Idiopathic granulomatous mastitis - a diagnostic and therapeutic dilemma

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Idiopathic granulomatous mastitis (IGM) is a relatively uncommon benign inflammatory condition of female breast mimicking other chronic inflammatory conditions and even carcinoma breast. Since its first description by Kessler and Wolloch in 1972,[1] only case reports and case series have emerged in the World literature. The affected patient is usually a lactating mother in her third decade (Range 11-80 years) or the one having had finished lactation.[2,3] Patient usually presents with unilateral painful breast mass with erythematous skin mimicking a breast abscess occasionally with discharging sinuses, nipple inversion and axillary lymphadenopathy. In 25% of cases, it is bilateral.[4]

The true prevalence of IGM is unknown. Baslaim et al found 1.8% of cases of IGM out of 1106 women with benign breast diseases.[5] The disease has been found worldwide and in all races, but there is a described predilection for Hispanic and Asian women.[5] In Asian population, it is to be specifically differentiated from tuberculous mastitis and bacterial abscess. Most of the cases might have been treated with anti-tuberculous therapy.

Though the exact cause is still unknown, various mechanisms have been proposed. These include chemical reaction associated with oral contraceptive pills, autoimmune phenomenon, atypical bacterial infection, and localized immune response to extravasated secretions from lobules. Conditions such as pregnancy, breast feeding, breast trauma, hyperprolactinemia with galactorrhea, and alpha-1-antitrypsin deficiency have been associated with an increased risk of IGM.[6] An association with local infection with Corynebacterium kroppenstedtii has recently been suggested but remains unconfirmed.[7] It has been proposed that extravasation of lactational material cause granulomatous inflammation and accumulation of lymphocytes and macrophages which initiates granuloma fornation and microabscesses. Granulomatous inflammation centered on lobules with lymphocytes, plasma cells, epithelioid histiocytes, multinucleated giant cells with neutrophils and superadded necrosis in 11%.[7]

Diagnosis of IGM remains a dilemma. Only histological diagnosis is confirmatory. Mammographic findings are usually ill defined mass, with asymmetric density. This can be seen in normal breast or other types of diseases, including cancer. Ultrasound appearance of IGM is hypoechoic isolated and sometimes multiple hypoecohoic masses with focal areas of mixed echogenicities with parenchymal deformity with or without lymphadenopathy.[8] These findings are also seen in breast carcinoma. Most specific MRI pattern is ring like areas of enhancement or nodular enhancement without mass effects or solid masses with fistulous tract to skin in few patients. These mixed features are also present in other breast conditions especially in tuberculous mastitis. Radiological diagnosis can play an adjuvant role but cytological or histological evaluation is must for accurate diagnosis.

FNAC has low specificity and sensitivity to differentiate IGM from other forms of mastitis and its usefulness is debatable but it is relatively cheaper and easily available. However, core biopsy or vacuum assisted biopsy usually gives diagnosis and helps in differentiating it from other conditions.[6,9,10] Recently, US-guided core biopsy has been used to obtain specimen with minimal disturbance of tissue architecture. Core biopsy is diagnostic in 75% of IGM patients who underwent US-guided biopsy. Surgical biopsy is most conclusive and ideal in granulomatous mastitis giving definite histological diagnosis and therapeutic as well. Role of PCR to rule out IGM has been reported.[11,12]

No definite treatment has been formulated for IGM

yet. The first step is to get histological diagnosis and exclude other forms of mastitis and malignancy to avoid inappropriate and unnecessary treatment. Mainstay of treatment is antibiotics and surgical drainage. Surgical excision gives better results and gives exact diagnosis. Surgical management includes incision and drainage, wide local excision and quadrantectomy. Wide local excision is considered to be the most effective with low recurrence. In all surgical procedures, cultures should be sent to rule out bacterial mastitis. Rarely, mastectomy is recommended for resistant or relapsing cases and in few cases, multiple surgeries are required.[12,13] Role of medical treatment is more in avoiding recurrence. Steroids can be used before and after surgery.[14] Infectious pathology must be rule out before introducing steroids otherwise condition gets worse. Besides usual side effects of steroids most of patients relapse after leaving steroids which can be as high 50%.[14] In resistant cases, role of methotrexate and azathoiprone is documented with variable response.[11,14]

IGM is usually a self-limiting disease. Patient's understanding is very important in this disease to minimize psychological trauma.[15] Women often get depressed due to chronic nature of disease and recurrences and may need antidepressants as adjuvants.[16] Regardless of therapeutic intervention, the condition may take about 6 to 12 months to resolve completely.

In summary, despite advances in diagnostic modalities and better understanding of histological features, the diagnosis as well as treatment of idiopathic granulomatous mastitis remains a dilemma. A high index of suspicion is required to diagnose such cases and differentiate them from other chronic infectious diseases especially tuberculosis and carcinoma of breast.

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