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Budd Chiari Syndrome Revealed by Spinal Cord Compression

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Abstract **Case Report**

Budd-Chiari syndrome results from obstruction of hepatic venous drainage, from the hepatic venules to the terminal part of the inferior vena cava, whatever the cause of the obstruction. It may be of primary or secondary origin. In our case, we report a case of Budd-Chiari syndrome with thrombosis of the terminal part of the inferior vena cava extending to the right suprahepatic vein, revealed by an unusual picture of functional impotence of all 4 limbs due to spinal cord compression caused by collateral venous circulation. Etiological assessment for budd chiari syndrome revealed a protein C deficiency, for which the patient was put on lifelong anticoagulation.

Keywords: Budd chiari, collateral venous circulation, functional impotence.

INTRODUCTION

Budd-Chiari syndrome or suprahepatic block is a rare entity characterized by obstruction of hepatic venous flow in the vascular space between the hepatic venules and the junction between the inferior vena cava and the right atrium, thus excluding upstream causes (sinusoidal obstruction syndrome) and downstream causes (cardiac causes), most often associated with congenital or acquired haemostasis disorders, sometimes with compression by a neighbouring expansive lesion or intraluminal invasion by a malignant tumour.

In this context, we report an unusual presentation of Budd Chiari syndrome in a 47-year-old patient, admitted to the Neurology Department for etiological assessment of functional impotence of all 4 limbs. The etiological work-up revealed Budd Chiari syndrome at the stage of portal hypertension responsible for nerve root compression by collateral venous circulation.

OBSERVATION

This 47-year-old patient was admitted to the neurology department to assess the etiology of subacute functional impotence of all 4 limbs, which had been evolving for a month and a half, associated with spinal pain and a sensation of electric discharge on flexion of the neck, with no sphincter disorders and no other associated extradigestive or digestive manifestations. All this evolving in a context of apyrexia and altered general condition. Clinical examination revealed a walking deficit with assistance, hypotonia and a predominantly distal motor deficit. The patient held the barré in the 2 upper limbs and the magazzini in the 2 lower limbs, with good tone in all 4 limbs, superficial hypoesthesia of the left lower limb with a spinal syndrome. Abdominal examination was unremarkable apart from epigastric venous circulation with homogeneous hepatomegaly with FH at 16cm.

Spinal MRI revealed multiple dilated vascular structures draining into the inferior vena cava, with foraminal extension and nerve root compression at D11, D12, L1 and L2. An abdominal CT scan confirmed the diagnosis of Budd Chiari syndrome, showing homogeneous hepatomegaly with thrombosis of the terminal portion of the inferior vena cava extending to the right suprahepatic vein. The portal trunk was patent, with portocaval venous circulation. Electromyography revealed axonal myeloradiculoneuritis in all 4 limbs. HIV, syphilis, HBV and CVH serology were all negative. The immunological work-up was negative, and cerebrospinal fluid aspiration was normal.

A diagnosis of Budd Chiari syndrome was made, and a protein C deficiency was identified as part of the etiological work-up. A FOGD was carried out, showing grade 1 esophageal varices without signs, for which he was put on non-cardioselvic beta-blockers for

primary prophylaxis and put on LMWH with relay to anti-vitamin K as etiological treatment for Budd Chiari syndrome, and put on symptomatic treatment with motor rehabilitation for his functional impotence.

DISCUSSION

The definition of Budd-Chiari syndrome recently drawn up by a group of European experts was validated at the last European Consensus Conference on Portal Hypertension [1]. and in the European recommendations of the EASL: Budd-Chiari syndrome results from obstruction of hepatic venous drainage, from the hepatic venules to the terminal part of the inferior vena cava, whatever the cause of the obstruction. Budd-Chiari syndrome is classified as either primary or secondary [1]. Secondary Budd-Chiari syndrome is defined as endoluminal obstruction by material of extravascular origin (benign or malignant tumour, abscess, parasitic or non-parasitic cyst) or by extrinsic compression. In other situations (thrombosis, stenosis), Budd-Chiari syndrome is primary. We report the observation of a Budd-Chiari syndrome secondary to thrombosis of the terminal part of the inferior vena cava extended to the right suprahepatic vein. The etiology of Budd-Chiari syndrome in this case was protein C deficiency. The originality of this observation lies in the existence of neurological symptoms linked to the development of voluminous epidural varices. Epidural varices are a rare entity. They can cause nerve or spinal cord compression, simulating a herniated disc [2]. The avalvular epidural venous system forms an anastomosed network around the vertebral elements coming into close contact with the dural sac and foramen, and drains into the venous cave and azygos systems. Epidural varicose veins are most frequently found in the lumbar region. Epidural varices are most often associated with degenerative lesions of the vertebral disc. They have also been described in some cases of obesity and pregnancy [3]. This type of complication has never been reported in isolated Budd-Chiari syndromes. On the other hand, a recent study reported 13 cases of symptomatic epidural varices secondary to inferior venous thrombosis of various etiologies [4]. In our case, the diagnosis was suspected by the appearance of neurological signs associated with signs of portal hypertension, and confirmed by MRI, currently considered to be the most effective examination [5].

CONCLUSION

Budd-Chiari syndrome is a rare entity, most often associated with congenital or acquired disorders of hemostasis. Diagnosis is based on imaging. Treatment of Budd-Chiari syndrome relies mainly on anticoagulants.

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