatous Mastitis

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A 31-year-old G0P1 female with recent miscarriage but otherwise no significant past medical history presented with left breast pain and fevers. She initially noticed a painful lump on her left breast a month prior to presentation and subsequently developed fevers and erythema of the overlying skin. Mammogram and ultrasound showed a "large ill-defined mass and associated architectural distortion" measuring at least 6cm with an enlarged axillary lymph node. Given concern for breast abscess, the patient underwent aspiration and was subsequently admitted. In the hospital, she was persistently febrile despite broad spectrum antibiotics. Cultures from the initial aspiration were negative, and the patient subsequently underwent operative incision and drainage (I&D). Intra-operatively she was noted to have a large phlegmon with innumerable tiny loculated purulent pockets. Both surgical cultures and universal PCR were negative. Surgical pathology showed "extensive lobulocentric non-necrotizing granulomatous inflammation with microabscess formation" with rare Gram-positive bacteria most consistent with cystic neutrophilic granulomatous mastitis.

Discussion

Idiopathic granulomatous mastitis (IGM), also referred to as idiopathic granulomatous lobular mastitis is a rare, benign, chronic inflammatory breast disease of unclear origins. It is a diagnosis of exclusion based on pathology after other etiologies including foreign body reaction, tuberculosis, sarcoidosis, fungal infection, and autoimmune diseases such as granulomatosis with polyangiitis and giant cell arteritis are ruled out.

A chart review of over four years of patients at two breast-imaging clinics in Texas found only 90 patients with biopsy-proven granulomatous mastitis.[1] Of these patients, 96.6% were women with a mean age of 35 years old at time of presentation. Interestingly, 88.9% of patients were Hispanic (our patient was not). The majority of women were at reproductive age, 92% were premenopausal or perimenopausal, 63.1% were pregnant within the previous 5 years including 9.5% who were pregnant at the time of diagnosis. Only 1.2% were nulliparous, 10.7% with pregnancy greater than 5 years prior, and 21.4% unknown; 17.2% were using hormonal contraceptives at the time of diagnosis, 39% were not using hormonal contraceptives, with 43.8% unknown. One patient had a personal history of breast cancer while 5.6% had a family history of breast cancer, compared to 7.74% in the general population;[2] IGM does not increase the risk of breast cancer. There has been an association between IGM and elevated prolactin levels, however, our patient's prolactin level was normal.[3]

The most common presentation was a palpable mass 66.7%, followed by unilateral breast pain in 27.8%. Examination is typically notable for palpable mass 83.3%, tenderness to palpation (83.3%), erythema (40%), nipple discharge (31%), and rarely fistula formation (4.7%). Of note, fever was not reported in this study. Initial imaging typically includes an ultrasound, which may show one or more breast masses (92.2%) with ipsilateral axillary adenopathy in approximately a quarter of patients (26.6%). Mammogram findings are more diverse ranging from asymmetry (42%) or mass (30%), to normal mammogram (22%).[1] Based on the clinical presentation and these radiographic findings, IGM may be mistaken as breast abscess or malignancy. Diagnosis is made based on pathology, such as from a core needle biopsy. Pathology typically shows "noncaseating granulomas centered on lobules, with or without associated microabscesses".[4] A subset of patients with IGM show a histologic pattern termed "cystic neutrophilic granulomatous mastitis" (CNGM) which has been associated with although not clearly caused by Corynebacterium species. CNGM is characterized as "distinct enlarged vacuoles within neutrophilic inflammation" with rare Gram-positive bacilli within a cystic space.[5]

The best treatment for IGM is uncertain. Largely, IGM is a self-limited condition that resolves spontaneously, with periods of waxing and waning, over several months average of approximately 6 months, but up to 20 months.[6,7] Approximately 10% of patients have recurrence of IGM.[6] NSAIDs can be effective for pain control. IGM complicated by secondary infection is treated with antibiotics and possibly drainage. If only Corynebacterium is recovered/suspected without known susceptibilities, then doxycycline or linezolid is suggested. Optimal duration of antibiotics is unclear. Furthermore, there is no clear evidence that treatment with antibiotics leads to faster resolution of IGM. For persistent or refractory symptoms, steroids (possibly intralesional) and/or methotrexate have been used in observational studies, but otherwise have not been rigorously studied.[8-10]

Patient Update

One unique feature of our patient's presentation was her fever. Fevers were not directly mentioned in the studies above, suggesting it is not a common feature among patients with IGM.

Given her fevers, there was a strong suspicion for super-infection despite the negative cultures obtained. Given the Gram-positive bacteria seen on pathology and the association of IGM and especially CNGM with Cornyebacterium, our patient was treated with four weeks of doxycycline. On follow-up, she was doing well without any fevers or breast pain, but did have some breast deformity from her surgical I&Ds.

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