ECRONICON OPEN ACCESS

EC GASTROENTEROLOGY AND DIGESTIVE SYSTEM

Case Report

Diffuse Hepatosplenic Angiofibromatosis with Fatal Outcome: A Case Report

Dalila Tagzout*, Imene Khedairia and Amar Tebaibia

Algiers Faculty of Medicine, Algiers University, El Biar Hospital, Algeria

***Corresponding Author:** Dalila Tagzout, Algiers Faculty of Medicine, Algiers University, El Biar Hospital, Algeria.

Received: July 29, 2021; **Published:** September 28, 2021

Abstract

We report the case of an adult patient in whom the diagnosis of diffuse hepatosplenic angiofibromatosis was revealed during exploration for abdominal pain. The diagnosis was made on histological evidence with immunohistochemistry. The progression was towards clinical and biological worsening with hepatocellular failure followed by hepatorenal syndrome without improvement on treatment progressing to death of the patient four months after diagnosis.

Keywords: *Hepatosplenic Angiofibromatosis; Hepatorenal Syndrome; Abdominal Pain*

Abbreviations

IHC: Immunohistochemistry; CT: Computed Tomography

Introduction

Hepatosplenic angiomas are common benign tumors with a good prognosis. However, diffuse hepatic angiofibromatosis is a severe form defined by the presence of cutaneous angiofibromatosis with involvement of at least two visceral organs [1], it is observed in newborns and manifests as an abdominal mass and a cardiac damage. This diffuse form of hepatic angioma is very rare in adults, with a phenotype often different from the neonatal form. We report the case of an adult patient in whom the diagnosis of diffuse hepatosplenic angiofibromatosis was made on histological evidence with immunohistochemistry (IHC).

Observation

49-year-old patient, with no particular medical-surgical history, smoking at a rate of 30 packs a year having presented, diffuse abdominal pain more important in the left hypochondrium, which has progressed for three months with progressive worsening in a context of deterioration of general condition with asthenia, weight loss not quantified, nocturnal fever. Clinical examination found an abdomen sensitive to palpation, grade 4 splenomegaly and left basal pleural fluid effusion, there were no other abnormalities noted, particularly dermatological. The cytological study of pleural puncture fluid found inflammatory pleural fluid predominantly lymphocytic. The biological assessment found normocytic normochromic anemia at 11 g/dl of Hb, hyperleukocytosis at 11,500/mm^3, thrombocytosis at 900,000 elements/mm^3, a biological inflammatory syndrome with VS at 72 mm the first hour and a CRP at 96 mg/l, fibrinogen at 4.7 g/l, electrophoresis of plasma proteins was in favor of a subacute inflammatory reaction. The hepatic assessment was discreetly disturbed with an alkaline

phosphatase level twice the normal, the rest of the hepatic assessment (ASAT, ALAT, Gama glutamyltransferase, bilirubin) was normal as well as the renal assessment and the blood ionogram. Abdominal ultrasound revealed heterogeneous hepatomegaly with heterogeneous splenomegaly without signs of portal hypertension, there is no dilation of the intra or extrahepatic bile ducts. The etiological assessment of hepatopathy (viral serology B and C, autoimmunity assessment, iron assessment) is negative, the HIV serology was negative, the tuberculosis assessment (with intra dermo reaction to tuberculin and bacilloscopy) was negative, the assay of tumor markers ACE, CA19.9, alpha foeto protein, PSA was normal. Thoraco-abdominal-pelvic computed tomography (CT) revealed heterogeneous enhancing hepatomegaly with scattered millimeter microdensities and a globular splenomegaly with a height of 188 mm with the presence of an aspect of diffuse splenic parenchymal necrosis; this necrotic, liquefied, strained spleen lifts the left diaphragm (Figure 1). Upper gastrointestinal endoscopy revealed an aspect of erosive erythematous pangastritis. There are no other endoscopic abnormalities. The histological study of gastric biopsies and systematic duodenal biopsies was in favor of mild gastritis and nonspecific duodenitis. Given the abnormalities in the blood count, a bone biopsy was performed, the histological examination with IHC with glycophorin, myeloperoxidase, CD34, CD20 and CD3 concluded in a hematopoietic marrow of richness 2 to 3 polymorphic, no signs in favor of 'chronic myeloproliferative neoplasia, no lymphomatous or carcinomatous tumor infiltration. The patient was put on analgesic treatment without improvement in clinical symptoms with worsening of the pain in the left hypochondrium. Faced with necrotic splenomegaly and under tension with uncontrolled abdominal pain, the therapeutic decision after multidisciplinary consultation: (Hematology, internal medicine, general surgery) was to perform a splenectomy. The histopathological study supplemented by the IHC of the splenic parenchyma revealed a cellular and vascular proliferation of moderate density accompanied by areas of extensive necrosis. The IHC found: smooth muscle actin positive, CD34 positive, H-caldesmone negative and HMB45 negative (Figure 2). These results were in favor of an angiomyofibroma without pejorative character and the appearance of the necrosis is more favor of infarction necrosis.

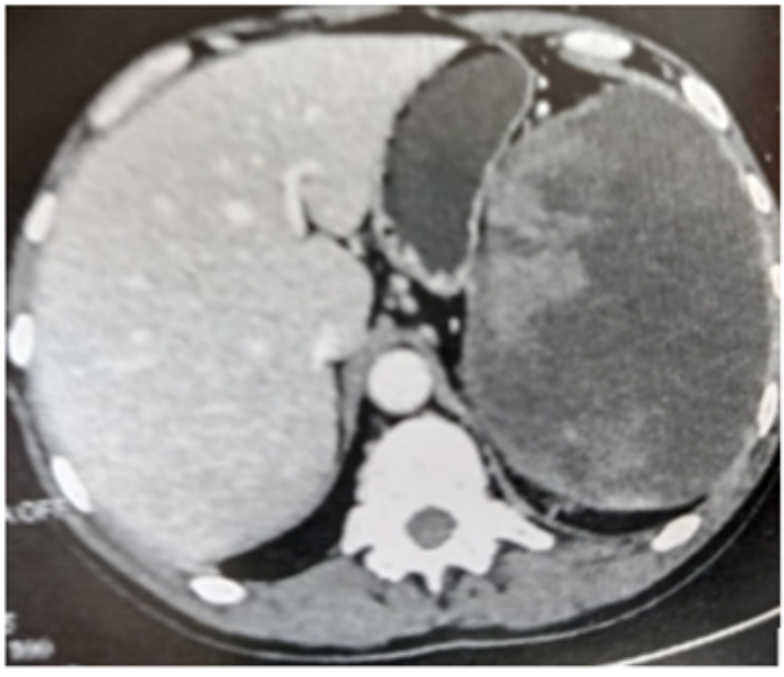

Figure 1: *Abdominal CT cut.*

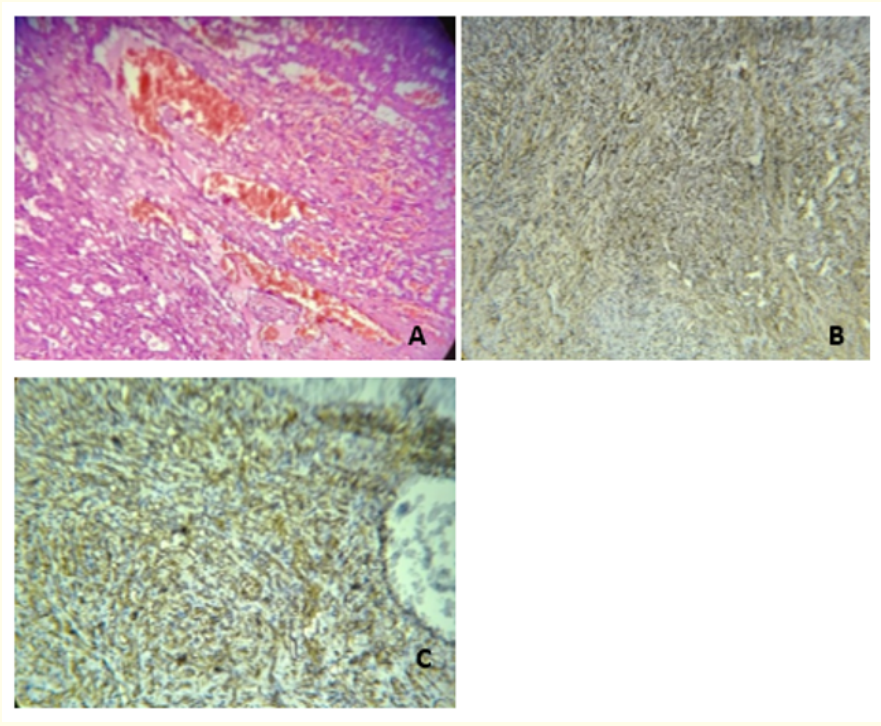

Figure 2: *Histological appearance of the splenic parenchyma. A: Moderate density cell and vascular proliferation with extensive patches of necrosis. B: IHC: positive smooth muscle actin. C: IHC: CD34 positive.*

The evolution was made towards the worsening of the clinical picture with the appearance of intense diffuse abdominal pain, the clinical examination found an ascites of great abundance, edema of the lower limbs, a fluid pleural effusion of average abundance, the evolution of the biological assessment was made towards a disturbance of the hepatic assessment with a cholestasis syndrome (alkaline phosphatase at 2 times the normal, Gamma glutamyl transferases at 2 times the normal, minimal increase in the level of conjugated bilirubin), hypo albuminemia at 22 g/l, with a prothrombin rate of 65%. The exploration of hepatic morphological abnormalities found negative viral B and C serologies, negative autoimmune hepatitis assessment. Thoraco-abdominal CT shows an enlarged liver with irregular contours dotted with microlesion taking the contrast to the arterial phase giving the liver a punctuated appearance with persistence of the heterogeneous appearance at the different phases of the injection with peritoneal effusion of average abundance free without sign of carcinoma and on the thoracic level a bilateral pleural effusion of small abundance on the right and moderate abundance on the left without pericardial effusion. The biochemical analysis of the ascitic fluid found a transudate at 11 g/l, the bacteriological examination with culture was negative and the cytological examination was unremarkable. The cardiac ultrasound found undilated cardiac chambers, a 68.8% ejection fraction and minimal pericardial detachment. A scano-guided hepatic biopsy puncture was performed, finding a hepatic parenchyma greatly altered by a poorly limited vascular lesion, dissociating the hepatic parenchyma, made of blood capillaries of variable size with clean walls with a regular endothelium, hyperplastic in places, the hepatocytes show degenerative and regenerative aspects, an objective IHC complement: actin-smooth muscle positive, CD34 positive; result in favor of diffuse hepatic angiofibromatosis. The patient was put under symptomatic treatment with analgesics, albumin supplementation, diuretic treatment but without improvement of symptoms and ascites and worsening of the hepatic insufficiency TP at 35% and factor V at 30%. The progression was to refractory ascites requiring several evacuating ascites punctures and then to hepatorenal syndrome without any improvement leading to the death of the patient.

Discussion

Hepatic angioma is the most common benign tumor of the liver occurring at any age, while diffuse hepatic hemangiomatosis is a clinical form occurring mainly in children and can be life-threatening [1]. In addition, hepatic hemangiomatosis is very rarely seen in adults. Our patient presented a diffuse hemangiomatosis with two hepatic and splenic localizations proven histologically with the presence of an increase in endothelial vascular proliferation taking the specific staining in IHC allowing a diagnosis with certainty and eliminating other hepatic tumor proliferations such as hemangioendothelioma or l ' angiosarcoma. Indeed, there is no specific clinical or radiological sign of diffuse hemangiomatosis and the differential diagnosis with other rare vascular tumors of the liver is based only on histological examination with IHC [2]. The prognosis is not yet clear, especially for the isolated hepatic form, for our patient the evolution was rapid with worsening of the clinical symptoms and the appearance of complications of hepatocellular insufficiency with refractory ascites and hepato-renal syndrome progressing to the death of the patient. The progression of hepatic angiofibromatosis to death from hepatic failure has been reported in the literature [3].

Conclusion

Diffuse hepatosplenic hemangiomatosis in adults is a rare entity to recognize, the diagnosis with certainty is based on histological examination with immunohistochemistry which makes it possible to rule out other tumor proliferations. The prognosis remains bleak. Our case illustrates the seriousness of this tumor variety with hepatic and splenic localization leading to hepatic insufficiency and its complications and then to the death of the patient.

Bibliography

1. Lopriore E and Markhorst DG. "Diffuse neonatal haemangiomatosis: new views on diagnostic criteria and prognosis". *Acta Paediatrics* 88 (1999): 93-97.

2. Bishop PR., *et al*. "Radiological case of the month. Hepatic hemangiomatosis". *The Archives of Pediatrics and Adolescent Medicine* 154 (2000): 743-744.

3. Eun Hui Kim. "Diffuse Hepatic Hemangiomatosis without Extrahepatic Involvement in an Adult Patient". *Korean Journal of Radiology* 9 (2008): 559-562.

Volume 8 Issue 10 October 2021