Other full case

Idiopathic granulomatous mastitis: a great imitator?

Annabel M Ruiter, Irene L Vegting, Prabath W B Nanayakkara

Department of Internal Medicine, VU University Medical Center, Amsterdam, The Netherlands

Correspondence to Prabath W B Nanayakkara, p.nanayakkara@vumc.nl

Summary

The patient in this case was seen by numerous doctors for a long time before the diagnosis was made because, right from the start, an abscess or a cancer of the breast was suspected. She was first seen by her general practitioner (GP); thereafter, she went to two consultant physicians and a surgeon in a regional hospital and eventually an idiopathic granulomatous mastitis (IGM) diagnosis was made by a specialist in internal medicine in a university hospital. When the diagnosis IGM was made, the patient was treated with steroids and made a complete recovery. Although the incidence of IGM is not high, the GPs are likely to see these patients initially and should be aware of the existence of this disease, which may spare the patient unnecessary consultations, diagnostics tests or even mastectomy.

BACKGROUND

Idiopathic granulomatous mastitis (IGM) is an inflammatory disease of the breast with an unknown aetiology. However, granulomatous mastitis can mimic many conditions and is often misdiagnosed. Patients are mostly first seen by a general practitioner (GP) and thereafter referred to many specialists and undergo many tests before the diagnosis is made. Therefore it is important for GPs and first and second line specialists to recognise the symptoms and to have some knowledge of this condition. We ourselves had great difficulty in starting prednisolone in this patient because, in spite of the extensive evaluation, we were not certain whether the symptoms still could be due to an infection.

CASE PRESENTATION

A 33-year-old Turkish woman presented for a second opinion to our hospital with an extremely tender left breast. The

complaints started 6 months ago with an expanding sore swelling in her breast. After a few weeks she developed numerous fistulas around her areola with production of yellowish, granular pus. She had no fever, weight loss, night sweats, joint pains, easy bruising, cough or dyspnoea on exertion. She had also developed painful, red spots on her lower legs, a few months ago, which disappeared after some time. She also complained of pain in her small joints, without redness or swelling, for which she was taking diclofenac 50 mg.

When presented to our hospital we saw a tired 33-year-old woman with numerous fistulas, filled with pus, in her left breast. The whole breast was extremely painful and examination was almost impossible but we could feel an extremely tender diffuse swelling of around 10×4 cm.

Her medical history was uneventful. Her youngest child was 6-year-old and she had breast fed for a few days. She

Figure 1 Photograph of breasts showing several pus-filled fistulas of the left breast.

was not pregnant at presentation. There was no family history of breast or other cancers. She presented to her GP about 6 months before presentation who diagnosed a breast abscess and prescribed five courses of augmentin but her condition deteriorated. She was referred to a regional hospital and underwent a detailed diagnostic workup. The diagnosis was unclear and she was referred to us.

INVESTIGATIONS

On examination she had no fever and an extremely tender mass about 10×6 cm that could be palpated on the base of her left breast. There were around four fistulas around her areola, which discharged yellowish granular pus when pressuring the breast (figure 1). We could also see many healed scars around her areola.

Her laboratory results were erythrocyte sedimentation rate 70 mm/h, C reactive protein 74 mg/l, haemoglobin 6.8 mmol/l, white blood cells $40 \times 10^9/l$ (differentiation: eosinophils $2 \times 10^9/l$, neutrophils $53 \times 10^9/l$, lymphocytes $37 \times 10^9/l$, monocytes $8 \times 10^9/l$), creatinine 62 $\mu\text{mol/l}$, angiotensin converting enzyme level 12 U/l, aspartate transaminase 18 U/l, alanine transaminase 11 U/l and bilirubin 4 $\mu\text{mol/l}$. Microscopic examination of the urine was normal. Antinuclear antibodies were negative, classical antineutrophil; cytoplasmic antibodies were negative; rheumatoid factor was negative; and anticyclic citrullinated peptide was negative.

An ultrasound-guided needle aspiration was performed and histopathology showed a granulomatous inflammation with large numbers of neutrophils and multinucleated giant cells. No acid-resistant rods were found. Repeated cultures of the pus showed no growth. PCR for *Mycobacterium*

tuberculosis, *Norcardia*, actinomycosis and eubacterial-PCR were negative.

An ultrasound examination showed an inhomogeneous hypoechoic mass located mostly at the left upper quadrant but spread over the whole breast with multiple cavities filled with a fluctuating fluid—possibly pus. A T1-weighted MRI of the breast was made and an extensive mastitis was seen. There were several well-defined pus collections and fitful staining implying malignant disease. Mammography comparing left to right showed an irregular parenchyma at the left side and some nodular thickening. At the right side there were no abnormalities.

DIFFERENTIAL DIAGNOSIS

IGM is a diagnosis per exclusion; other causes should be excluded before diagnosing IGM. Differential diagnosis of IGM: breast cancer, abscess, infection, tuberculosis, sarcoidosis and idiopathic granulomatous mastitis.¹

TREATMENT

A half year after presentation to our hospital, 14 months after presentation to her GP, a diagnosis of IGM was made and the patient was put on prednisone 60 mg a day for 2 weeks, which was tapered to 20 mg a day for 6 weeks. With the introduction of prednisolone there was a dramatic reduction of the pain and swelling of her breast. Within 2 months after initiating the treatment there was no evidence for any indurations of fistulas. Patient was put on 2.5 mg prednisone a week. In 5 months prednisolone was tapered off but her symptoms recurred. The patient also developed a typical 'moonface' and put on a few pounds. We then increased the dose of prednisolone to 20 mg a day for 2 weeks and then 10 mg a day and methotrexate 5 mg a week was added.

OUTCOME AND FOLLOW-UP

Eighteen months after initiating the methotrexate, prednisone was stopped and methotrexate was continued for another 10 months. There was a complete normalisation of the mammography. At the moment, after 3 years without prednisone and 2 years without methotrexate, the patient is still asymptomatic.

DISCUSSION

Our patient came to us with a painful swelling in her left breast with numerous fistulas producing a yellowish material. One year after presentation to her GP, IGM was diagnosed. IGM is an uncommon benign chronic disease of unknown origin; it represents 1.6% of all breast diseases seen in the second line.^{2–6} IGM is characterised by the presence of granulomas and neutrophils. Its clinical manifestations mimic numerous diseases like sarcoidosis, Wegener's disease, tuberculosis, bacteria, mycobacterium and a breast malignancy.^{3 5 7 10 11} Because of the unknown aetiology and the resemblance of symptoms with other diseases, IGM is mostly a diagnosis per exclusion. The average time before the right diagnosis is approximately 7–12 months (14 months in our case).^{7 10}

Often ineffective cures of antibiotics are given under the suspicion of infection or an abscess and occasionally mastectomy or chemotherapy is performed when confused with a malignancy of the breast. Therefore, it is important that thorough investigation is performed through a fine needle aspiration or biopsy to exclude all possible causes and to set the right diagnosis. In a large study with 2053 patients to identify infiltrating breast lesion, the sensitivity of fine needle aspiration under ultrasound guidance was 95%, the specificity 86% with a positive predicting value of 0.86 and a negative predicting value of 0.73.¹² Corticosteroids are the first line of treatment if possible in combination with methotrexate as low doses of methotrexate have a corticosteroid-sparing effect.^{4 7 10 13} Recurrent disease rates can be up to 50%.^{3 13}

Because of the beneficial effects of corticosteroids, it is believed there is an autoimmune component in IGM.^{4 7 8 10 13} and it seems to be that there is a correlation between IGM and (multi)parous women with a history of breast feeding.^{3 4 8–11}

Learning points

- ▶ GPs should keep in mind IGM in their differential diagnosis.
- ▶ Other conditions must be excluded with cultures and a biopsy to provide the patient the best treatment as possible and to prevent unnecessary interventions.
- ▶ Treatment with corticosteroids, if possible in combination with prednisone, should be preferred above a surgical excision to treat IGM. Long-term follow-up should be considered for at least 12–24 months.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Kamal RM**, Hamed ST, Salem DS. Classification of inflammatory breast disorders and step by step diagnosis. *Breast J* 2009;**15**:367–80.
2. **Nazer MA**. Idiopathic granulomatous mastitis. A forgotten clinical diagnosis. *Saudi Med J* 2003;**24**:1377–80.
3. **Diesing D**, Axt-Fliedner R, Hornung D, *et al*. Granulomatous mastitis. *Arch Gynecol Obstet* 2004;**269**:233–6.
4. **Kim J**, Tymms KE, Buckingham JM. Methotrexate in the management of granulomatous mastitis. *ANZ J Surg* 2003;**73**:247–9.
5. **Molckovsky A**, Fitzgerald B, Freedman O, *et al*. Approach to inflammatory breast cancer. *Can Fam Physician* 2009;**55**:25–31.
6. **Baslaim MM**, Khayat HA, Al-Amoudi SA. Idiopathic granulomatous mastitis: a heterogeneous disease with variable clinical presentation. *World J Surg* 2007;**31**:1677–81.
7. **Bes C**, Soy M, Vardi S, *et al*. Erythema nodosum associated with granulomatous mastitis: report of two cases. *Rheumatol Int* 2010;**30**:1523–5.
8. **Elsiddig KE**, Khalil EA, Elhag IA, *et al*. Granulomatous mammary disease: ten years' experience with fine needle aspiration cytology. *Int J Tuberc Lung Dis* 2003;**74**:365–9.
9. **Kuba S**, Yamaguchi J, Ohtani H, *et al*. Vacuum-assisted biopsy and steroid therapy for granulomatous lobular mastitis: report of three cases. *Surg Today* 2009;**39**:695–9.
10. **Larsen LJH**, Peyvandi B, Klipfel N, *et al*. Granulomatous Lobular Mastitis: Imaging, Diagnosis and Treatment. *AJR* 2009;**193**:574–81.
11. **Schamajuk G**, Genovese MC. First report of idiopathic granulomatous mastitis treated with methotrexate monotherapy. *J Rheumatol* 2009;**36**:1559–60.
12. **Kuo YL**, Chang TW. Can concurrent core biopsy and fine needle aspiration biopsy improve the false negative rate of sonographically detectable breast lesions? *BMC Cancer* 2010;**10**:371.
13. **Marriott DA**, Russell J, Grebosky J, *et al*. Idiopathic granulomatous lobular mastitis masquerading as a breast abscess and breast carcinoma. *Am J Clin Oncol* 2007;**30**:564–5.

This pdf has been created automatically from the final edited text and images.

Copyright 2010 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Ruiter A M, Vegting I L, Nanayakkara. Idiopathic granulomatous mastitis: a great imitator?. *BMJ Case Reports* 2010;10.1136/bcr.03.2010.2844, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow