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(CASE REPORT)

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Congenital Portosystemic Hepatic Fistula: A case report

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Abstract

Congenital portosystemic fistula (CPF) is a congenital malformation defined by one or more communications between a hepatic portal venous system and a systemic venous cava. Other terminologies exist in the literature, in particular for extrahepatic communications, such as "Abernethy malformation". It may be associated with other malformations or syndromes. Because of this non-physiological vascular communication, PFC can lead to the development of extrahepatic complications: hepatic encephalopathy and pulmonary vascular diseases (hepato-pulmonary syndrome or pulmonary hypertension). It can also lead to hepatic complications with the appearance of benign or malignant tumors and biological and metabolic disorders.

We report the case of a newborn with a portosystemic hepatic fistula discovered during neonatal jaundice with cutaneous angiomatosis.

Keywords: Fistula; Congenital; Portosystemic; Abernethy; Neonatal; Hepatic

1. Introduction

Portosystemic hepatic fistula (PHF) Abnormal communication between the portal venous system and the vena cava system. It is secondary to a defect of evolution or involution of the vitelline veins during embryogenesis. It was described for the first time in 1793 by John Abernethy. Its prevalence is estimated at 1/30,000 births [1,2]. FPS can be isolated without any clinical repercussions and closes spontaneously. and it can be associated with other malformations (cardiac; skeletal or genetic) [3].

2. Observation

This is a female neonate at d21 of life; from a so-called attended pregnancy; carried to term without incident. The delivery was by vaginal route; Apgar not specified, with notion of immediate crying; birth weight: 3kg; exclusively breastfed; mother aged 20 years, G1P1, without particular pathological history; infectious anamnesis was negative. No notion of consanguinity. Admitted for neonatal jaundice with diffuse neonatal hemangiomatosis. The history of the disease goes back to D3 of life with the installation of multiple bright red cutaneous lesions of variable size on all the body and increasing progressively in size associated with a cutaneous-mucosal icterus with normocoloured stools and urines; without palmar-plantar involvement; evolving in a context of apyrexia. The clinical examination showed a eutrophic, subicteric newborn on a pink background with the presence of several bright red maculo-papular cutaneous hemangiomas (about 30) ranging from the size of a pinhead to 1.5 cm distributed over the whole body; without signs of dehydration or malnutrition. The somatic examination was unremarkable, especially the cardiovascular and abdominal examinations; no clinically detectable malformations. The biological assessment showed a mixed hyperbilirubinemia with hepatic cytolysis and hyperammonia. The abdominal echo-Dopller showed a liver of normal size, with regular

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contours and multiple hypodense nodular lesions, fairly well limited, with regular contours, enhanced after injection of the contrast medium, the most voluminous ones located and measuring: segment II: 9.6x 9.3 mm and 7x10 mm; segment I: 8x6 mm with individualization of a communication between the right portal bronchus and the right suprahepatic vein. The portal trunk is of normal caliber measuring 6 mm in diameter. The diagnosis was made by an abdominal angioscanner which showed an intrahepatic portosystemic fistula associated with hepatic nodules to be characterized.

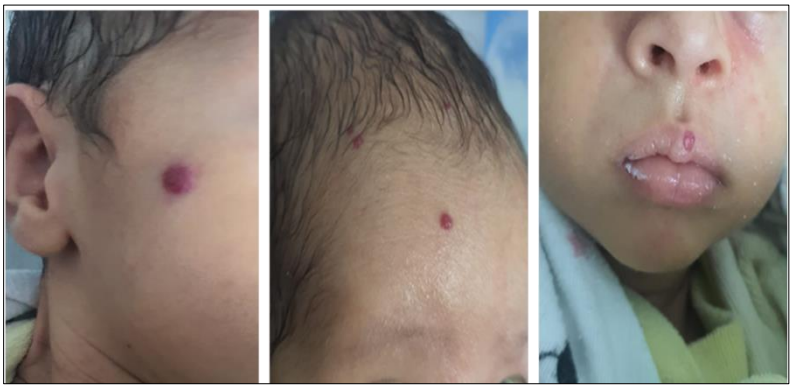

Figure 1 Multiple cutaneous angiomatosis associated with portosystemic liver fistula in our patient

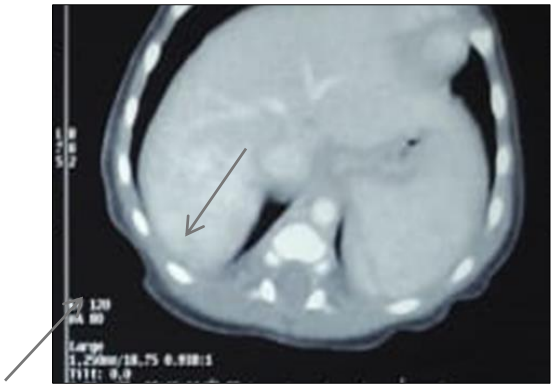

Figure 2 Intrahepatic portosystemic fistula associated with hepatic nodules

3. Discussion

Portosystemic hepatic fistula (PHF) is a developmental anomaly of the hepatic vasculature during embryogenesis. The hepatic outline emerges at the 4th SA. It is related to 3 major venous systems of the fetal circulation: vitelline veins; umbilical veins and cardinal veins. All 3 converge towards the canal of Aranthius (sinus venosus). At the end of the 6th SA; resulting from 4 intervitelline anastomoses which will form the portal vein. The canal of Aranthius connects the portal sinus to the inferior vena cava (IVC).

At birth, the umbilical vein and the duct of Aranthius close. The portal vein becomes the only afferent vein of the liver. Incomplete involution of one or more of these primordial vessels may result in abnormal communications between the portal system and the inferior vena cava in the form of a hepatic portal-systemic fistula.

Many classifications exist for PFCs and are either anatomical or surgical [6-7]. Historically, PFCs were first classified as extrahepatic (Abernethy malformation) and intrahepatic. In recent years, new classifications have been proposed to provide the necessary information suitable for optimal therapeutic management. The Bicetre surgical classification describes four types of PCF taking into account the cave termination of the shunt [8]. Later, Kanazawa proposed to expand this classification by adding a description of intrahepatic portal veins during balloon occlusion. Three subtypes were proposed: mild, moderate, and severe [6]. A combination of the Bicetre and Kanazawa classifications may be useful in selecting therapeutic approaches and management to improve patient outcomes [4].

A complete clinico-biological and radiological evaluation is necessary when congenital portosystemic fistula is suspected.

Discovery may be incidental antenatally during an antenatal ultrasound scan showing the fistula or its indirect signs. These include enlargement of the umbilical vein and/or inferior vena cava, and the absence of an abnormally large ductus venosus is also an indirect sign of CPF. Postnatally and at any age, it can be discovered during an abdominal ultrasound for another indication; during an abnormal neonatal screening for galactosemia; high concentrations of galactose can be present in the plasma of newborns without an enzymatic deficiency of galactose metabolism [5]; during the etiological diagnosis of a complication; or during a workup for an associated malformation.

The mode of diagnosis is variable according to age. Multiple cutaneous angiomas are present in 5% of cases. There is no hepatomegaly, but splenomegaly may be present even in the absence of portal hypertension [4]. Signs and symptoms in the neonatal period include jaundice and heart failure, conjugated hyperbilirubinemia, hypoglycemia, abnormal neonatal galactosemia screening, thrombocytopenia, hyperammonemia and coagulation abnormalities [6]. Later in life, complications such as liver nodules, hepatic encephalopathy; hepatopulmonary syndrome or pulmonary hypertension may reveal CPE. Similarly, CPE may be diagnosed and/or should be sought during the evaluation of malformations known to be associated with CPE, including: congenital heart disease, heterotaxy, polysplenia syndrome, chromosomal abnormalities, cutaneous or hepatic hemangiomas [9].

Systemic complications or signs/symptoms are estimated to occur in 30% of cases and the risk of developing them increases with age. In addition to the in-utero complications described above, complications of this non-physiologic communication between the portal system and the systemic system in the postnatal period may include chronic hepatic encephalopathy (CHE); cardiac complications: pulmonary arterial hypertension (PAH). Pulmonary: hepato-pulmonary syndrome (HPS). Hepatic nodules: the development of benign and regenerative hepatic lesions is well known and seems to be related to portal blood deprivation [6]. Focal nodular hyperplasia (FNH) lesions are the most common, but other benign and malignant tumors are common, including regenerative nodular hyperplasia (RNH), hepatocellular adenoma (HCA), hepatoblastoma (child), and hepatocellular carcinoma (child and adult) (HCC). [10]. The diagnosis of PFC could be made in the context of exploration of hepatic nodules. Metabolic complications: metabolic abnormalities associated with CPE include neonatal hypoglycemia, neonatal cholestasis, abnormal neonatal screening for galactosemia, hyperammonemia, unexplained coagulation disorders, increased serum bile acids, and rather accelerated growth disturbances in older children.

A complete biological and radiological evaluation is required in case of suspected PCF. The biological workup to be performed includes a hemostasis workup with a CBC and platelet count; serum bile acids, which are the witness of the porto-systemic passage of bile acids from intestinal absorption. Their measurement is an element of follow-up. Fasting and postprandial ammonia to look for arguments in favor of a hepatic encephalopathy, in fact hyper ammonia is both a witness and a monitoring element of PFC. Alpha-fetoprotein in case of hepatic nodules on imaging, screening for hepatocellular carcinoma. Glycemic cycle with glucose dextrometry For the assessment of neonatal hypoglycemia and hypergalactosemia [4-6-10].

There are several modalities for both diagnosing PCF and planning management: Hepatic and abdominal Doppler ultrasound is the key diagnostic test. It shows the presence of one or more abnormal communications between the portal system and the vena cava system without signs of portal hypertension. There is usually hypoplasia or even absence of visualization of the portal network downstream of the PCF. The examination will specify the location of the fistula, the size of the communication, the appearance of the downstream portal system, the appearance of the liver and in particular the presence of nodules. Abdominal ultrasound should be used to search for the presence of other vascular and/or visceral malformations [11]. Abdominal angioscanner with injection of contrast medium to specify and confirm the PCF, its location, its anatomical type. It also allows the detection of hepatic nodules. Hepatic magnetic resonance imaging (MRI) without and possibly with injection of contrast product to look for hepatic nodules. They will allow to specify the vascular anatomy and to search for other associated malformations. In MRI, the injection of contrast medium will only be performed in the presence of a nodule to try to characterize it [6]. Angiography with occlusion test and pressure measurement is performed either when the intrahepatic portal system is not visible on ultrasound or CT and/or MRI, or for therapeutic purposes to close the shunt. When the portal branches are not visible after opacification of the shunt, a balloon occlusion test of the shunt is performed. A portal pressure measurement is performed before and after balloon occlusion of the shunt to assess the tolerance of the shunt closure [7]. In case of hepatic nodules: a biopsy of the nodule and the healthy liver is necessary, with molecular biology typing of the hepatic nodule(s) and search for the beta-catenin mutation in the biopsied nodule [11-7]. Brain MRI and spectro-MRI look for signs of portosystemic encephalopathy, such as hypersignal of the lenticular nuclei on T1-weighted sequences; or associated cerebral vascular anomaly [6].

At the end of this assessment, the patient is classified as symptomatic or asymptomatic, and the anatomical shape of the fistula is specified: starting point, end point, lateral and terminal (intrahepatic network visible or not during the occlusion test).

Once the shunt has been identified and confirmed, the objective is to evaluate the risk-benefit balance of closing the shunt. Closure of the shunt must be systematically discussed. There are two situations: closure in symptomatic patients and preventive closure.

Therapeutic management consists of treating the symptoms and closing the fistula. In case of significant and/or symptomatic hyperammonemia: A low protein diet and/or hypoammonemic treatment (sodium benzoate, phenylbutyrate). In case of pulmonary arterial hypertension confirmed by right heart catheterization, initiation of targeted treatment of pulmonary arterial hypertension (endothelin receptor antagonists and/or phosphodiesterase 5 inhibitor and/or prostacyclin analogues) and monitoring of this treatment and response to drug therapy. In case of hepato-pulmonary syndrome with PaO2 < 60 mm Hg, nasal oxygen therapy is instituted [12].

PCFs can be closed in two ways: by endovascular interventional radiology or by surgery. The benefit of prophylactic PCF closure in the absence of subclinical or clinical signs or symptoms has not been demonstrated. Interventional radiology treatment is preferred when possible because of its lower morbidity and cost. Interventional radiology treatment consists of occluding the abnormal communication(s) with endovascular devices [13]. Surgical treatment is reserved for fistulas not accessible to radiological treatment.

Indeed, closure of the PFC results in: Restoration of intrahepatic portal flow; Regression or even disappearance of most hepatic nodules; Regression of the hepato-pulmonary syndrome; The effect of fistula closure on the evolution of PAH is less predictable but may lead to its stabilization [14]. Liver transplantation has become exceptional in case of failure of shunt closure or in case of associated liver pathology (intrahepatic fibrosis), malignant nodules.

Postoperative prophylactic anticoagulation is essential to avoid postoperative portal or mesenteric thrombosis. Post-operative monitoring by ultrasound-doppler: Checks the occlusion of the SPF and looks for a complication (vascular thrombosis, sign of portal hypertension). The follow-up workup includes the clinical elements mentioned above in the workup and biological elements: Complete hepatic workup, hemostasis, bile acids, fasting ammonia, alphafoetoprotein, hepatic ultrasound-doppler/ cerebral hepatic MRI if initially abnormal. Follow-up is performed at D21 after closure, 3 and 6 months, then one year or annually depending on the presence and follow-up of complications. If normal, liver and brain MRI should be done only once in the follow-up [6].

4. Conclusion

Portosystemic fistulas are rare but their incidence is increasing with the improvement of imaging techniques because they are better recognized. They remain exceptional and must also be specifically managed because they expose to possibly severe portal hypertension. Ante-natal diagnosis with appropriate follow-up can certainly limit the complications caused by SPF.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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