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CASE REPORT

Clue to the cause of portal hypertension: Look at the raindrops

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Abstract

Portal hypertension is a clinical syndrome characterized by splenomegaly, with or without hypersplenism, and esphagogastric varices. Non-cirrhotic portal hypertension (NCPH) belongs to the presinusoidal sub category of portal hypertension. We present the case of a young 35-year-old male who had presented with two episodes of upper gastrointestinal bleed. On endoscopy, he was found to have large esophageal varices. On general physical examination, there were diffuse hyperpigmented papules and nodules all over the body, with palmoplantar thickening. His liver function tests were normal. Computed tomography of the abdomen showed a dilated portal and splenic vein with splenomegaly and normal liver size and histopathology showed non-cirrhotic portal fibrosis (NCPF). He had history of intake of oral indigenous medications for skin lesions. On investigating, patient was found to have chronic arsenicosis, which was likely the underlying etiology for NCPF as well. Hence, while evaluating patients of NCPF, it is imperative to rule out the use of indigenous medications, especially in the Indian scenario.

Introduction

NCPH is known to occur in all parts of the world but is more prevalent in developing countries. It accounts for one third of cases of variceal bleed in India.[1] Multiple risk factors are known to be associated with NCPF. These include early and recurrent gastrointestinal infections, prolonged xenobiotic exposure, prothrombotic states and auto immune disorders. Arsenic has traditionally been used in Western as well as Indian medical systems for treating diseases like psoriasis, eczema and leukemia. Although this practice has been almost completely abandoned by the Western society, it is still prevalent in India. Chronic use of arsenic has been associated with development of non-cirrhotic portal fibrosis.[2,3] We present a case of NCPF with signs of chronic arsenic toxicity.

Case report

A 37-year-old man presented with a single episode of painless hematemesis. On examination, hyperpigmented macules were noted all over the body. Papular thickening of the palmar and plantar skin was observed to lead to a "raindrop appearance" (Fig. 1a,b). Furthermore, the patient gave a history of erythematous papules over extensor aspects of the limbs for 15 years. He had been taking indigenous medicine for the same, and the lesions had resolved gradually over 5 years. However, the patient had, by then, begun to develop new hyperpigmented patches as noted above. His upper gastrointestinal endoscopy demonstrated large esophageal varices with portal hypertensive gastropathy, for which band ligation was performed. Liver function tests showed normal values of transaminases and albumin suggesting preserved synthetic function, and workup for chronic liver disease was nonindicative. Complete blood counts demonstrated pancytopenia. Computed tomography of the abdomen showed a dilated portal and splenic vein with massive splenomegaly (Fig. 1c). Liver stiffness measurement by transient elastography was 10 kPa. Transjugular liver biopsy and estimation of hepatic venous pressure gradient (HVPG) was performed, and the HVPG was 13 mm Hg. Liver biopsy showed noncirrhotic portal fibrosis (Fig. 1d). Urine and hair arsenic levels were elevated ([683 μg/L (normal ≤ 50 μg/L) and 6.22 μg/gm (normal < 1 μg/gm], respectively). Skin biopsy was suggestive of psoriasis vulgaris with chronic arsenicosis.

Discussion

Non-cirrhotic portal hypertension is a heterogenous group of disorders which cause resistance at presinusoidal level, leading to portal hypertension. Multiple hypotheses have been proposed for explaining the pathogenesis of portal hypertension in cases of NCPF. The most widely accepted is the unifying hypothesis.[4] Unifying hypothesis states that recurrent infections leading to mild repeated microthrombotic events in peripheral branches of portal vein lead to NCPF. Patients with NCPF usually present with episodes of painless hematemesis and are found to have

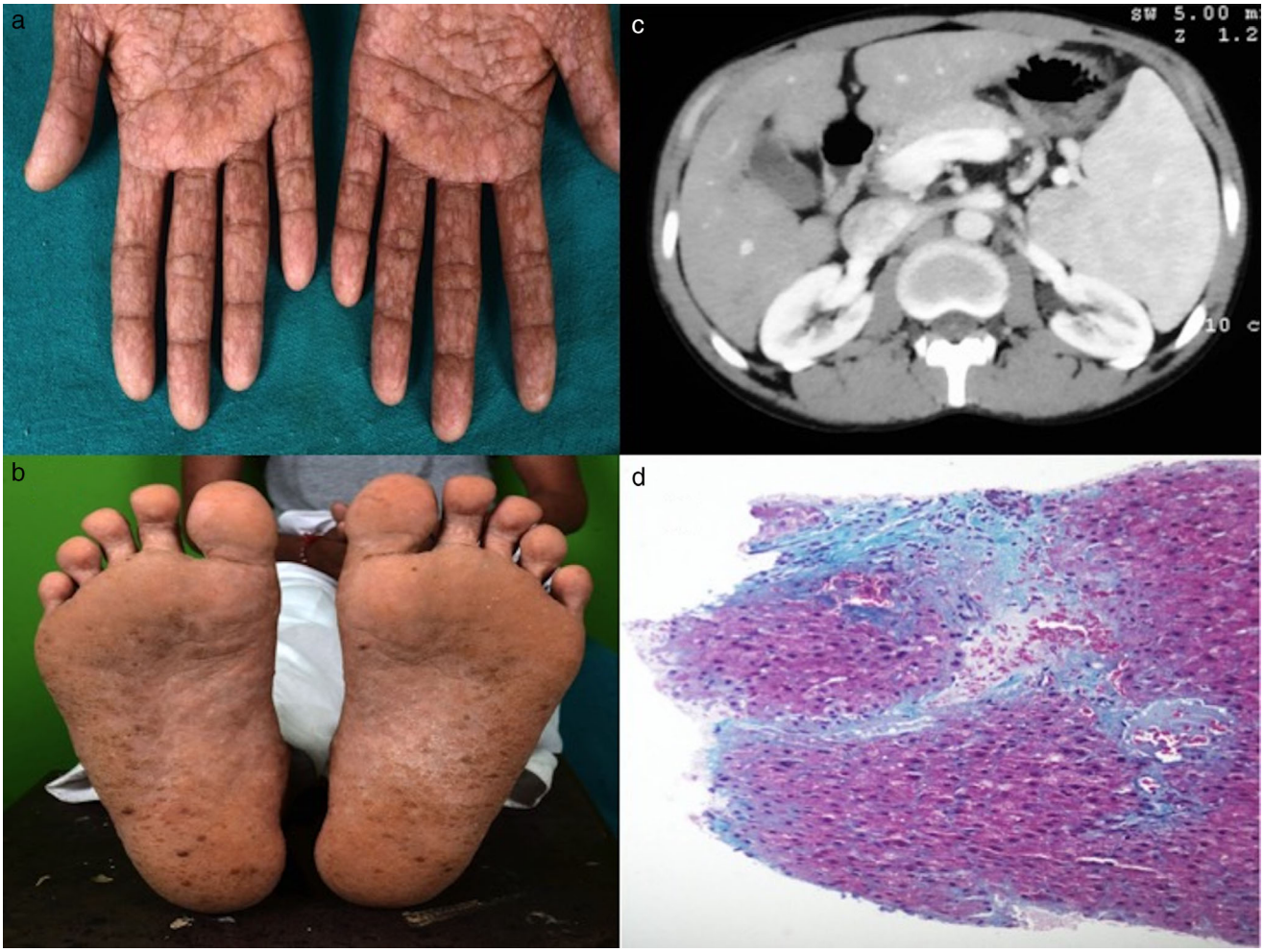

Figure 1 (a, b) Papular thickening of the palmar and plantar skin giving a "raindrop appearance." (c) Computed tomography image showing dilated splenic vein with splenomegaly. (d) Liver biopsy showing noncirrhotic portal fibrosis.

splenomegaly. Although liver functions are preserved in these patients, they may later go on to develop parenchymal atrophy and decompensation. NCPF is diagnosed based on clinical findings and radiological features. Doppler USG is the first line investigation. Liver is typically normal in size, outline and echotexture. Spleen is enlarged. splenoportal axis is dilated and patent. Portal vein is thickened (>3 mm) with echogenic walls. Narrowing occurs at the level of second or third degree intra hepatic radical. This leads to a "withered tree" appearance.[1] HVPG is significantly lower in NCPF (mean 9.1 mm Hg) as compared to patients with cirrhosis (mean 25.8 mm Hg).[5] Treatment of NCPF consists of measures taken to prevent gastrointestinal bleed. This includes endoscopic, radiological and surgical interventions. Steps should be taken to identify the underlying etiology and treat accordingly. Arsenic-containing compounds continue to be prescribed by complementary alternative medicine systems in India. In one study of 248 cases of chronic arsenic toxicity from West Bengal, 190 cases had hepatomegaly. Of the 69 patients who underwent liver biopsy, 91.3% showed a change in NCPF.[6] Hence, one must look into the use of indigenous medicines in cases with portal hypertension of unknown etiology.

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