LESSON OF THE MONTH

Pneumococcal septicaemia in a patient with systemic lupus erythematosus

A R Mackenzie, R B S Laing, A G MacDonald, C C Smith

Case report
A thirty eight year old woman with insulin dependent diabetes mellitus and systemic lupus erythematosus (SLE) was admitted with a two day history of fever, rigors, and diarrhoea. SLE was diagnosed at the age of 18 years when she presented with joint pain, a positive antinuclear antibody (ANA), and severe autoimmune thrombocytopenia, which was treated with corticosteroids. At no stage of her illness did she have evidence of renal, central nervous system, muscle or skin involvement. She had also suffered four miscarriages in association with anticardiolipin antibodies. There was no history of arterial or venous thrombosis and her disease had been inactive for a number of years.

When reviewed as an outpatient, two months before admission, she was well with no clinical manifestations of SLE. ANA was positive (1:160), extractable nuclear antigen screen negative, C3 141 g/l (88–201), C4 23.1 g/l (16–47), DNA binding 31 U/l (0–7), anticardiolipin antibody was 30 gpl units (<10) and lupus anticoagulant was not assayed. Three sets of blood cultures all grew Streptococcus pneumoniae. Stool culture on admission grew no enteric pathogens and her diarrhoea was thought to be secondary to the bacteraemic illness. Chest and sinus x rays were normal. An abdominal ultrasound scan revealed that the spleen was too small to visualise.

She was treated for septic shock with oxygen, intravenous colloid expansion, and intravenous cefotaxime pulses with an insulin infusion and made a full recovery over the next five days. She was discharged from hospital while receiving oral amoxycillin and was subsequently started with long term prophylactic nephophenoxymethyl penicillin and vaccinated against Streptococcus pneumoniae, Neisseria meningitidis, and Haemophilus influenzae type b.

Discussion
Impaired splenic function in SLE patients was first described in 1979 and was followed by reports of splenic atrophy. The clinical implications of these complications were highlighted by a series of case reports describing fatal or near fatal septicaemia in patients with SLE and hyposplenism. Splenic dysfunction is suggested by the presence of Heinz and Howell-Jolly bodies in the red cells, as occurred in our patient, and should be considered in patients with a thrombocytopenia or monocytosis on the blood film. The regular examination of peripheral blood films may permit early detection of hyposplenism but the finding of a normal full blood count two months before our patient’s admission highlights the comparatively short period over which hyposplenism may develop. Furthermore, this patient’s inactive disease (as suggested by her normal DNA binding titre and complement values) did not preclude the development of hyposplenism.

The mechanisms by which hyposplenism develops in SLE are uncertain. Splenic infarction has been described in association with the
antiphospholipid syndrome and thrombosis has been suggested as a possible mechanism for hyposplenism in SLE.

A recent review of published reports revealed 13 SLE patients with sepsis with pneumococcal sepsis in six and a salmonella septicaemia in one. All of these patients were female, nine had hypocomplementaemia and only two of the septicaemic patients survived. The duration of SLE ranged from two months to 14 years.

This patient’s SLE activity was thought to be low because of normal values of complement, DNA binding, and lymphocytes. The C reactive protein is often normal in SLE but would be expected to rise in bacterial sepsis. Our patient’s clinical presentation, coupled with her positive blood cultures, put the diagnosis of sepsis beyond doubt and her normal C reactive protein might be explained by the fact that this acute phase reactant can take 48 hours to rise in response to infection.

Loose stools or frank diarrhoea are common in septicaemia but the clinical features of this case suggested that sinusitis was the primary focus of infection. Enteric infection such as Salmonellosis must always be excluded in such patients.

Recent British guidelines for patients with an absent or dysfunctional spleen have not included patients with SLE. In view of the potentially fatal complications, we would suggest that patients with SLE who have any features of hyposplenism on blood film should be actively considered for pneumococcal prophylaxis—that is, pneumococcal vaccination and long term therapy with phenoxyethyl penicillin. The Haemophilus influenzae type b vaccine should also be given and vaccination against Neisseria meningitidis should be considered. As the response to pneumococcal vaccination may be better in patients with normal splenic function than in those with hyposplenism or asplenism and as this case shows the difficulty in identifying those most at risk of hyposplenism, it might be argued that all patients with SLE, 5% of whom are likely to develop hyposplenism, receive pneumococcal vaccination.

The lesson

- Hyposplenism is a predisposing factor to septicaemia in patients with SLE.
- Hyposplenism is frequently found in clinically inactive SLE.
- Regular monitoring of the blood film is required to detect early hyposplenism and start appropriate prophylaxis against infection.

Pneumococcal septicaemia in a patient with systemic lupus erythematosus

A R Mackenzie, R B S Laing, A G MacDonald and C C Smith

Ann Rheum Dis 1997 56: 403-404
doi: 10.1136/ard.56.7.403

Updated information and services can be found at:
http://ard.bmj.com/content/56/7/403

These include:

References
This article cites 10 articles, 4 of which you can access for free at:
http://ard.bmj.com/content/56/7/403#BIBL

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.

Topic Collections
Articles on similar topics can be found in the following collections

- Immunology (including allergy) (5144)
- Connective tissue disease (4253)
- Systemic lupus erythematosus (571)
- Clinical diagnostic tests (1282)
- Degenerative joint disease (4641)
- Musculoskeletal syndromes (4951)
- Pain (neurology) (883)
- Radiology (1113)
- Radiology (diagnostics) (750)

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/