Case Report No. 2

Case report of surgical management of a giant cavernous malformation of liver

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Abstract

Hemangiomas are the most common benign tumors of the liver. They are generally asymptomatic, but giant hemangiomas can lead to abdominal discomfort, bleeding, or obstructive symptoms, and spontaneous rupture which is rare but has a catastrophic outcome if not promptly managed. The effectiveness of liver resection to treat such symptoms has varied in published reports. The management of giant (⩾5 cm) hemangioma of the liver remains controversial. The aim of this case report was to assess the influence of tumor size on postoperative outcomes after hepatectomy in patients with giant hemangioma of the liver.

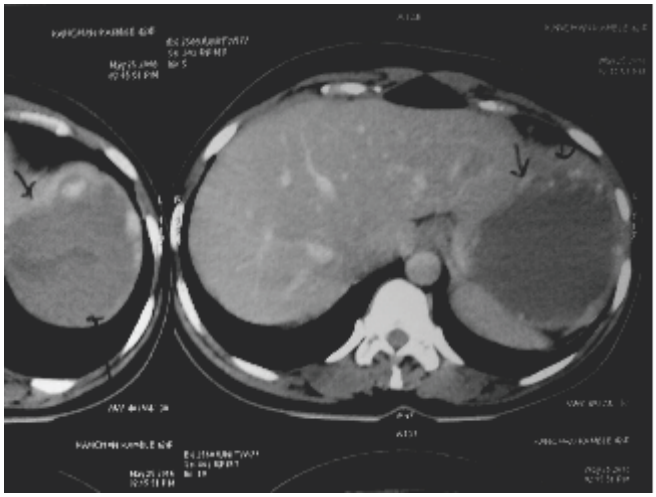
Introduction : Hemangiomas are the most common benign tumor of the liver with an estimated prevalence of 0.4% to 20%.[1] Cavernous liver hemangiomas (CLH) are congenital vascular malformations that grow by ectasia—rather than hypertrophy, hyperplasia or neoplasia—therefore resulting in microscopic, and often macroscopic, cavernous spaces. CLH range in size from a few millimeters to over 20 cm; those over 5 cm referred to as giant hemangiomas.[2]

We report a case that is particularly interesting given the rare presentation of this fairly common tumor, and expand on indications for treatment and surgical decision-making.

Case report

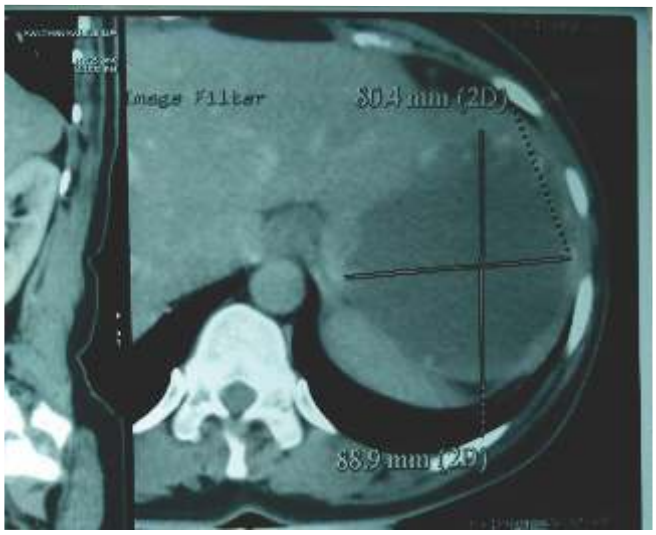
A 42-year old woman with vague complaints of hyperacidity since one and half month. She was complaining of diffuse abdominal pain since several months and there was no history of any functional sign. On physical examination the patient was slightly pale without jaundice; bilateral lower extremities were free from edema.

Figure 1. Contrast CT showing a large well circumscribed hypodense SOL in segment II & III of the left lobe of liver with large exophytic component bulging inferiorly.



The abdomen was mildly distended and slightly painful. There was no ascites and no collateral venous circulation. Laboratories investigations revealed a mild anemia ,liver function tests were normal. Hepatitis B and C virus markers were negative and alpha feto-protein level was in normal value. Ultrasonography examination revealed a huge hyperechoic mass (about 8x5 cm)well defined in subdiaphramatic region abutting the posterior capsule of spleen. Computed tomography scan showed a huge hypodense mass of the left lobe of the liver with nodular peripheral enhancement measuring about 89x80x54 mm in size closely related to posterior pole of spleen.

Figure 2. Contrast CT showing dimensions of hemangiomatous growth



With the diagnosis of giant hemangioma (GH) established the goal of treatment was to remove the mass while preserving the majority of uninvolved liver.

Figure 3:Surgical specimen— revealing a giant cavernous hemangioma measuring 8x5cm .The specimen weighed 1500 g.

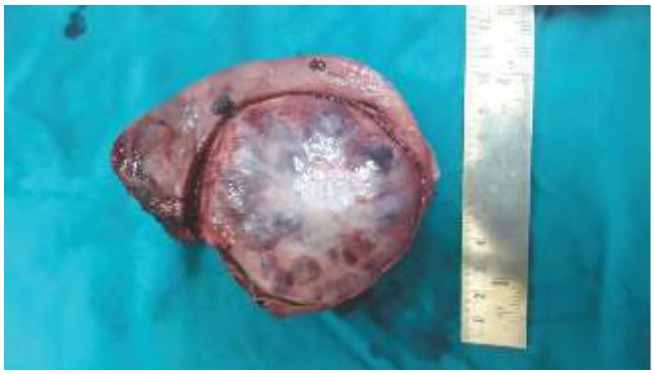
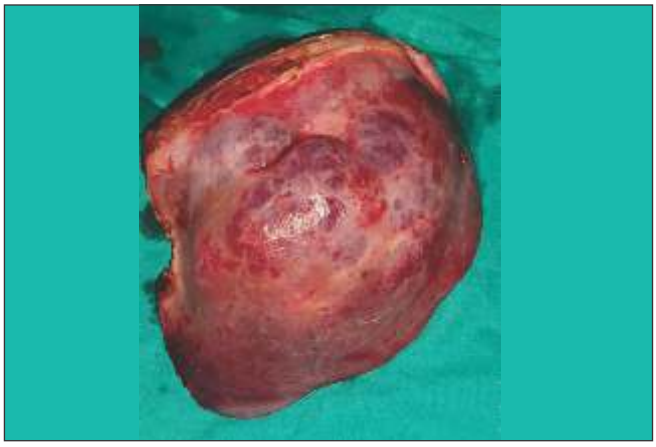


Figure 4: Gross specimen on cut section revealed mass which was soft, spongy inconsistency



Laboratories liver investigations were normal

We performed liver resection with left hepatectomy(segment 2&3) , she passed a smooth post operative period with 1.5 L blood loss during operation which was replaced.

She was discharged on fifth post operative day.

The histopathology result confirmed the diagnosis of giant cavernous hepatic hemangioma.

Discussion : Giant Hemangiomas of the liver are uncommon and rare incidence was found to be 2%; They are more common in the right lobe of the liver than in the left lobe[3]. A giant hemangioma is considered when its size is more than 4 cm.[4] Most of them remain stable and no symptom over time. But some of them may become symptomatic and the most common symptom was abdominal pain or discomfort.

The lesion is predominantly found in female, about 4.5 times higher than the male.

With these unusual clinical and imaging characteristics, the differential diagnosis was broader and included concern for an underlying neoplasia (i.e., hepatocellular carcinoma or cystadenoma/ cystadenocarcinoma). Additional clinical, imaging and laboratory information were used to narrow the diagnosis so that appropriate treatment could be established; in the context of no underlying liver disease, with the pattern of peripheral nodular and progressive enhancement of a solitary lesion, normal tumor markers and lack of any distant disease, the preoperative diagnosis of a giant CLH was confirmed

It is not frequent that large hemangiomas sequester and destroy platelets causing consumptive coagulopathy, symptomatic thrombocytopenia, known as Kasabach Merritt syndrome.[5] The modalities used to aid in the diagnosis of hepatic hemangiomas include ultrasonography; dynamic contrastenhanced computed tomography; nuclear medicine studies using technetium (Tc) 99m-labeled rbcs, magnetic resonance imaging, hepatic arteriography and digital subtraction angiography.[6]

In general, the finding on ultrasonography of a suspected hemangioma should be diagnostically

integrated with CT scan or MRI to insure a correct diagnosis.[6,7]

Right upper quadrant pain or fullness is the most common complain. In some cases pain is explained by thrombosis and infarction of the lesion, hemorrhage into the lesion, or compression of adjacent tissues or organs. In other cases pain is unexplained.[6]

For the symptomatic patients it is advocated that they should undergo a thorough evaluation to find out any other cause for the symptoms. If the hemangiomas are the cause of pain, these patients are candidates for surgical treatment. The size of lesion is not a criterion for resection during management of giant liver hemangioma. However the smaller giant hemangioma also can cause symptomatic manifestation.

In our case the indication for surgical resection was: rapid increase in size; hyperacidity and discomfort for long duration .high risk of thrombosis inside the tumor; high risk of rupture with hemoperitoneum.

After surgery majority of patients surgical treatment can achieve the best result of symptom relief.

The size and location of a lesion will influence the surgeon's decision to perform either a segmental resection or an enucleation of the hemangioma. In general, both of two methods (surgical resection & enucleation) are safe and well tolerated by patients .Besides surgical resection there are other modalities available to manage symptomatic hemangioma in the literature. Arterial embolisation is an option in such circumstances. Branches of hepatic artery can be embolized with polyvinyl alcohol and other substances.[8,9]

Other techniques were mentioned in the literature

- Surgical ligation of feeding vessels: transhepatic compression sutures using polytetrafluoroethylene Pledgets and selective ligation of large feeding vessels have been described. In one case this technique successfully reduced intratumoral shunting that otherwise would have led to intractable cardiac failure.[10]
- Hepatic irradiation: with a dose of 30 Gy in 15 fractions over 3 weeks has been reported to produce complete regression of hepatic hemangioma with minimal morbidity.[11]
- Orthotopic liver transplantation: is occasionally offered to symptomatic patients with large or diffuse lesion. Several cases have now been reported in the literature.[12]

Finally, for huge symptomatic giant hemangioma the management by combination of both non-surgical and surgical modalities may reduce the size of lesion and make surgical procedure easier.

Of the various treatment options for giant hemangiomas, surgical treatment, including resection and enucleation, provides the only consistently effective outcome with satisfactory results[13]

Because of variations in size, location, and number of tumors, the surgical strategy should be decided on a case-by-case basis

Hemostasis is important for resection of a giant hemangioma . The larger the size and the greater the number of tumors, the more difficult it is to achieve hemostasis. Huge hemangiomas, multiple giant hemangiomas, and hemangiomatosis frequently require challenging operations

Conclusion : When the appropriate indications are present ,major liver resection is a reasonable and safe alternative, with excellent outcomes when performed in the appropriate setting.

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