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Case report

Surgical management of portal hypertension in Felty's syndrome: A case report and literature review☆

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***Background/Aims*: Bleeding esophageal varices are a common complication of portal hypertension in patients with underlying liver disease. Often patients with hepatic cirrhosis have hypersplenism with thrombocytopenia and leukopenia. Felty's syndrome is a disorder where patients with rheumatoid arthritis develop splenomegaly, neutropenia, and on rare occasions, portal hypertension without underlying cirrhosis.**

***Methods*: We present a case of a patient with portal hypertension secondary to Felty's syndrome and discuss the importance of recognizing this condition since the treatment of choice is surgical management with splenectomy. A review of the literature and underlying liver histologic features are discussed.**

***Results*: Medical and surgical management of patients with Felty's syndrome is different from those with portal hypertension due to cirrhosis.**

***Conclusion*: Splenectomy is the treatment of choice for complications of portal hypertension in patients with Felty's Syndrome.**

***Keywords*: Varices; Portal hypertension; Splenectomy**

1. Case report

A 53-year-old Caucasian male presented to the emergency department with hematemeis. His medical history was significant for rheumatoid arthritis, benign prostatic hypertrophy, GERD, hyperlipidemia and COPD. His medications included etodolac, fenofibrate, rabeprazole, tiotropium, hydroxychloroquine and occasional over the counter NSAIDs. He denied drug or alcohol abuse and had no prior episodes of gastrointestinal bleeding. Family history was pertinent for his father having died with idiopathic cirrhosis. The physical examination revealed resting tachycardia, lower extremity petechiae, epigastric tenderness, splenomegaly, and hemoccult positive stool. Abnormal laboratory tests showed a hemoglobin of 11.1 g/dl and platelet count of 73,000/μl. Liver profile and coagulation times were normal.

An upper endoscopy was performed revealing grade 3 esophageal varices (Fig. 1A) extending from the gastroesophageal junction (40 cm) to 28 cm with stigma of recent bleeding and a mosaic appearance was seen in the gastric mucosa consistent with portal gastropathy (Fig. 1B). Variceal ligation was performed with a

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Abbreviations: GERD, gastroesophageal reflux disease; COPD, chronic obstructive pulmonary disease; NSAID, nonsteroidal antiinflammatory drugs; RA, Rheumatoid arthritis; NRH, nodular regenerative hyperplasia; IPH, idiopathic portal hypertension; H&E, hematoxylin and eosin.

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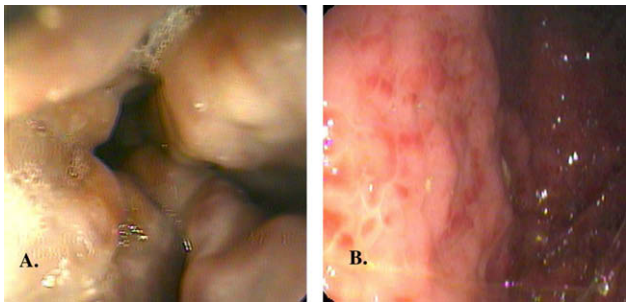

Fig. 1. Photographs taken during initial upper endoscopy revealed (A) grade 3 esophageal varices with stigmata of recent bleeding and (B) congestive portal gastropathy.

banding device. Abdominal venous ultrasound revealed patent portal and superior mesenteric veins and normal hepatic. Liver biopsy was performed and revealed macrosteatosis, mild portal inflammation, focal mild fibrous expansion, normal sinusoids, and no evidence of cirrhosis (Fig. 2). Transhepatic wedge pressures were performed and were consistent with significant portal hypertension (the sinusoidal pressure, i.e., difference between the mean right hepatic wedge pressure and the mean right atrial pressure was 11 mm Hg; the mean wedge pressure was 18 mm Hg). The patient was referred to surgery for splenectomy. Splenic artery

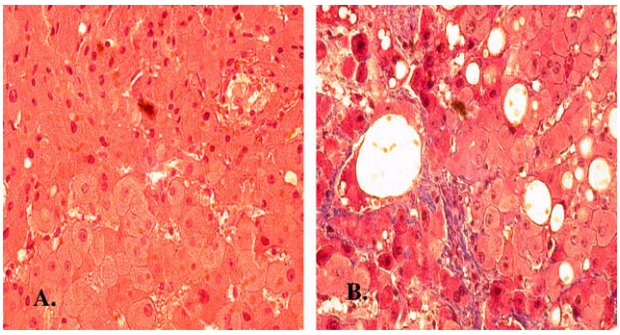

Fig. 2. Histology sections of the liver revealed macrosteatosis, mild portal inflammation, and normal sinusoids on the hematoxylin and eosin stain (A) and focal mild fibrous expansion without evidence of cirrhosis on the trichrome stain (B).

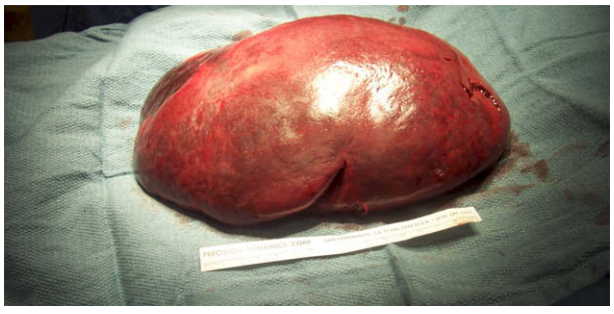

Fig. 3. A massively enlarged spleen was removed at surgery weighing 1875 g and measuring 12.2 × 1.8 × 2.5 cm.

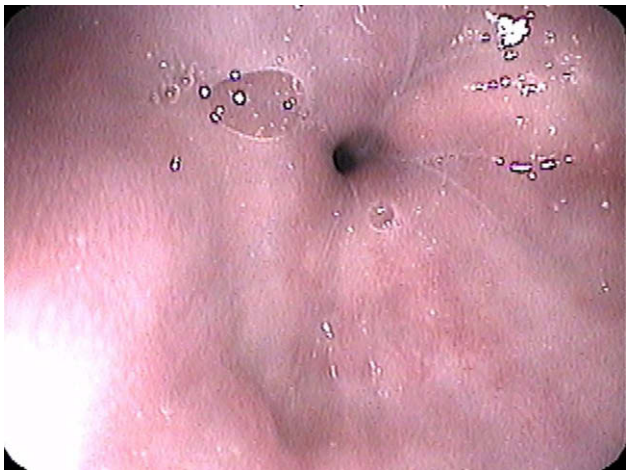

Fig. 4. Six months post-splenectomy repeat upper endoscopy showed a normal distal esophagus with complete resolution of esophageal varices.

embolization was performed preoperatively. At surgery the liver appeared normal and a 1875 g (12.2 × 1.8 × 2.5 cm) spleen was removed (Fig. 3). Follow-up endoscopy post-operatively revealed no evidence of esophageal varices or portal gastropathy (Fig. 4).

2. Literature review

2.1. Felty's syndrome overview

Felty's syndrome, a well-described triad of neutropenia, splenomegaly, and rheumatoid arthritis (RA), has made multiple appearances in the literature since it was described in 1924 [1]. Less than 1% of patients with rheumatoid arthritis (RA) have Felty's syndrome, and these patients frequently have severe joint destruction, a higher occurrence of rheumatoid nodules, lymphadenopathy, hepatic pathology, vasculopathy, leg ulcers and other recurrent infections. A high percentage of those with Felty's syndrome (78–90%) have the HLA-DR4 antigen (compared to 70% of typical RA patients and 30% of controls), suggesting a genetic component to this syndrome [2–4].

2.2. Liver histology in Felty's syndrome

Several authors have reported varices in association with Felty's syndrome, with varying underlying liver pathology, most frequently nodular regenerative hyperplasia (NRH) [5–12]. Other liver abnormalities found on biopsy include: diffuse lymphocytic infiltration in the sinusoids and Kupffer cell prominence, periportal fibrosis, macronodular cirrhosis, and "nodules" without fibrosis or lobular disruption [5]. No correlation between transaminase abnormalities and histology could be made [5]. In 1974, Blendis et al. found what seemed to be a pattern in Felty's associated liver pathology: they reported 5 patients with Felty's syndrome who had hepatic nodular