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Clinical Image

When the Liver is the Victim and Not the Culprit: A Challenging Case of Non-Cirrhotic Portal Hypertension

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A 79-year-old male previously admitted to a long-term care for cognitive impairment was transferred to our center for a first occurrence of ascites, pleural effusion and edema in lower limbs. His past medical history revealed hypertension, diabetes and two previous strokes with residual dementia, without known malignancies or liver diseases. Alcohol assumption and serologies for hepatitis were unremarkable. Platelets count were within limits, serum protein electrophoresis showed monoclonal component of 1.1 g/dl IgA/Lambda type in beta zone and increased beta2 globulins (27,9%). Urinalysis was negative for Bence-Jones proteinuria. Ascitic fluid analysis showed normal cell count and serum-ascites albumin gradient was > 1,1 g/dl, consistent with portal hypertension.

Computed tomography of the abdomen (Figure 1) revealed no hepatosplenomegaly, no lymphadenopathy, or sign of peritoneal carcinosis. The main portal vein was patent, excluding pre-hepatic portal hypertension. Transthoracic echocardiography showed normal chambers and no pericardial effusion, excluding post-hepatic portal hypertension.

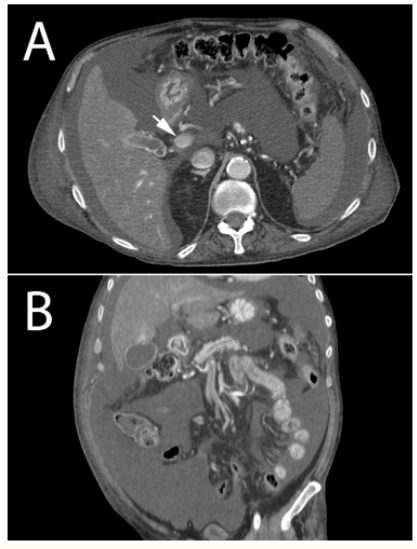

Figure 1: *Axial (panel A) and coronal (panel B) contrast-enhanced CT images of the abdomen showing diffuse ascites, but normal liver size and margins (B) and normal spleen size (A). The white arrow depicts patent portal vein in contrast-enhanced portal-venous phase image (A).*

The patient rapidly developed jaundice, renal and respiratory failure and eventually died.

The autopsy revealed no macroscopic signs of cirrhosis. Surprisingly, heart, kidney and above all liver and spleen samples (Figure 2) stained positive with Congo red, consistent with amyloid deposition.

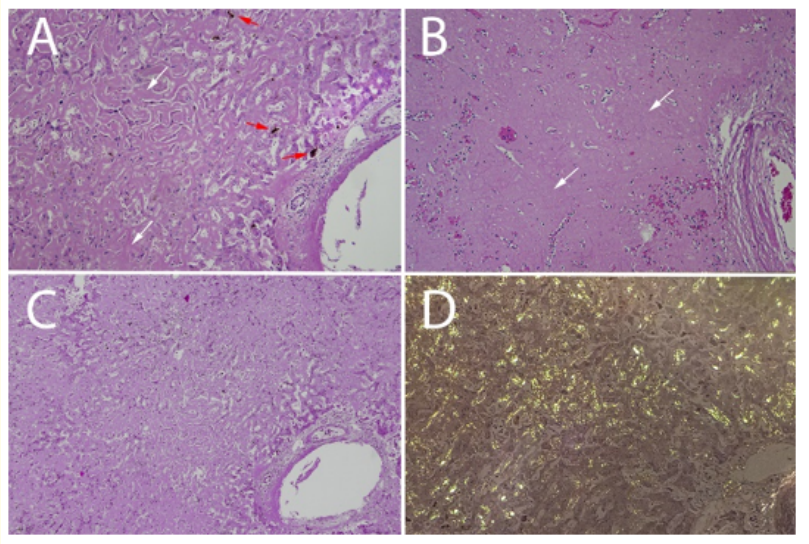

Figure 2: *Panel A (original magnification 20× Hematoxylin and eosin stain): Liver sample showing almost complete architectural subversion, from sinusoidal and peri-vascular accumulation of pale eosinophilic, amorphous, hyaline, acellular material (white arrows). Bile plugs in dilated canaliculi (red arrows) are evident in the specimen and no significant peri-portal or sinusoidal fibrosis was seen.*
Panel B (original magnification 20× Hematoxylin and eosin stain): Spleen sample showing diffuse parenchymal accumulation of eosinophilic, amyloid substance (white arrows).
Panel C (original magnification 10× Hematoxylin and eosin stain): Liver sample at lower magnification with evident normal portal tract in the lower right corner, but diffuse peri-sinusoidal accumulation of eosinophilic material.
Panel D (original magnification 10×): Congo red staining confirming massive hepatic amyloid deposits with diffuse apple green birefringence appearance under polarized light.

Primary amyloidosis frequently involves the liver, but rarely causes overt portal hypertension [1]. In this case, systemic amyloidosis caused sinusoidal portal hypertension due to a massive infiltration of amyloids in liver and spleen.

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Bibliography

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