

Non-Cirrhotic Portal Hypertension in Children at the Kamenge University Hospital Center (Burundi): About One Case

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ABSTRACT

Non-cirrhotic portal hypertension is a rarely described condition in children. It occurs mainly in young adults. Significant splenomegaly exists long before diagnosis. The occurrence of complications constitutes the main mode of discovery of the disease. We report the case of a 12-year-old patient, with a history of splenomegaly since the age of 2 years, in whom the diagnosis of Banti syndrome was made following hypersplenism. occurred during follow-up for esophageal varices due portal hypertension syndrome.

Keywords

Portal hypertension, Hypersplenism, Child, Splenectomy.

finding of Banti's syndrome in a 12-year-old girl during follow-up for upper gastrointestinal hemorrhage.

Introduction

Non-cirrhotic or idiopathic portal hypertension or hepatoportal sclerosis, also called Banti's syndrome, is a rare entity in children. It has an unknown pathogenesis and generally affects young adults [1]. Affected subjects are in apparent good general condition. We note in these patients almost always one or more episodes of hematemesis, which are well tolerated, and a history of a mass in the left hypochondrium [2]. Banti syndrome, described for the first time by Guido BANTI in 1898 as a condition associating portal hypertension with splenomegaly and anemia in a healthy liver, is characterized by the presence of an increase in portal pressure secondary to sclerosis of the wall of the small intrahepatic portal branches, with a non-cirrhotic liver [3,4]. Hypersplenism is often found in these patients [5]. Splenectomy alone may be justified in the management of this syndrome. It makes it possible to treat splenomegaly and correct hypersplenism [6]. In order to show the importance of properly conducting investigations in the face of portal hypertension in children, we report a case of an incidental

Observation

This is a 12-year-old girl living in the north of Burundi in the MUYINGA commune, third of a family of four children all alive and in good health, with no particular history except splenomegaly of unknown etiology which had been discovered at the age of two. She had consulted the gastroenterology department of the Kamenge University Hospital Center in December 2019 for etiological assessment and management of an upper gastrointestinal hemorrhage. Her height and weight growth and her pubertal development were consistent with her age. She had normal vital signs, conjunctival pallor, and a slight umbilical eversion. Her abdomen was supple with splenomegaly extending into the pelvis.

Additional tests were carried out. The CBC showed hyperleukocytosis at 11,640/mm³, normocytic normochromic anemia with hemoglobin at 6.1 g/dl, thrombocytopenia at 38,000/mm³. The other tests carried out (Uremia, creatinine level, Blood sugar, Gamma GT, transaminases, alkaline phosphatases, HBsAg,

Antiviral hepatitis C antibodies, electrolyte panel, blood smear) were unremarkable. The Prothrombin was low at 42%, the INR was slightly increased at 1.79; the albumin level showed hypoalbuminemia at 20.58 g/L. Abdominal ultrasound showed moderate hepatic fibrosis without objective suspicious focal lesions, periportal fibrosis with portal flow presenting aliasing on color Doppler, signs of portal hypertension, splenomegaly, low abundance ascites, dilatation of the splenic veins. Esophagogastroduodenoscopy showed esophageal varices. An anatomopathological examination of the liver has not been done.

At this time, we concluded that there was a syndrome of portal hypertension with rupture of esophageal varices and probable bilharzia as the child came from a rural area. The child was put on praziquantel tablet 300 mg per day for three days, ampicillin 100 mg/kg or 1g every 8 hours by IV and Metronidazole 10 mg/kg or 125 mg by IV every 8 hours for 8 days, duphalac syrup, 8 transfusions of 500 CC of packed red blood cells, omeprazole 80 mg per 24 hours in 500 CC of G 5%, propranolol tablet 10 mg at a rate of 2 tablets per day, aldactone tablet.

A follow-up was organized. The assessments carried out since December 26, 2019 had consistently shown bicytopenia (leukocytes around 2000/mm³, platelets around 30,000/mm³). The esophagogastroduodenoscopy which should have been done in July was postponed for fear of creating a hemorrhage on severe thrombocytopenia. She will be referred to the pediatric department for management of bicytopenia. At this time, the examination always noted enormous splenomegaly and after reanalyzing the case (the history, the semi-urban living environment outside the rice-growing regions, the evolution of the blood lines in the CBC from February to July, the liver ultrasound showing periportal fibrosis without hepatic cirrhosis), the conclusion was hypersplenism due to Banti's syndrome. After multidisciplinary staff (pediatric hepatogastroenterologists, intensivists), a splenectomy was decided and carried out. In the postoperative aftermath, there was a large hematoma in the splenic bed, which was treated with a wait-and-see approach and analgesic treatment based on paracetamol at 500 mg every 6 hours for 3 days. The hematological assessment normalized after the splenectomy with CBC which showed white blood cells at 8000/mm³, hemoglobin at 11.5 g/dl, platelets at 256,000/mm³.

The patient was discharged on postoperative day 23 on penicillin V at a rate of 1g/day in two doses while awaiting administration of the pneumococcal vaccine pneumo 23 (the vaccine was not available at that time because it was not free, the family had to buy it) which was carried out one month after discharge from the hospital. The clinical and biological examination remained normal over 2 years of follow-up and there was no recurrence of digestive bleeding

Discussion

Idiopathic portal hypertension in non-cirrhotic liver is rare, it can be found anywhere in the world, but it is far more common in countries with limited resources. Its frequency is poorly known

but some countries such as India estimate its frequency at 30% of portal hypertension [1]. This entity is not very reported in children although manifestations such as splenomegaly can begin very early in childhood. It mainly affects young adults between the 3rd and 4th decades [7]. Walid L et al. described a case in Morocco in a 36-year-old woman [8]. The 3 cases reported by Mahomed SB et al. had an age varying between 22 and 41 years [6]. A female predominance is reported in the literature [6,9]. Our patient was also female but was younger compared to the cases reported in the literature.

Clinically, the beginning is marked by the appearance of an enlarged spleen, progressively evolving, of variable size, discovered during a systematic examination or following a feeling of heaviness and soreness of the left hypochondrium [6]. But patients most often consult at the stage of portal hypertension (PH). The examination reveals, in addition to splenomegaly, collateral circulation, ascites, edema, etc. Sometimes, the circumstances of discovery can be digestive hemorrhages due to rupture of esophageal varices, an anemic syndrome secondary to hypersplenism [8,9]. Our patient had a history of splenomegaly discovered since the age of 2 during routine consultations but which had not greatly worried the family. The consultation with the specialist will be done during a digestive hemorrhage during the rupture of esophageal varices 10 years later, therefore at the age of 12.

The notion of a history of geophagia or jaundice described in the literature [10] was not found in our patient. But what concerns the history of enterocolic infections mentioned by the same literature [10] although difficult to assert could not be formally eliminated either because they are common in our countries with limited resources. Ascites is rare but can be found in small amounts in Banti's syndrome after bleeding [2]. Our patient who came for ruptured esophageal varices had low levels of ascites.

The occurrence of hypersplenism made it possible to make the diagnosis of Banti's syndrome in our patient. The results of the additional examinations carried out were similar to the data in the literature. In this condition, biology shows pancytopenia confirming hypersplenism, while the hemostasis assessment and the liver assessment are not altered [10]. A somewhat low Prothrombin is noted in some cases, it was 42% in our case and 54% for the case of Walid L et al. [8].

Abdominal ultrasound often reveals dilatation of the portosystemic axis associated with thickening of the walls of the portal vein and its main branches with splenomegaly without abnormalities of hepatic morphology [11]. Abdominal Doppler ultrasound confirms the enlargement of the spleen, assesses the size of the liver, and allows exploration of the venous system. In our patient, the ultrasound observed moderate hepatic fibrosis without objective suspicious focal lesions, periportal fibrosis with portal flow presenting aliasing on color Doppler, signs of portal hypertension, splenomegaly, low abundance ascites, dilation of the splenic veins. Similar results are noted on ultrasound in the case of Walid L. [8] with a dilated splenic vein, heterogeneous dysmorphic liver

without suspicious focal lesion and splenomegaly.

Esophageal varices are frequently demonstrated in Banti's syndrome in 85 to 95% of patients with esophagogastroduodenoscopy [12,13]. Just like in our patient, they were found in all Mahomed SB patients [6]. The management of Banti's syndrome is based on a set of medical, instrumental and surgical means. It is mainly based on the treatment of hematemesis due to portal hypertension and the treatment of hypersplenism [14]. Ligation of esophageal varices associated with beta-blockers is often indicated to treat and prevent hemorrhage from esophageal varices as in the case of portal hypertension in cirrhosis [8]. Our patient had benefited from ligation of the esophageal varices and placement on beta-blockers initially but the occurrence of hypersplenism with severe thrombocytopenia and leukopenia required the use of laparotomy for splenectomy. This support is consistent with literature data. Surgical management is indicated in patients presenting with symptoms such as hypersplenism, severe anemia requiring repeated blood tests, transfusions or repeated episodes of splenic infarction. Splenectomy can be performed by laparotomy or laparoscopy [15]. In the study by Mahomed SB, all patients underwent splenectomy associated with ligation of esophageal varices with maintenance treatment with beta-blockers and the results were satisfactory [6]

Common complications of splenectomy include bleeding [15]. In our patient, the postoperative course was marked by the occurrence of a large hematoma in the splenic bed, the treatment of which was monitoring and analgesic treatment. Vaccination against hemophilus influenzae type B, influenza, pneumococcus, meningococcus, tetanus/pertussis/diphtheria is recommended at least 14 days before elective surgery and 14 days after surgery otherwise taken preoperatively [8]. In our patient, it was not possible to obtain all the vaccines and within the recommended time frame, only the pneumococcal vaccination was administered. But the short and medium term evolution was favorable in our patient as the literature states that postoperative recovery and prognosis are excellent [8]. Furthermore, although the natural history of Banti's disease is poorly understood, it appears to have a prolonged benign course with a very good prognosis [3].

Conclusion

Banti's syndrome should be considered in children when faced with a picture of portal hypertension with chronic splenomegaly without known liver pathology. A good history, imaging tests, biological tests and a fibroscopy can lead to the diagnosis. Surgical treatment based on splenectomy improves the prognosis.

References

1. Cazals-Hatem D, Hillaire S, Rudler M, et al. Obliterative portal venopathy: Portal hypertension is not always present at diagnosis. *J Hepatol*. 2011; 54: 455-461, Epub 2010.
2. Aikat B, Bhusnurmath S, Chhuttani P, et al. The pathology of noncirrhotic portal fibrosis: A review of 32 autopsy cases. *Hum Pathol*. 1979; 10: 405-18.
3. Banti G. Splenomegalie mit leberzihhose beitrage zur Pathologischen. *Anat Allgemeinea Pathol*. 1889; 24: 21-33.
4. Datta D. Non-cirrhotic portal fibrosis (idiopathic portal hypertension in India). *J Assoc Physicians India*. 1976; 24: 511.
5. Boyer JL, Sengupta KP, Biswas SK, et al. Idiopathic portal hypertension. Comparison with the portal hypertension of cirrhosis and extrahepatic portal vein obstruction. *Ann Intern Med*. 1967; 66: 41-68.
6. Mohamed Said Belhamidi, Salah Eddine Hammi, Mohamed Bouzroud, et al. Role of splenectomy in the treatment of non-cirrhotic portal hypertension: about 3 cases. *The Pan African Medical Journal*; 2017; 28: 84.
7. Krasinskas A, Eghtesad B, Kamath P. Livertransplantation for severe intrahepatic non cirrhotic portal hypertension. *Liver transpl*. 2005; 11: 627-634.
8. Walid L, Rania EG, Ilyasse Y, et al. La splénectomie dans le traitement du syndrome de Banti: A propos d'un cas. *Int J Adv Res*. 2021, 9: 270-275.
9. Faik M. le traitement chirurgical du syndrome de Banti. *Médecine du Magreb*. 1998; 67: 6-8.
10. Benkhalifa H. Splénomégalies méditerranéennes. Étude clinique de 47 cas et comparaison avec un groupe de cirrhose avec splénomégalies. *Méd Chir Diges*. 1986; 15: 227-234.
11. Gürkaynak G, Yildirim B, Aksoy F. et al. Sonographic findings in noncirrhotic portal fibrosis. *J Clin Ultrasound*. 1998; 26: 309-313.
12. Chawla Y, Dilawari J. Anorectal varices and their frequency in cirrhotic and non-cirrhotic portal hypertension. *Gut*. 1991; 32: 309-311.
13. Sarin S, Govil A, Jain A, et al. Prospective randomized trial of endoscopic sclerotherapy versus variceal band ligation for esophageal varices: influence on gastric varices and variceal recurrence. *J Hepatol*. 1994; 26: 826-32.
14. Ravenna P. Banti syndrome (fibrocongestive splenomegaly): Definition, classification and pathogenesis. *Archives of Internal Medicine*. 1940; 66: 879-879.
15. Maurus CF, Schäfer M, Müller MK, et al. Laparoscopic versus open splenectomy for nontraumatic diseases. *World Journal of Surgery*. 2008; 32: 2444-2449.