CASE REPORT

Multidetector CT of pancreatic hemangiopericytoma

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Abstract

Hemangiopericytoma is an aggressive, highly metastatic tumor of the soft tissues and meninges. Metastases have been reported in the liver, lungs, bones and rarely other organs. To the best of our knowledge, 18 cases of pancreatic metastases have been reported, but none have been described in the radiology literature. We report a case of hemangiopericytoma with metastasis to the pancreas and describe the CT imaging features of hemangiopericytoma with a review of the literature.

Keywords: pancreas; hemangiopericytoma; metastasis; CT.

Introduction

Hemangiopericytoma (HPC) is a rare mesenchymal tumor derived from pericytes of Zimmerman[1]. Theoretically, the tumor can occur wherever there are capillary beds, but most commonly occurs in the soft tissues of the lower extremity, pelvis or retroperitoneum. Intracranial HPCs are less common than soft tissue HPCs and are generally associated with the meninges. In the past, intracranial HPCs have been considered a variant form of aggressive meningioma (angioblastic meningioma). Regardless of its location, the tumor is considered aggressive with a high rate of local recurrence and metastasis. Metastases have been described in the liver, lung, bones and rarely other locations.

The radiologic appearance of HPC, primary or metastatic, is non-specific. The tumor is hypervascular with a well developed vascular network not dissimilar from hepatocellular carcinoma or other hypervascular metastases. Several reports have described features that are frequently seen in these tumors but no distinguishing features have been identified.

We report a case of metastatic hemangiopericytoma with pancreatic involvement. Rare cases of pancreatic metastases of HPC have been previously described in the English literature, but to the best of our knowledge, this is the first case report of pancreatic metastasis of HPC in the radiology literature.

Case report

A 67-year-old woman with a history of recurrent meningeal hemangiopericytoma presented for abdominal CT with symptoms of nausea, abdominal pain and a palpable abdominal mass. Approximately 12 years earlier, the patient had undergone primary resection of a meningeal hemangiopericytoma. This had subsequently recurred and required repeat craniotomy 3 and 8 years after primary resection. A repeat MR of the head approximately 8 months before presentation demonstrated a third recurrence in the operative bed. Given the patient’s abdominal symptoms, CT of the abdomen was obtained at an outside institution prior to repeat craniotomy. This revealed an enhancing mass in the head of the pancreas.

Based on this finding, a dedicated CT of the pancreas was performed at our institution according to our standard arterial and venous pancreas protocol. Following ingestion of Volumen oral contrast material and injection of 125 cc Ultravist 370 with a 50 cc normal saline ‘chaser’, helical CT was obtained with 40 and 65 s delays using a 64 slice GE Lightspeed VCT scanner (GE Medical Systems; Milwaukee, WI). Scanner settings for this protocol are: kVp = 120, mA = 500, 40 mm collimation, 0.675 mm slices at a pitch of 1.375:1. Images were reconstructed into 2.5 mm slices, and coronal and curved reformats were performed to better define the anatomy prior to surgical resection.
CT revealed a well circumscribed, enhancing 6.3 x 4.9 cm mass arising from the anterior inferior aspect of the pancreatic head (Fig. 1). Small areas of low attenuation centrally were presumed to represent necrosis. No calcification was evident. Numerous large supplying arteries arising from the gastroduodenal and superior mesenteric arteries were noted on the parenchymal phase imaging (Fig. 2). The biliary duct was mildly dilated but there was no significant pancreatic ductal dilatation. No definite evidence of encasement of the adjacent vasculature was noted and no other foci of metastatic disease were appreciated in the abdomen.

Fine-needle aspiration of the mass was performed and histopathology revealed a neoplasm of high-grade spindle cells, similar in appearance to the patient’s prior meningeal tumors, compatible with metastatic hemangiopericytoma. Immunohistologic staining confirmed this diagnosis with CD34 and vimentin positivity and cytokeratin negativity.

Prior to repeat craniotomy, resection of the pancreatic mass was performed. The patient underwent a pylorus preserving Whipple with resection of a 5 x 5 x 6.4 cm mass. Surgical pathology revealed a firm, well circumscribed tan mass with central necrosis and hemorrhage. There was no vascular invasion, involvement of the pancreatic or biliary ducts, or extension of the tumor through its capsule.

**Discussion**

Hemangiopericytoma (HPC) is a rare, aggressive tumor that primarily involves the soft tissues and less commonly the meninges. The tumor is more common in men and typically occurs in the fourth or fifth decade[1]. HPC is considered locally aggressive with high rates of local recurrence and metastases. Local recurrence rates of up to 90% and metastatic rates of up to 33% have been reported[2,3]. The most common sites of metastases described in the literature are the liver, lungs and bones, with reports of metastases to other organs being much less common. To the best of our knowledge, 18 cases of

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**Figure 1** CT images from the described case demonstrating metastatic hemangiopericytoma involving the pancreas. During the parenchymal phase (A) the mass appears well defined and heterogeneously enhancing. Multiple, prominent feeding vessels surround the mass. Subsequent venous phase imaging (B) further defines the mass which is clearly delineated from the pancreatic parenchyma and has central areas of low attenuation consistent with necrosis. The adjacent duodenum is compressed by resultant mass effect. Curved axial reformatting in the venous phase (C) and curved coronal reformatting in the parenchymal phase (D) better define the position of the mass relative to the pancreatic head. The mass again appears well defined and associated mass effect leads to pancreatic ductal dilatation and compression of the second portion of the duodenum. Numerous dilated, feeding vessels are seen as foci of high attenuation throughout the mass on the parenchymal phase image (D).
pancreatic metastases from primary intracranial HPC have been described in the English language literature\[1/C15116\]. The exact means of metastases is not yet understood. Lymph node metastases appear incredibly rare, indicating that tumor cells may be hematogenously disseminated by entering damaged vessels at the time of surgical resection [16].

Given the rarity of these tumors, few data are available defining prognostic factors which can be used to guide future therapy. Histologic grade has not been shown to correlate with the risk of metastasis [17]. Additionally, recurrences and metastases have been demonstrated to occur as long as 20 years after the primary resection [15]. Because of the significant risk of local recurrence and high rates of metastatic disease, some authors have recommended close follow-up with cross-sectional imaging [16].

The imaging findings of hemangiopericytoma are similar regardless of whether the tumor is intra- or extracranial. Alpern et al. described a small series of primary abdominal HPCs and described CT findings of a lobulated mass with enhancing solid components, cystic areas and speckled calcification as ‘suggestive of, but not specific for, hemangiopericytoma’ [19]. Calcifications and cystic areas were less commonly found in other series [20]. The finding of a highly vascular mass appears common among all reports. HPCs are frequently described to have well developed vascular networks with a dense capillary blush on angiography and intense enhancement on CT.

Given that these findings are non-specific, a differential diagnosis of hypervascular pancreatic masses should be considered. The primary differential diagnoses include: neuroendocrine tumor, renal cell carcinoma metastasis [21,22], intrapancreatic portal cavernoma [23], and intrapancreatic accessory spleen. Differentiation among these lesions is primarily based on the clinical history. Renal cell metastases occur in patients with a history of a primary renal lesion. Neuroendocrine tumors are predominately identified in patients with von Hippel Lindau or endocrine abnormalities such as hyperinsulinemia [22,24], but can also occur sporadically. Functioning neuroendocrine tumors typically present as small enhancing lesions while non-functioning tumors are usually larger at presentation. Intrapancreatic portal cavernoma occurs in the setting of portal venous occlusion and appears as a tangle of engorged collateral vessels within the pancreas [23]. Intrapancreatic accessory spleen cannot be differentiated on the basis of history but are primarily located in the tail of the pancreas and should have an enhancement pattern which follows that of the spleen [25].

In addition to hypervascularity, the vast majority of HPCs have been described as either well circumscribed or encapsulated without invasion of the adjacent organ parenchyma [1,6,9,10,20,26,27]. These features were observed in the case described herein. Rare cases in the literature have been described to have a more invasive profile [28,29]. In a previous report of the CT findings of hepatic metastasis of HPC, Shin et al. described an ill defined mass with areas of sclerosis and minimal contrast enhancement [28]. These characteristics appear to be the exception rather than the rule, and it should be noted that MR examination of the same lesion revealed a well circumscribed mass which did not appear infiltrative.

**Conclusion**

The prospective identification of hemangiopericytoma on CT, whether primary soft tissue or metastatic intracranial, will remain difficult despite multiple case reports of this entity. The tumor is rare and the imaging findings are non-specific and similar to the findings of hypervascular primary and secondary tumors. However, in the setting of a patient with a known primary HPC, a hypervascular/enhancing, well encapsulated lesion in distant organs, such as liver, lung or pancreas, as described in this case, should be considered to possibly represent metastatic disease.

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