CASE REPORT

Idiopathic granulomatous mastitis, erythema nodosum and bilateral ankle arthritis in an Iranian woman

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SUMMARY

Here we report a case of idiopathic granulomatous mastitis (IGM) associated with erythema nodosum (EN) and ankle arthritis. The skin, joint and mammary symptoms improved with corticosteroid. Coincidence of granulomatous mastitis, EN and arthritis is a rare feature.

BACKGROUND

Idiopathic granulomatous mastitis (IGM) is a term that has been proposed for a granulomatous inflammatory process of the breast characterised by the presence of non-caseating granulomas confined to breast lobules, in which no microorganisms are found. The cause of this uncommon lesion is obscure but it is suggested that the disease may be immunologically mediated. It can simulate malignancy clinically and on mammography, ultrasound and MRI.[1] On the other hand, erythema nodosum (EN) is an acute, painful, erythematous, nodular eruption. EN may be associated with numerous conditions. The occurrence of EN as a response to medications and to tuberculin skin testing in patients with a positive reaction suggests that a type IV delayed hypersensitivity reaction may play an important role.[2] In this paper, we aimed to present a woman with IGM accompanied with EN and oligoarthritis, which were successfully treated with corticosteroid. EN should be kept in the mind as an extramammary manifestation of IGM. Coincidence of these diseases supports the theory that there is an autoimmune component in the aetiology of IGM.

CASE PRESENTATION

A 40- year-old woman presented a 2-month history of a painful lump in her left breast, and 2-week history of tender, red, plaques on both shins and arthralgia affecting both ankles. All her children were breast-fed. There was no nipple discharge. She had not taken the contraceptive pill or any other medication. On physical examination she was febrile (her body temperature was 38°C) and we noted a firm mass 8×5 cm in her left breast with overlying skin erythema and mild retraction of the nipple (figure 1). Dark reddish plaque skin lesions were found over both legs (figure 2), and arthritis involving both ankles. There was no lymphadenopathy.

INVESTIGATIONS

Her blood test results showed the following: white blood cells 12×10^3/µl, neutrophils 80%, lymphocytes 20%, platelets 400×10^3/µl and an erythrocyte sedimentation rate was 70 mm/h. Her blood culture revealed no growth, while her chest radiography was unremarkable. A fine needle aspiration cytology (FNAC) was performed. No organisms were seen on Gram, periodic acid-Schiff (PAS) and Ziehl-Neelsen stainings. Microscopic examination revealed aggregates of epithelioid histiocytes with reniform-to-oval nuclei and a moderate amount of cytoplasm (figure 3). In addition multinucleated giant cells and other inflammatory cells including neutrophils and lymphocytes were seen. No caseous necrosis was seen. Tuberculin skin testing was negative.

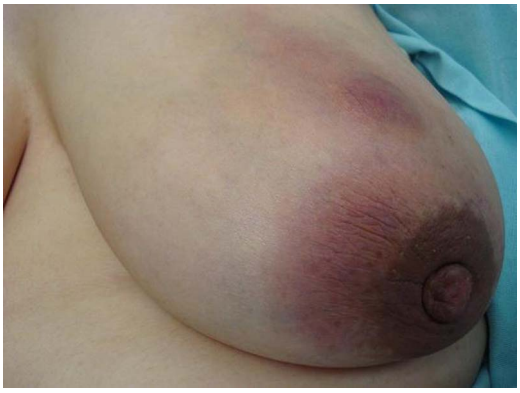

Figure 1 Photograph shows redness of the breast and mild nipple retraction.

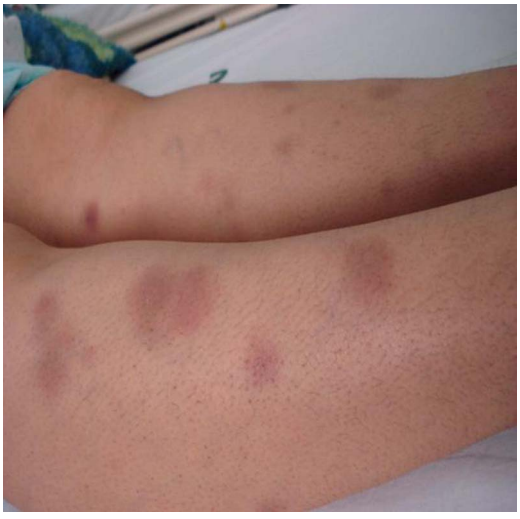

Figure 2 Dark reddish plaque skin lesions over both legs.

To cite: Binesh F, Shiryazdi M, Bagher Owlia M, *et al*. *BMJ Case Reports* Published online: [*please include* Day Month Year] doi:10.1136/bcr-2012-007636

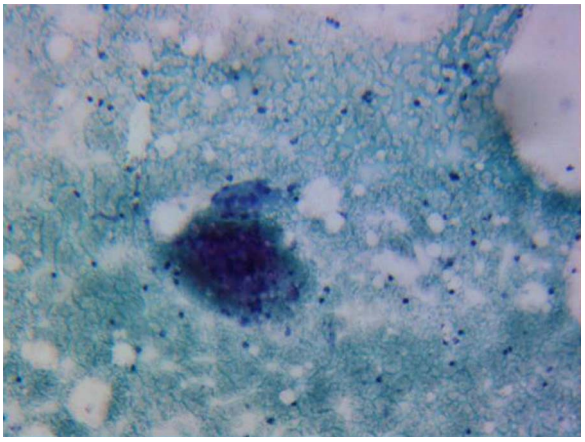

Figure 3 Smear shows aggregates of epithelioid histiocytes with multinucleated giant cells (Papanicolaou stain ×10).

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of IGM including specific infections, vasculitis, sarcoidosis and foreign body reaction should be excluded.[3 4]

TREATMENT

The patient received 4 mg dexamethasone three times a day for 1.5 days. After that dexamethasone was discontinued and she received 30 mg prednisolone, which after 1 week was halved and slowly was tapered.

Treatment with corticosteroid produced a rapid resolution of the fever, EN, breast symptoms and arthritis.

OUTCOME AND FOLLOW-UP

One month follow-up showed that the breast mass gradually has become smaller.

DISCUSSION

Granulomas in the breast are caused by a wide variety of diseases, all present in less than 1% of the breast biopsies.[5] Two types of GM are identified: idiopathic and specific. Specific granuloma may be seen in tuberculosis (TB), mycotic infection and parasitic infestation. IGM is a rare inflammatory disorder, which is characterised by the presence of non-caseating granuloma within the breast lobules and absence of any organisms. IGM was first described as a specific entity by Kessler and Wolloch.[6] It was further elaborated by Cohen in 1977.[5] The cause of this uncommon lesion is obscure.[6 7] It usually presents as a mass, nearly always in young parous women, and is often related to recent pregnancy. Our patient was 40 years old. Clinically, the lesion can simulate carcinoma. Although the lobulocentricity of the granulomas and the presence of neutrophils should raise the possibility of IGM, other causes of granulomatous inflammation (such as infections, sarcoidosis and reaction to foreign materials) should always be excluded. EN is the most frequent clinico-pathological variant of the panniculitides. The disorder is a cutaneous reaction consisting of inflammatory, tender, nodular lesions, usually located on the anterior aspects of the lower extremities. EN was originally described in 1798 by Willan.[8] A review of the literature reveals that the list of aetiological factors which can lead to EN is long and varied, including infections, drugs, malignant diseases and a wide group of miscellaneous conditions.[9] Association of IGM with EN, a rare case, at the first time, presented in 1987 by Adams.[10] After that some cases of IGM with EN and or oligoarthritis and episcleritis have been presented from other countries and EN has been diagnosed as an extramammary manifestation of IGM. Weber *et al*[11] reported a case of GM associated with EN and oligoarthritis. There are also two case reports from Iran which are suggestive of such associations.[12 13] Cemal Bes *et al*[14] and Nohira *et al*[15] have also described two cases of IGM and EN. Association of GM with EN and arthritis supports the theory that there is an autoimmune component in the aetiology of IGM. In endemic TB regions such as our country, a painful breast mass with cutaneous manifestation of EN is clinically relevant to determine a diagnosis of breast TB. Diagnosis should be confirmed by histopathological findings, culture as well as molecular detection of *Mycobacterium tuberculosis* using PCR. Our patient underwent FNAC because FNAC of breast lump is accepted worldwide and is an established method of choice to determine the nature of breast lump. It is cheap, safe and highly accurate method for diagnosis of breast lump preoperatively to avoid undue surgery and inconvenience during biopsy. In addition aspirated culture can also be used for ancillary testing.[16] In case of our patient, no organisms were seen on Gram, PAS and Ziehl-Neelsen stainings and the bacterial, fungal and mycobacterium cultures of the aspiration specimen were all negative. Since the chest x-ray was normal and Mantoux test was negative, TB or sarcoidosis was excluded. Sarcoidosis constitutes one common aetiological factor in adult patients with secondary EN and granulomatous inflammation of breast in some countries.[17] A chest x-ray should be performed in all patients to rule out pulmonary diseases as the cause of the cutaneous and breast diseases. There is no definite accepted treatment for IGM. In 1980, a short course of high-dose prednisone (60 mg/day) was recommended.[18] In the current case, treatment with corticosteroid produced a rapid resolution of the fever, EN, breast symptoms and arthritis. Thereafter the breast mass gradually became smaller. In conclusion, IGM may rarely be associated with EN. Association of GM with EN and arthritis supports the theory that there is an autoimmune component in the aetiology of IGM. Tissue biopsy or cytological examination remains the gold standard for diagnosis.

Learning points

- Erythema nodosum should be kept in the mind as an extramammary manifestation of idiopathic granulomatous mastitis (IGM).
- Coincidence of these diseases supports the theory that there is an autoimmune component in the aetiology of IGM.
- A chest x-ray should be performed in all patients to rule out pulmonary diseases such as tuberculosis or sarcoidosis as the cause of the cutaneous and breast diseases.
- Tissue biopsy or cytological examination remains the gold standard for diagnosis.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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