LESSON OF THE MONTH

Pneumococcal septicaemia in a patient with systemic lupus erythematosus

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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. 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 phenoxymethyl penicillin and vaccinated against *Staphylococcus 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 thrombocytosis 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 functional asplenia, seven of whom developed 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 phenoxymethylpenicillin. 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 hyposplenic or asplenic, 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.

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