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Case Report

Giant Hepatic Hemangioma Causing Multiple Pressure Effects in a Teenage Girl: A Case Report

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Abstract: Hemangioma, as is known as the most common benign primary tumour of liver, is usually asymptomatic. They are usually less than 3 cm and can be solitary or multiple in number. Asymptomatic hemangiomas of liver need no surgicel intervention and masterly inactivity is suggested. But the rare symptomatic ones are vulnernable for various dreadful sequelae and hence operative intervention is indicated. We here report a case of 16 yr old girl presenting with symptoms of intermittent small bowel obstruction and Inferior venecava compression due to the presence of a Giant Hepatic Hemangioma.

Keywords: Giant Hepatic Hemangioma, Liver, Hemangioma, Wedge resection of liver, Non anatomic resection of liver, Subacute Intestinal obstruction, Inferior Venecava Syndrome

INTRODUCTION

Being the most common primary benign lesion of liver, hepatic hemangiomas are usually less than 3cm in size, a significant proportion of which are asymptomatic [1-4]. Their prevalence are approximately 1-20% in general population and 7 % in autopsies [5-8]. Their incidence is higher in women, as a result of influence of sex hormones on their growth [9].

Regarding the terminology Giant Hepatic Hemangiomas are referred to hemangiomas of liver of size more than 4cm and they represent 10 percent of all hepatic hemangiomas [10-15]. In 40 % of the patients with giant hepatic hemangiomas, symptoms have been reported of which abdominal pain is the most common followed by pressure symptoms like early satiety, nausea, vomiting, cholestasis, or even cough [16-19]. Rare presentations are spontaneous rupture presenting as chylous ascites and hemoperitoneum. Others are Kasabach Meritt syndrome in which hepatic hemangioma is associated with thrombocytopenia and intravascular coagulation [20-22]. Even rarer complications are associated with GHH like intestinal obstruction, Budd-Chiari syndrome and inferior venecaval syndrome.

Here we report a case of giant hepatic hemangioma in a 16 year old girl with both subacute intestinal obstruction and IVC syndrome occurring simultaneously. Presence of multiple compressive symptoms in the same patient with GHH is something which is very less discussed in literature. Our study aims to present such a rare case with its rarest complications to our readers.

CASE HISTORY

A 16 year old girl presented to a District Hospital with the complain of pain and abdominal distension with nausea, vomiting and menstrual irregularity for last 6 months. The patient was apparently normal 6 months back when she felt heaviness in the lower abdomen which gradually increased over the last 3 months. The patient reported a loss of weight of about 10kg during the last 3 months. Abdominal examination clinically revealed a mass in the central abdomen and encroaching the hypogastrium. CT imaging demonstrated a very large solid cystic abdomino-pelvic lesion with relatively small solid cystic mass in adnexa suggestive of bilateral ovarian neoplasm and a well defined hypodense wedge shaped lesion in the right lobe of liver. Blood counts were normal. Midline laparotomy was carried out left salpingo-oophorectomy was done for left ovarian haemorrhagic cyst which was measuring 6 cm * 6 cm. A huge vascular SOL was found to be originating from the right lobe of liver which showed bloody content on aspiration. Due to the vascular nature of the tumor and lack of experienced surgeon, the operation was concluded a patient was referred to our centre after her

discharge with the diagnosis of Giant Hepatic Hemangioma. The patient on admission had features of subacute intestinal obstruction such as recurrent vomiting, abdominal pain with distension and occasional constipation. She too had features of Inferior venecaval syndrome like swelling of lower limbs with palpitation on lying on back or right side. These symptoms were relieved on sitting or standing and on lying on the left lateral position. At our centre blood tests were done which showed Hemoglobin- 11gm/dl, total leukocyte count-11,300/cumm, platelet-2.5 lacs/cumm, INR-1.2, Bilirubin Total/direct- 0.9/0.2 mg/dl, AST-25 iU/l, ALT- 27 iU/l, Alk Ph- 591 iU/l, albumin 3.3 gm/dl, AFP- 0.89 iU/ml. Other tumor markers along with hepatitis profile was normal. Triple phase MDCT revealed a large well defined heterogeneously enhancing solid mass of 19.2cm * 11.2cm * 20.4cm noted in the right lobe of liver extending from right side of the abdomen below upto the right iliac fossa compressing both the small bowel loops and infrahepatic IVC (Figure 1).

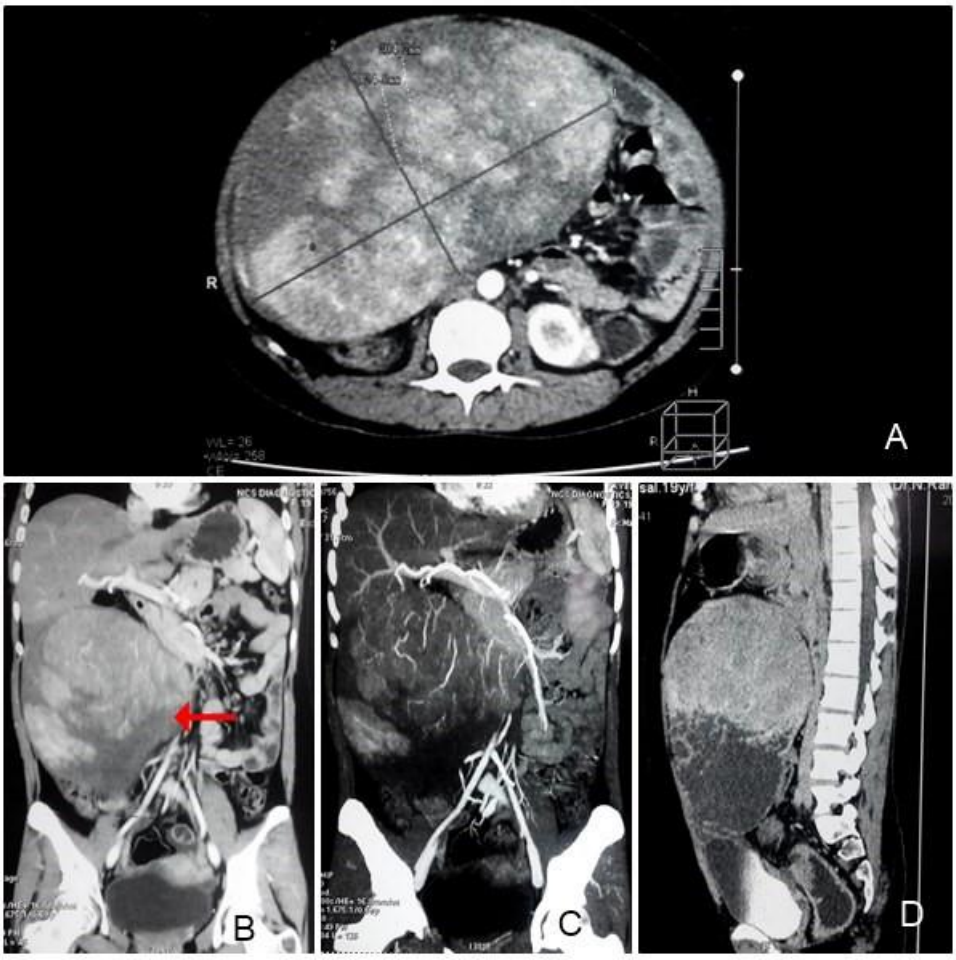

Fig-1: CTscan WA showing the GHH in different planes A-Axial B,C-Coronal D-Sagittal

After a period of 6 weeks we proceeded for operative intervention. Preoperatively the patient weighed 52kg. A Chevron incision was made. A huge mass continuous mass with the inferior surface of right lobe of liver (Segment 5 and 6) was found encroaching the hypogastrium and right iliac fossa. After adhesiolysis in the pelvic cavity due to adhesions caused by previous surgical intervention, the huge mass was delivered out of the abdomen (Figure 2).

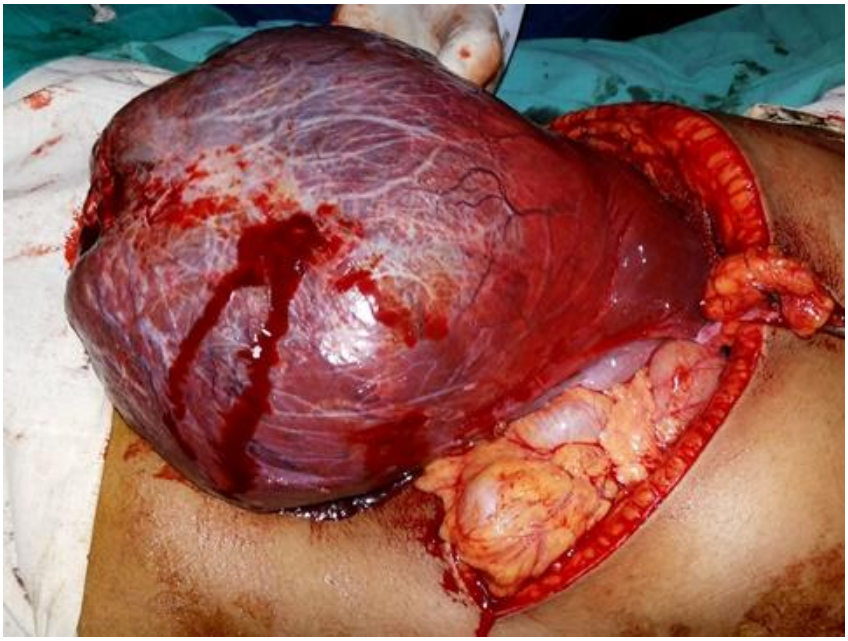

Fig-2--Delivering the GHH out of the abdomen

There was no infiltration in any adjacent bowel or great vessels. Cholecystectomy performed and a wedge resection of the exophytic growth was carried out by means of a harmonic scalpel taking a 2 cm healthy liver tissue from Segment 5 and 6. The defect was primarily sutured with catgut over a surgical (Figure 3).

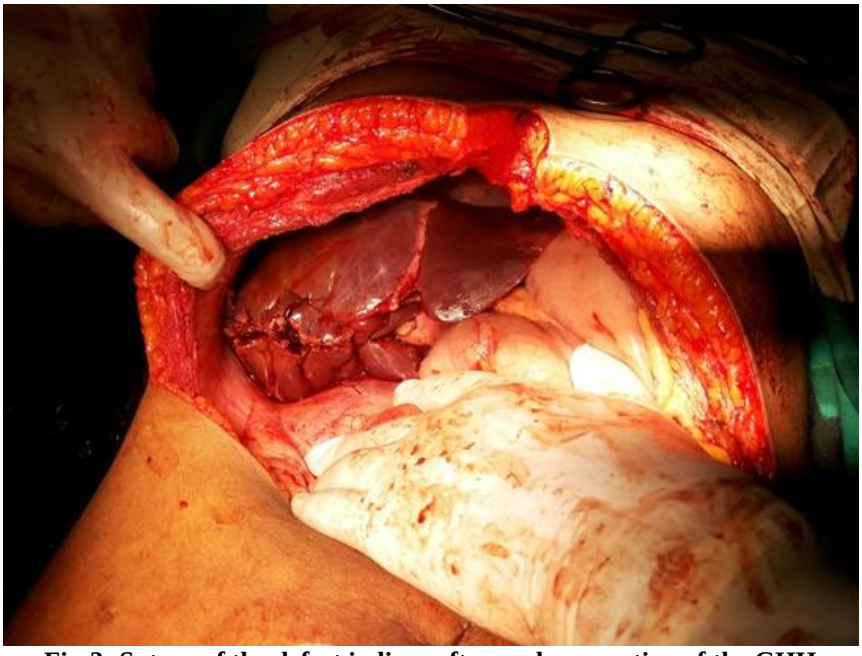

Fig-3: Suture of the defect in liver after wedge resection of the GHH

The procedure took nearly 1 hour with approximately 400ml of blood loss. The specimen weighed 12 kg, solid in consistency with soft and cystic degenerations at places on cut section (Figure 4).

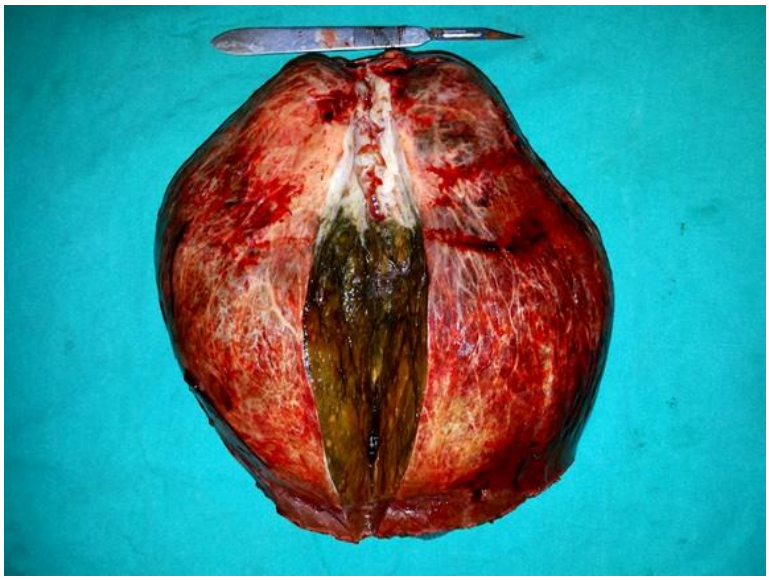

Fig-4: Cut section of the specimen

Histopathology confirmed hemangioma of liver. Post operatively the patient had an uneventful course and her symptoms were totally relieved. On 1 month and 6 month follow up, the patient was absolutely normal on the basis of physical examination, biochemical and radiological investigations.

DISCUSSION

Hemangiomas are hamartomas caused by proliferation of vascular endothelial cells over time. Hemangiomas though are found most commonly in the skin of head and neck, are common in all surface and organs of the body including liver. Conventional hepatic hemangioma are smaller in size and do not cause any symptoms. They are more common in women between 30 yrs to 50 yrs. The etiology is still controversial, as the relation between female sex hormone and HH is unclear in literature [25-28].

Most hepatic hemangiomas are incidentally diagnosed during radiological tests done for different other complaints. The manifestations are based on 2 factors - the size and the location of the lesion. 40% of the hemangiomas which are more than 4 cm and 90% of the hemangiomas more than 10cm diameter are symptomatic.

Most frequent symptom of GHH is pain which are caused due to stretching of Glissons capsule, thrombotic transformation of hemangioma, necrotic degeneration and intratumoral hemorrhage. Other features of Gastric outlet obstruction like nausea ,vomiting, epigastric fullness can be present [11, 14]. Rare complications which have been mentioned in earlier case reports are Kasabach Meritt Syndrome causing thrombocytopenia, microangiopathic haemolytic anemia and consumptive coagulopathy [20-22]. Clinical findings of Budd-Chiari syndrome and Inferior Venecaval Syndrome may develop due to pressure effect on the hepatic veins and inferior venecava [29]. Complications of intestinal obstruction can be acute or subacute. It is due to the compression of small bowel loops by the enlarged mass. Complete obstruction is uncommon due to the peristaltic activity and the elastic distensibility of the bowel.

Diagnosis can be made by radiological investigations. Ultrasonography is the initial investigation showing homogenous hyperechogenicity. Confirmation can be done by triphasic Computed tomography (CT) and Magnetic Resonance Imaging (MRI). In case of diagnostic uncertainty , Tc 99m labelled RBC SPECT is of immense help. PET is used to differentiate between heterogenous hepatic hemangioma and Angiosarcoma [23]. Angiography is the gold standard in the diagnosis of hemangioma having invasiveness as its major disadvantage. It is of diagnostic help when the diagnosis of hepatic hemangiomas from other malignant tumors could not be ruled out [5].

Close monitoring is recommended for small and asymptomatic hepatic hemangioma [13, 14, 29]. Whereas for its giant variant which are mainly symptomatic there are a lot of treatment options. They are enucleation, hepatic resection- non anatomic wedge resection and anatomic resection, hepatic artery ligation, transcatheter embolisation, and recent therapy with Radiofrequency ablation (RFA) [24]. Other recent advances are systemic corticosteroid and Interferon treatment [30]. Liver transplantation is the last option.

The indications of surgical treatment can be summarised as tumor size more than 5cm, peripherally located, persistent abdominal pain, suspicion of malignancy and compressive symptoms like gastric outlet obstruction, portal hypertension, obstructive

jaundice, intestinal obstruction, inferior venecaval syndrome, etc., [11, 14].

Previously liver resections were preferred mode of surgical treatment. With the modern technologies and advancement of surgical principles, an evolution took place in the surgical management of giant hepatic lesions like hemangiomas and other benign tumors. A paradigm shift towards more conservative approach of surgery was implemented. Enucleations and non anatomic wedge resections were carried out and non randomised studies has proven it as safer, quicker, having the advantage of less blood loss and decreased rates of morbidity and mortality. These were not the best options for centrally located lesions, though Pringle manoeuvres at appropriate intervals proved to be effective in these situations. Total hepatic vascular exclusion with extended hemihepatectomy [13, 14] are the suitable approaches for multiple hemangioma and large centrally placed hemangioma not involving more than 70% of liver. If more than 70% involvement is present or when there is hepatic hilum involvement causing unresectable disease, then liver transplantation is the only available appropriate approach [13, 30].

CONCLUSION

Hemangiomas are benign tumours of the liver for which close observation is recommended and surgical resection is indicated for symptomatic relief of complicated and giant hemangiomas and in which diagnosis is uncertain. Enucleation and hepatectomy are the surgical options of which the former is ideal. Giant Hepatic Hemangiomas cause multiple mass effects of which two rare complications have been mentioned in this report. Though they cause devastating sequaelae if not treated , a simple wedge resection can ameliorate the symptoms and prevent the patient from life threatening complications. Hence sound knowledge regarding the behaviour of rare giant tumors and their remedies must be known to a surgeon.

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