

## Cystic Mesenchymal Hamartoma of Liver – Case Report With Review of Literature

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### Case Report

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**Abstract:** Mesenchymal hamartoma of liver is the second most common benign liver tumor in children after hepatoblastoma. First described in 2006 as multicystic biliary hamartoma, it generally presents as large multicystic mass in less than three years of age. We report one such case in a 3 year old male child who came with history of fever and rash and was incidentally diagnosed with a cystic mass in the liver. Partial hepatectomy was done which on microscopy revealed epithelial structures in loose connective tissue stroma along with cystically dilated spaces lined by columnar and flattened epithelial cells, typical of Mesenchymal Hamartoma of liver. We are presenting this case because of its rarity.

**Keywords:** Hamartoma, Mesenchymal, Liver Cyst, Liver tumor.

### INTRODUCTION

Mesenchymal Hamartoma is a benign tumor of liver seen in pediatric age group and constitutes 8 % of all tumors in this population and 80% tumors present within first two years of life [2]. They mainly affect the right lobe of liver mostly around hepatic capsule close to falciform ligament [3]. The pathogenesis of mesenchymal hamartoma still not clear but is said to occur due to failure in normal development of embryonic fetal liver [1].

Mesenchymal hamartoma is characterized by the presence of overgrowth of mesenchymal or connective tissue of varying maturity and tendency for cystic formations. Basically connective tissue overgrowth is the most striking feature along with cysts separated by fibro-myxoid stroma only 140 cases have been reported in literature [4].

### CASE REPORT

A three year old male child presented with complaints of history of fever and rashes all over the body, on and off since one month which responded to conservative treatment. Liver function test (LFT), alpha-fetoprotein and other hematological investigations were within normal limits. Viral markers were negative. Ultrasonography abdomen revealed a cystic mass in left lobe of liver- 10 x 5.9 cm.

On CT abdomen and pelvis hepatomegaly was seen with large, well-defined, multi-loculated, variable sized, cystic lesion involving segment -IVA & IVB

measuring 7.5 x 7.3 x 7.2 cm. Kidney and other organs were normal. Based on this, clinical diagnosis of complex cyst of Liver was made. Partial hepatectomy was done.

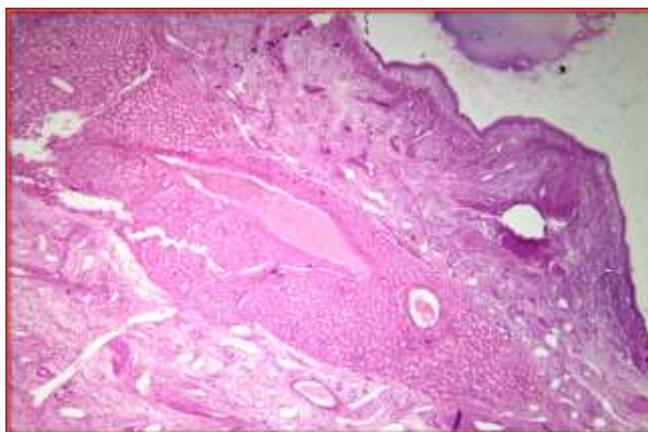
Grossly we received a subtotal liver specimen measuring: 8 x 7 x 3 cm. External Surface was grey-brown and congested with multiple cystic lesions separated by fibro-fatty tissue. Cut surface showed multi-loculated cysts, ranging from 0.5cm to 2 cms and yellow colored serous fluid oozed out of these cysts. There were no solid areas, necrosis or hemorrhage. Normal liver tissue was not seen (Figure 1).



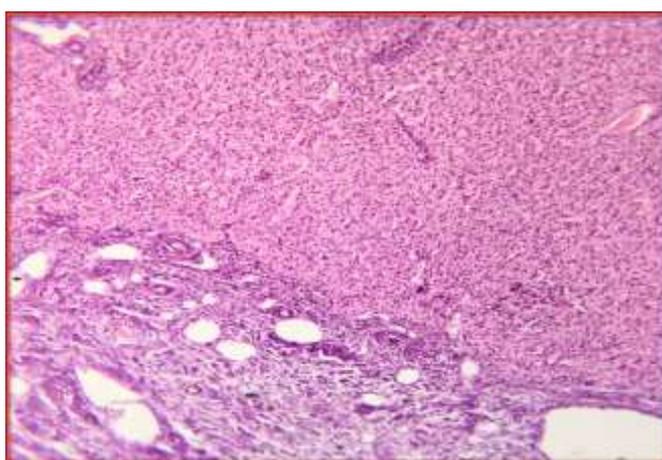
**Fig-1:- Gross showing partial hepatectomy with multiple cysts separated by thick fibrous septae**

Microscopy showed loss of normal liver architecture along with disorganized bile ducts and hepatocytes in loose fibrotic stroma. Cystically dilated spaces lined by bile duct epithelium, hepatocytes

arranged in small plates and nodules at the periphery showing early fatty change was also seen. Diffuse mixed inflammatory infiltrate was also noted (Fig 2 and 3).



**Fig-2:-H& E, 10X showing cysts admixed with hepatocytes and fibrosis**



**Fig-3: H& E, 10 X displaying mesenchymal tissue and hepatocytes**

Presence of disorganized bile ducts, hepatocytes in loose fibrotic stroma, cysts lined by columnar to flat cuboidal cells and fibro-myxoid tissue comprising of stellate and spindle cells between cysts

lead to the diagnosis of hamartoma of liver. Based on above histopathological features diagnosis of Mesenchymal Hamartoma of liver was made.

## DISCUSSION

Mesenchymal hamartoma was first reported in 1903 by Maresch but the term was given by Edmondson in 1956 [1]. Earlier it was called as cavernous lymphangio-adenomatoid tumour, cystic hamartoma, benign mesenchymoma [1]. 80% lesions are seen within two years of age and others within five years of age with male to female ratio of 2:1[5]. Laberge *et al.* had reviewed twelve cases out of which eight fetuses survived to term after diagnosis of mesenchymal hamartoma was made in third trimester [5].

Thirty cases have also been reported in adults between age group of 19 to 69 years of age [5]. Most are seen in newborn to five years of age mean age being 16 months similar to our case in which child was 3 years old [9]. The males are more affected in pediatric population whereas female in adult population[5]. Pathogenesis given by Edmson was that lesion might result from failure of normal development of embryonic fetal liver or maybe due to degenerative changes of accessory lobe [1]. Delner *et al.* Packard and Palmer thought that it was developmental anomaly of portal connective tissue during fetal life [1].

It is said that most of the patients are asymptomatic while some may present with right upper quadrant mass, respiratory distress, fever or abdominal distension similar in our case patient presented with fever and unexplained rashes all over the body [1]. It is reported that laboratory investigations like Liver function test (LFT) are usually within normal limit except slight raise in alpha-fetoprotein. In our case both LFT and AFP were within normal limits [1]. CT and USG suggest the presence of complex mass containing areas of low attenuation separated by solid septae and stroma similar in our case [6].

Grossly they are well circumscribed multiloculated mass which have myxoid areas with fluid filled cyst [7]. Microscopically typical lesions show both epithelial and mesenchymal components with branching bile ducts of various sizes and shapes in loose myxoid stroma with myofibroblast like cells, dilated veins and lymphatics along with extramedullary hematopoiesis. In our case showed all these features [7]. These lesions should be differentiated from other liver masses like hemangio-endothelioma and cavernous haemangioma which contain vascular spaces lined by immature plump endothelial cells [7]. In our case vascular spaces were not seen.

Other differential diagnosis of mesenchymal hamartoma were first ciliated hepatic foregut cyst which shows four layered cyst wall with respiratory epithelium along with smooth muscle layer, second bile duct hamartoma which have peri-portal small clusters of dilated bile ducts, often angulated in fibrous stroma with intraluminal bile, third polycystic liver disease shows greater replacement of hepatic parenchyma by

cystic areas lined by columnar to cuboidal epithelium outside the epithelium, cyst walls consist of collagenous connective tissue, associated mostly with polycystic kidney disease and fourth was epithelial mesenchymal hamartoma composed of epithelial and mesenchymal component due to absence of all these features in our case these diagnosis were ruled out [7].

Recent cytogenetic studies have shown rearrangement at chromosome 19 in mesenchymal hamartoma as well as undifferentiated embryonal sarcoma. However undifferentiated embryonal sarcoma is composed of sarcomatous mesenchymal component nuclear pleomorphism and mitotic activity which clearly separates these from hamartomas [5].

It is said that positivity for IHC markers like CK-7, vimentin, smooth muscle actin and desmin is seen while CK-20 is negative in these lesions. However, histopathological appearance is enough for the diagnosis in absence of malignant features [7]. It is reported that treatment is surgical resection and this condition has excellent long term prognosis. In the present case also patient is uneventful post-surgery [8].

## CONCLUSION

Mesenchymal hamartoma is a rare liver tumour, with no specific symptoms and should be kept in the differential diagnosis of cystic liver mass, especially in children less than 5 years of age.

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