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Hepatopulmonary syndrome: an incidental radiological finding in a child with signs of portal hypertension

Síndrome hepatopulmonar: achado radiológico incidental numa criança com sinais clínicos de hipertensão portal

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Keypoints

What is known

– Hepatopulmonary syndrome is a rare condition, affecting children with portal hypertension/chronic liver disease. It is an important cause of morbidity, demanding early recognition of signs/symptoms and prompt diagnosis, including imaging studies, to allow for adequate treatment.

What is added

– We present an atypical case study of highly-symptomatic children due to portal hypertension, showcasing all the characteristic imaging findings of accompanying abdominal consequences, and a simultaneous subclinical intrapulmonary vascular dilation, resulting in hepatopulmonary syndrome.

A 10-year-old boy was transferred to our hospital from his native country, Guinea-Bissau, due to sustained pancytopenia, ascites, hepatosplenomegaly, and grade III esophageal varices of unknown origin. Symptoms started when he was 8 years old, presenting with an increased abdominal perimeter, and a palpable liver and spleen. Over the last two years, he suffered repeated episodes of hematemesis, requiring multiple blood transfusions. Two months before admission, he began to show periorbital and lower limb edema.

At our hospital, a physical examination added the presence of subicterus. Peripheral oxygen saturations were normal (SpPO2 > 98%). The ultrasound showed a liver of normal dimensions (9 cm) and heterogeneous texture, splenomegaly (16 x 8 cm), ascites, and decreased main portal vein (MPV) peak systolic velocity (15 cm/s), suggesting portal hypertension (Fig. 1). An abdominal computed tomography (CT) scan also revealed portal vein cavernoma (20 mm) and normal permeability in all arterial segments evaluated. There were also varices in splenic (spleno-renal shunt), inferior mesenteric, and superior rectal veins (Fig. 2).

A thoracic CT suggested the presence of esophageal varices. In this exam, a nodular image on the lower lobe of the left lung was discovered. It was in the subpleural region, with a bi-lobulated shape and slight contrast retention (Fig. 3). A serpiginous structure visualized inside the nodule, suspected of corresponding to a vessel, was better characterized by ultrasound, confirming the vascular nature of the lesion (Fig. 4). Endoscopy showed esophageal varices (grade III), gastric varices (GOV1 and GOV2), and congestive gastropathy, and some were simultaneously treated with elastic ligation.

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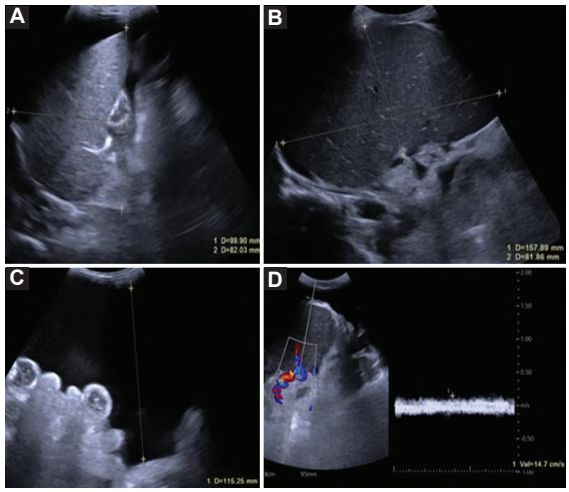

Figure 1. Ultrasound showing normal dimensions of the liver (9cm) with heterogeneous texture **(A)**, splenomegaly **(B)**, and ascites **(C)**, associated with decreased main portal vein peak systolic velocity **(D)**.

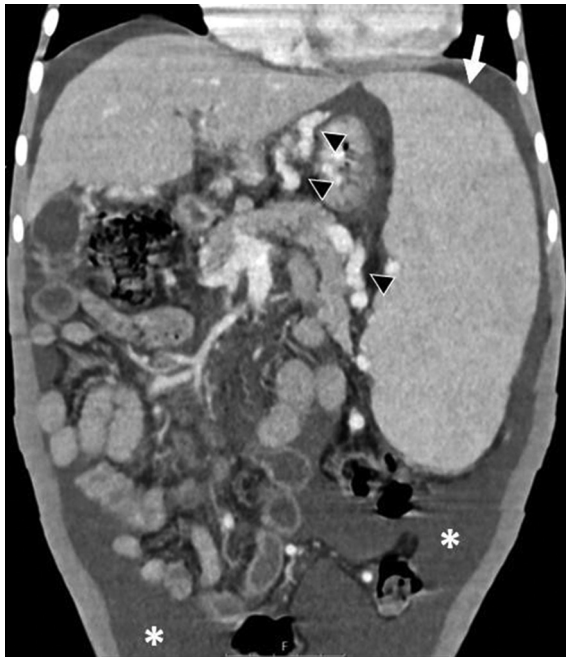

Figure 2. Abdominal coronal computed tomography with ascitis (asterisk), splenomegaly (arrow) and perigastric and perisplenic varices (arrowheads).

Throughout hospitalization, the patient remained free from respiratory symptoms. His serial SpO2 measurements were stable and he had no need for supplemental oxygen. During the follow-up, the patient remained

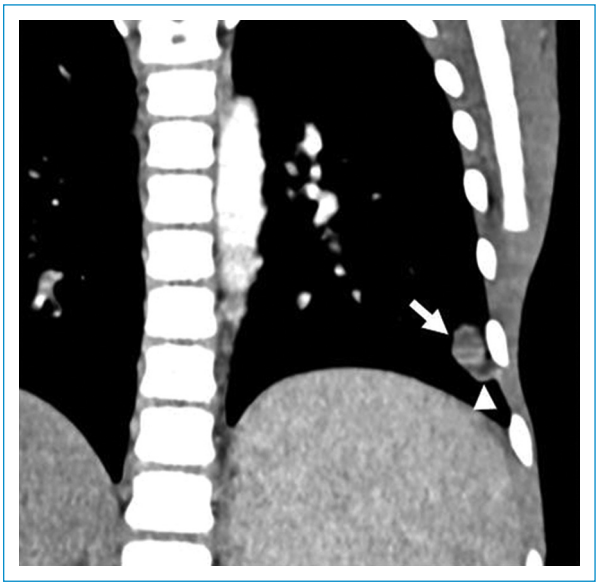

Figure 3. Thoracic coronal computed tomography with a nodular image on the inferior left lower lung lobe, in the subpleural region, with a bi-lobated shape and slight contrast retention (arrow). There was a serpiginous structure visualized inside the nodule (arrowhead), suspected of corresponding to a vessel.

free from any additional complications, and the current status involves awaiting liver transplantation.

Hepatopulmonary syndrome (HPS) is characterized by the triad of abnormal arterial oxygenation caused by intrapulmonary vascular dilation (IPVD) in the setting of liver disease, portal hypertension, or congenital portosystemic shunts. The estimated prevalence of HPS among children with chronic liver disease is around 4-5%[1-3]. Subclinical HPS occurs when there is IPVD without hypoxemia[4].

The pathophysiology of HPS is still unclear, but evidence suggests it occurs owing to an excess of endogenous vasodilators such as nitric oxide (NO) and endothelin (ET-1)[5]. Symptoms are related to the underlying liver disease and oxygenation impairment, when present.

The diagnosis of HPS is clinical, and the presence of a shunt is usually confirmed by pulmonary scintigraphy, a diagnostic imaging modality. Excluding other causes for a shunt and conditions that present with hypoxemia is also essential.

Liver transplantation remains the only treatment for HPS[5-7]. In the pre-transplant evaluation of pediatric patients, ultrasound, and mainly CT scans, are essential imaging tools, providing crucial information about the liver, other abdominal structures, and vascular anatomy. These radiological modalities may also demonstrate

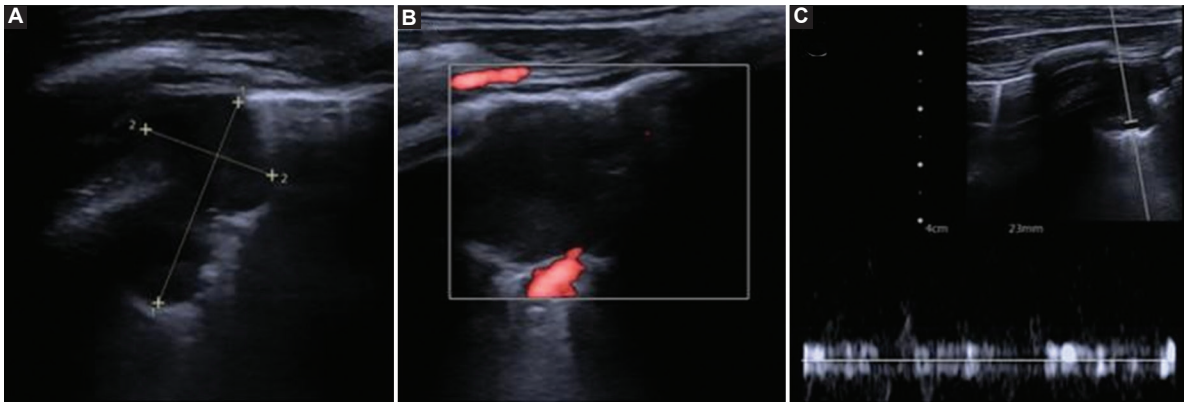

Figure 4. A-C: ultrasound of the lung nodular structure, confirming the vascular nature of the lesion.

some suggestive findings of HPS in this setting. In our patient, HPS diagnosis was made incidentally, in an atypical scenario, from images obtained during the evaluation of portal hypertension in a child without hypoxemia.

Early recognition of HPS is fundamental since it is an urgent criterion for listing patients for liver transplantation, and early action may lead to a shorter recovery period following the procedure.

Funding

None.

Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

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