Breast vasculitis in association with breast gigantism in a pregnant patient with systemic lupus erythematosus

D J Propper, D M Reid, L Stankler, C J Eastmond

Abstract
A 24 year old woman with systemic lupus erythematosus (SLE) developed widespread necrotic skin ulceration and gigantism of both breasts during an exacerbation of SLE in the last trimester of her second pregnancy. Over the remainder of the pregnancy the ulceration was only controlled by high dose corticosteroids. After parturition, however, it was possible to reduce the steroid dose without recurrence of the ulceration.

Cutaneous vasculitis is a well recognised feature of systemic lupus erythematosus (SLE), but there are no reports of such vasculitis affecting the breasts. We describe a patient with SLE who developed florid cutaneous vasculitis and gigantism of both breasts during pregnancy.

Case report
A 21 year old woman presented with a six month history of symmetrical polyarthritis. There was no other significant history, and the clinical abnormalities were confined to a symmetrical arthritis affecting the knees and the proximal interphalangeal joints of both hands.

Results of investigations were as follows: erythrocyte sedimentation rate 40 mm in the first hour, antinuclear antibody positive (1/128), rheumatoid factor negative, DNA binding 53 U/l (normal <10 U/l), C3 88 U/l (normal 70–180 U/l), C4 13.6 U/l (normal 14–70), LE cells were present in the peripheral blood, haemoglobin 113 g/l, white cell count 6.5 x 10^9/l. Results of urea, electrolytes, creatinine, transaminases determinations, and urine analysis, electrocardiography, joint and chest radiography were all normal.

A presumptive diagnosis of SLE was made, and treatment with indomethacin was started, with clinical benefit. One year later the patient became pregnant. During the third trimester her arthritis deteriorated, and she developed Raynaud’s phenomenon and digital infarcts. In addition, her breasts enlarged from a measurement before pregnancy of 91 cm to 132 cm. Treatment was continued with indomethacin, and no steroids were given. Subsequently, a full term healthy baby was delivered. Hydroxychloroquine 200 mg/day was then started, with marked clinical improvement. Over the post-partum period her breasts gradually returned to their size before pregnancy.

Eighteen months later hydroxychloroquine was stopped as she wished to conceive, and five months later she became pregnant. By 26 weeks’ gestation her breasts were again considerably enlarged, and had developed widespread confluent areas of necrotic ulceration (figure). At the time there was no clinical evidence of active SLE, though serological results were as follows: erythrocyte sedimentation rate 130 mm in the first hour, DNA binding 61%, C3 110 U/l, and C4 10.2 U/l. Anticardiolipin antibodies were not detected in serum samples obtained throughout pregnancy. Cultures of swabs taken from the ulcers were sterile.

Treatment consisted of prednisolone 60 mg/day for three weeks and topical paraffin gauze, with which the ulcers began to heal. It was not, however, possible to reduce the dose of prednisolone below 30 mg/day without an exacerbation in the ulceration. For the remainder of pregnancy treatment was therefore continued with prednisolone 30 mg/day, and the ulcers continued to heal.

At 32 weeks’ gestation she developed a perforation of the cartilaginous nose septum. Further examination showed that the ears, nose, and throat were normal, as were the skull and sinuses as shown by radiography. Antineutrophil cytoplasmic antibodies were not detected in the serum. Histological examination of a nasal sepal biopsy specimen showed hyperkeratotic squamous epithelium and non-specific inflammatory granulation tissue, but no evidence of vasculitis or giant cells. There were no further complications during the remainder of the pregnancy, and she delivered a healthy baby weighing 2.8 kg at term.

After parturition the breast ulceration resolved fully, and over the next four months it was possible gradually to reduce the dose of prednisolone to 5 mg/day without any recurrence in the ulcers. Her breasts, however, remained markedly enlarged, and one year later she underwent bilateral mammoplasty. Breast histology showed intralobular fibrosis, but no evidence of vasculitis. Since that pregnancy her SLE has been quiescent.

Discussion
Skin ulceration is a well recognised feature of SLE, and is a manifestation of cutaneous vasculitis. There are, however, no reports of breast ulceration in association with SLE. In fact, reports of breast involvement during the course of connective tissue diseases are rare and confined to patients with either Wegener’s granulomatosis, polyarteritis nodosum, or giant cell arteritis. Although our patient did sustain a nasal perforation, there were no other features...
Breast enlargement and vasculitic lesions at 26 weeks' gestation.

suggestive of Wegener's granulomatosis. Moreover, nasal perforation is a rare but reported complication of SLE, and there were sufficient clinical and serological manifestations to fulfil American Rheumatism Association criteria for SLE.

Although a skin biopsy was not performed, the ulceration was thought to be vasculitic in origin on the grounds of macroscopic appearance, the pattern of response to corticosteroids, and the serological evidence of active disease. The observation that high doses of prednisolone were required for continued healing during pregnancy, but could be reduced in the post-partum period, suggests that pregnancy had some role in the cause of the ulceration.

It is widely believed that pregnancy can exacerbate SLE, though this has been disputed. The marked deterioration of disease during both pregnancies together with subsequent quiescent disease supports a pregnancy induced exacerbation as the cause of her ulceration. It is likely that the breast enlargement contributed to the severity of the ulceration as cutaneous vasculitis can be exacerbated by local stress or trauma.

Breast vasculitis in association with breast gigantism in a pregnant patient with systemic lupus erythematosus.

D J Propper, D M Reid, L Stankler and C J Eastmond

doi: 10.1136/ard.50.8.577

Updated information and services can be found at:
http://ard.bmj.com/content/50/8/577

These include:

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/