ANSWER TO PHOTO QUIZ (PAGE 163)

A SPLEEN LIKE YOU'VE NEVER SEEN?

DIAGNOSIS

X-ray and CT scan of the chest showed extensive small-sized calcifications of the spleen (figure 1).

Various autoimmune disorders can cause splenic abnormalities, like splenomegaly and splenic infarction. SLE is a chronic multisystemic autoimmune disease, in which several organs and tissues are damaged by pathogenic autoantibodies and immune complexes. Abdominal involvement of SLE can occur in virtually any organ within the abdominal cavity, although only renal involvement integrates diagnostic criteria.[1] Rupture, splenomegaly, infarction, infections, and atrophy of the spleen have been recognized in patients with SLE. Rapid enlargement of the spleen in a lupus patient should raise concern for the possibility of lymphoma.[2]

Splenic calcifications have been reported in SLE and in various other diseases such as rheumatoid arthritis, systemic sclerosis, amyloidosis, sickle cell anaemia, anthracosilicosis, lymphoma, infections (histoplasmosis, tuberculosis, brucellosis, candidiasis, *Pneumocystis jirovecii*), trauma and coeliac disease.[3-5] Despite this wide range of causes, splenic calcifications still remain rare and reports on prevalence are not available. In our case, these other causes of splenic calcifications like infections (negative HIV testing, interferon gamma-release assay – tuberculosis negative), sickle cell anaemia, lymphoma and environmental causes were ruled out from the history, clinical examination and laboratory findings. Segmental splenic infarction associated with lupus anticoagulants and anti-cardiolipin antibodies can also result in splenic calcification, however were negative in our patient. Whether splenic calcification can predispose to hyposplenism remains unclear.[3] It may precede autosplenectomy and hyposplenism, possibly emphasizing the importance of pneumococcal vaccination.[6]

The guideline on hyposplenism from the 'Dutch National Institute for Public Health and the Environment' recommends vaccination and antibiotics for patients with sickle cell disease, splenic infarction and radiation of the spleen. In other diseases able to cause functional hyposplenism (table 1) like SLE, this is less clear. Peripheral blood smear without Howell-Jolly bodies does

Table 1. *Diseases with risk of functional asplenia (adapted from the guideline Asplenia, Dutch National Institute for Public Health and the Environment)*

	Diseases
Cardiac	Congenital cyanotic heart diseases
Intestinal	Celiac disease* Inflammatory bowel diseases (mainly colitis ulcerosa)
Liver	Cirrhosis, with or without portal hypertension* Chronic active hepatitis
Haematological	Sickle cell disease* Other haemolytic anaemia with extreme haematopoiesis Primary thrombocythemia
Auto-immune	Vasculitis (splenic infarction)* Systemic of discoid lupus erythematosus* Rheumatoid arthritis
Infiltrating	Amyloidosis Sarcoidosis
Vascular	Splenic artery occlusion Splenic vein thrombosis Coeliac artery thrombosis
Other	Graft versus host disease Stem cell transplantation* High dosed steroids Radiation of spleen (Hodgkin's disease)* HIV infection with low CD4-cell count

* One of more common causes of functional asplenia

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not exclude functional hyposplenism, and ultrasound and CT imaging is inefficient to judge splenic function. Spleen scintigraphy could be conclusive, but is invasive. So, in these cases, the advice to treating medical specialists is to not routinely provide vaccination and antibiotics to all patients with potential hyposplenism, but to individually assess each case in consultation with an infectious diseases consulant.[7]

DISCLOSURES

The authors have no conflicts of interest to declare.

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