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W. F. NG, P. COHEN1, A. HEPBURN, S. HAMDULAY, M. CARPANI2 and J. C. MASON

Rheumatology Section, Eric Bywaters Centre, Imperial College London, 1Department of Histopathology, Charing Cross Hospital and 2Gastroenterology Unit, Hammersmith Hospital, London, UK
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Correspondence to: W. F. Ng, Department of Rheumatology, Ealing Hospital, Uxbridge Road, Middlesex UB1 3HW, UK.
E-mail: faingl@doctors.org.uk


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Pneumococcal sepsis after autosplenectomy in a girl with systemic lupus erythematosus

Sir. Autosplenectomy is infrequently described in systemic lupus erythematosus (SLE) [1, 2]. Asplenia is associated with high mortality due to Streptococcus pneumoniae infection [2–6]. In this case report, we describe a girl who survived pneumococcal sepsis and meningitis. Our observation supports the application of pneumococcal vaccine in SLE patients.

A 15-yr-old girl was treated with prednisolone (5 mg/48 h) and fosinopril due to SLE complicated by nephritis and arterial hypertension. After years of inactive disease she was admitted with fever, headache, cough and tachypnoea.

The diagnosis of SLE was established at the age of 6yr. Subsequent manifestations included lupus nephritis with nephrotic syndrome, Sjögren’s syndrome, pancreatitis, haemolytic anaemia and arterial hypertension. There was no history of anticardiolipin antibodies or thrombosis. Treatment consisted of corticosteroids, mexitoxetane followed by cyclophosphamide and azathioprine.

The patient was seen 3 months earlier with no clinical manifestations of SLE. At that time the ANA titre was 1:640, antibodies to double-stranded DNA were slightly elevated and complement levels were within the normal range.

On admission, physical examination revealed tachypnoea, a depressed level of consciousness and cold, lilac-livid coloured fingers and toes. There was no stiffness of the neck. The blood pressure was decreased and the girl was anuric. Body temperature was elevated to 38.8°C. The clinical presentation was compatible with sepsis.

Laboratory tests showed thrombocytopenia, leucopenia, neutropenia, highly increased C-reactive protein, elevated creatinine, ASAT and ALAT, as well as abnormal coagulation tests, indicating intravascular coagulation. The ANA titre was 1:640 and antibodies to DNA were 52 U/l (normal range <75 U/l). Antiphospholipid antibodies were undetectable. Complement levels were normal. The blood film showed anisocytosis and Howell–Jolly bodies. Streptococcus pneumoniae could be cultivated from blood and cerebrospinal liquor.
The chest X-ray showed no infiltration. Computed tomography scanning of the paranasal sinus revealed sinusitis. Fundoscopy was normal, without signs of elevated intracranial pressure. Cerebrospinal fluid opening pressure was not measured.

The spleen was undetectable by ultrasound scan. MRI of the abdomen showed a thin strand of tissue in the area of the spleen (Fig. 1A and B).

The patient was treated for septic shock and respiratory failure in the intensive care unit for 9 days; artificial ventilation was necessary for 4 days. Blood pressure stabilization required dopamine and epinephrine. Appropriate antibiotics were given. During the first 2 days fever was continuously elevated to about 38.8°C, followed by 3 days with spiking fever. After 3 critical days there was a gradual improvement in clinical condition. She recovered fully and was discharged from hospital with prophylactic penicillin treatment. She has been vaccinated with 23-valent pneumococcal vaccine and the pneumococcal antibody titre increased to 469 mg/l.

SLE typically involves multiple organ systems. The immune system can show variable abnormalities [7]. It has been suggested that SLE patients demonstrate an increased risk of infection even if they are not treated with immunosuppressants [7]. Possible factors increasing this risk are abnormalities in the complement system, including complement receptors, deficiency of mannose-binding lectin, impaired chemotaxis and phagocytosis of macrophages and polymorphonuclear cells, abnormal T-cell-mediated cytotoxicity, and functional asplenia [7].

The association of functional asplenia and SLE was first described in 1979 [1]. Functional asplenia occurred in approximately 5% of SLE patients [2]. Asplenia may result in impaired elimination of microorganisms and immune complexes and in a decrease in immunoglobulin M serum levels. This leads to increased susceptibility to pneumococcal and salmonella sepsis.

Howell–Jolly bodies, spherocytes and target cells in the red cell population, thrombocytosis and monocytosis are suggestive of splenic dysfunction [3, 4]. The mechanism by which hypoplasenism or asplenism develops in SLE is rather unclear. Some authors postulate splenic infarction caused by the antiphospholipid syndrome and thrombosis as a possible mechanism, others suggest vasculitis with silent ischaemia or lymphocyte depletion caused by the presence of a circulating lymphocytotoxin [2, 8, 9].

Fifteen SLE patients with functional asplenia, of whom seven developed sepsis caused by *S. pneumoniae* (six cases) and by *Salmonella* species were reported in one review [4]. All were female, aged between 9 and 44 yr, and only two survived. None of them had received pneumococcal vaccination.

Recommendations from the case are as follows: (i) early vaccination with the 23-valent or the pneumococcal conjugate vaccine is mandatory in patients with asplenia; (ii) regular monitoring for Howell–Jolly bodies is necessary to detect splenic dysfunction; and (iii) adequate anti-infectious prophylaxis in patients with splenic dysfunction is recommended.

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R. **Hu¨hn**, H. **Schmeling**, C. **Kunze** and G. **Horneff**

Department of Paediatrics, Asklepios Clinics Sankt Augustin, Germany

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Correspondence to: G. Horneff, Department of Paediatrics, Asklepios Clinics Sankt Augustin, Arnold Janssen Str. 29, D-53757 Sankt Augustin, Germany.

E-mail: g.horneff@asklepios.com


In the histopathological sections there was a moderate lymphocytic infiltration in the lobules of the panniculus and fibrocytes invading the lobules from the fibrous septae. Work-up at that time also included antinuclear antibodies (ANA), rheumatoid factor (RF), C-reactive protein (CRP), anti-double-stranded DNA, anticytochrome-C oxidase, anti-Scl-70 and cryoglobulins, which were all negative. A complete blood count was normal. With these findings and lack of any systemic manifestations, a diagnosis of Rothmann–Makai syndrome was established. The patient received an 8-month course of steroids. The erythema resolved and the patient was left with two slightly atrophic areas in both calves. No relapse has occurred. History also revealed Hashimoto’s thyroiditis diagnosed at the age of 14 yr and the onset of alopecia areata at the age of 20 yr.

The patient has developed progressive increase in her leucocyte count [(8–12) × 10⁹/l] with a normal distribution and platelet count [(450–600) × 10⁹/l] over the past 8 yr. When seen in our clinic she was feeling well. Except for obesity, the physical examination was unremarkable. Haemoglobin was 14 g/dl with a normal distribution. A peripheral blood smear revealed anisopoikilocytosis and numerous Howell–Jolly bodies, indicating functional hyposplenism. Computed tomography revealed a small spleen and no flow signals could be derived by colour Doppler measurements from the spleen.

Rothmann–Makai syndrome is a rare form of Weber–Christian disease, usually seen in adolescent and middle-aged women [1]. Subcutaneous nodules develop during the course of the disease whereas systemic manifestations are absent. Multiple painless nodules appear throughout the muscle and subcutaneous tissues. The primary pathological process is lobular panniculitis, which passes through the same three stages as Weber–Christian disease. In the first stage, there is acute inflammation of the fat lobules with fat cell degeneration accompanied by an infiltrate of neutrophils, lymphocytes and macrophages. The second stage is characterized by many foamy histiocytes, and the infiltrate is discretely localized to the fat lobules. Finally, the foam cells are replaced by fibroblasts. The first two histological stages correspond to clinically apparent induration, while in the third stage atrophy of the skin may develop. There are no diagnostic laboratory findings. Corticosteroid treatment does not prevent new lesions or seem to alter the course of the self-limited disease.

Infiltrative, inflammatory or thromboembolic processes in the parenchyma of the spleen can cause a functional loss of the organ. This phenomenon is called functional asplenia and occurs as a complication, especially in sickle cell disease, lupus erythematosus (SLE) and after bone marrow transplantation [2–4]. Functional asplenia has complicated the course of autoimmune diseases other than SLE, such as candidiasis endocarditis syndrome and alopecia areata [5].

This is the first report of functional asplenia occurring in the setting of Rothmann–Makai syndrome. The aetiology and pathogenesis of Rothmann–Makai syndrome is still unknown. Christian–Weber disease often occurs in patients with autoimmune disorders [6–8], as is the case with our patient, reinforcing the view that Rothmann–Makai syndrome, a variant of Christian–Weber, may also have an autoimmune basis.

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A. Psyrri and T. Economopoulos

2nd Department of Internal Medicine, Attikon University Hospital, Athens, Greece

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Correspondence to: A. Psyrri, Attikon Hospital, Rimini 1, Haidari, Athens, Greece. E-mail: Diamando.Psyrri@yale.edu