Case Reports

Congenital Mesenchymal
Hamartoma of Liver

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Mesenchymal hamartoma of the liver is a rare tumor like lesion of childhood(1,2). It is most common in children under five years of age and rarely manifests at birth (2). The tumor is easily resectable and has an excellent prognosis (1-3). Fatal hemorrhage in mesenchymal hamartoma is unknown though marked vascularity of the tumor may be responsible for the hemorrhage. Herein we report a case of mesenchymal hamartoma occurring in a newborn child which was complicated by hemorrhage resulting in death.

Case Report

A 34 weeks gestation, male child weighing 1950 g was born to a 20 years old primigravida by assisted delivery following premature labor after an unsupervised but uneventful pregnancy. He was asphyxiated and had a low Apgar score requiring ventilatory support. His abdomen was markedly distended by a soft tissue mass which was occupying almost the whole of the flank. Abdominal ultrasonography revealed a large cystic tumor rising from the right lobe of the liver with evidence of hemorrhage in the tumor. Within few hours after birth the child was noted to be pale with features of peripheral circulatory failure and a low hematocrit of 35%. Despite intravenous dextrose and fresh blood transfusion, the child had a cardio-respiratory arrest and died after 22 hours of birth. A complete autopsy was performed.

Pathologic Findings

The peritoneal cavity contained 25 ml of blood clots localized to the right hypochondrium and the right lumbar regions. The right lobe of the liver showed a large tumor mass measuring 5x4x2 cm. There was no impingement by this tumor on any of the large vessels or hilar structures. The outer surface was smooth and did not show any tear or rupture. Serial slicing through the tumor revealed an un-encapsulated but well circumscribed cystic lesion containing hemorrhagic fluid. The wall was finely trabeculated and showed small cystic areas with foci of hemorrhages.

Multiple sections were studied from the tumor which revealed variable morphology with a predominance of loose mesenchymal tissue containing numerous vascular channels lined by a single layer of endothelium resembling a cavernous hemangioma (Fig. 1). Many of the vascular channels contained fibrin thrombi while others were packed with red blood cells. Lymphatic channels were also noted, these were smaller in size containing pale eosinophilic fluffy fluid. Other tissue elements noted in the tumor were numerous ductal structures, islands of hepatocytes and foci of extramedullary hematopoises. The cysts lacked lining epithelium and their cavities were filled with blood. Foci of necrosis, calcification

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and hemorrhage were also noted within the loose mesenchyme. A diagnosis of congenital mesenchymal hamartoma of liver was made. The remaining liver parenchyma was unremarkable. A patch of sub-arachnoid hemorrhage was noted over the cerebellum. The other organs were grossly and microscopically normal.

Discussion

Mesenchymal hamartomas are rare tumor like lesions of the liver(1,2). They represent developmental anomalies rather than true tumor(1-3). More recently it has been suggested that the lesion is reactive and may result from an anomalous blood supply which leads to ischemia and cystic change(4). They manifest clinically in the first five years of life(1-3). Occasional cases have been reported in the newborns(2-5). Clinical manifestations are characterized by abdominal distension and a palpable abdominal mass in an otherwise asymptomatic infant(1-3). Morphologically the tumors are cystic with a predominance of mature mesenchymal tissue interspersed with bile ducts, islands of hepatocytes and lymphatic channels suggesting the possibility that they originate from the connective tissue of the portal tract(3). Radiographic scanning and histologic studies have rarely revealed any significant or abnormal vascularity in this tumor(1-3). One of the eight cases of mesenchymal hamartomas described earlier(5) showed marked vascularity and angiographic embolization facilitated the surgical resection of the tumor.

The tumor in our case was richly vascular with numerous ectatic channels embedded in a background of loosely cellular mesenchyme. In many areas the lesion resembled a cavernous hemangioma and many of these vessels contained fresh thrombi. Bile ductal structures and islands of hepatocytes were interspersed in between them. Hemorrhage was grossly evident within the cystic cavities in the more solid areas and outside the tumor in the peritoneal cavity. The development of pallor and peripheral circulatory failure within few hours of birth and a low hematocrit in the absence of any evidence of bleeding from other organs or major vessels suggests that the hemorrhage in the tumor may have been responsible for the above features. The hemorrhage was also noted on ultrasonography. At autopsy there was no tear or rupture of the capsule. However, the fact that the tumor was large and richly vascular comprising of thin walled, poorly supported vessels and also that trauma caused by a difficult delivery may have been responsible for rupture of the vessels with subsequent intratumoral and intraperitoneal hemorrhage. Furthermore, in a premature neonate with birth asphyxia, hypoxia may lead to vascular endothelial damage making these vessels more prone to rupture and damage. The large mass compressing the lungs could have significantly contributed to the respiratory distress in the child.

A case of mesenchymal hamartoma has been described in a new born child where the lesion was solid rather than cystic which was attributed to the age of the
child(2). On the contrary, the tumor in our case was grossly cystic. Another rare feature noted in our case was the presence of calcification around large areas of necrosis (2).

Mesenchymal hamartoma of the liver are easily resectable and have an excellent prognosis(1-3). This case highlights the unusual vascularity of this tumor which led to hemorrhage and death before any surgical intervention could be contemplated.

REFERENCES