Massive Splenomegaly and Hypersplenism in a Young Woman with Primary Sjögren's Syndrome

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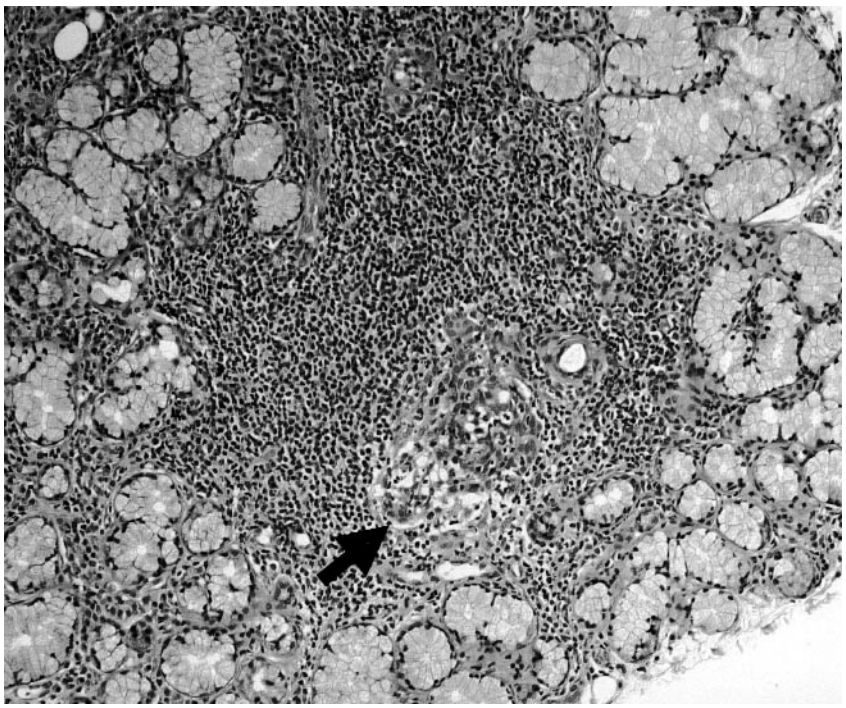
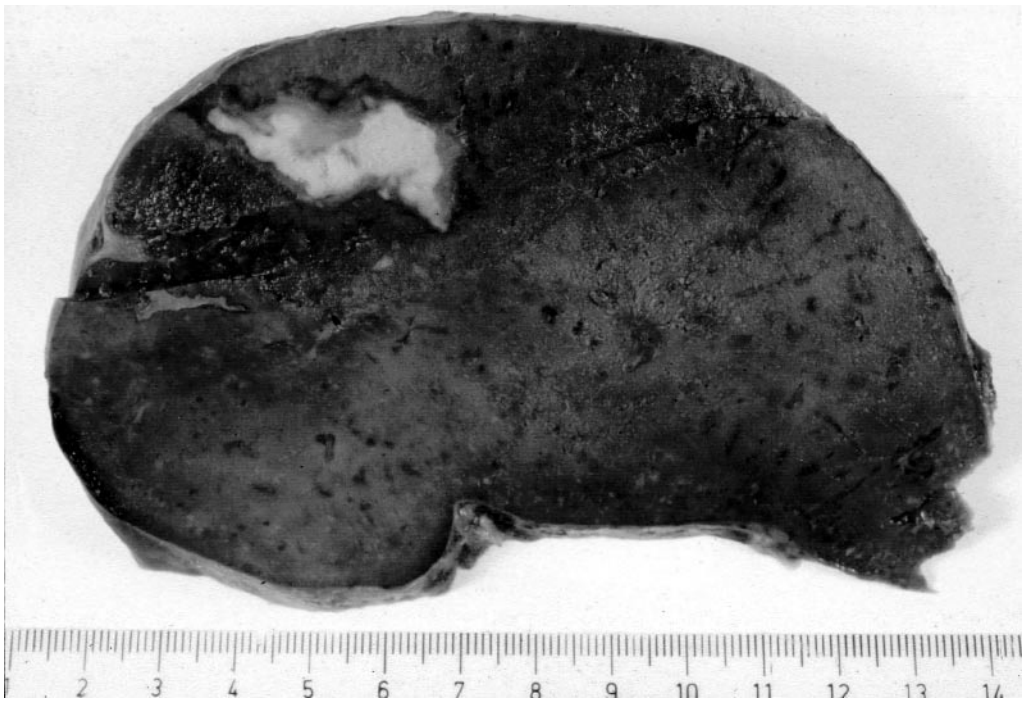
Splenomegaly is not a major clinical feature in patients with primary Sjögren's syndrome (SS) unless the disease is complicated by lymphoma. We describe a patient with primary SS, massive splenomegaly, and hypersplenism[1], but without lymphadenopathy[2]. She was recently admitted to our hospital with anemia (Hb 8.2 g/dl), thrombopenia (95/nl), and splenomegaly. Highly elevated autoantibodies (antinuclear antibodies, Ro/SSA, Ro/SSB, La/SSB, Scl-70) were also found. A biopsy of the salivary glands revealed multiple benign lymphoepithelial lesions (Figure 1A, arrow). An abdominal computer tomography scan showed massive splenomegaly with multiple infarcts. Histology of the bone marrow revealed hyperplastic erythropoiesis, granulopoiesis, and megakaryopoiesis. Splenectomy was performed (1) to rule out malignant lymphoma, (2) because the size of the spleen constituted a high risk for spontaneous spleen rupture and caused abdominal pain, and (3) because the patient had hypersplenism. The spleen weighed 1200 g and measured 27.5 × 14.5 × 6.8 cm (Figure 1B, section perpendicular to long axis with infarct). Histology showed hyperplasia mostly of the red but also of the white pulp with no morphological or immunohistochemical evidence of lymphoma infiltration (Figure 1C, CD20 stain), with extramedullary erythropoiesis and megakaryopoiesis (panel C, inset, CD 61 stain). After splenectomy, reactive thrombocytosis occurred and this was treated with aspirin, with the result that hemoglobin rose significantly.

This case report illustrates the rare association of primary SS with nonmalignant massive splenomegaly and hypersplenism. This patient benefited significantly from splenectomy.



A

B



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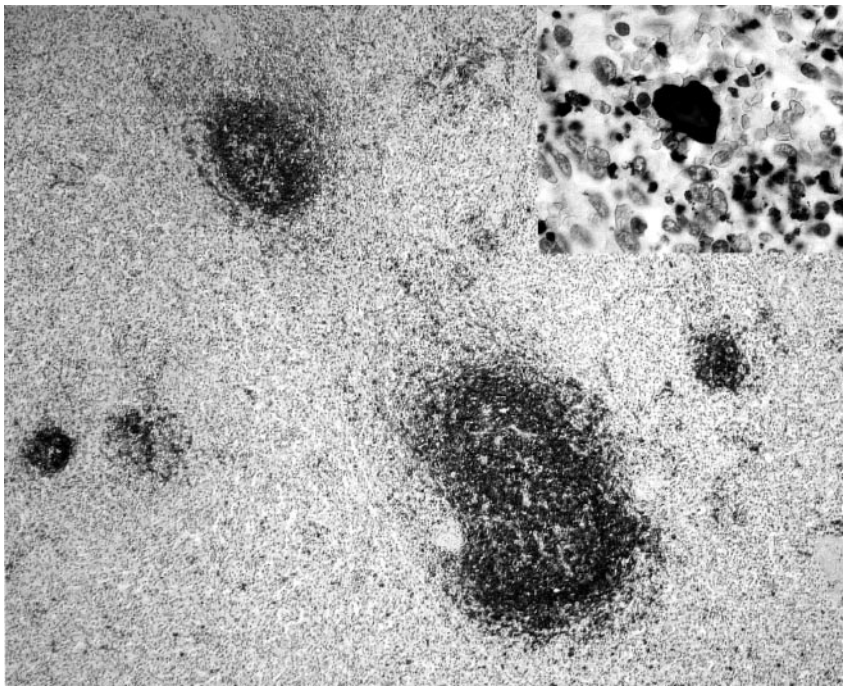

Figure 1. A. Multiple benign lymphoepithelial lesions of the salivary glands. B. Section of the spleen. C. Histology shows hyperplasia of the red and white pulp with no evidence of lymphoma (CD20 stain).

REFERENCES

1. Zittoun R, Debain P, James JM, Bilski-Pasquier G. Hematological manifestations and complications in Sjögren's syndrome. Sem Hop 1978;54:1011-20.
2. McCurley TL, Collins RD, Ball E. Nodal and extranodal lymphoproliferative disorders in Sjögren's syndrome: a clinical and immunopathologic study. Hum Pathol 1990;21:482-92.