A 44-Year-Old Woman with Abdominal Pain Giant Pedunculated Cavernous Hepatic Hemangioma

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Hemangiomas are the most common benign hepatic tumors in adults, but they are rarely pedunculated. To the best of our knowledge, less than 30 cases of giant pedunculated hepatic hemangiomas have been reported since 1985. This article focuses on a case of a woman who presented with mild epigastric pain and a large abdominal mass on imaging.

INTRODUCTION

Hepatic hemangiomas are the most common tumors of the liver with an estimated prevalence of up to 20%.¹ Liver hemangiomas tend to be small and mostly asymptomatic, rarely requiring further treatment due to their benign nature and tendency towards slow growth.² They consist of large blood-filled cavities lined by abnormal endothelial cells which are sustained by the hepatic arterial circulation.²,³ Hemangiomas mostly occur in women between the ages of 40-60 years and are often found incidentally on abdominal ultrasound, computed tomography (CT), or magnetic resonance imaging (MRI).¹,⁴ Although the cause for the development of hepatic hemangiomas is not known, there are a few reports suggesting that they may be congenital and possibly genetically linked.³

Cavernous hemangiomas are thought to arise as congenital vascular malformations that form from vascular ectasia rather than hypertrophy, hyperplasia, or neoplasia.² These collections of blood vessels range in size from 0.4 centimeters to as large as 40 centimeters.³ A hemangioma larger than 4 centimeters is classified as “giant,” with a pedunculated giant cavernous hemangioma considered a rare variant.¹ To date, there are only 26 reported cases of giant pedunculated cavernous hemangiomas described in the literature.¹⁵ A pedunculated hemangioma is defined as, “clearly extended beyond the border of the liver or if the center of the lesion was located outside of the expected margins of the liver.”⁶ ⁷ They most often present clinically with intense abdominal pain in over half of the cases and often require intervention due to the mass effect on adjacent organs, the potential for pedicle torsion, and the stretching of Glisson’s capsule.¹

CASE REPORT

A 44-year-old obese woman with a past medical history of iron deficiency anemia and abnormal uterine bleeding who presented to her primary care provider for intermittent, cramping epigastric abdominal pain usually within 30 minutes of her last meal. She noted a weight gain of approximately 35 pounds over the prior year as well as occasional nausea and acid reflux that was somewhat improved with a proton pump inhibitor. The patient denied fever, vomiting, constipation, and changes in bowel habits. An abdominal ultrasound revealed a large, heterogenous mass that appeared exophytic and seemed to extend from the left hepatic lobe or greater curve of the stomach (Figure 1A). Additionally, there was a 2.6 x 2.6 centimeter isolated lesion visualized within the right hepatic lobe demonstrating peripheral enhancement. An MRI of the liver was
performed that revealed classic findings of a hepatic hemangioma (Figure 1D/2C/2D). There was also a separate, small 2.3 x 2.9 centimeter lesion in the right lobe demonstrating peripheral enhancement. An MRI of the liver was performed that revealed classic findings of a hepatic hemangioma (Figure 1D/2C/2D). A radiolabeled/tagged red blood cell scan supported the diagnosis of hemangioma for both the right hepatic lobe and the large pedunculated left hepatic lobe lesion. Angiogram of the abdominal viscera was performed to evaluate sources of perfusion and to plan for resection (Figure 1B). Fine needle aspiration biopsy (FNAB) of the abnormal-appearing left hepatic lobe confirmed hemangiomatosis of this area.

Her case was presented at a multidisciplinary gastrointestinal and hepatobiliary tumor conference where the consensus decision was to proceed with surgical resection of the pedunculated mass and observation the smaller hemangioma. During the operation, the mass was exposed via a sub-costal incision extending to the left of the midline in order to maximize visualization of its margins. The operative team identified and ligated several branching vessels of the splenic artery that were supplying the mass as well as several large draining veins. An en-bloc resection of this mass was performed, which included removal of segments 1-4 as the mass extended well beyond the left lateral segments (segments 1 and 2) of the liver. A formal left hepatectomy was performed without complication and with minimal blood loss (<100ml). Figure 3A and 3B refer to the surgical and histologic specimens, respectively. The patient was discharged on the third post-operative day without further complications. At follow up in clinic, she reported that her abdominal pain had improved. The smaller hemangioma is being monitored with periodic ultrasounds.

MATERIALS AND METHODS

All imaging studies were performed at the University Medical Center in New Orleans from May-June 2016. The ultrasound images were obtained using a GE Logic E9 machine with a C1-6 transducer. Computed tomography images were obtained using a Philips iCT 256 model. We utilized two phases of contrast administration with Omnipaque 350 - a portal venous phase (65 seconds) and a delayed phase (3m) of intravenous contrast administration were used. Magnetic Resonance images were obtained using a Philips Ingenia 1.5T Model. Phases utilized for this study include: a Coronal T2 Fast spin single shot, an axial T1 70 second post contrast series and an axial T1 3 minute post contrast series with Magnevist 46.9%.
Figure 3: A) Intraoperative image of the hemangioma with a lateral portion of the left lobe of the liver. B) Low-power view showing a well-circumscribed lesion composed of cavernous vascular channels in a background of normal liver parenchyma and zones of sclerosis (hematoxylin-eosin, original magnification x2).

DISCUSSION

Hemangiomas of the liver can be seen in up to 20% of the population, with most cases being benign, asymptomatic, and managed with observation. Those that are symptomatic usually present with abdominal pain often the result of stretching Glisson’s capsule or compression of adjacent organs. Other symptoms may include nausea, vomiting, jaundice, early satiety, or hemorrhage. For large symptomatic and growing hemangiomas of the liver, surgical management is the preferred modality of treatment and typically includes enucleation or a wedge excision.

Pedunculated hemangiomas of the liver have a female predominance of 69% which is most likely attributable to increased estrogen exposure. There is also a predilection for the left hepatic lobe (74% of cases), likely due to the large surface area to volume ratio of the left hepatic lobe compared to the right lobe. Left lobe variants are particularly troublesome due to the risk of undergoing torsion and subsequent ischemia with resultant tissue necrosis.

Imaging characteristics of hepatic hemangiomas vary, but there are several consistent and classic findings that aid in diagnosis. On ultrasound, they are usually hyperechoic with a heterogeneous echotexture and well-defined borders, although a small percentage appear hypoechoic in a background of hepatic steatosis. Doppler images more often show no internal or peripheral vascularity or color flow, a sign more worrisome for hepatocellular carcinoma. CT findings for most hemangiomas depend entirely on the utilization of intravenous contrast as well as the phase in which the study is being performed. Non-contrast CT generally shows a hypodense, fairly well-circumscribed mass while contrast enhanced arterial phase images reveal a peripherally discontinuous pattern of enhancement. Images obtained in later phases usually show a pattern of enhancement that fills in from the periphery to the center. Ultimately, the enhancement pattern results in a lesion that is isodense, or hyperdense to the surrounding liver parenchyma and vascular structures. MRI findings of a typical hemangioma are homogenous on the T1-weighted series with a hypointense signal while the T2 signal is hyperintense, though not as intense compared to cerebrospinal fluid or hepatic cysts. MRI of the liver with intravenous contrast demonstrates a similar enhancement pattern to that seen on CT; specifically, there is a discontinuous, peripheral, and nodular enhancement that fills in centripetally on delayed phases. Nuclear medicine (SPECT) imaging typically demonstrates decreased radiotracer activity within hemangiomas on early perfusion images followed by increased activity on delayed blood pool images.

Our patient’s mass had some imaging findings that were classic while others were inconsistent. On ultrasound, it was hypoechoic with well-circumscribed borders in certain areas, but there was no definitive fat plane visualized between the stomach and the spleen. There was also no internal vascularity identified definitively, raising questions about the diagnosis of a benign hemangioma. On CT, this lesion possessed most of the classic CT findings of enhancement in a nodular, discontinuous manner. However, it’s unusually large size meant the tumor filled very slowly with blood, so slow in fact that the entire tumor did not fill entirely with contrast on delayed imaging. Nuclear imaging red blood cell scan showed a much more classic appearance of low activity on the initial phase of injection followed by increased activity on delayed blood pool phase images.
The preferred surgical management is enucleation as first line therapy whenever possible, followed by partial hepatectomy of the cavernous hemangioma. Enucleation is preferred over wedge excision due to the minimal blood loss, shorter operative time, and prevention of bile leakage that is a common complication of this operation.⁹ If the hemangioma is deep within the liver and occupying multiple lobes or undivided from Glisson’s capsule, wedge resection is the ideal treatment of choice.¹⁰ Wedge excision with radical liver resection was performed instead of enucleation in this case due to the large size of the hemangioma as well as adjacent parenchymal involvement of the left lateral lobes (segments 2 and 3).

For non-surgical candidates, treatment options may include hepatic arterial embolization, radiation, or a vascular endothelial growth factor (VEGF) inhibitor such as bevacizumab. When considering the large size of a giant cavernous hemangioma, hepatic arterial embolization is a potential pre-operative approach to consider. It has been shown to reduce intraoperative blood loss and shrink the tumor allowing for easier resection and shorter operative time.¹² Embolization alone is not recommended for giant symptomatic cavernous hemangiomas due to the risks of tumor necrosis with formation of a hepatic abscess.¹³ Radiation therapy is a non-invasive treatment modality; however, it is not recommended as a definitive treatment option for such lesions. A dose of 30Gy can be given over three weeks resulting in damage of endothelial and smooth muscle cells ultimately causing thrombosis of the arterioles with subsequent tissue necrosis. This reduces the size of the tumor and provides pain relief, but it is not without side effects. Due to the risk of liver toxicity and malignancy, it is generally recommended only as a last resort for pediatric patients with giant hemangiomas in the setting of uncontrollable heart failure or coagulopathy.¹⁴ Bevacizumab has been shown previously to be efficacious when combined with 5-fluorouracil (5-FU) to treat metastatic colorectal cancer; however, there is limited data on its use in the treatment of a giant cavernous hemangioma.¹¹ Mahajan et al. reported its efficacy in reducing the size of a hepatic hemangioma in a patient with invasive colorectal adenocarcinoma.¹⁴ This is the first documented response of a hepatic hemangioma to anti-angiogenic therapy. Further studies need to be performed in order to determine the utility of such agents in the future.¹⁴

CONCLUSION

Pedunculated giant cavernous hemangiomas are exceedingly rare hepatic tumors which can provide a diagnostic challenge due to frequent atypical imaging findings. This case report provides a detailed illustration of the radiographic appearance of a pathologically confirmed lesion across ultrasound, CT, MRI, and nuclear medicine exams. As for treatment modalities, there are multiple options for asymptomatic as well as symptomatic patients. Patients without symptoms are often observed, and those with symptoms who are surgical candidates are best treated with either enucleation or a wedge resection.

Therapy for non-surgical candidates involves hepatic arterial embolization, radiation, or bevacizumab in rare cases. Our case was unique in that the hemangioma was pedunculated, giant, involved multiple lobes, and caused increasing pain as it enlarged. For these reasons, surgical resection was chosen as the best definitive treatment option, with a true left hepatectomy resulting in the complete removal of this giant hemangioma. A thorough multidisciplinary, pre-operative clinical and radiologic evaluation is useful for management of such rare tumors of the liver.

REFERENCES


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