

# Spontaneous rupture of a giant hemangioma of the liver



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## Introduction

Hemangiomas are the most frequent benign tumors of the liver (2-7%) and are usually asymptomatic [1, 2] and incidental [3].

Most hemangiomas are less than 4 cm in diameter; larger hemangiomas are defined as giant [4].

Symptoms (abdominal pain and fullness) are mostly seen in lesions larger than 4-5cm and are due to a rapid enlargement or to an acute thrombosis with inflammation. Clinically they can be identified by a palpable upper abdominal mass. Rare but severe complications are: Kasabach-Merritt syndrome [5, 6], typically found in children, jaundice [7] and rupture. This last one is the most severe and can occur as a consequences of trauma, biopsy or spontaneously [8, 9]. Spontaneous rupture and intraperitoneal bleeding [10] occur in 1-4% of hemangiomas and have been described in about 30 [11, 12] cases in the international literature with a high mortality (about 60%).

Although the risk of rupture of a cavernous hemangioma is minimal, this complication is the principal indication for surgery.

The first description of a ruptured hemangioma in an autopic case has to be attributed to Van Haefen in 1898 [13]. Karp reported the first attempt of resection but the patient died on the tenth post-operative day. Finally in 1935 Tinker reported the first case successful surgically treated [13, 14].

## Abstract

*Hemangiomas are frequent benign tumors of the liver. Symptoms (abdominal pain and fullness) are mostly seen in giant lesions. Rupture is the most severe complication, can occur spontaneously, with intraperitoneal bleeding, in 1-4 % of hemangiomas and has been described in about 30 cases in the international literature with a high mortality (about 60%).*

*This complication is the principal indication for surgery. Although spiral CAT scan and MR are actually the most efficacious imaging methods for study of liver hemangiomas, after Echography, emergency techniques that allows a simultaneous therapeutic approach – as is angiography – are preferable. Trans-arterial embolization (TAE) is in fact useful to stop bleeding and then to perform a safer surgery. A successful embolization can delay the surgical resection of the hemangioma for the time necessary to recover from the hemodynamic distress.*

*Aside from the success of angiographic approach, surgery remains mandatory, effective in stopping the bleeding and in preventing re-bleeding or other complications of TAE such as abscess, fever, etc..*

*Intraoperative echography currently is the best method to identify vasculo- biliary anatomy and to perform a correct resection.*

*The absence of risk factors for spontaneous rupture of liver hemangiomas, makes this event unpredictable. The best treatment for non-ruptured hemangiomas is still controversial but surgery is usually limited to symptomatic tumors larger than 10 cm.*

**Key words:** Hemangiomas, liver, complications, angiography, surgery.

Rupture of giant hemangioma occurs at all ages with a high incidence in newborns (28%) [11].

A relationship between symptoms and rupture is not evident.

An accurate and timely diagnosis of this condition entails a multidisciplinary approach.

## Material and methods

A 43 years old white male, cigarette smoker, with an history of duodenal ulcer, gastro-esophageal reflux disease,

gallstones and colonic diverticular disease, presented to our outpatients Clinic complaining of constant epigastric discomfort, fullness, and upper abdominal pain exacerbated by food for at least three months.

Physical examination revealed a palpable liver mass extended to the right flank.

Tumor markers were absent or normal. Viral hepatitis markers were absent too. Also des-g-carbossiprothrombin was normal as were the routine liver function tests.

Ultrasound (US) (Fig. 1) demonstrated a large hepatic tumor (18 cm diameter) of the right lobe. The medial surface of the mass was contiguous to the middle hepatic vein. The presence of hyper- and hypo-echoic areas were suggestive of a giant cavernous hemangioma with large areas of necrosis and internal hemorrhage. The left lobe was hypertrophic and occupied the entire left hypocondrium.

The patient was placed on the waiting list for hospitalization, further evaluation and surgery, however a week later he came back with signs and symptoms of acute abdomen.

The patient was in acute distress, with severe tenderness of the abdominal wall.

Laboratory findings showed RBC count of  $2.8 \times 10^5$  /mm<sup>3</sup>.

WBC count of  $12 \times 10^3$ , Hb 7.5 g/dl, Ht 24%, Arterial pressure was 105/70 mmHg, pulse rate 106 /min, CVP 6 cm H<sub>2</sub>O, pCO<sub>2</sub> 42 and p O<sub>2</sub> 58.

Abdominal X-rays showed some small air-fluid levels but no free intra-peritoneal gas.

US showed that the mass was larger in diameter (22 cm) compared to the prior week before as were the internal necrosis areas; the left liver and the spleen were normal. Also about 1-liter of free fluid was detected in the abdomen.

After intravenous resuscitation and appropriate type and crossing, the patient underwent angiography (Digital

Subtraction Angiography – DSA-) that demonstrated the liver tumor being the source of the hemorrhage, with a vascular pattern suggesting an hemangioma (Fig. 2). Angiography also showed absence of cirrhosis in the left lobe of the liver, and the distal-flow was normal.

Trans - catheter Arterial Embolization (TAE) was not possible because of the presence of an abnormal arterial vascular pattern (right hepatic artery originated from the Superior Mesenteric Artery).

The patient was taken to the Operating Room and a bilateral subcostal laparotomy was carried out. One liter of blood and some clots were aspirated from the abdominal cavity. The right lobe was almost entirely occupied by a large soft bluish lumpy tumor, bleeding from its inferior surface. The Pringle's manoeuvre (58 min duration, interrupted for 5 minutes after the first 30 min.) stopped the bleeding from the inferior surface of the hemangioma. Intra-operative US allowed a good definition of the limits of the neoplasm.

A standard right hepatectomy including part of the first segment was performed. The Kelly clamp-fracture of the parenchyma was carried out after ligation of right branches of portal vein and hepatic artery. After the resection of the dorsal attachment of the vena cava, the right hepatic vein was divided extraparenchymally by Endo-GIA stapling. No significant bleeding or other operative complications were observed. One Penrose drain was placed in the abdomen before closing.

The postoperative course was unremarkable, and the patient was discharged on the twelfth postoperative day.

The patient returned to his normal working activity. The operative specimen grossly presented a large cavernous hemangioma (Fig. 3) and the specimen's angiography showed the site of the bleeding (Fig. 4). Histologically the neoplasm showed areas of fibrosis and wide hemorrhagic areas, especially in the lower surface.



Fig. 1: Ultrasound: large hepatic cavernous hemangioma (18 cm diameter), with large areas of necrosis. The medial surface of the mass is contiguous to the middle hepatic vein. The left lobe is hypertrophic and occupies all the left hypocondrium.



Fig. 2: DSA (Digital Subtraction Angiography) of celiac artery: the right hepatic artery is increased in caliber and branches of 3<sup>rd</sup> and 4<sup>th</sup> order are partially stretched. In the surroundings, clusters of vascular knots are present.

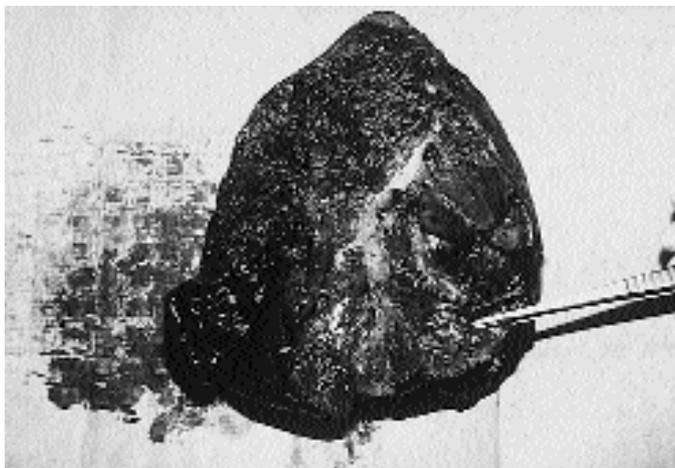


Fig. 3: The operative specimen grossly presented a large tender bluish cavernous hemangioma that occupied almost entirely the right lobe.

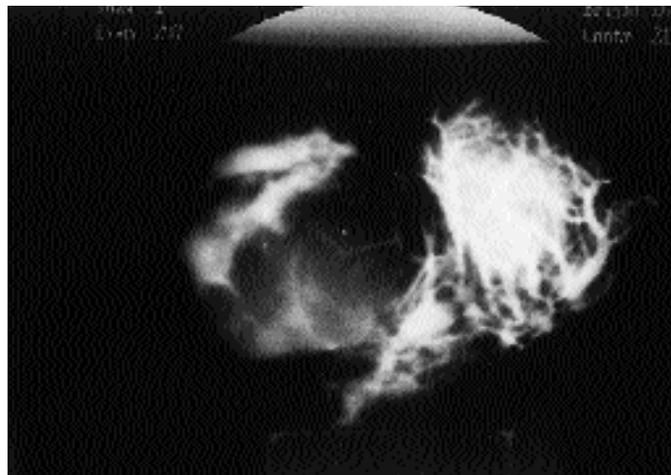


Fig. 4: Specimen arteriography: large bleeding area with a very large gap on the profile of the organ.

Chronic inflammation of the gallbladder was present. The 20 month follow up did not show recurrence or symptoms.

### Discussion

Intraperitoneal bleeding from the liver is a severe event. It is frequently due to the rupture of a hepatocellular carcinoma in cirrhosis but very rarely could be caused by the rupture of a hemangioma, this is usually observed after needle or open biopsies or after traumas; cases of rupture in patients with hypertension or on anticoagulation therapy have been reported in literature [12]. The rarest form is spontaneous rupture: 32 cases have been reported in literature. In the 28 cases reported by Yamamoto this event occurred frequently in newborns (8/28), probably due to birth traumas; in adults it is more frequent in young females than in males (1:6).

The mechanism of the rupture as well as the etiology of the hemangioma is unclear. All Authors agree with the benign nature of the lesion; enlargement is usually due to intra-tumoral bleeding without a proliferative pattern.

The literature demonstrated that the risk of rupture is higher when the size of the hemangioma is greater than 10 cm and is localized at the inferior surface of the liver or involving nearly all the right lobe.

The spontaneous rupture of the hemangioma is linked with high mortality rates (60-70%) so that high priority must be given to signs and symptoms in patients with risk of rupture.

Although giant hemangiomas are usually symptomatic – upper abdominal pain, vomiting, weight loss, jaundice and liver failure, and rarely associated with fever, chills or Kasabach – Merritt syndrome – none of these symptoms are clearly predictive of rupture.

Rupture declares itself with signs and symptoms of acute abdomen – upper abdominal pain and shock –, but sometimes the clinical presentation can be subtle.

US is the first diagnostic procedure and allows the differential diagnosis with HCC ruptured, other liver (metastasis, trauma, etc.) or abdominal bleeding lesions.

In fact the diagnosis is not easy. Yamamoto reported a preoperative misdiagnosis in 13 cases in which the major symptom was abdominal pain: peritonitis due to appendicitis or perforated ulcer or other causes in 6 patients (46.2%); ectopic pregnancy in 4 (30.8%); intra-abdominal bleeding in one (7.7%); uterine rupture in one (7.7%). In these series the correct diagnosis was formulated in only one case (7.7%); this data confirm the observation of Schumacker [11, 16].

CT scan is useful for avoiding misleading conclusions when the presence of the hemangioma is not already known [17]. Angiography is indicated when hemangioma is known and it is possible to attempt a trans arterial embolization, (TAE). This technique is life saving in critical patients [18] and is very useful in patients with ruptured hemangioma providing stable hemodynamic conditions. In this case angiography can define the vascular supply of the lesion and can achieve hemostasis using TAE [19].

The efficacy of the procedure in these conditions has not been demonstrated because of the small number of cases reported in literature and treated with such technique [11, 20], however in most series TAE is considered an important step in the management of ruptured hemangioma. The advantages of TAE in these circumstances are the definition of the exact extension of the tumor and the control of hemorrhage, but can be complicated by ischemia of the liver or infection. TAE is not always feasible because of abnormal vascular supply,

as in the case we report; in these situations emergency surgery is mandatory.

Also when TAE is successful surgery remains mandatory but is not an emergency procedure. In fact, liver resection must be performed after the complete re-establishment of haemodynamic conditions and before the onset of complications such as re-vascularization and re-bleeding or sepsis. When the patient is emergently taken to operating room before completion of the diagnostic work up, intra-operative US is a useful tool to better define the borders of the neoplasm. US is also useful to guide the surgical resection.

Transversal laparotomy is the approach of choice. Superficial or pedunculate tumors, and the ones confined to the 2nd and 3rd segments can be removed by enucleation without inflow occlusion. Larger or deeply located tumors must be treated by regulated resection along anatomic planes [21].

Vascular exclusion could be accomplished by the Pringle's manoeuvre, but resection of a tumor located in dangerous areas (central or near the vena cava) requires the inferior vena cava to be dissected and encircled with vessel loops as well as the suprahepatic one in preparation for hepatic vascular exclusion.

Monitoring of the patient must be carefully carried out, preferably using a Swan-Ganz catheter.

Postoperative morbidity and mortality are due to complications such as continual bleeding, sub-phrenic abscess, infected hematoma or bilioma and liver failure, and are substantially the same as for major liver surgery.

## Conclusions

The rupture of a giant hemangioma is a non-predictable dramatic event in which surgery is necessary. In all other cases surgery is indicated only if there are symptoms related to the existence of a giant hemangioma or when the mass is rapidly growing.

When the rupture occurs, the management protocol is controversial as well as for non-ruptured giant hemangiomas. Spiral volumetric CT scan and Gadolinium-MRI, which are actually considered the best imaging methods for space-occupying lesions of the liver, have only a minor role in emergency [17].

In emergency US and angiography are preferable.

TAE has been shown to be able to achieve a good hemostasis in a variety of bleeding lesions of the liver – traumas, biopsies, A-V fistulas- and has also been used for the treatment of liver hemangiomas.

TAE, when successful, can obtain, beyond the hemostasis, a reduction in the size of the mass; also if TAE is not successful angiography can be useful to define the limits of the mass as in the case here reported.

The success of the embolization can delay the resection of the hemangioma, but only for the time necessary to recovery from the shock.

Surgery remains mandatory, in fact it is the only method that can prevent re-bleeding.

## Riassunto

Gli emangiomi sono tumori benigni del fegato di frequente riscontro. I sintomi (dolore e tensione addominale) sono osservabili nelle lesioni giganti. La rottura è la complicanza più grave e può presentarsi spontaneamente, con sanguinamento intraperitoneale, in 1-4% degli emangiomi e tale complicanza è stata descritta in circa 30 casi nella letteratura internazionale con una mortalità assai elevata (circa 60%). Questa complicanza rappresenta una indicazione assoluta all'intervento chirurgico. Sebbene la TAC spirale e la RM siano allo stato attuale i mezzi diagnostici per immagini più efficaci per lo studio degli emangiomi epatici dopo l'ecografia, in urgenza, vanno preferite quelle tecniche, come l'angiografia, che consentono anche un approccio terapeutico.

La TAE è infatti molto utile per fermare l'emorragia e consentire un intervento chirurgico più sicuro.

Il successo dell'embolizzazione può dilazionare l'intervento resettivo consentendo di ripristinare l'equilibrio emodinamico del paziente.

Indipendentemente dal successo dell'embolizzazione la chirurgia resettiva rimane mandatoria allo scopo di perfezionare l'emostasi, prevenire un risanguinamento o altre complicanze della TAE come ascessi.

L'ecografia intraoperatoria è attualmente il mezzo migliore per identificare l'anatomia vascolo-biliare ed eseguire una resezione corretta. L'assenza di segni e sintomi predittivi di rottura spontanea di un emangioma epatico rende questa complicanza imprevedibile.

Il trattamento degli emangiomi epatici non complicati è tutt'oggi oggetto di controversie sebbene la chirurgia trovi ampia indicazione in quelli sintomatici e in quelli di dimensioni superiori ai 10 cm.

Parole chiave: emangioma, fegato, complicanze, angiografia chirurgia.

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Commento

Commentary

Prof. Ercole CIRINO

*Originale e particolarmente interessante è, a mio avviso, il lavoro sulla rottura dell'emangioma gigante sia per la rarità dell'evento che per le problematiche di ordine diagnostico e terapeutico che tale neoplasia epatica ancor oggi offre al chirurgo specie in urgenza.*

*Il tema è stato affrontato rigorosamente dagli autori con dovizia di particolari e immagini iconografiche di ottima qualità. Questo è certamente un capitolo complesso in cui è indispensabile un approccio interdisciplinare.*

*Utile quindi proporre ancora una volta in letteratura questo argomento della chirurgia generale e di urgenza.*

*The paper regarding the giant hemangioma of the liver is very original and interesting for the rarity of this complications as well as the problems that the benign liver neoplasm still present to the surgeon in order to the gold standard of the diagnostic and surgical procedure, specially in emergency. The approach of the authors to this argument has been very good with good figures and scientifically rigorous.*

*This chapter is complex and the interdisciplinary approach is mandatory.*

*Very useful to present to international literature again this aspect of the general and emergency surgery.*

*Autore corrispondente:*

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