MESENCHYMAL HAMARTOMA OF THE LIVER

Report Of A Rare Case *

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As mesenchymal hamartoma of the liver is a rare tumor of infancy and childhood, a fibroadenomatous hamartoma of the liver, primarily bile cell, observed in an infant will be reported and the possible mechanism of its genesis will be briefly discussed.

Case Report

A. Z., an eleven month old female infant, was admitted to the department of pediatrics, Zeynep - Kamil Hospital with complaints of abdominal swelling and watery diarrhea. No abnormality or illness had been noted until the present condition, and the family history was noncontributory.

Physical examination revealed an acutely ill infant with a large distended abdomen and protruding umbilicus due to ascites and an abdominal tumor. The lower border of the tumor, which filled the entire right side of the abdomen, was easily palpable while the upper border was not.

Laboratory observations were as follows : Hb. 8.5 gm per 100 ml, red blood cells 3,650,000 per cubic millimeter, white blood cells 9,500 per cubic millimeter with a differential of: neutrophiles 56 %, lymphocytes 38 % and monocytes 6 %. Urinalysis and chest roentgenogram revealed no abnormality except the elevation of the right cupula of the diaphram up to the level of the 7th rib (Figure 1). The flat roentgenogram of the abdomen showed an enormous tumor of homogenous appearance filling the right side of the abdomen (Figure 2).

The patient was taken into the oxygen tent immediately and parenteral fluids were begun. Because of the rapid deterioration of the general condition, an exploratory laparotomy was performed on the second day after admission.

On surgery a large solitary tumor was found attached to the hilar region of the liver by a broad base reaching as far as the upper border of the pelvic fossa, and two liters of free ascites were found in the peritoneal cavity. The tumor was separated from the liver and the margins of the freed surface of the liver were sutured together. The patient died ten hours later, and post-mortem examination was not permitted.

The tumor removed by operation was an eight cm spherical firm mass with a well encapsulated appearance. The cut surface displayed a bulging lobulated

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appearance with many small cysts measuring less then 0.1 cm in diameter. There were also translucent, and yellow-green 0.2-0.3 cm areas (Figures 3 and 4).

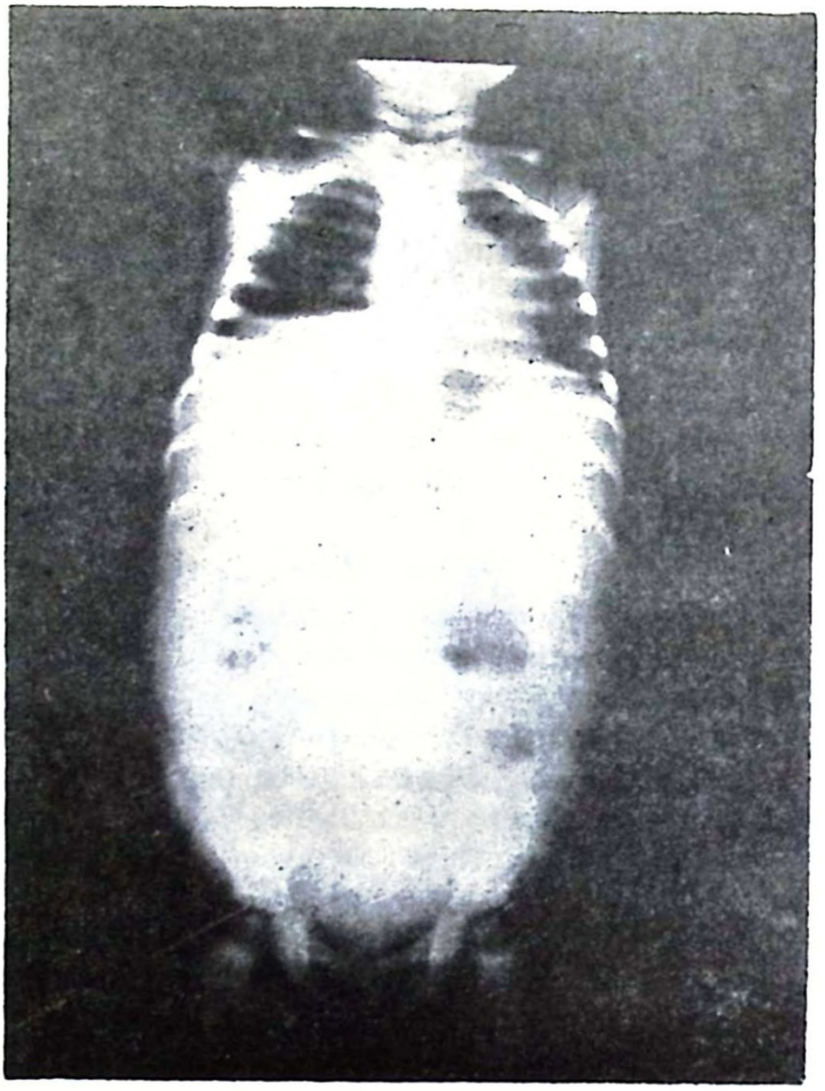

Figure 1. Roentgenogram showing elevation of the right cupula of the diaphram.

Microscopical examination revealed small groups of liver parenchymal cells arranged in cords without the usual structure. There were large areas of a cellular myxomatous-like connective tissue containing many bile ducts and capillary vessels arranged in a haphazard fashion. Growths of the pericanalicular connective tissue invaginated into the lumen of the bile ducts which were surrounded by one or two layers of cubical epithelium of an immature character reminiscent of the typical appearance of a mamary fibroadenoma (Figure 5).

Infiltration of polymorphonuclear leucocytes, lymphocytes and plasma cells were observed, dense in the pericanalicular zones (Figure 6).

Direct communications between the bile ducts and the liver parenchymal cells were detected.

Van Gieson's and Gomori silver impregnation stains revealed a young connective tissue with collagen and precollagen fibrils and reticulin fibers in and around the walls of the bile ducts and blood channels respectively.

Figure 2. X-ray showing tumor on the right side of the abdomen.

Figure 3. Tumor with its broad base.

Figure 4. Glistening semitranslucent sectioned tumor - elevated lobules have central cystic spots of a hollow appearance.

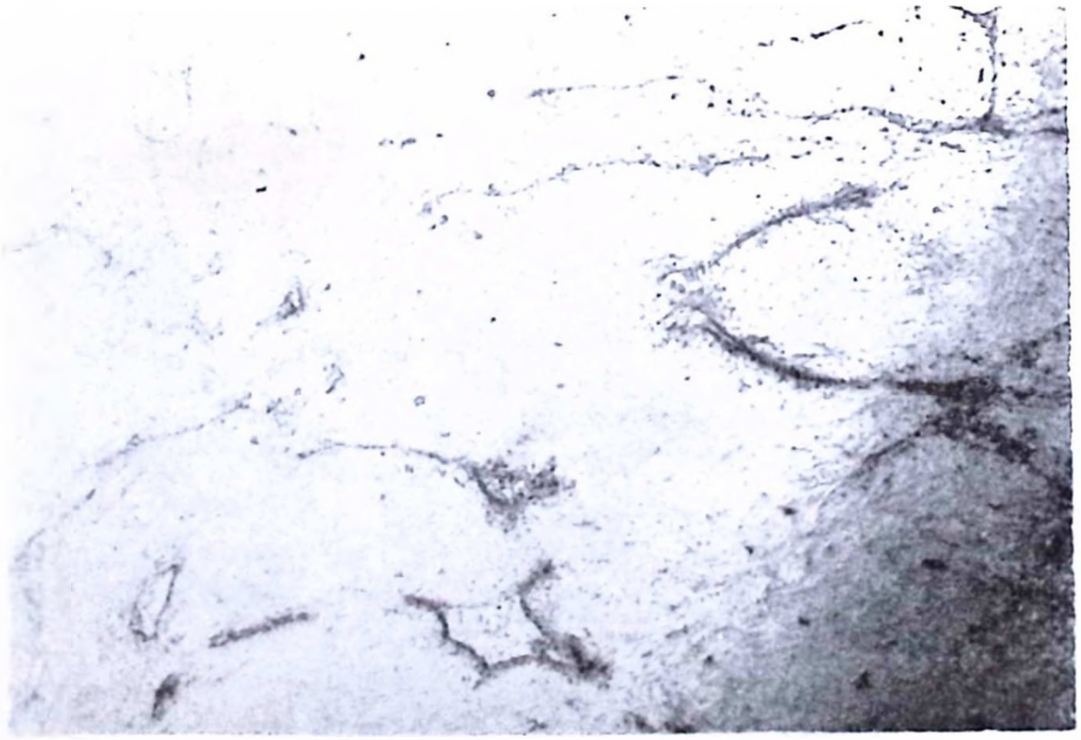

Figure 5. Bile ducts compressed into narrow channels by the growing young connective tissue, forming invaginations into some of the larger ducts. (x 60)

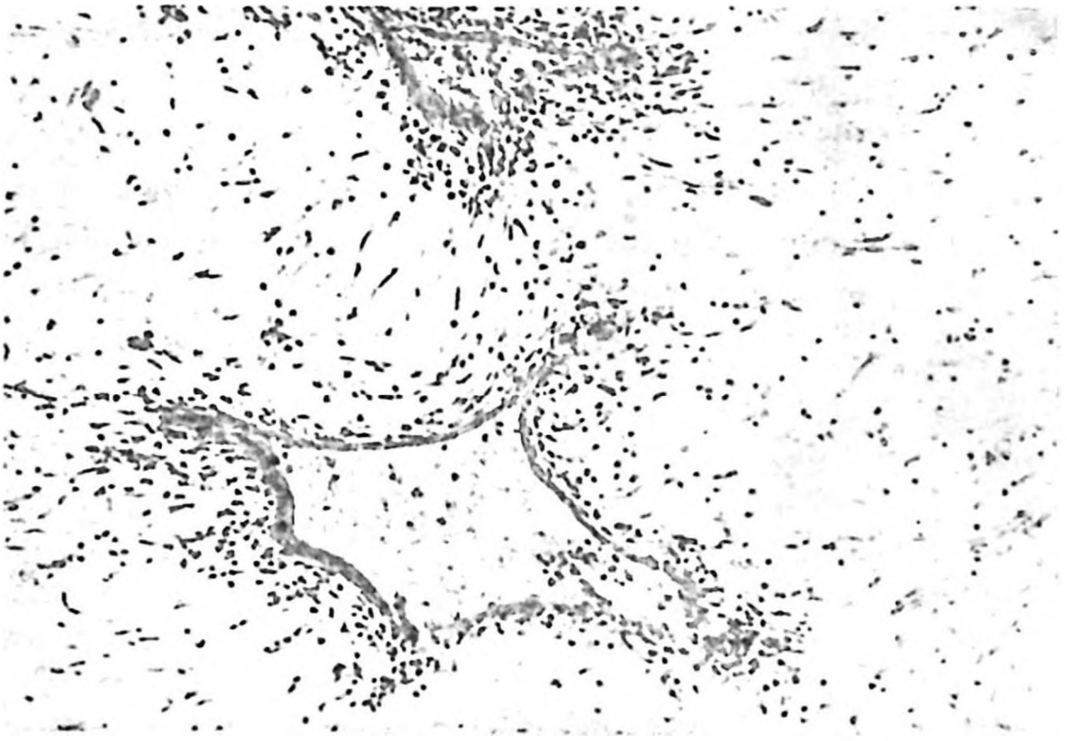

Figure 6. Young connective tissue loosely infiltrated by leucocytes, denser in the periphery of the bile ducts formed by immature epithelial cells. (x 100)

Discussion

The case is interesting because it illustrates a rare cause of ascites in children and a condition which should be differentiated from the tumors and other diseases of the abdominal organs and cavity such as mesenteric cysts and intestinal duplication.[8] In the present case, ascites developed very rapidly (four to five days) contrary to relatively slow development in previously reported cases.

Mesenchymal hamartomas of the liver are rare in the literature.[1 - 10] Some of these tumors presented a microscopic picture closely resembling that of a cirrhotic liver, with dense collagenous connective tissue bands containing wide venous channels and proliferating bile ducts, between the predominant areas of parenchymal cells. The tumors were either pedunculated or sessile. In the diagnoses the most consistent observation was an enlarged abdomen. However the likelihood of it being an hepatic tumor is seldom considered because other conditions are better statistical possibilities.

Three sources are thought of as the origin of intrahepatic bile ducts :

a) Liver parenchymal cells derived from the pars hepatica of the hepatic diverticulum of the foregut.[11, 13, 14]

b) Mesenchyma or mesothelium of the septum transversum into which the pars hepatica of the hepatic diverticulum grows, the latter also contributing to the formation of the bile duct epithelium.[15]

c) Secondary bile duct plate, a prolongation of the anlage of extrahepatic bile ducts.[16]

There is experimental evidence in favor of the hypothesis that the connective tissue surrounding the venous channels acts as organizer and promoter in the differentiation of bile duct epithelium.[17] In a recent study on the embryonal development of intrahepatic bile ducts, Kössling [14] concluded that the differentiation begins from the hilar region of the liver where a narrow meshed network of biliary ducts are formed by the liver parenchymal cells surrounding the connective tissue around the branches of the umbilical and left omphalomesenteric veins. From there the differentiation progresses deep into the liver. In a further stage of development, the network of bile ducts regresses in many sites, leaving behind a duct system parallel to the vein.

In the light of the morphology of the bile duct, three kinds of malformations are conceivable in the genesis of these hamartomas:

a) A remnant of the multipotent liver blastema with subsequent hamartomatous growth and differentiation into liver cells on the one hand and bile ducts on the other.

b) The failure of the anastomosing network of biliary ducts to regress in some parts of the liver maintaining the earlier stage of development and, growing to some extent, though independently, of the normally developed liver regions.

c) A hamartomatous growth of blood vessels causing an excessive growth and abnormal differentiation of otherwise normal liver cells into bile ducts. The causes of these are unknown but the lesions are benign and prognosis is better than usual in intra-abdominal tumors of children such as Wilms[2] or neuroblastomas. The exact cause of both, in our case, is unknown because of the absence of autopsy examination.

Summary

A case of mesenchymal hamartoma of the liver associated with a rapidly developing ascites in an eleven month old female infant is presented. Rapid enlargement of the abdomen and signs referable to a disturbance of the gastrointestinal tract were the first symptoms

of the condition in the previously perfectly healthy appearing infant. Microscopically the lesion consisted largely of loose myxomatous connective tissue of fibroadenomatous structure with bile ducts and scattered areas of hepatic cells.

It is stressed that tumors of the liver should be considered in differential diagnoses of abdominal syndromes.

Hypotheses in the genesis of the hamartoma are briefly discussed in the light of present knowledge of embryonic development of intrahepatic bile ducts.

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