

Cystic Mesenchymal Hamartoma of the Liver

— A Case Report —

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Mesenchymal hamartoma of the liver is an uncommon benign mass that appears as a large abdominal mass almost exclusively in the first two years of life.

This is a case report of a cystic mesenchymal hamartoma of the liver occurring in a 15 months old boy.

CASE REPORT

This 15 months old boy was the product of a normal full term gestation to a 33-year-old multiparous mother.

His abdominal distension became first noted by his parents at the age of eleven months. During the next four months, the abdomen continued to increase in size. A large firm, nontender mass was palpated in the left lobe, and it virtually filled the right side of the abdomen and extended across the midline. There were no other pertinent findings. Laboratory data included the following: hemoglobin 10.5gm%; white blood cell count 12,300/mm³; neutrophil 35%; lymphocyte 45%; monocyte 13%; eosinophil 7%; protein 6.3gm%; albumin 4.0gm%; alkaline phosphatase 670 IU/L, SGOT/SGPT 15/7 IU/L and alpha-fetoprotein 8ng/ml.

The mass was avascular on angiography, and was partly cystic and partly solid on ultrasonography. It was attached to the inferior edge of the left lobe by a broad pedicle. Partial excision of two thirds of the mass was per-

formed.

Pathological findings

The mass weighed 210gm and measured 16 × 15 × 8cm. Cut surface showed that the entire parenchyma was replaced by a well circumscribed gray white nodular mass containing multiple glistening translucent cysts filled with clear serous fluid. The largest cyst was 4cm in diameter. The nodules were variable in size. Along the margin of the mass normal liver tissue could be recognized.

Microscopically, the tumor was well demarcated from the adjacent liver parenchyma. Numerous nests of liver cells were seen throughout the lesion. They were separated by either dense collagenous connective tissue that was often arranged around vessels or loose myxomatous tissue with many ducts and gland-like structures. The hepatocytes in liver cell islands showed morphologically normal appearance with occasional sinusoidal structures. Typical bile ducts with concentric fibrous tissue and irregular elongated or cuboidal epithelial cells with surrounding fibrous connective tissue were also seen. Sections from cystic portions represented loose collagenous membrane lined by flattened cells. In areas, cystic structures containing fuzzy eosinophilic material and lined by endothelial cells were seen. These areas were strongly suggestive of focal lymphangioma.

COMMENT

Cystic mesenchymal hamartoma of the liver is a rare



Fig. 1. A huge tumor mass in the left lobe of the liver is seen (Picture taken during the operation)

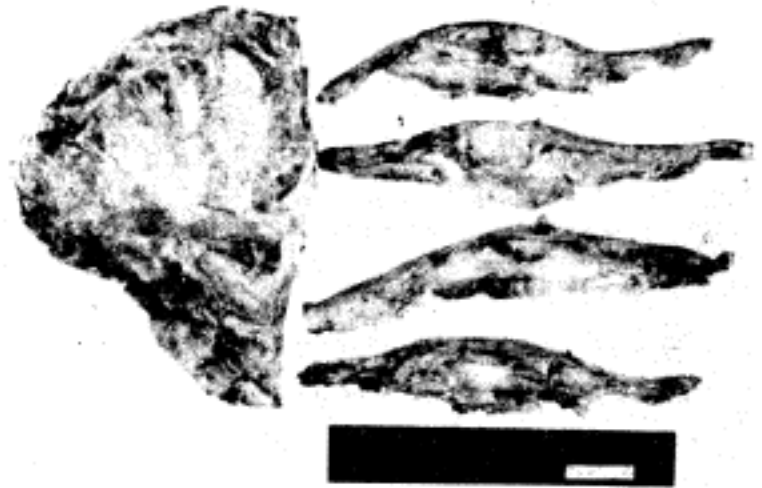


Fig. 2. Resected specimen showing inner and cut surfaces of the tumor.



Fig. 3. Photomicrograph of the tumor showing proliferated mesenchymal tissue with intervening epithelial structure. Liver cells are seen around this mesenchymal mass. Note also dilated lymphatic spaces in the right corners. (H&E $\times 40$)

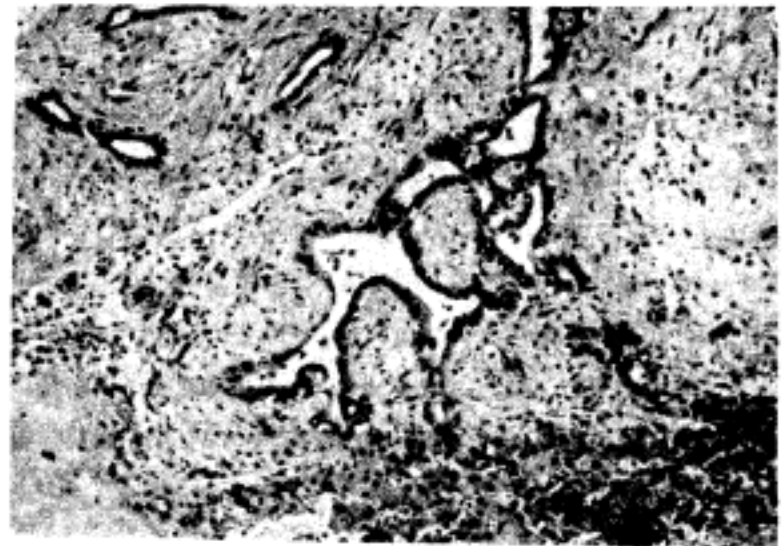


Fig. 4. In the solid portion of this tumor there are many bile duct-like structures lined by a single cuboidal epithelial cells. Note also liver cell islands (below.) (H&E, $\times 100$)

tumor that occurs almost exclusively in the first two years of life. Until 1975 approximately 27 cases has been reported in the literature¹⁾. In Korea a case of mesenchymal hamartoma of the liver in a 20 months old girl was successfully treated by surgical resection, and this case seems to be the only case described²⁾.

Although it was originally thought that this lesion was not a true neoplasm but rather some kind of developmental anomaly of mesenchyme³⁾, subsequent ultrastructural observation demonstrated the well-differentiated nature

of the epithelial elements⁴⁾. Therefore, hamartomatous nature of both epithelial and mesenchymal elements were suggested. In that sense it may be better to call this lesion as hamartoma instead of mesenchymal hamartoma. There is a hypothesis that this lesion arises from the connective tissue of the portal tracts at some point in their development.

It is interesting to note that minor component of liver cells and bile ducts has been present in all reported cases. Liver cells may be present as solitary cords or in larger irregular groups. Dehner et al¹⁾ suggest that the islands

of hepatocytes are there by entrapment and probably do not represent an integral part of the hamartomatous process.

Focal lymphangiomatous element seen in our case is interesting in the sense that one of the hypotheses of the genesis of this tumor is development from a lymphangioma of the liver⁴⁾.

Regardless of the exact cause of the tumor, this lesion behaves as a benign mass and carries a better prognosis than the usual intra-abdominal tumors of childhood. The treatment should be surgical removal of the mass without necessarily including adjacent normal liver.

SUMMARY AND CONCLUSION

A typical example of congenital cystic mesenchymal hamartoma of the liver is reported in a 15 months old boy who presented with abdominal distension at eleven months of age.

The tumor was in the left lobe and was a large circumscribed nodular mass measuring 16cm in maximum dimension. Numerous cysts were seen on the cut surface that were interconnected to form a large gray white mass which was well demarcated from the surrounding liver. Microscopically, the tumor was characterized by both epithelial and mesenchymal proliferation, epithelial being bile ducts and hepatocytes.

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=국문초록=

간의 선천성 낭성 간엽성 과오종(1증례보고)

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지제근 · 이해경 · 박귀원 · 김우기

15개월 남아에서 관찰된 간장의 낭성 간엽성 과오종의 1예를 보고 하였다. 환자는 생후 11개월부터 복부팽만으로 시작되어 점차로 심하여졌으며 수술로 간종괴 거의 전부를 절제하였다. 크기는 16×15×8cm였다.

종괴는 육안적으로 회백색으로 많은 낭성구조를 가지는 큰 종괴였으며 주위와의 경계는 명확하였다. 현미경적으로 간세포의 섬들과 더불어 담관의 증식이 있었고, 이와함께 점액성 조직 즉 활동성 섬유 모세포증식과 부종 그리고 약간의 맥관조직의 증식이 관찰되었다.