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Case Report

Perivascular Epithelioid Cell Tumor of the Falciform Ligament with Recruitment of the Paraumbilical Vein

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Abstract

We report a case of a perivascular epithelioid cell tumor occurring in the falciform ligament of an otherwise healthy 32-year-old woman. The mass was hypervascular with central necrosis on computed tomography (CT) and contrast-enhanced ultrasound. Our case is unique as CT and ultrasound revealed a paraumbilical vein arising from the expected location in the fissure for the falciform ligament and terminating in the superior aspect of the mass. The vascular anatomy of a recanalized paraumbilical vein, typically associated with portal hypertension, was complete with drainage of blood from the inferior aspect of the mass through the inferior epigastric veins to the femoral veins. The patient had no evidence of liver disease or portal hypertension.

Keywords: Falciform ligament, perivascular epithelioid cell tumor, recanalized paraumbilical vein

Introduction

Perivascular epithelioid cell tumors (PEComas) are rare mesenchymal neoplasms composed of perivascular epithelioid cell-distinctive epithelioid cells that express both melanocytic and smooth muscle markers. Histologically, PEComas are composed of epithelioid cells in nested architecture, with nests being surrounded by thin-walled capillaries. A small subset can have predominant spindle cell morphology. Most are sporadic though a small subset can be associated with tuberous sclerosis. PEComas have a wide anatomic distribution and can occur in the visceral organs (e.g., lung), the deep body cavity (e.g., retroperitoneum), as well the peripheral soft tissues.[1,2]

An extremely rare anatomic site for P is the falciform ligament. Folpe *et al*. first described the pathologic features of six consultation cases in 2000.[3] Imaging findings of a single additional case were reported by Handa *et al*. in 2016.[4] To

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the best of our knowledge, the case presented here is unique as ultrasound and contrast-enhanced computed tomography (CT) demonstrated a paraumbilical vein arising from the expected location in the fissure for the falciform ligament terminating in the superior aspect of the mass. The vascular anatomy of recanalized paraumbilical veins, which are typically associated with portal hypertension, was complete with drainage from the inferior aspect of the mass through the inferior epigastric veins to the femoral vein. Despite the similarity to the vascular anatomy frequently seen in paraumbilical veins associated with portal hypertension and cirrhosis, our patient had no history, imaging, or pathologic evidence of chronic liver disease, leading us to hypothesize that this vascular tumor recruited the paraumbilical vein itself.

Case Report

The patient is a 32-year-old female who presented to her primary care physician with worsening nausea and abdominal bloating in the setting of chronic abdominal discomfort for several years. Initial abdominal ultrasound showed a round, vascular, hypoechoic, solid mass with smooth borders in the epigastric region and no other abnormalities. Outside magnetic resonance (MR) imaging showed a 5.1 cm × 4.8 cm × 4.1 cm well-circumscribed, heterogeneous, enhancing lesion with areas of necrosis in the right upper abdomen that appeared to abut the gallbladder, pancreas, and second portion of the duodenum [Figure 1]. The mass was not easily appreciated on physical examination despite a thin body habitus (patient body mass index of 22.3 kg/m^2). Contrast-enhanced CT of the chest, abdomen, and pelvis further characterized the mass, again raising suspicion of involvement of the head of the pancreas and/or second portion of the duodenum. There was significant internal vascularity, and a large, paraumbilical vein traversing the falciform ligament and terminating in the anterior/superior aspect of the mass was found. In addition, arising from the inferior aspect of the mass were the dilated inferior epigastric veins which terminated as expected in the common femoral veins at the groin [Figure 2]. There was also a 6 mm hyperenhancing focus in the hepatic segment 4B; however, liver morphology and attenuation were normal otherwise. There was no imaging evidence of cirrhosis and no varices were seen elsewhere in the abdomen. Focused abdominal ultrasound with contrast was performed and re-demonstrated the mass, which enhanced intensely starting at 5 s and demonstrated central areas of necrosis [Figure 3]. The study was carried out for 4 min; no washout was seen. Based on her imaging studies, the differential diagnosis included gastrointestinal stromal tumors, extrapancreatic gastrinoma, metastatic hepatocellular carcinoma, gastrointestinal Schwannoma or neurofibroma, gastrointestinal carcinoid, and gastrointestinal lymphoma. Due to the hypervascular nature of the mass and surrounding vascularity, an anticipated core biopsy was not pursued.

The patient underwent exploratory laparotomy. On carefully entering the abdomen and palpating the mass, it was able to be easily delivered while attached to the abdominal wall, with no involvement of any intra-abdominal organs. Further dissection revealed that the mass was arising from the falciform ligament [Figure 4], with a large paraumbilical vein attached superiorly to the mass and enlarged inferior epigastric veins coursing from its lower aspect. A stapler was used to transect

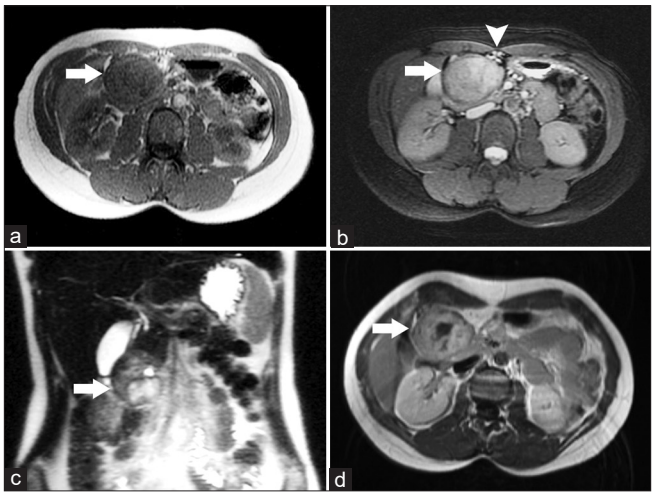

Figure 1: Magnetic resonance imaging of the abdomen without and with contrast: Precontrast axial T1-weighted (a), axial T2-weighted 2D fast imaging employing steady-state acquisition with fat saturation (b), coronal T2-weighted single shot fast spin echo (c), and postcontrast axial T1-weighted (d) images show the mass (arrows). Note prominent vessels (arrowhead) better evaluated on computed tomography scan. The mass is heterogeneous on T2 single-shot fast spin echo sequence and shows heterogeneous post contrast enhancement

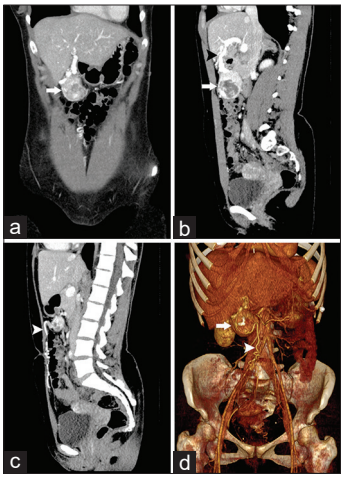

Figure 2: Contrast-enhanced computed tomography scan: Coronal (a), right parasagittal (b) and mid sagittal (c) and 3D volumetric (d) images show an avidly enhancing mass (arrows) with central necrosis. The mass is intimately associated with a large paraumbilical vein (black arrowhead). Draining veins coursing toward the periumbilical area are also seen (white arrowhead) in a pattern like that seen with a recanalized paraumbilical vein associated with portal hypertension. Note continued drainage inferiorly in the anterior abdominal wall through what likely represent the inferior epigastric veins. There is no evidence of liver disease. 3D image shows the relation of the mass to the surrounding structures, and prominent veins extending superiorly and inferiorly

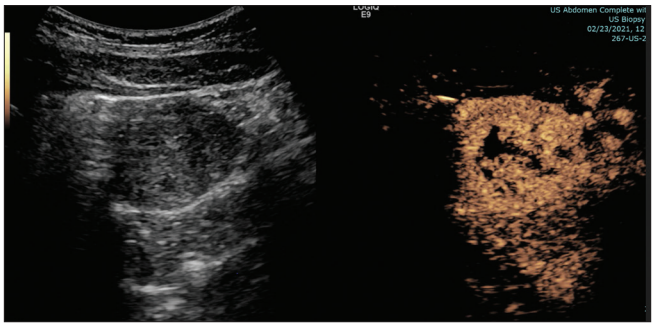

Figure 3: Contrast-enhanced ultrasound: Gray scale image (left) shows a solid mass to the right of midline in the upper abdomen. It was located immediately beneath the rectus abdominis muscle and did not appear to arise or invade any of the surrounding organs. Contrast-enhanced image taken at 25 s after injection shows intense peripheral enhancement and lack of perfusion centrally suggesting necrosis. Confirming computed tomography findings, the recanalized paraumbilical vein was found intimately associated with the mass (not shown). Enhancement persisted for the entire 4 min of the examination; washout was not seen

both these vascular pedicles and remove the mass *en bloc* with the falciform ligament. Intraoperative ultrasound was used to locate the previously seen 6 mm mass in the segment 4B of the liver, and a wedge resection of this area was performed. The abdomen was closed in layers, with extra care to avoid the remaining patent collateral vessels.

Pathologic evaluation revealed PEComa of the falciform ligament and hemangioma in the liver. The tumor was composed of epithelioid to spindle cells with eosinophilic to clear cytoplasm [Figure 5a]. The tumor cells were diffusely positive for smooth muscle actin [Figure 5b] and hydroxymethylbutyrate 45 [Figure 5c] and showed patchy positivity for melan A. There was a single mitotic figure present. Based on size >5 cm, it was classified as having uncertain malignant potential. There was a recanalized vein seen [Figure 5d]. The background liver did not show any significant pathological change, and there was no fibrosis (cirrhosis) or nodular regenerative hyperplasia seen which could have led to portal hypertension.

Discussion

Only one case with imaging of a PEComa involving the falciform ligament has been published previously, and paraumbilical vein recruitment was not described in that case report.[4] The six consultation cases described by Folpe *et al*.[3] dealt only with pathology, and no description of adjacent vascularity was provided. Given the complete lack of evidence of liver disease (including biopsy), we can only hypothesize that the recruitment of the paraumbilical vein seen in our patient was tumor induced.

The umbilical vein carries oxygenated and nutrient-rich blood from the placenta to the fetus during pregnancy. The right umbilical vein disappears early in the prenatal development, whereas the left umbilical vein drains into the newly developed left portal vein. Within a week of birth, the umbilical vein of

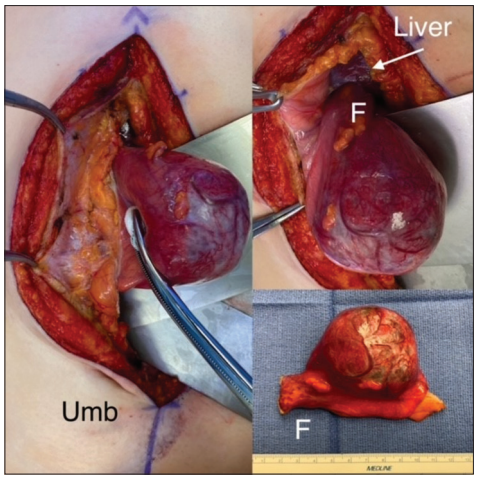

Figure 4: Gross surgical specimen demonstrating a mass arising off the falciform ligament. F: falciform ligament UMB: Umbilicus

the neonate is obliterated and is replaced by the ligamentum teres which extends to the transverse fissure joining the falciform ligament. Under increased pressure, typically, as the result of portal hypertension due to cirrhosis, there may be recanalization of the paraumbilical veins with hepatofugal blood flow to the epigastric veins.[5] Rarely, spontaneous hepatic vein-to-paraumbilical vein shunt in a patient with Budd–Chiari syndrome has been reported.[6]

There are only two reported cases of paraumbilical vein recanalization without cirrhosis or portal hypertension, both involving patients with acute pancreatitis and portal vein thrombosis.[7,8] These patients did not have cirrhosis or underlying liver disease but clearly had abnormal portal vein flow dynamics, which likely resulted in opening of the paraumbilical veins. We hypothesize that in our patient, opening of the paraumbical vein was a result of recruitment of the paraumbilical veins by this highly vascular tumor. In his description of PEComas, Folpe *et al*. described the vasculature as "dense and elaborate."[3] In the pathological evaluation of Thway and Fisher,[1] they further stated that the cells in PEComas are typically arranged around blood vessels and appear to form the vessel wall, often infiltrating the smooth muscle of small-to-medium–sized vessels.

The imaging findings of PEComa are nonspecific and have shown to be highly variable. A study of 32 CT and 15 MR cases of confirmed PEComa suggests that the diagnosis should be entertained in a normal liver with a solitary hepatic mass demonstrating a well-defined border, heterogeneous enhancement, and T1-hypo/T2-hyperintensity.[9] These findings have been further supported in the literature and expanded upon to include the somewhat characteristic recruitment of dysmorphic vasculature.[4] The imaging findings in the presented case largely recapitulate those in the referenced literature.

PEComas are generally benign but can have malignant potential. Criteria that predict malignant behavior include

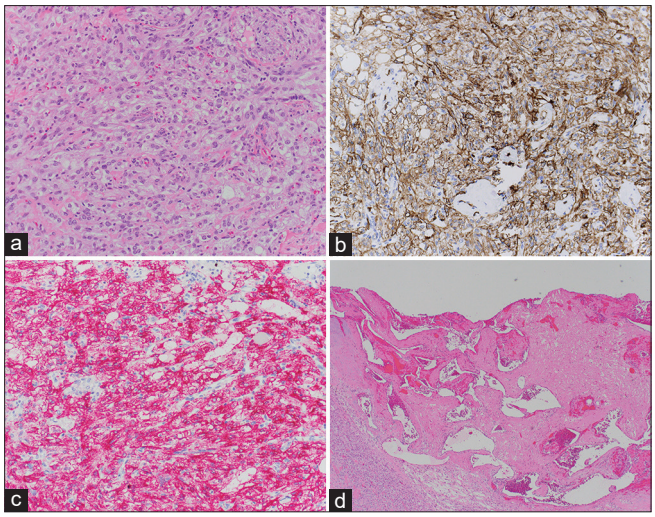

Figure 5: Tumor with spindle and epithelioid cells and clear to eosinophilic cytoplasm (H and E, ×200, a). Diffuse smooth muscle actin positivity (SMA × 200, b). Diffuse hydroxymethylbutyrate 45 positivity (HMB45, ×200, c). Recanalized vein (H and E, ×40, d)

size >5 cm, infiltrative growth pattern, high nuclear grade or cellularity, mitotic rate of >1/50 high-power field, necrosis, and vascular invasion. Tumors are classified as benign when there are <2 high-risk features and size <5 cm, uncertain malignant potential with size >5 cm with no other high-risk features or nuclear pleomorphism/multinucleated giant cells only, and malignant when two or more high-risk features are present.[10]

It is interesting to further speculate whether tumor growth and transformation toward malignancy were related to paraumbilical vein recruitment. In retrospect, the patient does recall "stomach problems" as a child with multiple doctor visits. However, these records were unavailable to us, so we were unable to determine whether a tumor may have been present at that time.

In summary, we report a case of a PEComa of the falciform ligament that appears to have recruited the paraumbilical and inferior epigastric veins, perfectly mimicking what is seen in patients with portal hypertension.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given consent for images and other clinical information to be reported in the journal. The patient understands that her names and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

Dr. William W. Tseng , an editorial board member at *Journal of Cancer Research and Practice*, had no role in the peer review process of or decision to publish this article. The other authors declared no conflicts of interest in writing this paper.

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