

Spontaneous Rupture of Spleen in Immunoglobulin Light Chain Amyloidosis: A Case Report and Review of Literature

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ABSTRACT

Rupture of spleen in absence of trauma is very rare and can occur in certain pathologic conditions. Amyloidosis involves spleen and splenomegaly has been reported in 4 to 13% of patients; however, splenic rupture has rarely been reported in amyloidosis. Here, we report a case of spontaneous splenic rupture in a patient with immunoglobulin light chain amyloidosis.

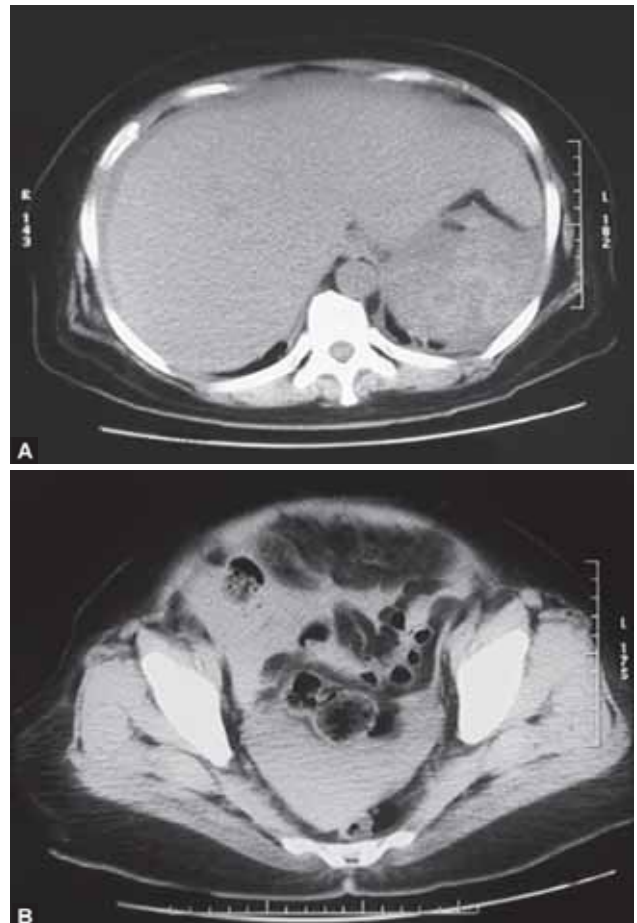
Abbreviations: AL: Immunoglobulin light chain; CT: Computed tomography.

Keywords: AL amyloidosis, Spleen rupture, Kappa light chain, Plasma cells.

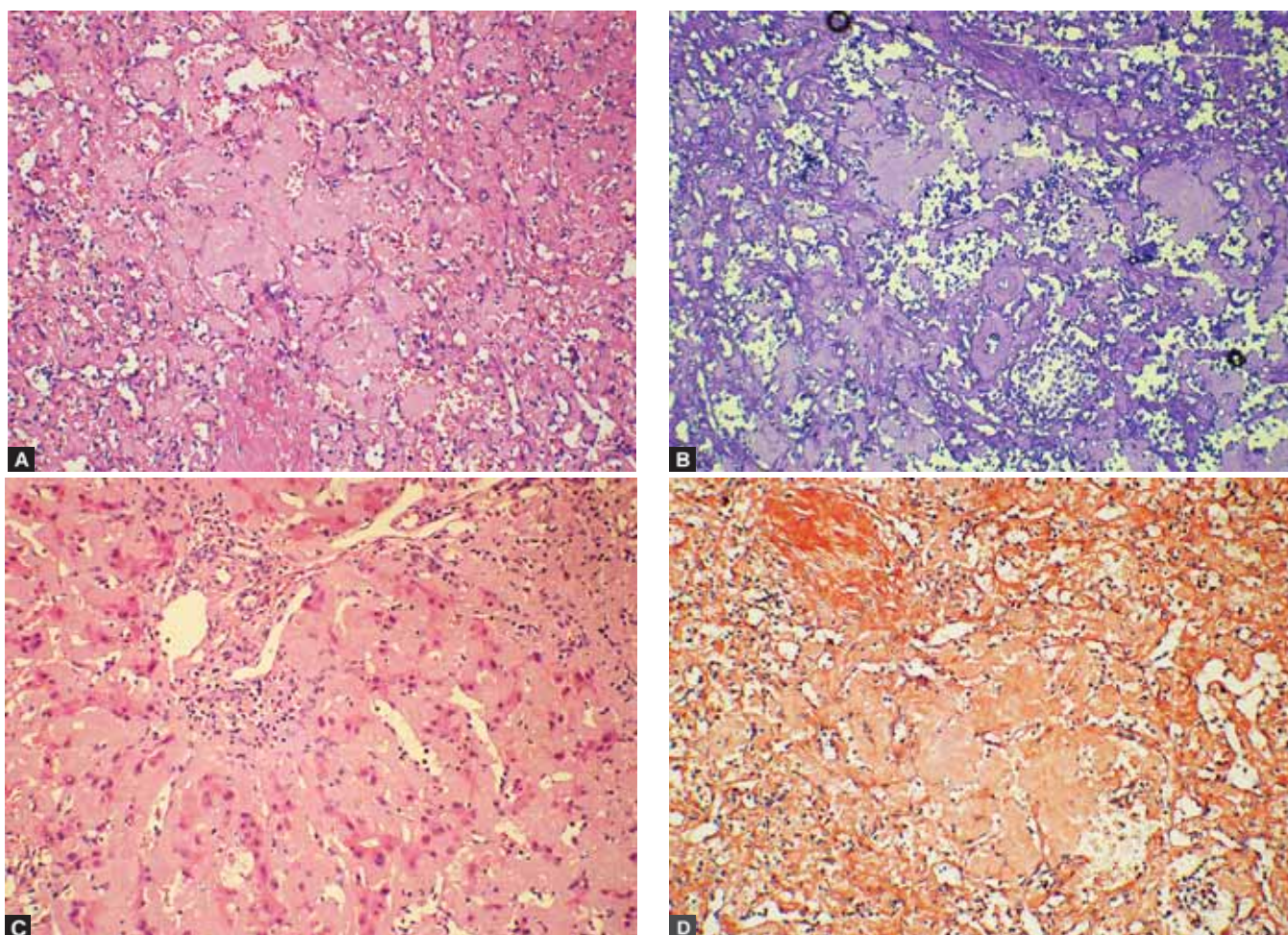
CASE REPORT

A 55-year-old female presented to the emergency room with complaints of pain abdomen and postural syncope of 6 hours duration. On examination, she was pale and tachypneic; pulse rate was 130/min and blood pressure was 80/60 mm Hg with a postural fall of 20 mm Hg. Computed tomography (CT) of abdomen was done which revealed hepatosplenomegaly with heterogeneous attenuation within the upper pole of spleen, having internal hyperdense areas representing hemorrhage, along with left subphrenic hematoma and hemoperitoneum (Figs 1A and B). Emergency laparotomy was done which revealed splenic and intraperitoneal hematoma. She underwent splenectomy and 2.5 liters of blood was evacuated from the abdominal cavity. Peroperative liver biopsy was done as liver showed firm consistency. During the postoperative phase she recovered well.

The liver biopsy (Figs 2A and B) and splenic biopsy (Figs 2C and D) specimens were suggestive of amyloidosis and she was subsequently evaluated for the etiology of amyloidosis. Her serum and urine electrophoresis were normal. Urine for immune fixation electrophoresis revealed monoclonal presence of kappa light chains in the form of two oligoclonal bands (Fig. 3) and urine for Bence Jones proteins was negative. Serum light chains assay revealed markedly elevated kappa light chain. Bone marrow biopsy showed 13% plasma cells with mild dyshemopoietic



Figs 1A and B: (A) Hyperdense area with heterogeneous attenuation within spleen suggestive of hemorrhage, (B) high-density collection in pelvis suggestive of hemoperitoneum



Figs 2A to D: (A) H&E stained section from the spleen showing amorphous pink extracellular material obliterating normal architecture, (B) methyl violet stain showing light pink possibility for amyloid, (C) H&E stained section from the liver showing amorphous pink extracellular material within the sinusoids obliterating normal architecture, (D) Congo red stains showing light pinkish orange possibility for amyloid

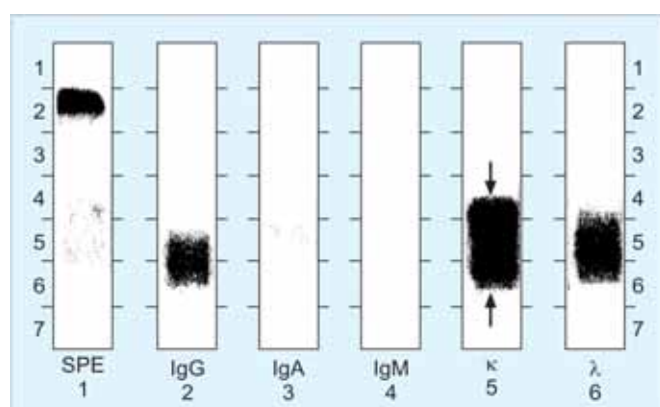


Fig. 3: Urine immune fixation showing presence of kappa light chains in the form of two oligoclonal bands

changes and was positive for amyloid. Skeletal survey and bone scan were normal. A final diagnosis of primary amyloidosis with kappa light chain disease was established.

DISCUSSION

Spontaneous splenic rupture is very rare and poorly defined in literature. It was reported to be resulting from an 'incident

without external force'.^{1,2} The first case of spontaneous splenic rupture was reported in 19th Century.^{3,4}

Spleen can rupture in the following circumstances: Trauma to a diseased spleen, trauma to a normal spleen, spontaneous rupture of a diseased spleen and spontaneous rupture of a normal spleen.⁵ Spontaneous rupture rarely occurs in a histologically proven normal spleen and such cases are regarded as 'true spontaneous rupture'. Spontaneous rupture usually occurs in a diseased spleen and is called 'pathologic spontaneous rupture'.

According to the criteria described by Orloff and Peskin, splenic rupture should be considered 'spontaneous' only if it occurs in the absence of trauma in a spleen unaffected by intra- and perisplenic diseases and in patients free of diseases that could involve the spleen.⁶ True spontaneous rupture of the spleen is extremely rare. However, in the current literature, splenic rupture is commonly categorized as spontaneous when it occurs without trauma, whether or not the rupture is pathologic.

Thus, according to the strict criteria our patient had 'pathological spontaneous rupture' of spleen. The incidence

of spontaneous rupture of the spleen in amyloidosis is not known and is thought to be very rare. Histopathologically, amyloid may usually be found in blood vessel walls or in the connective tissue of the capsule and trabeculae even in cases with no naked-eye abnormality.⁷ Amyloid angiopathy has been implicated as the cause of splenic rupture in AL amyloidosis. Coexistent coagulation abnormalities also contribute to splenic rupture.

Splenic involvement in amyloid can result in functional hyposplenism. This is seen in up to 24% of patients.⁸ Hyposplenism correlates poorly with degree of splenic involvement with amyloid and has been shown to be a poor prognostic factor. Other diseases, which can cause pathological spontaneous rupture, include infectious mononucleosis, malaria, sarcoidosis, Gaucher's disease, hematologic malignancies and connective tissue disorders.⁹

Our patient had primary amyloidosis with kappa light chain disease, which is uncommon as compared with the lambda light chain disease. Lambda chain class predominates over kappa chains in AL amyloidosis by a ratio of 2:1.¹⁰ These combinations make it the first such case report in literature.

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