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CASE REPORT

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The Exploration of Hepatoportal Sclerosis in Pediatrics

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ABSTRACT

Hepatoportal sclerosis, now categorized under the term porto-sinusoidal vascular disease, is a condition leading to non-cirrhotic portal hypertension and posing a substantial risk of variceal bleeding. This payer aims to elucidate this phenomenon through a case study involving a patient displaying manifestations of non-cirrhotic portal hypertension with hemorrhagic indications. The objective is to describe the clinical, radiological, and histological features, as well as the complications associated with this pathology, which is infrequently documented in the pediatric population.

ARTICLE HISTORY

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Introduction

Hepatoportal sclerosis is a seldom - discussed condition in pediatric case. It is characterized by the sclerosis of portal areas without cirrhosis and with permeable portal and hepatic vein [1]. Various terms, such as idiopathic non cirrhotic portal hypertension, nodular regenerative hyperplasia, benign intrahepatic portal hypertension and incomplete septal cirrhosis, have been used interchangeably to describe HPS. The use of multiple synonyms underscores the intricate and diverse nature of the disease, potentially contributing to an underestimation of its prevalence [2]. This case report aims to delineate the clinical, biochemical, radiological, histological, and evolutionary aspects of this condition, is rarely described in children

Case Presentation

A 4-year-old male child was admitted due to abdominal distension and upper gastrointestinal bleeding. The patient had no significant personal or family medical history, and consanguinity was absent. Physical examination revealed appropriate growth for age, an enlarged spleen (8.5 cm), and a liver size of 18 cm. Collateral circulation was observed on the abdominal skin, and no associated skin manifestations were noted.

Ultrasound examination showed granular liver parenchyma, periportal hyperchogenicity, and patent inferior vena cava and hepatic veins. The portal vein appeared enlarged and thrombosed with hepatomegaly and a multiple nodular lesions in abdominal CT scan, figure (1,2). Laboratory results indicated a hemoglobin level of 10.6 g/dl, a white blood cell count of 9290/mm3, a platelet count of 437,000/mm3, a prothrombin level of 120%, an activated partial thromboplastin time of 62.2 s (reference range 25-35 s), and an international normalized ratio (INR) of 1.49. Blood glucose, renal function, and electrolytes were within normal ranges,serum albumin was at 46g/l. Aspartate aminotransferase was 72 UI/L and alanine aminotransferase was at 68 U/l. Total bilirubin was 4 mg/l, gamma-glutamyl transpeptidase was at 1038UI/L and alkaline phosphatase activities was at 444 U/l.(Table 1)

Table 1: Biological Results of the Patient

Laboratory analysis	Results	Normal values
Hemoglobin	10,6 g/dl	12-15 g/dl
White blood cell	9290/mm^3	4000-10000/mm^3
Platelet	437000/mm^3	150000-450000/ mm^3
Prothrombin level	120%	70-140%
Activated partial thromboplastin time	62,2 S	25-35 S
International normalized ratio	1,49	-
Aspartate aminotransferase	72 UI/l	8-50 UI/l
Alanine aminotransferase	68 UI/l	7-45 UI/l
Total bilirubin	4 mg/l	<10 mg/l
Serum albumin	46 g/l	30-60 g/l
Gamma-glutamyl transpeptidase	1038 UI/l	6-29 UI/l
Alkaline phosphatase	444 UI/l	169-372 UI/l

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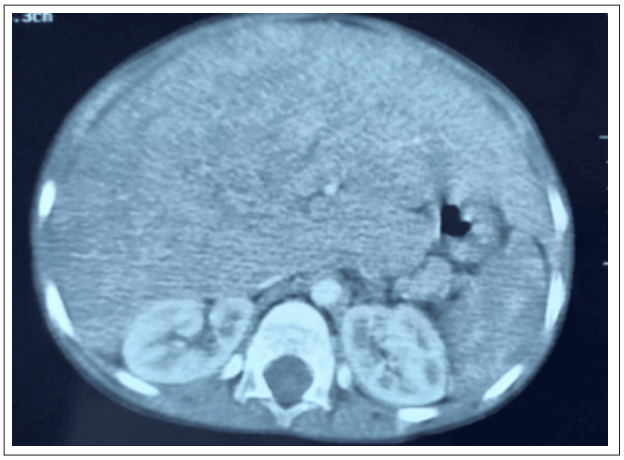

Figure 1: Hepatomegaly With Multiple Nodular Lesions

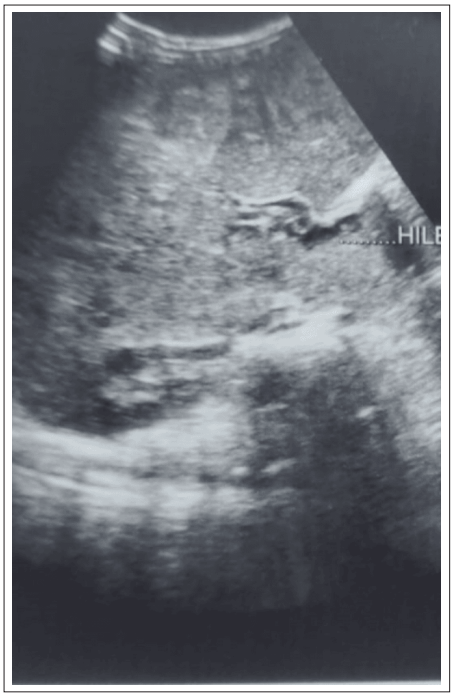

Figure 2: Portal Vein Sclerosis

The etiological investigation included tests for serology of hepatotropic viruses , serum copper, ceruloplasmin,sweat chloride test iron, transferrin saturation, ferritin,blood and urine amino acide, alpha1-antitrypsin. Autoantibodies related to autoimmune hepatitis were negative including anti-

nuclear antibodies, anti-smooth muscle antibody, anti-liver-kidney microsomal type-1, and there was no hypergammaglobulinemia.

In the presence of portal hypertension without alterations in liver tests, especially following the identification of portal vein thrombosis on subsequent ultrasound, a thrombophilia investigation was conducted. This included assessments of protein C and S levels, antithrombin III activity, heterozygous factor V Leiden mutation, anti-cardiolipin antibodies, and Beta-2 glycoprotein 1 antibodies. The results were within normal limits, except for hyperhomocysteinemia of 47.9 umol/l (range 6.6-17.8 µmol/l). Dried blood spot analysis yielded negative results. Endoscopic examination of the upper gastrointestinal tract revealed grade II esophageal varices with red signs and no evidence of hypertensive gastropathy. Echocardiography detected a thrombus in the right atrium.

A percutaneous liver biopsy revealed subcapsular fibrosis and vascular alterations. The portal system demonstrated patency, with no evidence of cavernous transformation. A wedge liver biopsy exhibited a well-preserved liver architecture, showcasing normal appearances of , hepatic arteries,portal tracts, and bile ducts. Fibrous bands were observed and it extends from the internal capsule to the parenchyma and from portal areas to adjacent parenchyma. Subendothelial fibrous thickening was noted in portal vein branches, leading to thickened vessel walls and irregular luminal contours. Abnormal portal vein branches protruding from portal tract borders were also identified. (Figure 3 (a,b,c,d)). Despite the initiation of anticoagulant treatment, the patient's condition deteriorated, leading to a severe secondary pulmonary embolism due to right atrial thrombosis, ultimately resulting in death.

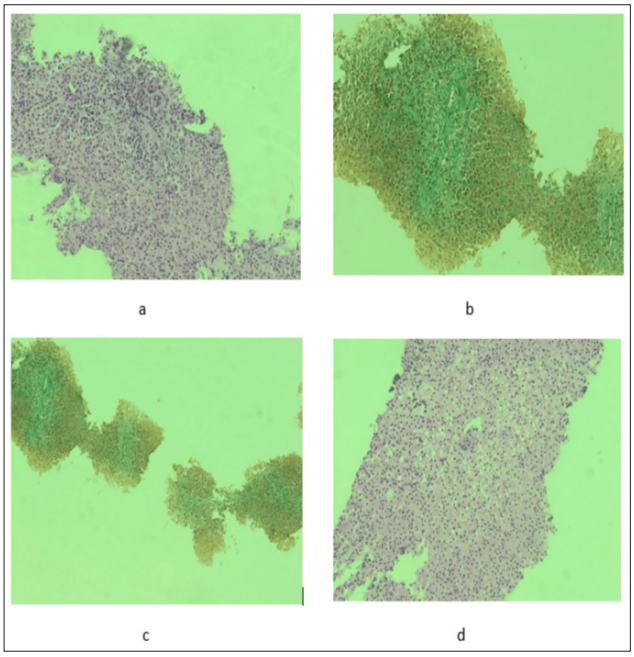

Figure 3:

A: Intraparenshymal Fibrosis with HES (X10),

B: Bridges of Sclerosis Between Portal Areas Clearly Seen With Trichroma Stains (X20)

C: Intraparenchymal and Periportal Sclerosis With Trichroma Stains (X10)

D: Herniation of Portal Veins HES (X10)

Discussion

Hepatoportal sclerosis (HPS) is a sclerosis in the portal areas whithout cirrhosis.the disease is frequent in developping country; in the litterature, There is limited data about HPS in children. Exposure to chemical agents has been implicated such as azathioprine, vinyl chloride, copper sulfate, and arsenic. an etiology was proposed in litterature of Intestinal bacterial infections with repeated septic embolization of the portal circulation [1-4]. Diarrheal episodes in infants,early childhood umbilical sepsis in newborn and bacterial infections are likely to cause a portal pyemia and pylophlebitis, leading to thrombosis, sclerosis, and obstruction of small- and medium-sized portal vein radicals [1]. Patients with hepatoportal sclerosis typically

present with upper gastrointestinal bleeding, often attributed to the rupture of esophageal varices, and report abdominal discomfort associated with splenomegaly [1-3]. Hepatic functions are generally preserved [1].

nodular transformation of the liver with extensive portal and sub-hepatic fibrosis has been noted in a small group of patients with ascites and hypoalbuminemia [1,4]. in our case the patient exhibited features of portal hypertension with normal liver function.

The coagulation abnormalities observed in these patients, including variations in protein C, S, and antithrombin III activities, antiphospholipid antibodies, and mutations in factor II and factor V Leiden, along with hyperhomocysteinemia, suggest a subclinical hepatic involvement affecting clotting factors that rely on the liver. This leads to a prolonged INR without significantly affecting the partial thromboplastin time [5]. Our patient exhibited hyperhomocysteinemia, a condition linked to the development of right atrial thrombosis and an increased risk of vascular diseases, including thrombosis in various blood vessels.

Research indicates that elevated homocysteine levels may contribute to endothelial dysfunction and foster a prothrombotic state. Further investigation and research are essential to understand the potential role of hyperhomocysteinemia in the context of hepatoportal sclerosis and its associated complications, such as portal vein thrombosis [1].

in biopsy specimens from individuals with hepatoportal sclerosis the histopathological examination typically reveals an abberrant portal veins whithin portal tracts causing by a sclerosis or obliteration of portal vein branches [6]. Characteristic features include pseudonodule formation, septa extending from portal areas into neighboring tissues, and extension of fibrosis into the hepatic parenchyma from subcapsular areas [7]. The biopsy specimen from our patient exhibited most of these characteristic finding.

The primary approach for treating hepatoportal sclerosis is endoscopic sclerotherapy. This procedure, typically performed in about six sessions, has proven to be effective in eliminating varicose veins, especially in pediatric cases. Sclerotherapy boasts a high success rate, successfully controlling approximately 95% of episodes involving upper gastrointestinal hemorrhage (UGI) [8]. Additionally, considering prophylactic propranolol administration may be a viable complementary therapeutic strategy. However, in situations involving hepatocellular failure and associated complications, liver transplantation remains a critical intervention to ensure optimal patient outcomes [1,8].

The typical course of hepatoportal sclerosis in children can vary significantly. In certain instances, the condition may exhibit a stable trajectory with minimal impact on liver function, allowing affected children to lead relatively normal lives without significant complications. However, it is crucial to emphasize that the course can differ markedly among individuals, and potential complications such as gastrointestinal hemorrhage due to the rupture of esophageal varices and the risk of thrombosis need to be considered [1,9]. On the other hand, the coexistence of fibrosclerosis and portal fibrosis could contribute to the deterioration of liver synthesis, necessitating contemplation of liver transplantation. Substantial portal fibrosis and phlebosclerosis may play a role in the impairment of parenchymal and posterior hepatic synthetic functions. Continuous monitoring and appropriate management are essential to evaluate the progression of hepatoportal sclerosis and address any emerging complications in pediatric cases [9-11].

Conclusions

Hepatoportal sclerosis is a rare etiology of portal hypertension in childhood, it is primarily observed in adults, represents a rare etiology of portal hypertension in childhood. The pathogenesis of this disease is not clearly known, we need a multi-center studies and registries to elucidate and understand the characteristics of hepatoportal sclerosis (HPS) in children. Rigorous research and clinical studies are imperative to unravel the underlying mechanisms linking hyperhomocysteinemia and hepatoportal sclerosis in the pediatric population. A nuanced comprehension of this correlation will enhance diagnostic protocols and therapeutic strategies for young patients grappling with this uncommon medical entity.

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