

Pancreatic Metastases from a Hemangiopericytoma of the Leg: A Case Report¹

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Hemangiopericytoma of the pancreas has rarely been described, and its radiological findings have never been described in the radiological literature. We report a case of a metastatic hemangiopericytoma involving the pancreas in a 48-year-old woman. CT, MR, and angiography showed three, well-demarcated, heterogeneously enhancing masses with necrosis and hemorrhage in the pancreas.

Index words : Pancreas

Pancreatic neoplasms

Tomography, X-Ray Computed

Magnetic resonance (MR)

Hemangiopericytoma

Hemangiopericytoma is a rare mesenchymal tumor originating from the pericytes of Zimmermann that are normally present in the basal membrane of the capillaries and postcapillary venules. The function of the pericytes is not well understood, although they appear to be involved in the regulation of vessel caliber. As all capillaries are enveloped by pericytes, hemangiopericytomas can be found virtually anywhere in the body (1, 2). However, this neoplasm more commonly affects the soft tissues of the extremities, pelvis, and retroperitoneum. Pancreatic involvement of a hemangiopericytoma is extremely rare. We report a case of a metastatic hemangiopericytoma involving the pancreas and its radiological findings on CT, MR imaging including MR angiography, endoscopic retrograde cholangiopancreatog-

raphy (ERCP), and conventional angiography.

Case Report

A 48-year-old female was referred to our institution for further examination and treatment of a palpable abdominal mass. The patient complained of vague abdominal discomfort and dyspepsia for a duration of four months but without weight loss. Twenty years ago, the patient had undergone surgery for varicose veins in the left leg; 10 years later, the patient underwent excision of a new mass at the previous surgery site in the left leg. The pathological diagnosis of the new mass was a hemangiopericytoma. A physical examination showed a palpable, non-tender mass in the right upper quadrant of the abdomen. The results of routine laboratory tests, including measurement of the levels of serum amylase, lipase and bilirubin, and tumor markers, including carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA 19-9), were within normal ranges. An abdominal CT demonstrated three masses arising from the pancreas head and body. The longest diameters of the

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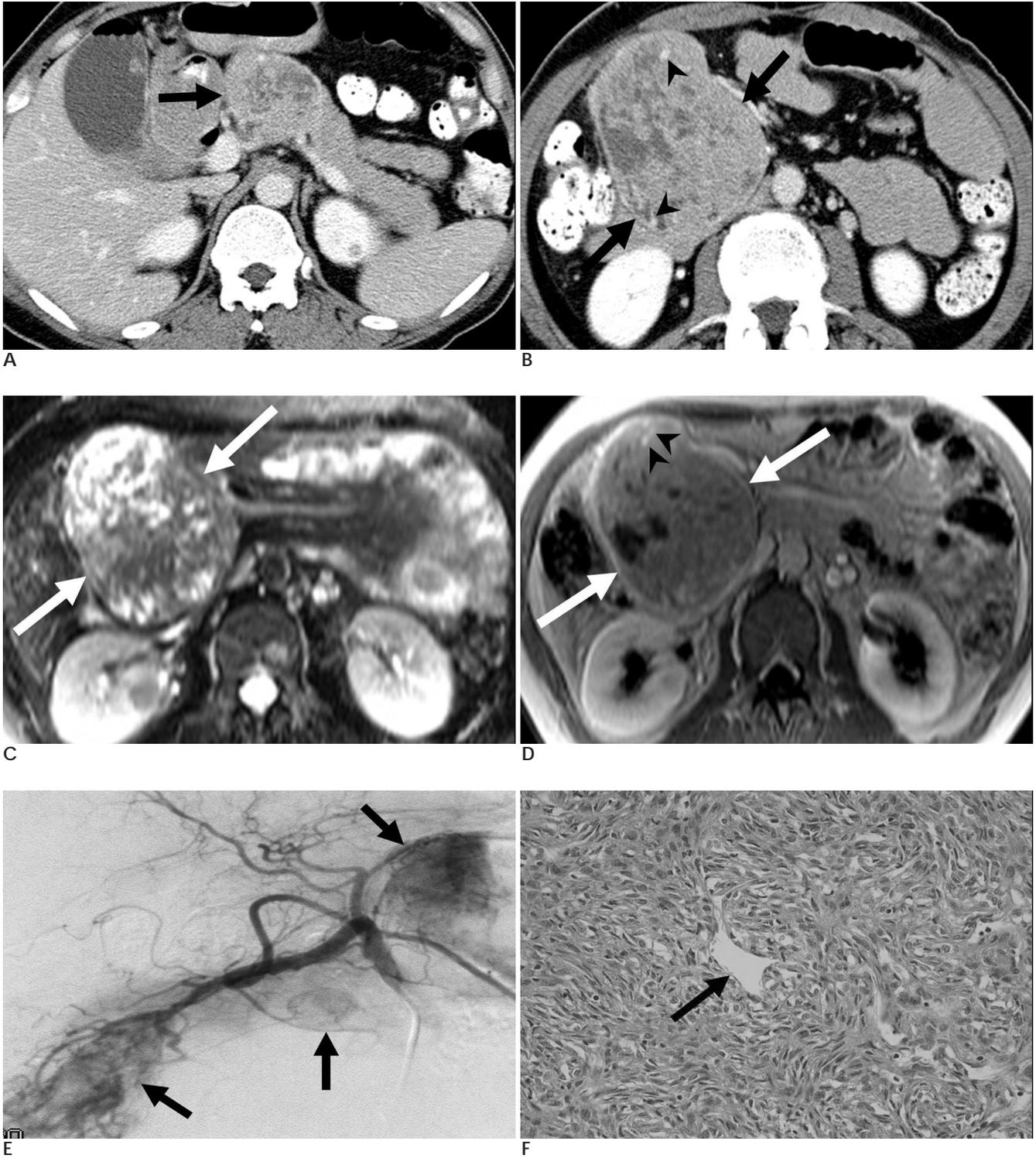


Fig. 1. Metastatic pancreatic hemangiopericytomas in a 48-year-old woman.
A, B. Contrast-enhanced axial CT scans show two, well-margined, heterogeneously enhancing masses (arrows) with low-attenuation portions, indicating necrosis or cystic degeneration as well as intratumoral vessels (arrowheads in **B**) in the pancreatic head and body.
C. On a T2-weighted MR image, the mass (arrows) of the pancreas head shows heterogeneous, intermediate- and high-signal intensity and some bright signal intensity, indicating cystic degeneration or necrosis.
D. On an axial gadolinium-enhanced T1-weighted MR image, the mass (arrows) in the pancreas head shows heterogeneous enhancement with nonenhancing portions, indicating necrosis or cystic degeneration, as well as intratumoral vessels (arrowheads).
E. Conventional angiography shows three hypervascular masses (arrows) with intratumoral vessels in the pancreas.
F. A photomicrograph of the histological specimen shows thin-walled, endothelial-lined, ramifying vascular channels (arrow). Reticulin and collagen fibers enmesh the tumor cells outside the vascular structure (Hematoxylin and eosin staining, $\times 400$).

masses were 9 cm, 7 cm and 2 cm, respectively. On unenhanced CT, the three masses showed a similar attenuation to the normal surrounding pancreas parenchyma with multifocal, low-attenuation portions without calcifications. On contrast-enhanced CT, the masses were well-demarcated and showed heterogeneous enhancement with multifocal, low-attenuation portions and some intratumoral vessels (Figs. 1A, B). The common bile duct was mildly dilated without pancreatic duct dilatation (Fig. 1A). There were no enlarged lymph nodes in the abdomen. ERCP revealed the upward displacement of the distal common bile duct and mild dilatation of the intra- and extrahepatic bile ducts. Compared with the normal pancreas parenchyma, the masses showed heterogeneous, iso- and low-signal intensity on T1-weighted MR images and demonstrated heterogeneous, intermediate- and high-signal intensity and some bright signal intensity portions, representing necrosis or cystic degeneration, on T2-weighted MR images (Fig. 1C). On gadolinium-enhanced MR images, the masses showed heterogeneous enhancement with intratumoral vessels and some nonenhancing portions, indicating necrosis, cystic degeneration or old hemorrhage (Fig. 1D). MR angiography and conventional angiography showed the hypervascularity of the masses (Fig. 1E). The patient underwent a pylorus-preserving pancreaticoduodenectomy and total pancreatectomy with a splenectomy. On the gross specimens, the masses were well-demarcated and firm. The cut surfaces of the masses were yellowish with partial hemorrhage and necrosis. Histology of the specimen showed thin-walled, endothelial-lined, ramifying vascular channels. Reticulin and collagen fibers enmeshed the tumor cells outside of the vascular structure (Fig. 1F). By immunohistochemical staining, CD 34 and CD 99 were positive. The final histopathological diagnosis of the masses was as hemangiopericytomas. Two years following the surgery, several new masses were detected in the left psoas muscle and in both kidneys. The patient underwent excision of these masses that were also diagnosed as hemangiopericytomas. Two years after the second surgery, a follow-up CT showed multiple, recurrent masses in the left kidney, pelvic mesentery, liver, and lung.

Discussion

An hemangiopericytoma is a rare mesenchymal tumor that accounts for less than 2% of all mesenchymal tumors. It was first described and named by Stout and

Murray in 1942 (2). It may occur at all ages, but is most common in the fifth and sixth decades. It is equally common in men and women (1, 2). Although there is no definitive pathological grading system to determine the malignancy potential, the prominent mitotic activity, necrosis, hemorrhage, and increased cellularity are all suggestive of a malignancy. The prognosis of this tumor is also related to its size. The relative 10-year survival rate of patients with tumors larger than a median size of 6.5 cm in the longest diameter is 63 %, whereas the 10-year survival rate of patients with tumors smaller than 6.5 cm in the longest diameter is 92% (1).

The most common site of a hemangiopericytoma is the lower extremity, especially the thigh. In a series of 106 patients reported by Enzinger and Smith (1), 37 tumors (35%) affected the lower extremity and 26 tumors (25%) were found in the pelvis or retroperitoneum. Of the other 43 patients in that study, 17 tumors were found in the head and neck region, 15 tumors involved the trunk, and 11 tumors were located in the upper extremity (1). Most abdominal hemangiopericytomas arise in the retroperitoneum, omentum or intestinal wall (3). An hemangiopericytoma originating from the pancreas is very rare. On reviewing the English language literature, we found four cases of a primary pancreatic hemangiopericytoma (4) and six cases of a metastatic pancreatic hemangiopericytoma that had arisen from an intracranial hemangiopericytoma (5). Unfortunately, there was only a description of the radiological findings without images of the pancreatic hemangiopericytoma in these ten case reports.

In this case, metastatic hemangiopericytomas of the pancreas appeared as well-demarcated, hypervascular masses with heterogeneous enhancement and multifocal cystic portions indicating necrosis, cystic degeneration or old hemorrhage. Goldman et al. (6) and Alpern et al. (7) reported that retroperitoneal hemangiopericytomas appeared as large, lobulate, enhancing, soft-tissue masses with cystic low-attenuation zones consistent with necrosis, hemorrhage or cystic degeneration. Alpern et al. (7) reported the presence of speckled calcifications in a retroperitoneal hemangiopericytoma. The incidence of calcification in hemangiopericytoma varied from one (1%) of 106 cases on plain radiography to five (71%) of seven patients on CT (1, 7). On MR imaging, a pelvic hemangiopericytoma showed intermediate to low signal intensity on T1-weighted images and intermediate to high signal intensity on T2-weighted images. On gadolinium-enhanced MR images, the solid portion of

the tumor showed strong enhancement (8). Shin et al. (9) reported that a hepatic hemangiopericytoma was sharply delineated and the extent of liver involvement could be more accurately evaluated on MR images. On angiography, hemangiopericytomas appeared as very hypervascular tumors as there was a clear increase in the number and caliber of arteries and veins of the tumor. Therefore, embolization might be considered preoperatively because of the extreme vascularity of hemangiopericytomas (6). The radiological features of hemangiopericytomas seem to be similar in the present study and in previous reports.

The differential diagnosis of a well-demarcated, hypervascular pancreatic mass with cystic portions should include a non-functioning islet cell tumor as these tumors show strong enhancement with cystic degeneration and necrosis on contrast-enhanced CT. Non-functioning islet cell tumors share similar radiological findings with the findings of hemangiopericytomas. A more common hypervascular metastasis such as a renal cell carcinoma should be included in the differential diagnosis of multiple hypervascular masses in the pancreas, because metastasis tends to repeat the imaging pattern of the primary tumor (10).

In conclusion, a metastatic hemangiopericytoma in the pancreas appears as a well-demarcated, hypervascular mass with cystic degeneration or necrosis. A correct radiological differential diagnosis distinguishing it from a nonfunctioning islet cell tumor or other hypervascular

metastasis remains difficult, if the diagnosis or history of the primary extrapancreatic hemangiopericytoma is unclear.

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