Giant Right Liver Hemangioma associated with Kasabach-Merritt Syndrome in an Adult Patient

Case Study

Introduction: Liver hemangiomas are often asymptomatic and diagnosed incidentally. Kasabach-Merritt syndrome (KMS) or consumptive coagulopathy is a rare but life-threatening complication of liver hemangioma occurring during observation. Surgery is an appropriate treatment option in such condition and coagulation usually returns to normal after surgical excision. We herein report a case of giant right liver hemangioma with Kasabach-Merritt syndrome treated surgically with literature review.

Case Presentation: A 36 –year old woman with a giant liver hemangioma (20 cm) discovered three years ago, who presented to emergency department for pallor and fatigability and no abnormalities were found on physical examination. After excluding hematologic diseases, a Kasabach-Merritt syndrome associated with giant liver hemangioma had been retained.Csoagulation disorders returned to normal after successful surgical resection of lesion by performing a right hepatectomy.

Conclusion: Resection is an appropriate and effective surgical procedure to treat giant liver hemangioma associated with Kasabach-Merritt syndrome.

Keywords: giant liver hemangioma,consumptive coagulopathy , surgical resection

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Introduction

Hemangiomas are the most common benign tumor of liver. Most liver hemangiomas are asymptomatic, small (< 4cm) and diagnosed incidentally [1]. A liver hemangioma is qualified giant when it has a diameter greater than 10 cm. Asymptomatic liver hemangioma is managed conservatively. However symptomatic or complicated lesion justified the surgical management [2,3].Kasabach-Merritt syndrome is a rare complication of liver hemangioma and it presents as hemolytic anemia, thrombocytopenia, prolonged prothrombin time, and hypofibrinogenemia. Surgical treatment is an appropriate therapeutic option for such condition and coagulation usually returns to normal after surgical removal .We report a case of giant right liver hemangioma associated with Kasabach-Merritt syndrome treated surgically with literature review.

Case Presentation

A 36 –year old woman presented to emergency department for pallor and fatigability. A giant liver hemangioma was discovered incidentally during pregnancy three years ago that it was managed conservatively.At admission, the patient was pale but not icteric and no abnormalities were found on the physical examination. Laboratory testing revealed that blood count and liver function tests results were WBC: 2.99. 10^9 /L (4.0–10.0$10^9$ /L),Hemoglobin : 8.2 g/L (115–150 g/L),Platelets : 80.000/mm^3(110–320.) ALT : 18 m/L (0–40 m/L), AST: 21 m/L (0–42 m/L), ALP: 56 m/L (40–150) , GGT: 35 m/L (0–52 m/L),TB : 9.7mmol/L (5.0–21.0mmol/L) , DB: 4.8mmol/L (0.0–7.0mmol/L) , Fibrinogen :1.83, g/L (2.00–4.00, g/L , INR : 1.54 (0.85–1.50) , Prothrombin time : 18,2 sec (11–15). Hepatitis B virus and hepatitis C virus markers were negative, and α-fetoprotein level was 8 ng/dL(0–10 ng/dL).

As showed on Computed Tomography Scan, the lesion occupied almost all liver segments 5, 6, 7 and 8 and measuring approximately 20 x 12 x 8 cm without vessel compression (Fig. 1) .After excluding hematologic diseases such as hemolytic anemia, hemolytic uremic syndrome, , systemic inflammatory response syndrome and basing on laboratory results, a Kasabach-Merritt syndrome associated with giant liver hemangioma had been retained.

The hematologic abnormalities had been corrected before surgery by using packed red blood cell, platelet concentrate and fresh frozen plasma. The operative exploration found a huge reddish pink tumor with thin walls and occupying almost all the right hemiliver. The intra-operative decision was to perform a right hepatectomy. The right liver portal vein ant artery was clamped after liver helium dissection (fig.2) and parenchymal transection was performed using an ultrasonic dissection device. The tumor was dissected away from the the inferior vena cava (IVC) after exposure of the antero-medial surface of the IVC and ligation of several short hepatic veins. The right hepatic vein, the right portal vein and artery were the last vascular elements to be divided. The tumor had a length of 20 cm approximately (fig.3).The patient developed a right bloody pleural effusion which was resolved after thoracic drainage maintained during five days. Histological examination of operative specimen revealed a cavernous hemangioma. The coagulation and hematologic abnormalities returned to normal value 3 weeks after surgery (Table.1).

Discussion

Hemangiomas are one of the most common benign tumors of liver.According to their size, hepatic hemangiomas are classified into 3 types: small (<5 cm), large (5–10 cm), and giant

(>10 cm). Observation is justified in asymptomatic lesion and surgery is indicated in the presence of complications [4]. Consumptive coagulopathy or Kasabach-Merritt syndrome (KMS), described firstly by Kasabach and Merritt in 1940, is a rare and severe coagulation disorder associated with vascular malformations [5].The Kasabach-Merritt syndrome is characterized by thrombocytopenia, hemolytic anemia, and consumptive coagulopathy [6]. Surgical management remains an effective and curative treatment for complicated or symptomatic liver hemangioma [7]. Our patient underwent a right hepatic liver resection using a hanging manoeuvre to avoid difficulties and minimize risk of bleeding during liver mobilisation. Transfusion of three units of red blood cells was required because of preexistent anemia and operative blood loss (300 ml). Risk of operative bleeding is likely to be more related to hemangioma size (> 20 cm) [7]. Compression of major vessels surrounding the lesion may expose to high risk of uncontrolled severe bleeding and blood loss during operation. So, cell saver system is highly recommended to decrease blood transfusion rate in these patients. The liver resection procedure is more likely recommended to remove liver hemangioma associated with KMS because hemangioma often has an extremely greater size (>20cm) making liver mobilisation more difficult with high risk of bleeding. So preligation of both artery and portal vein decreases lesion size, facilitates liver mobilisation and thus reduces risk of bleeding. In addition, an extremely giant hemangioma can occupy entirely a hemiliver or more and performing anatomic liver resection will not lead to substantial loss of healthy liver parenchyma.Although surgery remains the radical treatment of liver hemangioma , other therapeutic options including transcatheter arterial embolization (TAE) and radiofrequency ablation can especially be considered in patients with high surgical risk [8-10].These therapies can be performed prior to surgery in order to reduce tumor size of extremely giant lesion [8-10**].** As reported, liver transplantation had the same effects as surgery in the treatment of Kasabach- Merritt syndrome associated with liver hemangioma [11,12]. However, liver donor is rare, and patient needs to take an immunosuppressive treatment for a long-term period after transplantation. Since 2008, oral propanolol have been used to treat hepatic hemangioma and it was largely used in combination with steroids in infant [13,14]. As demonstrated by published study results, the efficacy and safety of this therapy as a first treatment , had an positive impact on changing the classical therapeutic indications particularly in diffuse hepatic lesion (type 3) by obviating liver transplantation for many of these patients [14,15] . therefore , the usage of propanolol alone or in combination with steroids as first treatment line in infantile liver hemangioma resulted in decreasing the indications of surgical treatment options . On other hand and in adult patient , propranolol has

been used as a first therapeutic option for liver hemangioma in selective few patients with good results. However randomized prospective studies are highly recommended to evaluate the results and clarify the appropriate use of this agent in such condition [16,17]

Conclusion

In summary ,Kasabach-Merritt syndrome is an uncommon complication of liver hemangioma occurred in adult patient. Surgery is an effective therapeutic option and hematological abnormalities and coagulation disorders returned to normal values after surgical resection .In such condition; liver anatomic resection is a safer surgical procedure.

Disclaimer regarding Consent/Ethical Approval:

As per university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

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Table 1: per and postoperative results of blood tests

parameter	Preoperative value	Postoperative value(3 weeks)	Postoperative value(6weeks)
WBC	2.99. 10^9 /L (4.0–10.0$10^9$ /L)	4.30. 10^9 /L	6.80. 10^9 /L
Hemoglobin	8.2 g/L (115–150 g/L)	11.3 g/L	13.2 g/L
Platelets	80.000/mm^3(110–320.)	130.000/mm^3	240.000/mm^3
Fibrinogen	1.83, g/L (2.00–4.00, g/L	2.13, g/L	3.22, g/L
INR	1.54 (0.85–1.50)	1.35	1.10
Prothrombin time	18,2 sec (11–15).	15,1 sec	12,3 sec

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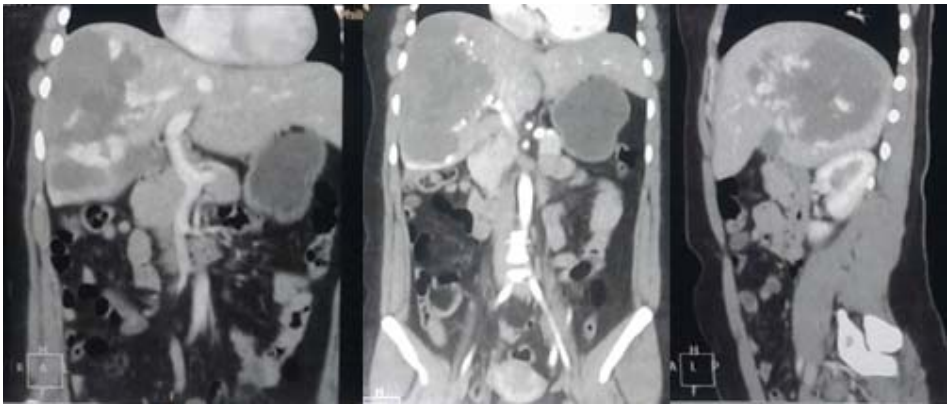

FIG.1: CT scan images of a patient with Kasabach-Merritt syndrome associated with giant liver hemangioma

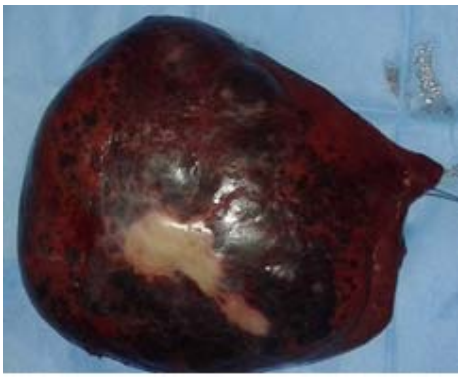

Fig.3: resected hemangioma

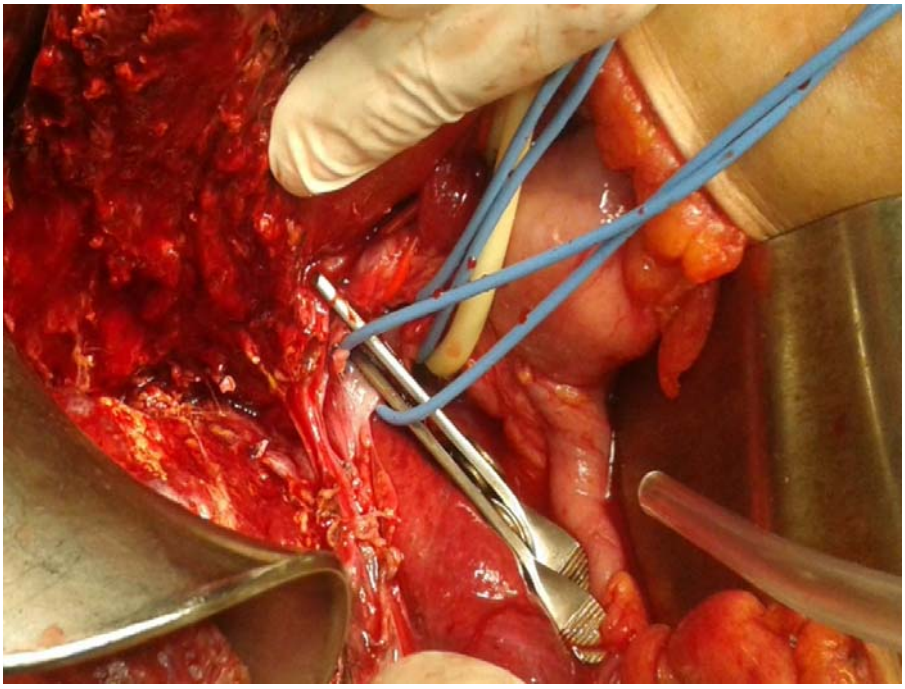

Fig.2: intraoperative view of clamped right portal vein and right hepatic artery