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Case Report

Giant Mesenchymal Hamartoma in Pediatric Patients

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Abstract

Mesenchymal hamartoma of the liver is a benign tumor with good prognosis that usually manifests in childhood before the age of two. Abdominal distension and the presence of a hepatic mass are the most common symptoms; anorexia, vomiting and signs of compression have been reported. The diagnosis is based on radiological data but above all on the results of the pathology. Only surgical resection can prevent recurrences and in some cases, liver transplantation is indicated. We report the case of a 16-month-old girl who was referred to as for surgical assessment of cystic mass in the right liver, with a respiratory recurrent symptomology. Imagery (CT scan MRI) has showed a large hepatic mass which compresses the hepatic pedicle and the vena cava. We performed a right hepatectomy after clamping the hepatic pedicle and the inferior vena cava. The postoperative follow-up were simple and disappearance of the preoperative symptoms.

Keywords: Mesenchymal hamartoma, Infant, Right Hepatectomy

Case Report

A 16 month old infant was referred to us for a surgical management of a large abdominal mass. The parents revealed that this infant was born from an uneventful pregnancy, weighing 3 kg at birth, breastfed up to the 4th month. The newborn was under inhaled treatment for bronchial asthma. At the age of 12 months and when hospitalized in a pediatric ward for an unexplained fever and abdominal distension, the ultrasound showed a heterogeneous cystic mass of the liver. The blood tests revealed anemia, hemoglobin electrophoresis showed heterozygous sickle cell anemia, hemoglobin S level at 38%. Clinical examination on admission, found a eutrophic infant, body weight and height were 10kg and 75cm. The patient had slight skin-mucous pallor, significant abdominal distension with, collateral circulation, palpable mass reaching the right flank and crossing the middle line, no splenomegaly, and no jaundice or haemorrhagic syndrome.

The initial blood tests was normal: White blood cells: 12,000, Hemoglobin: 9.4g / dl, Haematocrit: 28.7%, Platelets: 42000UL: aspartate transaminase 52U / L; alanine transaminase: 30U / L, GGT: 49U / L // Total Bilirubin 0.35 mg / dl, direct bilirubin 0.15mg / dl). Abdominal CT scan showed a heterogeneous mass of the liver (segments V, VI, VII and VIII). Multiple partitions enhanced by the contrast agent, oval with regular contours and sharp limits measuring 95X66X100 mm, compression the hepatic veins, the portal vein and the right kidney. MRI showed a large multilocular cystic mass involving the right liver, without signs of vascular invasion (Figure 1). Doubtful hydatid serology is an absolute contraindication for a biopsy puncture. In the presence of a large cystic mass of the right liver, compressing the hepatic pedicle, and the inferior vena cava, the strategy was to perform a right hepatectomy.

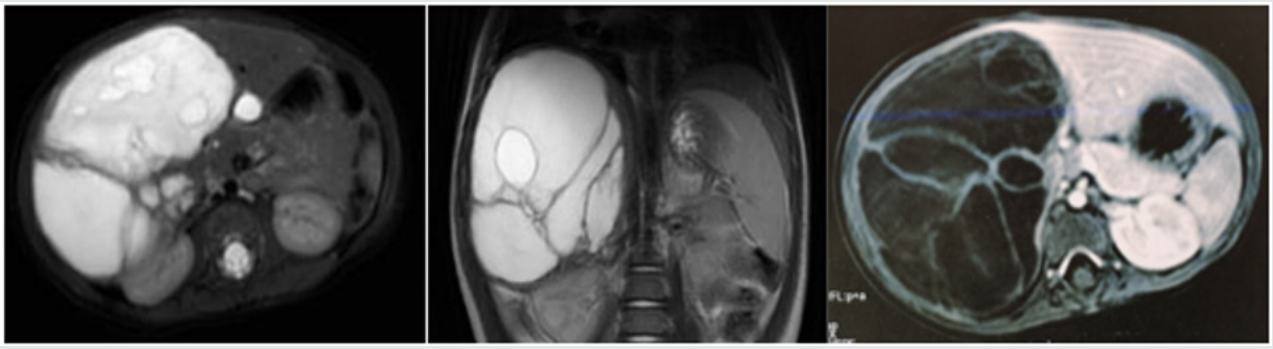

Figure 1: A. Abdominal CT scan showed a heterogeneous mass of the liver (segments V, VI, VII and VIII). Multiple partitions enhanced by the contrast agent, oval with regular contours and sharp limits measuring 95X66X100 mm, compression the hepatic veins, the portal vein and the right kidney.
B. MRI showed a large multilocular cystic mass involving the right liver, without signs of vascular invasion.

After the Pringle maneuver, we have done the following: approach to the inferior vena cava below the liver, individualization of the bile ducts, clamping of the hepatic pedicle and inferior vena cava (total clamping time 08 minutes). A right hepatectomy was performed after ligation of the artery right hepatic, right portal vein, right bile duct, and right hepatic vein. (Figure2) The duration of the surgery was 45 minutes; the loss of blood was estimated at 30ml, no transfusion, wall closure without drainage. The post-operative in the hepatobiliary unit was simple; the patient was released after 72 hours. The hepatic enzymes were high in the immediate post-operative phase and became stable in two weeks. The pathology confirmed the diagnosis of mesenchymal hamartoma (Figures 2B & 3). The last checkup was carried out at 09 months and the evolution of the patient was favorable. Respiratory signs disappeared, and there was a significant improvement in the patient's behavior who has become more active to the satisfaction of her parents.

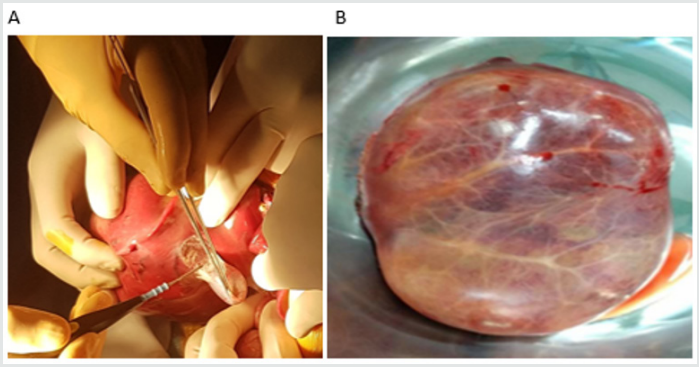

Figure 2A: Operative Procedure: Right Hepatectomy.
Figure 2B: Hepatic Tumors 16x11x7cm Giant Pseudocystic Mesenchymal Tumors (14x10 cm).

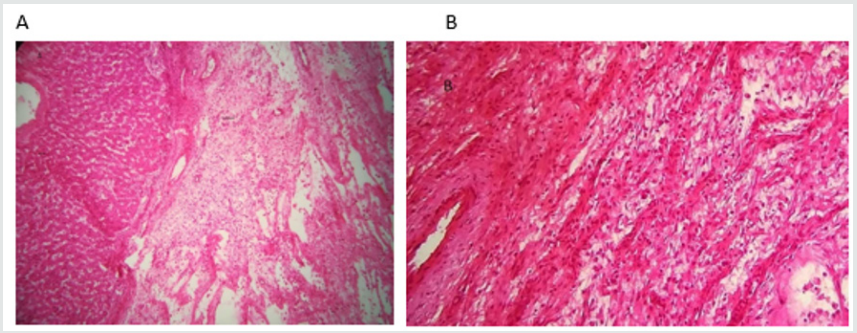

Figure 3:

A. Hepatic parenchyma sits from a well-separated tumor formation by a fibrous pseudo-capsule.

B. HM: Low-atypical fusiform cell ranges settle on a myxoid background punctuated by mononucleous inflammatory elements and congestive blood vessels. The hepatocytic epithelial contingent is minimal.

Discussion

Mesenchymal hamartomas (HM; also known as pseudocystic mesenchymal tumor, giant cell lymphangioma, bile cell fibroadenoma, and cavernous lymphagiomatoid tumor) of the liver represent the second common benign tumor in children, especially in girls, and is often discovered during the first two years [1,2]. The etiology of MH has not yet been determined. One theory suggests a developmental abnormality that stems from a malformation of the ductal plate, while another theory that is gaining ground, proposes that MH is a true neoplasm. Recent cytogenetic studies have shown balanced translocations involving chromosomes 11, 13, 15 and 19; flow cytometry studies have revealed aneuploidy [3-5]. MH are often asymptomatic, discovered during a routine radiological examination or for other pathology in the form of a cystic mass. Abdominal distension and the presence of hepatic mass are the most common symptoms [6,7]; cases of anorexia, vomiting and signs of compression such as jaundice and even in some cases compression of the vena cava have been reported [2]. Sometimes the mass will quickly increase in volume due to the rapid accumulation of fluid in the cystic spaces. Rare but often serious respiratory complications are responsible for asthma attacks and respiratory distress. About 75% of MH in the liver develops in the right lobe. Liver function and Alpha Foetoprotein (AFP) are only useful for excluding other diseases such as hepatoblastom [6-9].

The imagery highlights a large, well-circumscribed mass that may contain cysts of different sizes. Ultrasound and computed tomography show a multicystic mass with septations as in the case of our patient [10-14]. It is often question of very large liver lesions either from the inside or from the growth of the normal parenchyma, in most cases associating several cysts of 4 to 7 cm each, containing clear to yellow liquid and solid areas , consisting of loose myxoid stroma containing irregularly dilated ducts bordered by cuboid epithelia.

Often there are irregular islets separated from hepatocytes. The cysts are surrounded by a layer of loose mesenchyme with no identifiable epithelial lining and appear to be linked to the mesenchyme itself [15]. The myxomatous stroma contains doubled epithelial and non-epithelial cysts [1]. If there is any doubt about the possibility of a malignant tumor, a biopsy can be performed, as such, a diagnosis can lead to a change in the management of larger tumors which might otherwise be difficult to resect [9,10].

The practice of observation alone while awaiting spontaneous regression should be discouraged in view of reports of malignant transformation into undifferentiated embryonic sarcoma [15-18]. The risk of malignant transformation into indifferent sarcoma implies a systematic surgical resection [15,16]. Only surgical resection can prevent recurrences. The treatment consists of complete surgical resection with clear margins which may require a lobectomy or a non-anatomical resection [6,7]. If the tumor is considered unrespectable, some authors have recommended nucleation or marsupialization [7,19,20]. The insufficiency of the remaining liver is considered to be the main reason for the failure of the surgery [19]. Some authors have demonstrated the feasibility of ALPPS in infants with a huge hepatic tumor, especially mesenchymal hepatic hamartoma, due to the rapid regeneration of the future remaining liver [21]. Others believe that liver transplantation is a safe surgical option for pediatric patients with unresectable giant mesenchymal hamartoma, who generally have no other therapeutic alternative [22].

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