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Spindle Cell Tumor of the Portal Vein

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Abstract

The authors describe the first reported case of spindle cell tumor of the portal vein (SCTPV) surgically excised in a 28-year-old woman. The tumor had caused portal vein obstruction with extensive collaterals. A side-to-side mesocaval shunt was created to reduce portal hypertension. Six months later the SCTPV could be excised. Radiation therapy (4,500 cGY) was given 2 months later. Although the tumor recurred in the liver 4 years later, the patient is alive and free of symptoms 6 years after the initial diagnosis.

Keywords

portal vein sarcoma; extensive collaterals; portal hypertension

INTRODUCTION

Primary extrahepatic portal vein tumors are exceedingly rare. There have been only two cases reported in the literature to our knowledge. One was a leiomyoma which was partially excised, and another was a leiomyosarcoma which could not be surgically removed [1,2]. We report here a case of spindle cell sarcoma of the portal vein which was surgically resected after a mesocaval shunt.

CASE REPORT

A 28-year-old woman had been complaining of lower abdominal fullness and aching pain for 6 months. She underwent an ultrasonography of her abdomen and pelvis which revealed bilateral adnexal masses ($7.5 \times 5.5 \times 9$ cm on the right and $9 \times 10 \times 8$ cm on the left) and a large mass ($3.0 \times 4.7 \times 2.7$ cm) obstructing the portal trunk with extensive collateral circulation with splenomegaly. The computed tomographic (CT) scan demonstrated a normal size liver without parenchymal lesions, but the portal vein trunk was markedly enlarged with intraluminal obstruction and extensive collateral vessels. A fine needle aspiration biopsy of the portal vein trunk mass revealed spindle cell sarcoma.

Exploratory celiotomy was carried out at another medical center. The bilateral ovarian masses turned out to be endometriomas, which were excised successfully. The tumor of the portal vein trunk appeared to extend into the liver, and it also involved the bifurcation of the superior mesenteric vein and the splenic vein. There was significant portal hypertension with splenomegaly. No definitive surgical therapy of the portal vein tumor was attempted. She

recovered from this operation quickly, but she noticed increasing abdominal girth and leg swelling after the operation.

Hepatic angiography and endoscopic retrograde cholangiography were added to assess the extent of the portal vein tumor. Both the hepatic arterial and biliary systems were not involved by the tumor.

The patient was transferred to University of Pittsburgh Medical Center for excision of the portal vein tumor. However, the first attempt at removal was unsuccessful because of severe portal hypertension and extensive collateral vessels around the tumor. There were 8 L of ascitic fluid evacuated during the operation. A side-to-side mesocaval anastomosis was created using an iliac vein homograft to reduce portal hypertension. Although the decompression of the mesenteric venous system was well achieved by the shunt, that of the splenic venous system was inadequate due to the invasion of the tumor into the splenic vein. The operation was terminated there, hoping that the mesocaval shunt would decompress the splenic venous hypertension by collateralization in a few months, when another attempt at the resection was planned.

Her ascites and edema subsided gradually, and the patient returned to work within a month without significant symptoms, which delayed the planned surgery for 6 months. At this time the angiography confirmed the patency of the mesocaval shunt and good decompression of the entire portal venous system. The CT scan revealed no significant progression of the portal vein tumor for the last 6 months. Subsequently the portal vein tumor was excised. The tumor was 7 cm in the greatest diameter, and displaced, but did not involve either the common duct or the hepatic artery. The tumor extended into the splenic vein and the superior mesenteric vein, and both veins were doubly ligated and divided well beyond the tumor. However, the tumor also extended into the right and left portal vein branches in the hepatic parenchyma. Consequently, both portal branches were traced deep into the hepatic parenchyma, pulled out as much as possible, and ligated and divided. Although the gross tumor was removed from the hepatic parenchyma, the resection margins were inadequate.

The patient recovered from this operation and returned to work within a month with the only complication being a bile leak from the small biliary branches, which ceased to drain without consequences. Radiation therapy (4,500 cGy in 25 fractions) was administered to the hepatic hilum 2 months later. Annual CT scan had not shown any recurrence of the tumor until 4 years later, when a mass in the periphery of the right lobe of the liver was discovered. Needle biopsy of this lesion revealed spindle cell sarcoma which was most compatible with metastasis from the original portal vein tumor.

The progression of the tumor has been very indolent, and the patient is free of symptoms and able to perform normal activities more than 6 years after the initial diagnosis.

Pathology

The pathologic specimen consisted of a Y-shaped portion of vein wall containing a multinodular, fleshy mass 7.0 cm in the greatest dimension (Fig. 1). The mass was within the vessel and the adventitia of the vessel formed the outer portion of the tumor in areas.

On microscopic examination, the tumor cells were arranged in fascicles, accompanied by some storiform areas (Fig. 2A,B). Other regions of the tumor demonstrated a more myxoid appearance. The individual cells contained plump, blunt-ended fusiform nuclei with small to inapparent nucleoli (Fig. 2C). Endothelial cells were also seen with capillary formation throughout the tumor. Scattered erythrocytes and small mononuclear cells were seen, especially in the more myxoid areas. One mitotic figure was seen in 135 high-powered

microscopic fields. Tumor was noted to be present at the margin of resection on the liver side. A portion of cells were positive when stained with antibody to smooth muscle-specific actin (alpha actin).

DISCUSSION

Tumors of blood vessels are rare. The most frequent are sarcomas of the inferior vena cava [3,4]. One case each of leiomyoma and of leiomyosarcoma of the portal vein have been reported [1,2]. The lesion described above appears to be a low-grade spindle cell tumor. The present case had some evidence of smooth muscle differentiation in the form of smooth muscle actin positivity and the morphology of the spindle cells by light microscopy. However, the pleomorphism and mitotic activity present in the malignant forms of smooth muscle tumors were not observed in this case. One may argue that the lesion represents a low-grade leiomyosarcoma, but the extremely low mitotic index does not lend histologic support for this concept.

Typical leiomyomas appear as solid proliferations of smooth muscle cells in most cases. Myxoid variants do occur, often in association with more typical areas of smooth muscle differentiation.

Nonneoplastic tumorous proliferations of mesenchymal tissue have also been described within blood vessels. Patchefsky and Enzinger [5] reported a variant of nodular fascitis termed intravascular fascitis. These multinodular growths primarily affected vessels of the extremities and had a mean diameter of 1.5 cm. Treatment consisted of local excision. Two of nine patients had local recurrences at times ranging from 3 months to 2 years following surgery.

The location and size of the tumor in the present case suggest that this lesion does not fit into the category of intravascular fascitis. Additionally, the finding of a metastatic spindle cell lesion in the liver 4 years following surgery is consistent with a neoplastic origin of the mass.

Excision of this tumor in the presence of severe portal hypertension would not have been possible if we had not chosen to create a mesocaval shunt. In view of the prolonged survival of this patient to date, such an approach appears to be a reasonable option in the management of tumors of the portal vein.

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Fig. 1.

The gross photograph of spindle cell tumor of the portal vein. The multinodular tumor has been partially sectioned, showing solid areas with foci of hemorrhage. The lower portion of the specimen shows an area of tumor undergoing cystic change.

Fig. 2.

A: Low power microscopic photograph of the tumor. Loose fascicles of cells form a storiform pattern in areas (H&E). $\times 100$. **B:** Scattered endothelial channels are also present throughout much of the lesion. Scattered mononuclear cells are also observed (H&E). $\times 250$. **C:** The spindle cells show plump nuclei with little heterogeneity. Free erythrocytes are also seen within this area (arrows) (H&E; $\times 400$).