

Laparoscopic management of cystic mesenchymal hamartoma of liver

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Abstract:

Cystic mesenchymal hamartoma are rare benign tumours of liver of uncertain aetiology. Such tumours are accessible for laparoscopic surgery. In the current report, we discuss a case of a 4-year-old female child diagnosed to have cystic mesenchymal hamartoma of liver that was managed laparoscopically. The clinical, radiological and pathological features of the same are discussed.

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Introduction

Mesenchymal hamartoma of liver are rare benign tumours of childhood though few cases have been reported in adults too (1). Maresch reported the first case in 1903 (2). In 1956, Edmondson classified the tumours as mesenchymal hamartoma (3). It is usually a slow growing tumour, most common before the age of 2 years and composed of architecturally abnormal bile ducts in an uncommitted myxoid stroma (4).

Case History

A 4-year-old female was referred to us with abdominal distension since 2 months. There was no history of fever, jaundice, pain or any other gastrointestinal symptoms. On examination, there was a palpable lump about 15 cm x12 cm in the right hypochondrium, non-tender, firm in consistency, smooth surface.

Complete blood count, liver function test and coagulation profile were normal. AFP was 1.2. USG showed well defined multi-septate round lesion in right lobe of liver having mixed echotexture with central cystic areas. CECT abdomen showed a multi-septated cyst about 12 cm x 15 cm in segment V, VI, VII of liver.

Patient underwent laparoscopic partial excision of cyst wall with marsupialization using harmonic scalpel. Intra-operative blood loss was approximately 30 cc and operating time was 90 minutes. Postoperative course was uneventful. Liquids were started, 6 hours post surgery and gradually full diet was given. Drain output was approximately 30 cc per day, serous. Drain removed on third post operative day.

Histopathology of cyst wall showed collagenous tissue with oedematous myxoid material with low columnar epithelium lining the cyst wall confirming the diagnosis of cystic mesenchymal hamartoma (Fig. 1-6).



Fig. 1: Large multi-septate cystic collection



Fig. 2: Large cystic mesenchymal hamartoma arising from the liver



Fig. 3: Cystic hamartoma being aspirated to evacuate the contents



Fig. 4: Cyst wall being excised with harmonic scalpel



Fig. 5: Complete excision of cyst wall



Fig. 6: Marsupialization completed

Discussion

Mesenchymal hamartoma of liver is a rare primary benign tumour that occurs during infancy and childhood though few cases have been described in adults too. Majority of cases present before 2 years of age though ranges from new born to 5 years. It commonly originates from the right lobe of the liver. Recent evidence suggests the possibility of a true neoplasm such as translocation involving chromosome 19q13.4, rare incidences of malignant transformation and aneuploidy in flow cytometry. It is most commonly cystic but solid forms are also seen. Edmondson proposed that the mesenchymal hamartoma arises from a mesenchymal rest that becomes isolated from normal portal triad architecture and differentiates independently. The tumour grows along the bile duct

and may incorporate normal liver tissue. Most of the cases remain asymptomatic while others are detected incidentally with symptoms such as upper quadrant mass with or without pressure symptoms. Rapid enlargement is due to fluid collection within the cyst. Serum AFP is usually normal but may be elevated in solid variety.

Ultrasound is the most common investigation for diagnosis followed by CT which usually demonstrates single usually large, fluid filled mass with fine internal septations and no calcification or vascular invasion with cord like wall enhancement.

Histopathology is suggestive of overgrowth of mesenchymal stroma and proliferation of architecturally abnormal bile ducts with or without cystic changes, associated with periductal collaring of stromal cell with hepatocytes. AFP levels correlated with amount of hepatocytes in the tumour.

Complications are because of rapid increase in size which results in mass effect from bulky tumour causing pressure on IVC or other hollow organs or intraperitoneal rupture or torsion of pedunculated cyst. Rarely malignant degeneration of cyst has been reported.

Differential diagnosis includes non-parasitic cysts, vascular lesions like hemangioendothelioma and cavernous hemangioma, other benign tumours of liver like FNH, adenoma, teratoma, Caroli's disease, hepatic abscesses, metastasis or polycystic liver disease.

The treatment of cystic mesenchymal hamartoma is surgery. There are four surgical options and the procedure chosen must be individualised. Options include enucleation, excision of hamartoma, marsupialization of cyst and formal hepatic lobectomy. Excision and marsupialization is recommended in all patients with very low surgical morbidity and mortality and excellent long term prognosis. Death may occur without surgical treatment because of rapid growth and mass effect, hence early intervention is advisable.

Conclusion

These tumours are immensely suitable for laparoscopic excision or marsupialization which may become the standard of management for these tumours.

References

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