

Mesenchymal Hamartoma of the Liver

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Abstract—Mesenchymal hamartoma of the liver is an uncommon benign lesion that appears as a large abdominal mass, almost exclusively in the first 2 years of life. It has been believed that this lesion is not a true neoplasm but rather a kind of hamartomatous lesion arising from the connective tissue of the portal tracts at some point in their development. Minor components of liver cells and bile ducts are considered a result of entrapment.

We report a case of typical cystic mesenchymal hamartoma in a 17-month-old boy who first presented with an abdominal mass at 7 months of age. The tumor was located in the right lobe, 17 cm in maximum diameter, and was largely cystic and partly solid. The cysts were multiloculated and lined by a single layer of tall columnar partly ciliated epithelium, and the irregular admixture of mature liver cell islands and intervening loose myxoid stroma was evident. Electronmicroscopic study revealed that the lining epithelium was the same as that of the bile duct, and the liver cell element was so mature that it couldn't be differentiated from normal hepatocytes. The stroma cell resembled fibroblast.

Key Words: *Mesenchymal hamartoma, Liver, Cystic tumor, Congenital tumor, Hamartoma*

INTRODUCTION

Mesenchymal hamartoma of the liver is a primary benign lesion that occurs almost exclusively during infancy and childhood. It is undoubtedly congenital, and most cases are clinically suspected when other causes of cystic enlargement of the liver are excluded (Grases et al, 1979; Johnson, 1968). Histologically loose edematous stroma is the main element of the tumor, cystic-lining epithelium being minor in the development of the organs or tissues.

We report a case of typical cystic mesenchymal hamartoma in a 17-month-old boy with electronmicroscopic study, and its histogenesis is reviewed.

CASE REPORT

A 17-month-old boy was transferred to Seoul National University Children's Hospital for further evaluation of an abdominal mass. He was born via C-section after 39 weeks of gestation, and the immediate postnatal life was uneventful. A mild abdominal distention was first detected by his parents at 7 months of age, and an abdominal mass could be palpated by 1 year of age. He was brought to several local clinics without being given a specific diagnosis. Physical examination on this admission revealed a well-developed baby with an enlarged liver. The surface of the liver was soft and

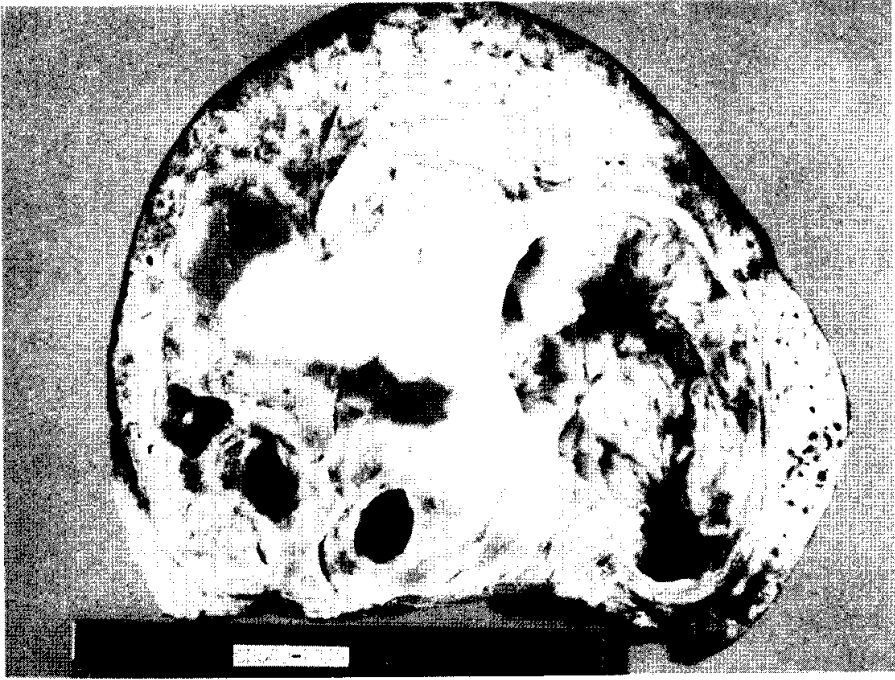


Fig. 1. Abdominal CT shows a huge cystic liver mass with smooth margined multiseptation.

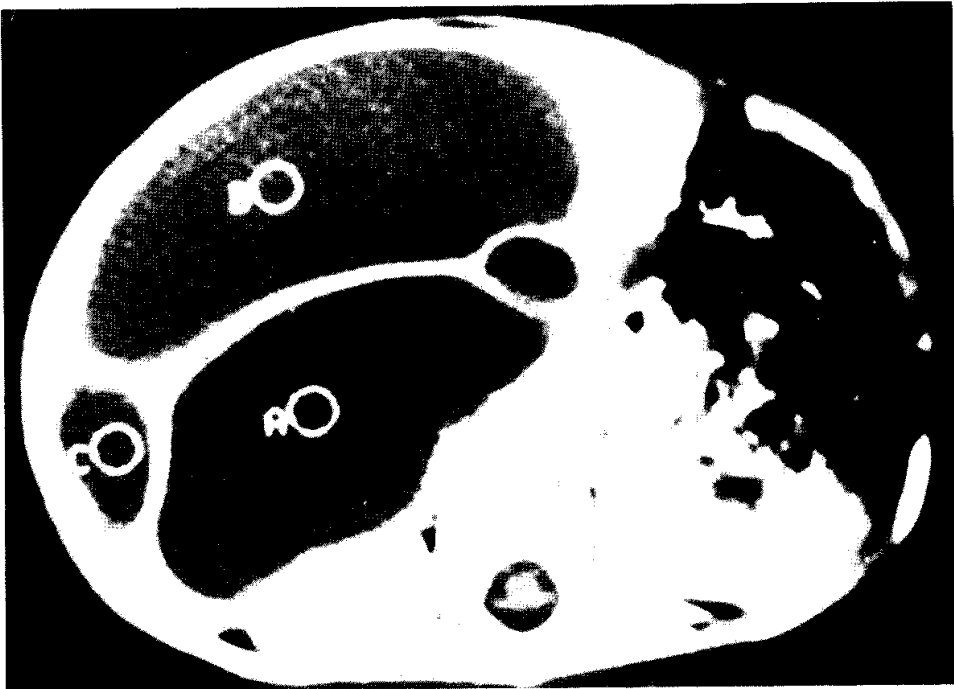


Fig. 2. Gross picture of the resected specimen shows well-demarcated largely cystic mass with irregular admixture of loose myxoid mesenchyme and hepatocytes islands.



Fig. 3. Microscopic view of the solid area. Loose myxoid mesenchymal tissue with an intervening epithelial structure is noted together with scattered liver cell islands (H&E x40).



Fig. 4. The cysts are lined by a single layer of partly ciliated columnar epithelium (H&E x400).

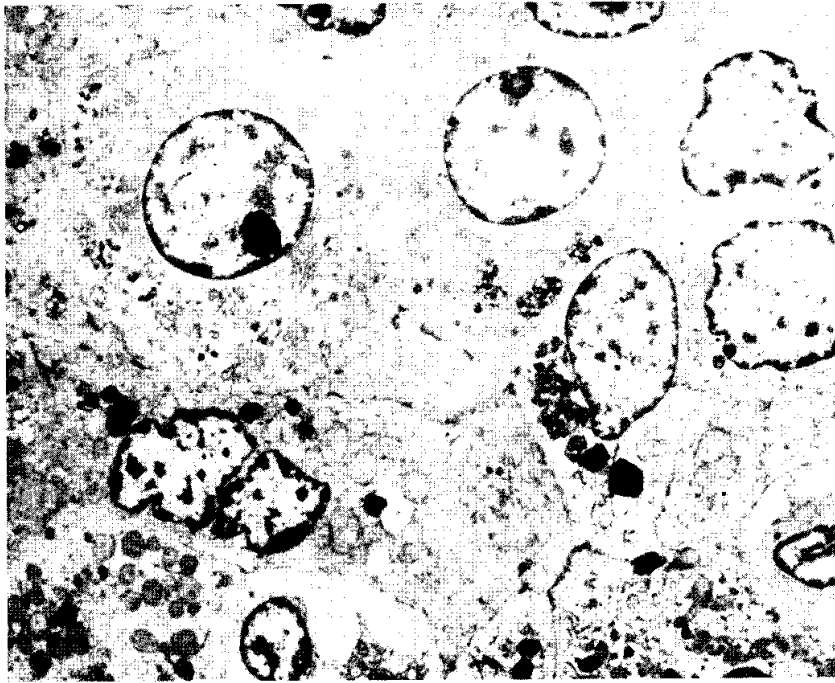


Fig. 5. Ultrastructural finding of entrapped hepatocytes. Note bile canaliculi between adjacent hepatocytes (Uranyl acetate-lead citrate, X2500).

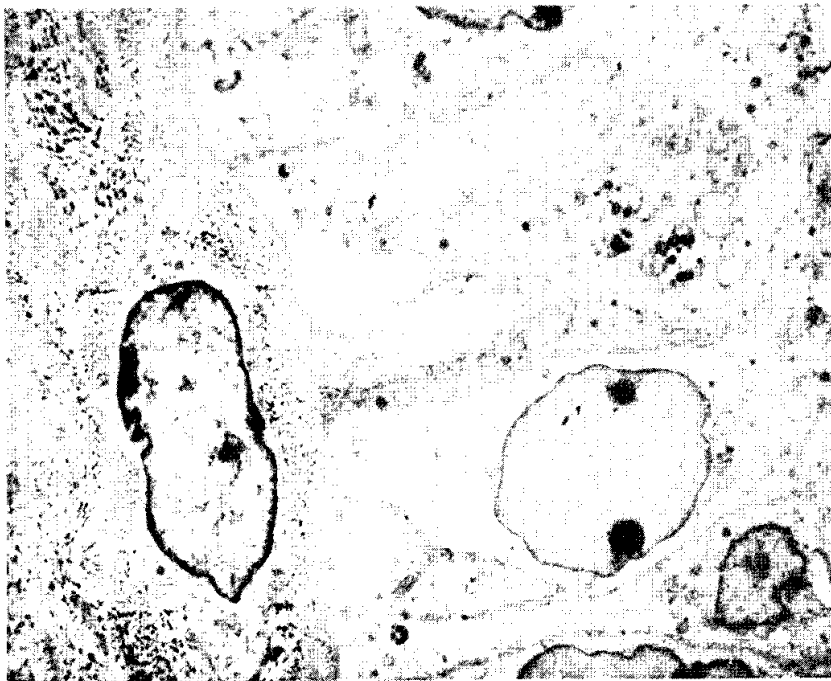


Fig. 6. Ultrastructural finding of lining epithelial cells of cysts. Short nonbranching microvilli are noted along the luminal border, and cytoplasmic interdigitation and cell junction are also found (Uranyl acetate-lead citrate, X3500).

smooth. Ascites was not present. Ultrasonography and CT revealed a huge multiseptate mass in the right lobe of the liver (Fig. 1). The septa of the locules appeared smooth. Laboratory data including liver function test, serum alpha-fetoprotein, carcinoembryonic antigen, and serology for echinococcosis remained within normal limits. Under a diagnosis of liver cyst, a right lobectomy was performed.

The resected specimen was the right lobe of the liver, measuring 17 X 12 X 8 cm. The entire lobe was replaced by a round mass. However, the lobe maintained its shape without a localized protruding mass. Cut sections showed a largely cystic (80%) and partly solid (20%) mass that was well-circumscribed from the normal liver parenchyme remaining intact around the mass along the periphery of the lobe (Fig. 2). The largest cyst measured 12 cm in diameter, and several other smaller thick-walled cysts were encountered. Some of the cysts were bile tinged. The solid portion showed yellow-tan liver parenchyme with an irregular admixture of round duct-like structures and loose myxoid stroma.

Microscopically, the cystic spaces were lined by a single layer of tall columnar partly ciliated epithelium (Fig. 4). Loose edematous myxoid stroma surrounded the spaces, but dense ovarian stroma-like area was absent. The solid area was composed of an irregular admixture of mature liver cell islands and loose myxoid stroma surrounding the tortuous branching bile duct-like structure (Fig. 3). Focal areas of lymphangiectasia were noted, but a tumor-like vascular lesion was not seen. The scattered hepatocytes were mature. Kupffer cells and sinusoidal development were good. Extramedullary hemopoiesis was found. The boundary between the tumor and normal liver parenchyme was free of any capsular structure; instead, several cords of liver cells were flattened.

Ultrastructurally, the loose myxoid tissue of the tumor consisted of mature collagen fibrils with interspersed fibroblasts. Most fibroblasts had a few mitochondria with moderately developed rough endoplasmic reticulum. The nuclei contained peripheral chromatin condensation. Nucleoli were inconspicuous. The duct-lining cells had short nonbran-

ching microvilli and were bound together by juxtaluminar tight and intermediate junctions (Fig. 6.) Some cytoplasmic interdigitations were noted between adjacent cells. The ductal structures were surrounded by a basal lamina. The hepatocytes were arranged in groups within the lesion, and their ultrastructural features were indistinguishable from those of normal hepatocytes (Fig. 5). The nuclei containing clumped chromatin usually had a nucleolus. Rough and smooth endoplasmic reticulum, mitochondria, and abundant glycogen were quite prominent in the cytoplasm. Typical bile canaliculi were noted between adjacent hepatocytes.

DISCUSSION

Mesenchymal hamartoma of the liver is a rare tumor that occurs almost exclusively in the first 2 years of life (Keeling, 1971). Since its pathologic description under the name of mesenchymal hamartoma by Edmondson in 1956, it has been called by several names such as lymphangioma, bile duct fibroadenoma, and cavernous lymphangiomatoid lesion. As the above terms imply, the pathological features of the hamartoma are somewhat variable. Moreover, some authors (Tate *et al.*, 1972) use the term hamartoma under the title of parenchymal hamartoma, which means focal nodular hyperplasia. As far as we are aware, none of the tumors has pursued an aggressive course (Gramlich, 1988; Grases, 1979).

It is interesting to note that a minor component of liver cells and bile ducts has been presented in all reported cases. Liver cells may be present as solitary cords or in larger irregular groups. Dehner *et al.* (1975) suggested that the islands of hepatocytes are there by entrapment and probably do not represent an integral part of the hamartomatous process. Our ultrastructural study confirmed the well-differentiated nature of the epithelial elements. The ductal structures in the loose mesenchyme or myxomatous tissue are formed and found together by cellular junction. These features are typical of what has been described for bile ducts. The hepatocytes, too, displayed normal ultrastructural features.

Although it is difficult to know the histogenesis

exactly, some suggestions can be made. The most appealing hypothesis is that this lesion arises from the hamartomatous growth of the connective tissues in portal tracts at some point in their development. The bile duct can be in such a situation, as a constant component of the lesion. The extent of the connective tissue proliferation and location in the liver would explain the differences that exist between the tumors. The islands of hepatocytes are there by entrapment and probably do not represent an integral part of the hamartomatous process. An alternative theory is that the hepatocytes are there by an aberrant inductive process between the derivatives of the primitive hepatic diverticulum as it extends into the septum transversum. In this way, the hepatocytes would represent an integral part of the mesenchymal hamartoma.

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= 국 문 초 록 =

간의 간엽성 과오종

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간에 발생한 간엽성 과오종은 대개 2세 이전에 나타나는 희귀한 양성 병변으로서 1956년 Edmondson에 의해 정확한 병리학적 기술이 있기 전까지는 여러 병명으로 불리어 혼동을 가져왔던 종양이다. 본 종양은 사람에서 간의 발생의 어느 과정에서 문맥부위에 있는 간엽조직의 과오종성 성장에 기인한다고 알려졌으며, 간세포군과 담관 구조물들이 병변의 일부로 많이 기술이 되어 있는데 이들은 종양의 일부라기보다는 포작의 결과라고 이해되어 왔다.

저자들은 17개월 남아에서 관찰된 간의 양성 간엽성 과오종 1예를 보고한다. 환아는 생후 7개월때부터 복부팽만이 시작되었고 증상은 진행되어 종괴가 촉진되었다. 간 우엽 절제술이 시행되었는데, 종괴는 육안적으로 회백색을 띠었으며 많은 낭성구조를 갖고 있었고 주위와의 경계는 명확하였으나 피막은 존재하지 않았다. 곳곳에 점액성 조직의 고형성 부위도 관찰되었으며, 현미경적으로 간세포의 섬들과 더불어 담관의 증식이 있었고 활동성 섬유모세포 증식과 부종이 관찰되었다. 커다란 낭성 구조물의 안쪽은 한층의 섬유성 원주세포로 피복되어 있었다. 전자현미경적 검사상 이들 세포는 담관세포와 유사하였고 간세포들은 정상과 같은 소견을 나타내었다.