

Splenic hypofunction in systemic lupus erythematosus

JULIANNE C. WILKINS CHILDS, DO

RAYMOND A. ADELIZZI, DO

MICHAEL B. DABROW, DO

NATHAN FREED, DO

Systemic lupus erythematosus is a relatively common autoimmune disease in the United States. Hyposplenism is infrequently described in patients with systemic lupus erythematosus. It is seen in up to 5% of patients and is thought to be caused by vasculitic changes within the spleen. The diagnosis of hyposplenism can be made easily by identifying Howell-Jolly bodies on a peripheral blood smear. The authors describe a patient with systemic lupus erythematosus associated with hyposplenism and discuss possible diagnostic and treatment implications.

(Key words: Systemic lupus erythematosus, hyposplenism, Howell-Jolly bodies)

Systemic lupus erythematosus (SLE) is a relatively common systemic autoimmune disease. The medical literature suggests that up to 50 per 100,000 US inhabitants in urban areas may be affected.¹ The majority (90%) of patients are women of childbearing age. The disease is also more prevalent in African Americans and Hispanic Americans than in whites. Hyposplenism, defined as a failure of the spleen to accumulate radiocolloid, has been described as an infrequent manifestation of SLE.²⁻⁷ Because hyposplenism leaves patients at

risk for overwhelming sepsis from encapsulated organisms, it is important to recognize the clinical association. Described here is SLE with associated hyposplenism diagnosed by the presence of Howell-Jolly bodies on a peripheral blood smear in a patient with coincidental β -thalassemia trait.

Report of case

A 32-year-old white woman of Mediterranean descent with a 5-year history of SLE was seen in the emergency department with a chief complaint of unremitting headache and neck stiffness. Because of a history of subarachnoid hemorrhage, a computed tomography scan of the head and lumbar puncture were performed. The findings of both were essentially normal.

The patient's medical history included a rash that had occurred within the previous 3 weeks and worsening anemia that were thought to be consistent, both clinically and by laboratory data, with a lupus flare despite treatment with corticosteroids. Therefore, the patient was placed on cyclophosphamide and prednisone immunosuppressive therapy.

A peripheral blood smear obtained in the emergency department revealed a few stomatocytes, moderate target cells, Howell-Jolly bodies, and microcytic cells. Laboratory values were as follows: hemoglobin level, 9.3 g/dL; hematocrit, 29.3%; white blood cell count, 14.7/mm³; platelet count, 282/mm³; mean corpuscular volume, 78 μ m³. Review of the patient's previous charts revealed peripheral blood smears with Howell-Jolly bodies as early as 1.5 years before this admission. The patient had no previous history of splenectomy. Serum iron studies revealed a normal iron level and total iron-binding capacity, with an elevated ferritin level. Hemoglobin electrophoresis revealed the HbA₁ to be 94.8% and the HbA₂, 5.2%. A liver-spleen scan revealed absence of a functioning spleen, and computed tomography (CT) of the abdomen confirmed the physical presence of the spleen. The diagnoses of hyposplenism secondary to SLE and coincidental β -thalassemia trait were made.

From the Department of Medicine, University of Medicine and Dentistry of New Jersey—School of Osteopathic Medicine, Stratford, where at the time this article was written, Dr Wilkins Childs was a hematology/medical oncology fellow; Dr Adelizzi is clinical associate professor of medicine; Dr Dabrow is clinical associate professor, hematology/medical oncology; and Dr Freed is chief, Department of Hematology and Medical Oncology. Dr Adelizzi is also director, Arthritis Center of South Jersey, Stratford, NJ.

Correspondence to Julianne C. Wilkins Childs, DO, 5 Teal Ln, Beesley's Point, NJ 08233-1926.

Discussion

Hyposplenism related to SLE has been reported infrequently in the medical literature. More commonly described is hypersplenism secondary to follicular hyperplasia or congestive splenomegaly leading to autoimmune hemolytic anemia or thrombocytopenia in some SLE patients. In select patients, splenectomy has been suggested to be of value in patients with these clinical manifestations.⁸ The mechanism of hyposplenism in these patients is thought to be on the basis of an active vasculitis within the spleen. Chronic vasculitic lesions with periarteriolar fibrosis or "onion skinning" and fibrinoid necrosis of vessel walls have been described as seen at autopsy in the spleens of patients with SLE.^{2,9} Splenic atrophy secondary to microinfarcts also has been described.^{2,5}

One study⁷ suggests that the prevalence of hyposplenism may be as high as 5% in patients with SLE. Just as in patients without lupus, overwhelming sepsis from organisms such as *Salmonella*⁴ species and *Streptococcus pneumoniae* have been reported in SLE patients with hyposplenism.^{4,5}

These data suggest that it may be wise to include peripheral blood smears in the follow-up of all SLE patients to establish the presence of Howell-Jolly bodies, the diagnostic indicator of hyposplenism. Howell-Jolly bodies are the remnants of RNA that are normally removed from mature red blood cells by a functioning spleen; hence, their presence indicates lack of splenic function. They are typically seen in patients with post-splenectomy or postsplenic infarction, that is, as in sickle cell disease. In light of the fact that many patients with SLE will be on immunosuppressive therapy, as was our patient, this follow-up procedure may be even more crucial.

As in any other patient with known hyposplenism, a pneumococcal vaccine should be administered to patients with SLE who have hyposplenism. One case report⁶ demonstrated measurable improvement in pneumococcal antibodies after injection, although this patient's splenic hypofunction appeared reversible after corticosteroid administration. More follow-up is needed regarding the effect of the vaccine in patients with SLE who have irreversible hyposplenism.

Of additional note, our patient had baseline exacerbations of anemia secondary to presumed hemolysis during active lupus flares. (Because this patient's spleen was hypofunctioning, the liver

and bone marrow are the end organs presumably involved in hemolysis.) Because of her microcytic indices, normal serum iron levels, Mediterranean family origin, and the presence of target cells on her peripheral blood smear, hemoglobin electrophoresis was performed. The hemoglobin A₂ level correlated with the presence of β -thalassemia trait. Interestingly, target cells, which typically are seen in patients with thalassemia, are also seen in patients with hyposplenism.

Comment

Hyposplenism can be a clinically relevant manifestation of SLE. Because up to 5% of patients with SLE may have hyposplenism,⁷ appropriate follow-up study should include serial peripheral blood smears for early indications of the typical diagnostic findings (Howell-Jolly bodies) of hyposplenism. To avoid the risk of overwhelming bacterial sepsis in these patients, it is prudent to administer polyvalent pneumococcal vaccine if hyposplenism is present. In the near future, studies looking at the prevalence of hyposplenism in patients with SLE and the effectiveness of the pneumococcal vaccine are warranted.

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