

Spontaneous rupture of a hepatic epithelioid angiomyolipoma: damage control surgery. A case report

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SUMMARY: Spontaneous rupture of a hepatic epithelioid angiomyolipoma: damage control surgery. A case report.

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Background. *Angiomyolipoma (AML) is a rare mesenchymal tumor composed by blood vessels, adipose tissue and smooth muscle cells in variable proportions. Although it is most often diagnosed in the kidney, this tumor may originate from any part of the liver. It is often misdiagnosed as hepatocellular carcinoma (HCC) or other benign liver tumor. We describe a case of spontaneous rupture of hepatic angiomyolipoma in a young woman, with evidence of internal hemorrhage and hemoperitoneum.*

Case report. *Liver tumor rupture is a rare but real surgical emergency. In our case it has been managed according to the trauma principles of the damage control surgery. At the time of the observation, the patient presented an instable condition, so the decision-making was oriented toward a less invasive first step of liver packing instead of a more aggressive intervention such as one shot hepatic resection.*

Conclusion. *Damage control surgery with deep parenchymal sutures of the liver and pro-coagulant tissue adhesives packing abbreviates surgical time before the development of critical and irreversible physiological endpoints and permits a more confident second time surgery. This surgical management concept helps to reduce the mortality rate and the incidence of complications not only in traumatic liver damages, it works very well in spontaneous liver ruptures as well.*

KEY WORDS: Angiomyolipoma - Damage control surgery - Liver lobectomy

Introduction

Angiomyolipoma (AML) is a rare mesenchyme-derived neoplasm that is primarily composed of adipose tissue, smooth muscle, and abnormal blood vessels, in variable proportion and is part of the perivascular epithelioid cell (PEC) tumor family known as PEComas (1, 2). Typical AML morphology have been reported in several sites outside the kidney and the liver, including lung, retroperitoneum, uterus, ovary, vagina, penis, spinal cord, bladder, bone, heart, nasal cavity, skin and colon. The epithelioid variant of AML is composed of a predominant or exclusive population of epithelioid cells. Since its first description by Ishak in 1976, more than 200 hepatic AMLs (HAMLs) have been reported, but very few reports of epithelioid AML of the liver have been found in the literature (2, 3).

This wide spectrum of morphological and histological appearance makes it difficult for a definitive diagnosis, therefore it is often misdiagnosed as hepatocellular carcinoma (HCC) or other benign liver tumors (1). Hepatic adenoma might be the most difficult benign tumor to distinguish from angiomyolipoma (4, 5). The tumor can be found in both males and females, with a female preponderance and about 5% to 13% of cases are associated with tuberous sclerosis. In these cases the hepatic lesions are frequently multiple and associated with renal angiomyolipomas. Usually HAMLs are solitary tumors. Most patients are asymptomatic and the tumors are often detected incidentally. It seems that hepatic AML is not associated with chronic viral liver disease.

The treatment for HAML remains controversial, because in the past, it has been considered as an entirely benign lesion and several authors advocated a conservative approach in the treatment of HAML (6-8).

However, dangerous complications such as late recurrence, malignant transformation, spontaneous rupture, disseminated intravascular coagulopathy and Budd Chiari syndrome have been reported in HAML (9). Therefore conservative management with close follow up is suggested in asymptomatic patients with small tumors (size < 5 cm), good

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compliance, negative viral hepatitis serology and when HAML is proven through needle biopsy (8).

Case report

A 25 years-old female came to the Emergency Room for sudden onset of abdominal upper-quadrant pain and hypotension, after two recent syncopal episodes. Physical examination demonstrated pallor, tachycardia, upper abdominal severe pain and tension, however associated with Glasgow Coma Score (GCS) 15. Fever, nausea, vomit, or weight loss were absent. Laboratory findings were suggestive of acute anemia (Hb 6 g/dL) and showed normal liver function parameters. Serum α -fetoprotein, CA19-9, CA125, CA15-3 and carcinoembryonic antigen were negative. Viral hepatitis serology was positive for HBcAb and HBeAb but negative for hepatitis C virus (anti-HCV) antibody. Her medical history was not significant, with no family history of tuberous sclerosis. Abdominal US examination revealed an irregular-shaped, poorly defined heterogeneous area in the left liver lobe. Fluid was detected in the upper abdominal compartments and especially in the sub-phrenic spaces, in the sub-hepatic space, and in the lesser sac. These reports were highly suggestive for a diagnosis of hepatic tumor with suspected pe-

ritoneal blood leakage. Meanwhile the patient's conditions got worse (GCS 10), for this reason she underwent straight to surgery.

A midline laparotomy was performed. We found a left liver lobe hemorrhagic mass with massive hemoperitoneum (2000 cc). At first the hemorrhage control was carried out by manual compression, followed by deep heparorrhaphy and pro-coagulant tissue adhesives placing on the liver surface. After plenty intra-abdominal toilette, one suction tubular drain (28 Fr) was placed in the left sub-hepatic area. Then the patient's abdomen was closed. The frozen section revealed a tumor mainly composed by atypical epithelioid cells. Mitotic figures were absent.

Postoperative contrast-enhanced CT, revealed a large oval-shaped hepatic mass, which measured 86x72 mm, involving the left liver lobe. Following the injection of contrast, the lesion showed heterogeneous enhancement in all arterial, portal, and late phases. The internal structure was inhomogeneous with both soft- and fat-tissue density areas (Figs. 1 and 2). After 48 hours of Intensive Care Unit (ICU) staying, the patient underwent again to surgery for a left-liver lobectomy (Fig. 3).

The postoperative period was uneventful and the dimission of the patient took place 9 days later.

Histology of the surgery specimen revealed a well-circumscribed nodular mass of 9 cm. The cut surface showed a yellow-colored mass. The tumor consisted of adult fat cells, smooth-muscle cells, and vasculature (epithelioid angiomyolipoma). The mitotic index was $< 1/50$ high-power fields and necrotic areas were present. Immunohistoche-



Fig. 1 - Plain CT: large hypo-attenuating lesion completely takes up the left liver.



Fig. 2 - Contrast-enhanced CT: peripheral enhancement in arterial phase with hypodense areas.



Fig. 3 - Surgical specimen of liver left-lobectomy for a 9 cm epithelioid angiomyolipoma.

mically epithelioid cells were positive for HMB-45 and MelanA but negative for S100 protein, Actin, CK(PAN), CK7, Desmin and MIB 1 2%.

Discussion

Spontaneously rupture is a rare and dangerous complication which may occur in fibrolamellar HCC and more rarely in hepatic AML. A tumor rupture followed by hemodynamic instability, is a surgical emergency and its treatment should be based on trauma principles. In this case we followed the damage control principles. The concept of damage control was introduced by Stone et al. in the 1980s and promulgated by Burch et al. in 1992 (10). The damage control surgery includes the first phase of control of hemorrhage, the second phase of resuscitation and stabilization in the intensive care unit for 24 h to 48 h and the third phase of re-exploration and definitive surgery. In the early 1990's the concept of damage control surgery revolutionized the world of trauma and dramatically changed how trauma surgeons operate. The concept focuses on abbreviated initial surgery, placing more emphasis on the body's metabolic responses and less on restoring anatomy to the pre-injury state. These concepts include minimizing time in the operating room, leaving the abdomen open and covered (laparostomy), and early re-warming and resuscitation in the ICU. This method is associated with signi-

ficantly survival advantages because it is directed toward the avoidance of hypothermia, coagulopathy, and acidosis that interact to produce a deteriorating metabolic situation and high mortality. Each of these life-threatening abnormalities exacerbates the others, contributing to spiraling cycle with cellular hypoxia and failure of the coagulation system.

Different surgical techniques allows the control of bleeding in the treatment of a number of liver injuries as well as massive abdominal trauma or hepatic parenchyma rupture from expanding tumors, such as Pringle maneuver, packing, resectional debridement, selective vessel ligation, and parenchymal sutures. Application of procoagulant tissue adhesives, fibrin sealants on the raw liver surface may also improve hemostatic control (10-13).

Conclusion

Control of liver hemorrhage due to a spontaneous rupture of hepatic epithelioid angiomyolipoma is a rare surgical emergency. Damage control surgery with deep parenchymal sutures and pro-coagulant tissue adhesives utilization, abbreviates surgical timing before the development of critical and irreversible physiological endpoints. This operative concept reduces the mortality rate and the incidence of complications (14).

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