

Mesenchymal Hamartoma of the Liver Associated with Discontinuity of the Vena Cava

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ABSTRACT

Primary liver tumors in children are infrequent, accounting for 0.5-2 % of all pediatric neoplasms. About two-thirds are malignant. The most common pediatric malignant liver tumours are, in order of frequency, hepatoblastoma, hepatocellular carcinoma, and embryonic sarcoma of the liver. Among benign tumors, the most frequent are hemangioendothelioma and mesenchymal hamartoma of the liver, and more rarely focal nodular hyperplasia and hepatocellular adenoma [1,2].

We report the case of an infant who was entrusted to us for the management of a large liver tumor. Radiology (CT scan and MRI), in addition to the elements in favor of a mesenchymal hamartoma of the right liver revealed a congenital anomaly associated, in this case, with the absence of the inferior vena cava in its intraabdominal portion. A right hepatectomy enlarged to segment IV was performed.

The right renal vein drained into the right suprahepatic vein was preserved. Note that this tumor represents 50% of the total weight of the patient, which explains the difficulties and constraints that hindered the development and quality of life of the young patient.

Keywords

Pediatric neoplasm, Mesenchymal Hamartoma, Discontinuity of Vena cava.

Introduction

Hepatic mesenchymal hamartoma is an abnormal hamartomatous growth of mesenchymal tissue, bile ducts and blood vessels in the liver. It is the second common benign neoplasm next to hemangioma, and it is the second most common benign liver tumor in children, accounting for about 5–8% of pediatric liver tumors [1], with 80% of these tumors presenting within the first two years of life.

Recurrences after conservative surgery are frequent and malignant transformation of the mesenchymal hamartoma of the liver remains possible. Cases of degeneration have been reported. These elements are in favor of radical surgery. Essentially a regulated hepatectomy is often indicated. Some cases of liver transplantation are also reported in the literature [1,2].

Observation

The patient is a 3-year-old infant from a full-term pregnancy, well monitored, delivered vaginally without neonatal incident who had a progressive increase in abdominal volume noted by the parents

since the age of 11 months. The physical examination objectified a soft mass at the expense of the right hypochondrium without collateral circulation in front, the lymph node areas were free, weight 18 kg, the blood test was normal apart from a high alpha-feto-protein level (6xN).

Abdominal-pelvic ultrasound showed the following: a voluminous multilocular hepatic mass with exohepatic development exerting a mass effect on the left and median suprahepatic veins which are laminated and permeable; right suprahepatic vein not visualized. Computed tomography (CT) revealed a bulky multi-septate cystic mass with exohepatic development, syndrome of discontinuity of the vena cava inferior to the height of the renal veins, with recovery by lumbar parietal collateral veins. Abdominal MRI objectified a voluminous cystic mass of segments (IV, V, VI, VII, VIII) suggestive of cystic mesenchymal hamartoma (Figure 1).

A right hepatectomy extended to segment IV carrying the mass was performed. The absence of the intra-abdominal vena cava was confirmed intraoperatively, the right renal vein drained in hepatic addition was respected during hepatectomy (Figure 2).

At the last control, 24 months after surgery, the little girl was in good health and has resumed a normal development, normal liver function. MRI did not objectify abnormalities, and the venous drainage was satisfactory. Macroscopically, the surgical specimen measured 35 cm long axis and weighed 09 kg (50% of the patient's total weight).

Histology: a well-limited tumor mass of 39X23X19cm, with encapsulated smooth surface, of renitent consistency, of grayish color. At the cut, we noticed a discharge of a greenish liquid, from a multilocular cavity, with smooth, whitish inner surface, with solidocystic areas dissociated by fibrous and hemorrhagic rearrangements. Microscopic examination revealed a benign cystic proliferation of anarchic architecture in abnormal quality, made of bile structures of variable shapes sometimes stretched with collar lumen, sometimes dilated and cystic, lined by a cubic epithelium, regular, without nuclear atypia. We noted the presence of clusters and spans of hepatocytes of normal morphology dissociated by fibrosis. The vascular component was also hyperplastic made of dilated and congestive vessels and capillaries. Additionally, there was a presence by sector of cystic cavities lined by a regular cubic epithelium sometimes pseudostratified surmounting a conjunctival

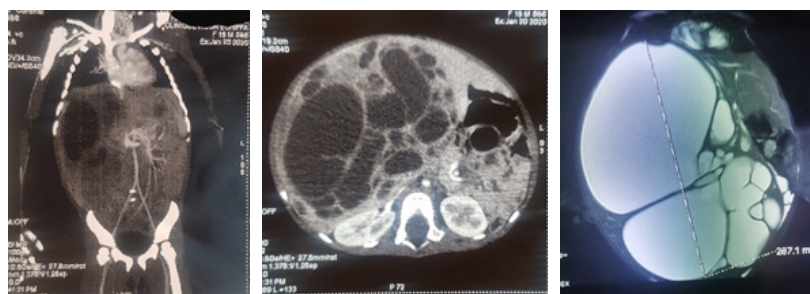


Figure 1: CT-Scan and MRI

Multi-septate cystic mass
Discontinuity of the vena cava inferior,
Lumbar parietal collateral veins.



Figure 2: Intraoperative aspects

Extended right hepatectomy

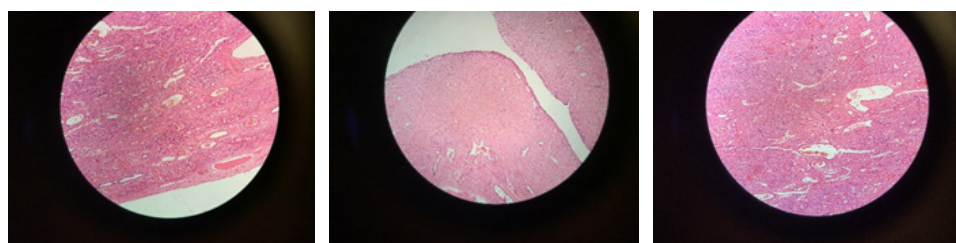


Figure 3: Histology

Cystic wall lined by a regular cubic epithelium overcoming a conjunctivo-vasculare tissue.

vascular tissue punctuated by mononuclear inflammatory elements (Figure 3).

Discussion

The mesenchymal hamartoma of the liver was definitively described in 1956 by Edmonson. This pathology usually occurs in pediatric patients with a median onset time of ten months and ranking second to infantile hemangioma, although a few cases have been reported in adults [2].

Histologically, the condition shows a stroma of fibromyxoid tissue with wavy cells and cystic spaces in the middle of the bile ducts and normal hepatocytes. Differential diagnosis includes other liver masses and infectious cystic lesions. Laboratory tests are usually normal, but in some cases, high levels of alpha-feto-protein are associated with poorly organized hepatocytes. This can be a source of confusion at the time of diagnosis, suspecting malignant liver masses such as hepatoblastoma.

The diagnosis may be suspected on clinical or radiological examination, but usually hepatic hamartoma is revealed by abdominal distension or mass. Other signs that have been reported include abdominal pain, anorexia, vomiting, and being overweight [3].

Calcifications can be objectified in the liver area on abdominal X-ray without preparation. Ultrasound, computed tomography, and MRI play a key role in preoperative diagnosis by showing a well-circumscribed multicystic tumor in the liver parenchyma. Cysts are often partitioned and rarely contain debris. On ultrasound, the presence of fine motile septa or parietal nodules is observed, the presence of hyperechoic cysts in the cyst is strongly suggestive of hepatic hamartoma [3]. Sometimes 1 or 2 large cysts predominate, as in our observation. In other cases, the cysts are very small, and the tumor appears as a solid echogenous mass [4,5]. Most of these tumors appear hypodense and hypovascularized on imaging. Solid nodules, septa and peripheral areas may show contrast after injection. Occasionally, large portal venous branches may cover the tumour, which can lead to confusion with a hemangioendotheloma [5].

In most cases, the diagnosis is made on imaging. However, when in doubt, a percutaneous or surgical biopsy may be suggested. Cytopuncture is of little use, since it rarely establishes a definitive diagnosis and does not formally exclude hepatoblastoma or malignant mesenchymal tumor. Certainty diagnosis is histological

and is usually easy due to the microscopic characteristics typical of the tumor [6].

The right lobe of the liver is implicated in 75% of cases [7]. These tumors are generally well circumscribed without an individualizable capsule and are surrounded by an irregular margin of compressed hepatic parenchyma. They have a dual solid and cystic component. They can be very large, sometimes reaching 20 to 30 cm in diameter and weighing up to 3 kg [8], like the case of our patient (09 kg). On the section, there are several cysts containing a gelatinous serous fluid, but never bile. Cysts do not come in direct contact with the bile ducts. They are separated by fibrous septa and surrounded by loose mesenchymal tissue containing tortuous bile ducts, blood vessels and hepatocyte islets [9].

Liver hamartoma should be differentiated from other liver tumours such as hemangioma, sarcoma, congenital and parasitic cysts [10] and rare cystic forms of some liver tumours (cystic hepatoblastoma, biliary cystadenoma, teratoma). An accurate diagnosis is essential. Some cases have been treated inappropriately for suspected hepatoblastoma [3] or hydatid cyst [11].

As a rule, hepatic hamartoma is not associated with other congenital anomalies. However, associations have been noted: congenital heart disease; poor intestinal rotation; esophageal atresia with or without annular pancreas; biliary atresia, myelomeningocele [8,11,12]. The absence of the infrarenal segment of the inferior vena cava associated with preservation of the suprarenal segment is a very rare association. In our patient, the absence of the inferior vena cava (VCI) discovered by the CT scan and MRI, did not influence the surgical procedure, the right renal vein was preserved.

The pathogenesis of hepatic hamartoma is still a matter of controversy. Some authors believe that this is a developmental anomaly due to excessive uncoordinated proliferation of the primitive mesenchyme. [13]. others assume that it is a true neoplastic process. Indeed, cytogenetic abnormalities have been found in some cases of hepatic hamartoma such as chromosomal translocations involving a rupture on the long arm of chromosome 19 (band 19Q13.4) [9], a region which involved in oncogenesis. Prolonged cell survival may be responsible for additional genetic mutations that explain the possibility of malignant degeneration of hepatic hamartoma or its association with certain malignancies, including undifferentiated embryonic sarcoma with poor prognosis [14,15].

In the past, non-radical resection of the tumour had been proposed as an acceptable management, but it is now clear that mesenchymal hamartomas share a common genetic discovery with undifferentiated embryonic sarcoma of the liver, breaking point 191q13.4. In addition, recent reports suggest, if possible, a radical surgical resection of the tumor since recurrence. Malignant transformation has been documented [9].

Surgical removal of the tumor is the treatment of choice. Progressive abdominal distension after tumor development can be fatal for untreated children. Complete resection of the lesion can sometimes be difficult, if not impossible, due to the involvement of vital structures [16].

Intra- and postoperative complications are rare. Fatal bleeding is exceptional [17], and bile complications are unusual and curable [18]. After complete resection, clinical and ultrasound monitoring is recommended for at least 5 years [19], but an excellent prognosis can be predicted from the outset. Several non-surgical therapeutic means have been reported (percutaneous drainage, chemotherapy, radiotherapy) [7].

However, these are only isolated cases and there are, so far, no rigorous studies proving their effectiveness. Some authors point out that these tumors can progress to spontaneous regression and therefore suggest that asymptomatic forms should not be treated [20]. However, the risk of malignant degeneration warrants a complete resection of the tumor [19].

Currently, the gold standard for the treatment of mesenchymal hamartoma is a complete surgical resection of the tumor to prevent local recurrence and long-term malignant transformation. However, cases of complex surgical management have been reported in the case of giant tumours [21]. In a series of 17 cases, Karpelowsky et al. mention intraoperative problems ranging from intraoperative death, bile duct injury, and accelerated recurrence after incomplete tumor resection that also died in surgery [22]. Liver transplantation is controversial in this condition, but in giant tumors, it is a valid therapeutic option. So far, three pediatric liver transplants have been reported for a non-breakable giant mesenchymal hamartoma following incomplete resection, Tepetes et al. [23]. Two cases in have been reported in adult patients with progressive hepatic failure and anterior partial hepatectomy, which had a satisfactory outcome.

In some cases, due to tumor expansion, various complications may result such as ascites, jaundice or congestive heart failure, engorged veins on the anterior abdominal wall and edema of the lower limbs and/or respiratory distress.

Conclusion

The mesenchymal hamartoma is a rare benign tumor in children. Its pathogenesis remains a matter of controversy. The diagnosis of certainty is histological and is usually easy to make from the typical microscopic features of the tumor. The treatment of choice is surgical resection and the prognosis is generally good.

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