Management of Giant Hepatic Hemangiomas-Case Series and review of literature

Amarjothi J M V, Naganath Babu O L, Villalan R, Jeyasudhahar J
Department of Surgical gastroenterology, Madras Medical College,
Rajiv Gandhi Government General Hospital, Chennai, Tamilnadu, India
Corresponding Author: Amarjothi J M V

Abstract

Giant hemangiomas (GH) are benign liver tumors which are more than 4 cm and can be frequently symptomatic. They may be associated with complications due to local compression, rupture or rare coagulopathy syndromes like Kasabach-Merritt Syndrome (KMS), which are indications for surgery. Surgery can be either enucleation or hepatic resection where enucleation is the preferred surgery of choice and hepatic resection reserved for indications like complete lobe involvement by the tumor. Enucleation the procedure of choice and is safer, quicker and associated with less morbidity than liver resection with certain exceptions like total replacement of a lobe.

METHODOLOGY

All cases of operated giant hepatic hemangiomas were retrospectively analysed and statistical analysis done using unpaired t test for test of significance. From 2007-2017, 17 cases of giant hemangiomas (M:F::1:16) were operated. The mean age of the patients was 41.6 years (22-60 years). The main symptoms included abdominal pain (N=16, 94.1%), and abdominal mass (N=14, 82.3%). Surgical removal of hepatic hemangiomas was performed due to intractable symptoms (12 cases), size increase (1 case), and liver failure (1 case) and uncertain diagnosis (3 cases). In all, 3 patients underwent enucleation of hemangiomas, while the remaining 14 underwent anatomical resection. The surgeries included right hepatectomy in 7, left hepatectomy in 3 and left lateral segmentectomy in 4 cases. One patient underwent preoperative angioboomobilisation as the tumor was very large (5.2 kg on post op measurement) and then followed by sorafenib. Unfortunately, patient did not tolerate sorafenib treatment and hence had to be taken for surgery expeditiously. One patient had features of KMS (Kasselbach-Merritt Syndrome) and had to be managed for the same.

The Pringle maneuver was done in 2 cases. Parenchymal transection was done using a combination of kellyclysis and diathermy in most cases. The mean blood loss and operative time for patients undergoing enucleation was 500 ml (range, 300-600 ml) and 146 min (range, 140-150 min) which was statistically less than that for patients undergoing anatomical resection at 746 ml (300-2000ml) and 193 min (140-420 min) (p<0.05). The mean hospital stay was also less for those undergoing enucleation (10.3 days, range 10-11 days) than anatomical resection (12.7 days, range 7-18 days) (p<0.05).

We observed one perioperative mortality inpatient with liver steatosis who had post operative liver failure, and two cases of major morbidity (bile leaks not requiring reoperation). All patients were completely cured of their symptoms and remain alive and in good health, without longterm complications, after a follow-up period ranging from 1 month to 10 years.

Key Words: Hemangioma, hepatectomy, Kasabach-Merritt, enucleation

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I. Introduction

Hepatic Hemangiomas are seen with prevalence ranging from 3% to 20% based on various autopsy series (1, 2,) and estimated incidence in population range from 0.6 to 7% [2] which is predominantly among females (3:1) [1] in the fourth and fifth decade.

The majority of hepatic hemangiomas are asymptomatic and diagnosed incidentally. Giant hemangiomas (GH) are defined as tumors larger than 4 cm which is taken as the limit as symptoms rarely appear unless the tumor exceeds the size of 4 cm (3,4). However it is thought the definition of giant hemangioma should be changed to a minimum size of 10 cm as lesions below this size limit are less likely to be symptomatic and those (< 5 cm) are less characteristic on imaging. Patients with a giant hepatic adenoma (GH) may have inflammatory features and a triad in elevated erythrocyte sedimentation rate (ESR), thrombocytosis and reversible hyperfibrinogenemia [6]. About 40% of hepatic adenoma exhibit growth with
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The symptomatic patient should be evaluated to rule out additional intra-abdominal diseases which may be seen in 42% of symptomatic hepatic hemangiomas [5], and in another study, 50% of GH patients remained symptomatic after surgery [6]. Symptoms commonly seen in GH include abdominal fullness or pain (due to haemorrhage within the hemangioma or in abdominal cavity), jaundice, Gastric Outlet Obstruction (GOO), hepatocellular carcinoma due to local compression, and cardiac failure from massive arterial-venous shunt [9,10]. Conventional grey-scale ultrasound is the most common first line investigation and CECT scan confirms the diagnosis in most patients. In case of equivocal CT, MR scan, with high sensitivity (98.1%) and specificity (92.98%) [13] can be used to diagnose hepatic GH as characteristic hyperintense lesion in the T2 phase with high accuracy. However, large GH may appear heterogenous further causing confusion in diagnosis [14]. It is reported that the diagnostic accuracy using a combination of MR scan and Tc99m-labelled red blood cell scan, increases the diagnostic accuracy further to 90–95% [15]. Conventional angiography shows typical “cotton wool” or “snowy tree” appearance after contrast injection. Still, a proportion of cases are inconclusive which may necessitate surgery. It is now agreed that liver FNAC can be done due to low incidence of complications and are done when radiologic study result and alpha fetoprotein testing are equivocal [16].

Conservative management and serial surveillance is preferred for vast majority of patients [17] as the complications rates of observation when compared with operative management is similar. Therefore, preoperative treatment of liver hemangiomata is indicated in symptomatic cases, or increase in size or complications and uncertain diagnosis due to giant hemangiomas like rupture and KMS and uncertain diagnosis in unique situations. In most studies, the most common indication for surgery was the presence of symptoms (18,19). Because the possibility of malignant transformation is so rare, it is unlikely to be a cause for surgery [20].

Described in 1940 [2], Kasabach-Merritt syndrome is a thrombocytopenic purpura characterised by profound thrombocytopenia, consumptive coagulopathy and intravascular haemolysis suggestive of disseminated intravascular coagulation (DIC), thought to be due to platelet activation when in contact with the abnormal endothelium of the hemangiomaleading to shearing stress and coagulopathy, which is reversed by resection of the tumour. In a study of 97 cases of symptomatic GH, 52.6% had nonsprontaneous rupture and was more frequent in patients less than 40 years with mean size of 11.2 cm (range 1-37 cm). Though massive bleeding occurred in 90.7%, overall mortality has come down ~35% [21]. Immediate surgical procedure is mandatory in unstable patients with a ruptured giant hemangiomata which can be treated with surgical enucleation and ancillary procedures like intermittent inflow vascular occlusion and temporary perihepatic packing [22].

Microscopically, HA consist of mostly dilated, disorganized blood vessels, separated by fibrous connective tissue with a variable fibrous tissue. 80% of hepatic HA are of the cavernous type which are larger than the less common capillary type and are more symptomatic [23].

Radiological procedures like transcatheter arterial embolization (TACE) are increasingly used in the treatment either as a preoperative temporising measure before parenchyma preserving surgery instead of massive resection or as definitive treatment. [24] Embolization should be selective [25] and the vascular interstices within the hemangiomata and the feeding arteriostems are obliterated with gel-foam or polyvinyl alcohol particles with the feeding artery by steel coils in some cases [26]. The most common complications of embolization are self limiting and include pain, pyrexia, leukocytosis, and anaemia. Postembolization pain though described frequently is rare [25]. TAE should be used in GH when the hepatic hemangiomata increases in size, or is symptomatic or features like hemangiomata Kasabach-Merritt syndrome, compression occur and there is high surgical risk [27].

The most preferred option for symptomatic giant hepatic hemangiomas is surgery. Several studies reported high morbidity of 10-27% and mortality of 2% after resection or enucleation of the hepatic HA [27,28]. Though Enucleation is safer, quicker and is associated with less recurrence than liver resection where recurrence is thought to be due to increased release of growth factors due to stimulus of initial resection [30]. Resection is still ideal for patients with uncertain diagnosis or total replacement of a lobe. Though enucleation and resection of lobe can be performed irrespective of tumour size, enucleation generally has shorter mean operative time, significantly shorter duration of hospital stay, decreased blood loss and bile leak as the plane of enucleation is a blood vessel - bile duct free interface between the liver and tumour [31].

Enucleation can also be applied successfully to large or centrally located lesions with the Pringle manoeuvre at appropriate intervals [32]. In a study of complications after surgery for GH, most complications were grade I, and both resection and enucleation are relatively safe with an acceptable complication rates and mortality rates (33,34) and are related to proximity to major vascular structures than size. In a long term study of 33 enucleation patients, surgery was successful in complete symptom control in 88
Laparoscopic resection is feasible in giant liver hemangioma especially in pedunculated GH in inferiorsegmentos of the liver. An array of thermal transection devices can be used as in our case series. Liver transplantation from both deceased and living donors is treatment of last resort for a very limited group especially unresectable lesions, multiple bilobar involvement and hepatic hilum involvement. (36,37)

Non operative pharmacological treatment has also been considered to shrink the tumor before surgery but their role is controversial. Propranolol, an oral non-selective beta blocker is effective for proliferative infantile HA [49], which may be due to multiple mechanisms like vasoconstriction, induction of apoptosis, down-regulation of angiogenic factors such as vascular endothelial growth factor (VEGF) and basic fibroblast growth matrix metalloproteinase 9 inhibitor (38,39). Very large GH can be treated by sorafenib for tumor shrinkage. Significant tumor shrinkage may occur as seen in one of our cases also. However, this tumor shrinkage must be counterbalanced by issues of non compliance and debilitating adverse effects which require treatment cessation. (40). It is still not clear whether the tumor shrinkage is due to the VEGF inhibiting effect of sorafenib.

Conclusion
Surgical approaches are the best for the management of symptomatic giant hemangiomas where the exact choice of surgery is further influenced by the involvement of liver and certainty of diagnosis. Non-symptomatic hemangiomas are best followed by surveillance. Liver transplant may be a last resort for a certain subset of patients. Non-pharmacological approaches have so far been not very promising in the management of giant hemangiomas.

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