

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

### **Abstract**

**Background:** Liver hemangiomas are often asymptomatic and diagnosed incidentally. Kasabach-Merritt syndrome (KMS) or consumptive coagulopathy is a rare but life-threatening complication of giant 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) which was incidentally discovered during pregnancy three years ago . The patient 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 excision of lesion by performing a right hepatectomy.

**Conclusion:** surgical excision is an appropriate and good surgical treatment option for Kasabach-Merritt syndrome complicating a giant liver hemangioma.

**Keywords:** giant liver hemangioma, consumptive coagulopathy , surgical excision

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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 5cm. asymptomatic giant liver hemangioma is managed conservatively. However symptomatic or complicated giant hemangiomas justified the indication of surgery [2-5 ]. Consumptive coagulopathy or Kasabach-Merritt syndrome is a rare complication of giant liver hemangioma .The most reported liver hemangiomas associated with Kasabach-Merritt syndrome are case report. Kasabach-Merritt syndrome presents as hemolytic anemia, thrombocytopenia, prolonged prothrombin time, and hypofibrinogenemia. Surgery is an indication for such condition and coagulation usually

returns to normal after surgical excision .We report a case of giant right liver hemangioma with Kasabach-Merritt syndrome treated surgically with literature review.

### Case Presentation

A 36 –year-old woman without history of disease, a giant liver hemangioma was incidentally discovered by ultrasonography examination performed during pregnancy three years prior to this presentation. The tumor size was about 20 cm and as it was an asymptomatic lesion, a conservative management was indicated. The patient presented to emergency department for pallor and fatigability. 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:  $2.99 \times 10^9 /L$  ( $4.0\text{--}10.10^9 /L$ ), 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  $\alpha$ -fetoprotein level was 8 ng/dL( 0–10 ng/dL).After excluding hematologic diseases such as hemolytic anemia, hemolytic uremic syndrome, and 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 using packed red blood cell concentrate, platelet concentrate and fresh frozen plasma. Computed tomography scan showed that lesion occupied almost all of liver segments 5, 6, 7 and 8 and measuring approximately 20 x 12 x 8 cm without vessel compression (Fig. 1) .A right hepatectomy was performed. A huge reddish pink tumor with thin walls was noted. It was highly vascular with easy bleeding. The right liver portal vein and 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 IVC after exposure of the antero-medial surface of the IVC. Several short hepatic veins were divided. Finally, the right hepatic vein, the right portal vein and artery were ligated. The tumor measured 20x10x7 cm ( fig.3 ). Histological examination revealed a cavernous hemangioma.The patient developed a right bloody pleural effusion which was resolved after thoracic drainage.

### Discussion

Hemangiomas are one of the most common benign tumors of liver. A liver Hemangioma is qualified giant when it has a diameter of more than 5 cm. Observation is justified in

asymptomatic lesion. Giant hemangiomas can become symptomatic and even may cause life-threatening complications such as mechanical complications, rupture and coagulopathy [6]. Consumptive coagulopathy or Kasabach-Merritt syndrome ( KMS) , reported firstly by Kasabach and Merritt in 1940 [7], is a rare and severe coagulation disorder associated with vascular malformations [8-10] , and uncommon in adults[11-14]. It is characterized by thrombocytopenia, hemolytic anemia, and consumption coagulopathy [10].In this present case, the patient had thrombocytopenia, prolonged prothrombin time, and hypofibrinogenemia, and severe hemolytic anemia indicating the severe impact of lesion on hematologic system and coagulation. The size of the majority of giant liver hemangioma with KMS reported in literature was greater than 20 cm [14-18].Moreover all reported cases of hemangioma associated with KMS were larger than 10 cm which speculating that consumptive coagulation disorder is closely associated with the tumor size.Surgery remains the only effective curative treatment option for complicated or symptomatic giant hemangioma[11,15,16,17,19].

Our patient underwent a right hepatic liver resection using a hanging manoeuvre to avoid difficulties and minimize risk of bleeding during liver mobilisation. Blood loss amount was 300 ml and blood transfusion was required (3 units of red blood cells ). Risk of hemorrhage during operation is to be more related to hemangioma size (> 20 cm). Also compression of major vessels surrounding the lesion exposes to the risk of uncontrolled severe bleeding and blood loss during operation. In such situation, cell saver system is highly recommended to decrease blood transfusion rate during operation. The patient developed a right bloody pleural effusion which was resolved after thoracic drainage .Furthermore platelets , red blood cells and coagulation returned to normal after surgery.

Giant liver hemangiomas can be safely removed by both resection and enucleation. Substantially, the choice between the two surgical procedures, enucleation or anatomic resection, depends on the location and size of lesion, complication, and the experience and skills of surgeon. Enucleation offers the benefit to be associated with reduced blood loss and transfusion and of lower operative morbidity [20-26]. However Liver resection procedure is more likely recommended to remove giant liver hemangioma associated with KMS because hemangioma often has an extremely grater size (>20cm) posing difficulties to mobilize liver and exposing to high risk bleeding. By contrast, after liver vascular exclusion by unilateral preligation of hepatic artery and portal vein, the lesion would become softer and smaller facilitating liver mobilisation and thus deceasing risk of bleeding. Additionally and in most cases, an extremely giant hemangioma occupied entirely or more than hemiliver and

performing anatomic liver resection will not lead to substantial loss of healthy liver parenchyma. Although surgery is the main treatment of giant liver hemangioma, other therapeutic options including transcatheter arterial embolization (TAE) and radiofrequency ablation can be considered especially for patients with high surgical risk [28-32,34]. These therapies can be performed preoperatively in order to reduce tumor size of extremely giant lesion, and thus decreasing the risk of blood loss and bleeding during surgery [28-32,34].

Good results have been reported with liver transplantation in the treatment of Kasabach-Merritt syndrome associated with giant liver hemangioma. Coagulation and platelets returned to normal values after transplantation [12,16,33]. However, liver donor is rare, and patient needs to take an immunosuppressive treatment for a long-term period after transplantation. The reported results of studies demonstrated that surgery for giant liver hemangioma associated with Kasabach-Merritt syndrome had the same effect as liver transplantation [11, 34]. Corticoids, radiotherapy and embolization alone had been used to treat giant liver hemangioma associated with Kasabach-Merritt syndrome [17,18,35]. The use of oral corticoids (prednisone) and radiotherapy were associated with no significant improvement in coagulation or platelets [17,18]. A short-term improvement in coagulation and platelets was showed after embolization. However, Further results of this treatment method should be evaluated to confirm long-term efficacy because the lesion is still existing [35].

In summary, Kasabach-Merritt syndrome is an uncommon complication of giant liver hemangioma occurred in adult patient. Hematological abnormalities and coagulation disorders returned to normal values after surgical excision. In such condition, liver anatomic resection may be a better surgical procedure than enucleation and liver transplantation for these patients.

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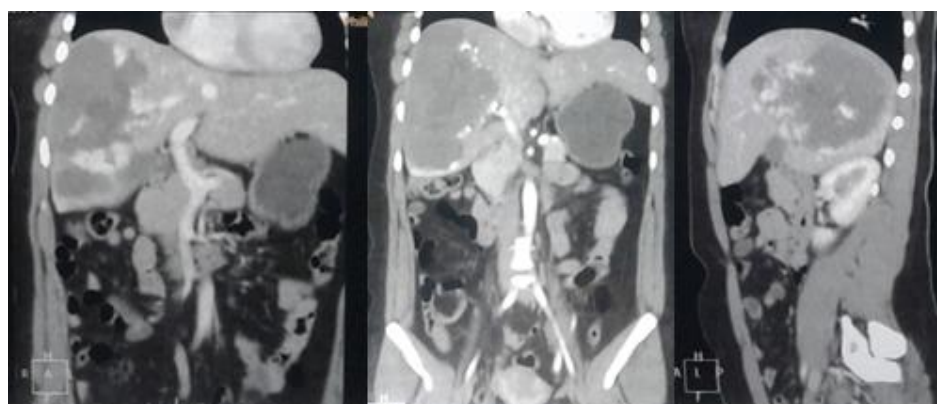
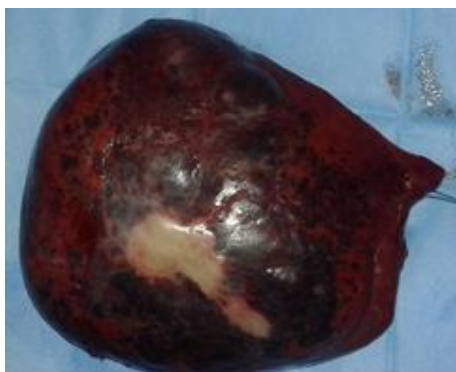


FIG.1: CT scan images of a patient with Kasabach-Merritt syndrome associated with giant liver hemangioma

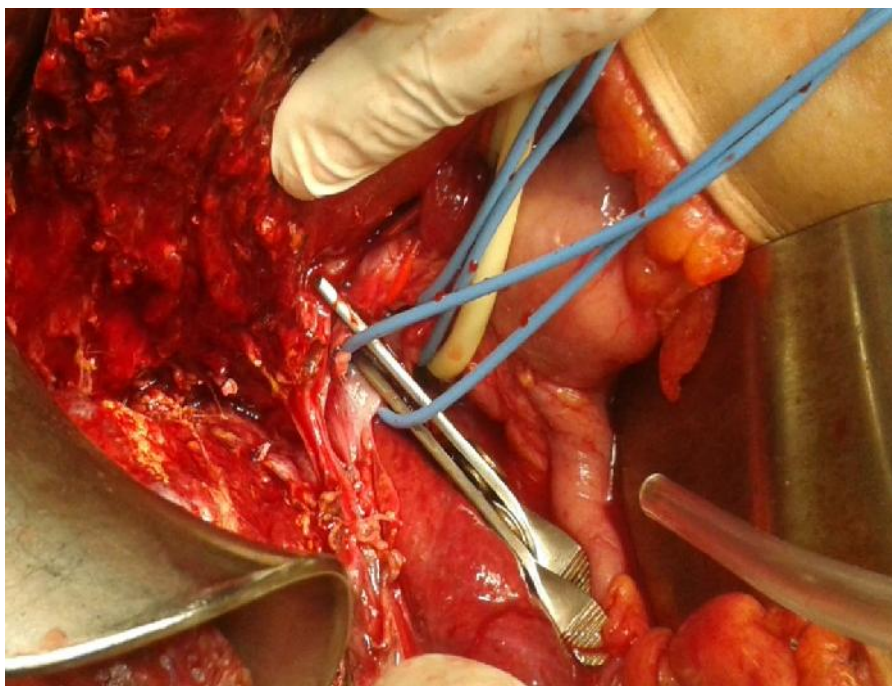
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235 Fig.3: resected hemangioma

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