Case report

Portal cavernoma in children : rare pathology with complex management about 3 cases

Abstract

We report 03 cases of portal cavernoma hospitalized in pediatric department. Through these observations, we will try to describe the different clinical and biological aspects and discuss the diagnostic and therapeutic approaches through a literature review. Somatic examination revealed a distended abdomen and hepatosplenomegaly, presence of collateral venous circulation. The diagnostic approach to portal cavernoma in children is based on careful clinical examination, including identification of symptoms such as gastrointestinal bleeding, hepatomegaly and signs of liver disease. The biological work-up showed bicytopenia (microcytic hypochromic anaemia with thrombocytopenia) with a normal blood and liver count. The long-term course of portal cavernoma in children depends on various factors, such as the presence of complications, response to treatment and medical and surgical management. The prognosis is generally better in children who receive regular medical follow-up and appropriate treatment in the event of complications.

Keywords : Portal cavernoma, abdominal trauma, hepatosplenomegaly, bicytopenia

Introduction :

Portal cavernoma is considered to be the sequela of obstruction of the portal vein, most often due to thrombosis, and is synonymous with chronic obstruction of the portal vein. The cavernoma is a network of new “collateral” veins in the same venous network, which develops after obstruction of the portal vein (obstruction of the portal vein by a clot) to circumvent this obstruction. With regard to the specific aspects of the incidence of portal cavernoma in Morocco, it is important to note that the prevalence of this pathology in children is still poorly documented. We report 03 cases of portal cavernoma hospitalized in pediatric department. Through these observations, we will try to describe the different clinical and biological aspects and discuss the diagnostic and therapeutic approaches through a literature review. The purpose of this article is to highlight the diagnostic and therapeutic methods of

portal cavernoma in children, in order to improve the overall management of these cases, allowing a better prognosis of patients and a better quality of life.

Case présentation

FIRST CASE :

M.T, (born 03/08/2020), the first of three healthy siblings with a history of prematurity at 34 weeks' amenorrhoea, DBP= 2000g, hospitalised at birth in the neonatal intensive care unit for jaundice (no history of umbilical catheterisation), no history of previous surgery or abdominal trauma. No family history of thromboembolism or thrombophilia.

Hospitalised at the age of 08 months for haemorrhagic syndrome with abdominal distension.the physical examination found pallor, abdominal distension with hepato splenomegaly with good psychomotor development and staturo weight.a biological check-up was carried out which showed pancytopenia, normal smear, negative coombs, low TP, reduced factor 5 dosage, cytolysis, normal albumin. Medullogram with no abnormalities, Viral serologies (CMV EBV parvovirus B19 HIV) negative, Vit B12- B9 dosage = normal, Screening for left-handed disease and Niemann pick on blotting paper = negative. A radiological work-up was carried out, Chest X-ray and cardiac ultrasound were normal**,** abdominal ultrasound showed hepatosplenomegaly with portal hypertension, a Doppler complement The absence of flow in the portal vein and visualisation of a collateral venous network corresponding to the cavernoma . An aetiological investigation was carried out including a thrombophilia work-up which included: Antithrombin III, protein C, protein S normal income assay, search for factor V leiden mutation, and mutation of prothrombin returned negative homocysteine dosage which is normal and anti phospholipid antibody and anti cardiolipin antibody returned negative. Follow-up Endoscopic examination: presence of stage 3 oesophageal varices. Treatment is based on: elastic ligation of the varices after haemodynamic stabilisation. Prophylaxis of bleeding due to portal hypertension with β-blockers.

SECOND CASE:

W.H, (born 02/09/2020), second of two siblings with a history of prematurity at 36 weeks of amenorrhoea, no history of hospitalisation at birth, no history of umbilical catheterisation. Hospitalized at 15 days of age for PNA/ uropathy malformative type RVU left grade 4, vesicostomized at the age of 01 year with notion of a single functional kidney. Operated at the age of 02 years and a half for testicular ectopia.

Fortuitous discovery at the age of 02 years 09 months an ultrasound shows a tortuous dilated venous laceration in the hepatic hilum corresponding to a portal cavernoma with thrombus in the portal trunk. Clinical examination was normal, good psychomotor and statural development, no hepatosplenomagia, no signs of haemorrhage or portal hypertension. The biological work-up was unremarkable, and an aetiological investigation was carried out with

an unremarkable thrombophilia work-up. In terms of treatment, the patient was started on low molecular weight heparin, followed by a 3-month course of VKAs. A follow-up Echodoppler showed a stable portal cavernoma and no portal thrombosis.

THIRD CASE :

H. i, (born on 28/07/2020), the second of two siblings with a history of prematurity at 30 weeks' amenorrhoea and a birth weight of 1000 g. She had been in intensive care for two and a half months for extreme prematurity and had undergone umbilical catheterisation. Admitted at the age of 02 years for abdominal distension. Somatic examination revealed a distended abdomen and hepatosplenomegaly, presence of collateral venous circulation. The biological work-up showed bicytopenia (microcytic hypochromic anaemia with thrombocytopenia) with a normal blood and liver count. An abdominal echodoppler supplemented by an angioscanner showed hepatomegaly and significant splenomegaly with an increase in the calibre of the splenic vein. the portal vein is not separated and is replaced by a venous laceration corresponding to the portal cavernoma. A thrombophilia test came back normal. An oesogastroduodenal fibroscopy showed grade 3 oesophageal varices. 3 sessions of oesophageal varices ligation and beta-blocker were administered.

Discussion

Portal cavernoma is a rare vascular malformation characterised by the presence of abnormal dilations of the veins of the portal system, forming vascular shunts. These shunts can cause hypertension in the portal venous system, leading to complications such as gastrointestinal bleeding. Portal cavernoma may also be associated with hepatomegaly and an increased risk of thrombosis. In children, this condition may be asymptomatic for many years, but the risk of serious complications warrants close monitoring and appropriate management.[1]

The epidemiology of portal cavernoma in children is fairly rare, with an estimated incidence of around 1 case per 100,000 children. Boys are slightly more affected than girls, with a sex ratio of approximately 1.5/1. The disease is often diagnosed in children aged between 2 and 12 years[2]. In a retrospective study of 75 cases of portal hypertension at the university hospital in Casablanca, portal cavernoma was observed in 15 patients [3] .In Europe, the incidence of new or chronic DVT in adults is estimated at 0.7/100,000 population/year and the prevalence at 10/100,000 population. In children, the diagnosis is most often made in the chronic stage of portal cavernoma. Some forms occur in the context of malformations, after umbilical vein catheterisation or omphalitis. Approximately 50% remain idiopathic (without cause) [4].

The diagnostic approach to portal cavernoma in children is based on careful clinical examination, including identification of symptoms such as gastrointestinal bleeding, hepatomegaly and signs of liver disease. Medical history, including family history of liver disease, is also assessed. In addition, biological tests are carried out to assess abnormalities in liver function. Medical imaging, including Doppler ultrasound, MRI and CT scan, is crucial to confirm the diagnosis and assess the extent of liver damage caused by portal cavernoma.[5]

Acquired causes of portal thrombosis in children include a wide range of medical conditions such as haematological diseases including sickle cell disease, leukaemia, haemolytic anaemia

and anticoagulant protein deficiencies [6], Viral infections such as CMV or Coxsackievirus, local and systemic inflammatory causes and local infections are also common causes of portal thrombosis. Depending on the context, Behçet's disease or celiac disease may be suspected. Paroxysmal nocturnal haemoglobinuria (PNH) is less common, [4] trauma, surgery and medical treatments such as chemotherapy. These factors can lead to alterations in the blood coagulation system, favouring the formation of clots in the portal vein. Identifying and treating these underlying conditions is essential to prevent portal thrombosis in children[7].

Congenital causes of portal thrombosis in children may include genetic abnormalities such as antithrombin, protein C or protein S deficiency. These deficiencies can lead to increased coagulation and increase the risk of clot formation in the portal system. In addition, factor V Leiden mutation is also an important congenital cause of portal thrombosis in children, affecting resistance to activated protein C and increasing the risk of thrombosis[8].

Complications of portal cavernoma in children mainly include GI haemorrhage, portal hypertension and liver complications, as well as other less common complications. These complications can occur acutely or be chronic, and require appropriate medical and interventional management to avoid severe and potentially life-threatening complications[9].

The medical management of portal cavernoma in children mainly involves treating complications such as digestive haemorrhage and portal hypertension. Children with GI haemorrhage should be stabilised and receive blood transfusions if necessary. Vasoactive drugs can be used to reduce pressure in the portal system[10].

The surgical and interventional management of portal cavernoma in children is a crucial step in the treatment of this disease. The main aims of surgery are to restore normal blood flow, prevent bleeding complications and reduce pressure in the portal vein. Surgical options include transjugular portosystemic intrahepatic shunt (TIPIS), elastic ligation of oesophageal varices and splenectomy. Radiological procedures such as embolisation or sclerotherapy of varicose veins can also be used to control bleeding. The choice of procedure will depend on the extent of the lesions, the presence of complications and the child's general condition[11].

The long-term course of portal cavernoma in children depends on various factors, such as the presence of complications, response to treatment and medical and surgical management. The prognosis is generally better in children who receive regular medical follow-up and appropriate treatment in the event of complications. However, some children may develop severe liver complications, such as cirrhosis, which may affect their long-term prognosis. Long-term studies are needed to assess the course of the disease in children with portal cavernoma and to identify the factors influencing prognosis more accurately[12].

Conclusion

Portal cavernoma is a major cause of portal hypertension in paediatrics. It is diagnosed by imaging and requires a high level of multidisciplinary management by paediatricians, radiologists and paediatric surgeons, enabling an early and appropriate decision to be taken, which can only be achieved by mastery of the various therapeutic tools. The prognosis depends on the quality of this management, which is often a problem in developing countries.

Ethical Approval:

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

Consent

As per international standards, parental written consent has been collected and preserved by the author(s).

Disclaimer (Artificial intelligence)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

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