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Pelvic retroperitoneal mass in a 36-year-old man

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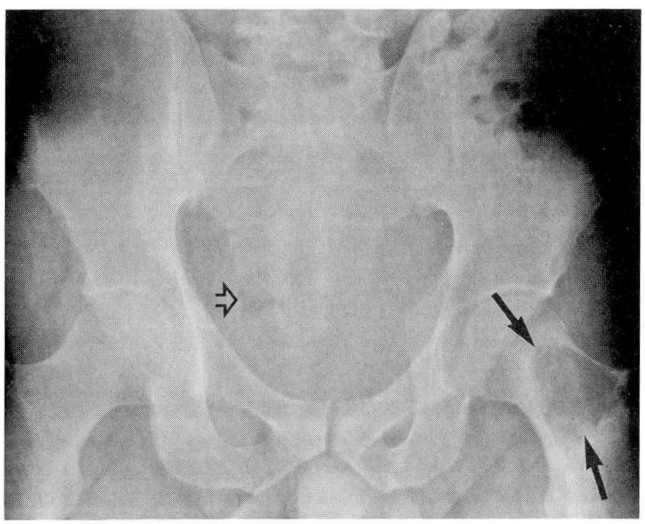

FIGURE 1. Pelvic radiograph demonstrates a well-defined lytic lesion of the left femoral neck (straight arrows). A small amount of gas is noted in the rectum which lies in the right hemipelvis (open arrow).

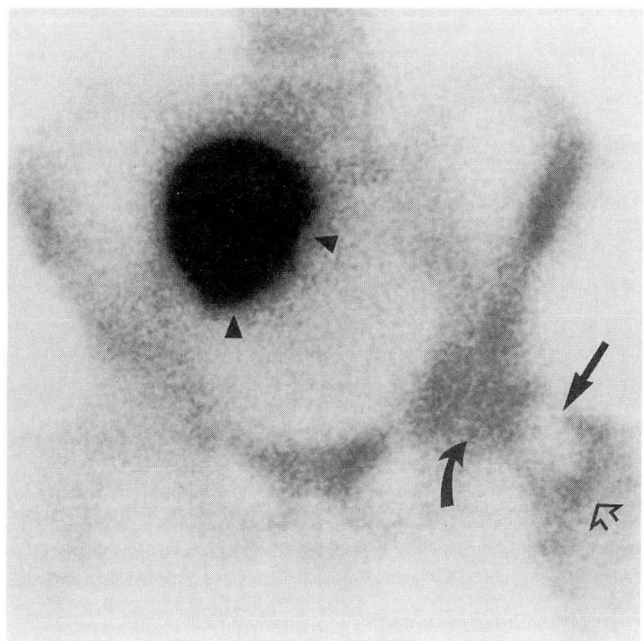

FIGURE 2. Technetium 99m radionuclide bone scan (delayed frontal image of the pelvis) demonstrates a photopenic lesion of the left femoral neck (solid arrow) surrounded by increased activity of the femoral head (curved arrow) and intertrochanteric region (open arrow). The urinary bladder is displaced superiorly and to the right (arrowheads).

A 36-year-old man complained of left groin and hip pain for 4 months. Physical examination was remarkable for suspected prostate enlargement and guaiac-positive stool. Laboratory studies revealed normochromic, normocytic anemia. Endoscopic gastroduodenoscopy demonstrated a duodenal bulb ulcer, gastritis, and duodenitis, likely related to the patient's use of nonsteroidal anti-inflammatory drugs for hip pain.

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Plain radiography of the pelvis and left hip demonstrated a well-defined lytic lesion of the left femoral neck (*Figure 1*). Delayed images of a three-phase radionuclide bone scan demonstrated the left femoral neck lesion to be photopenic surrounded by increased activity of the intertrochanteric region and femoral head, with displacement of the urinary bladder (*Figure 2*). Dynamic blood flow and blood pool images of the pelvis had demonstrated a large hyper-

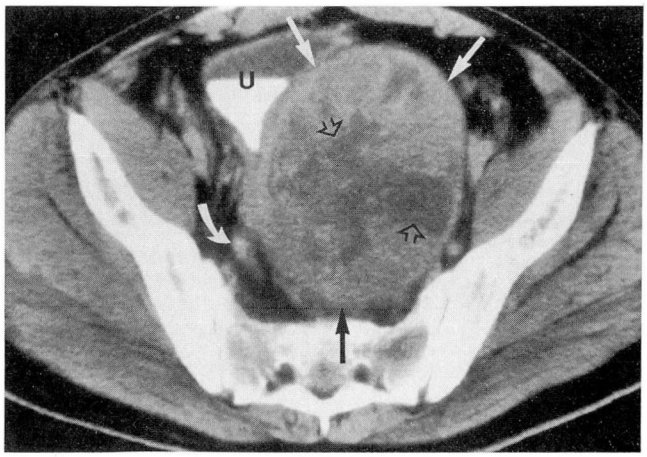

FIGURE 3. CT scan of the pelvis following administration of oral and intravenous contrast material reveals a large, enhancing, well-defined left retroperitoneal mass (solid arrows) which contains areas of low attenuation (open arrows). The urinary bladder (U) and rectosigmoid colon (curved arrow) containing contrast material are displaced by the mass.

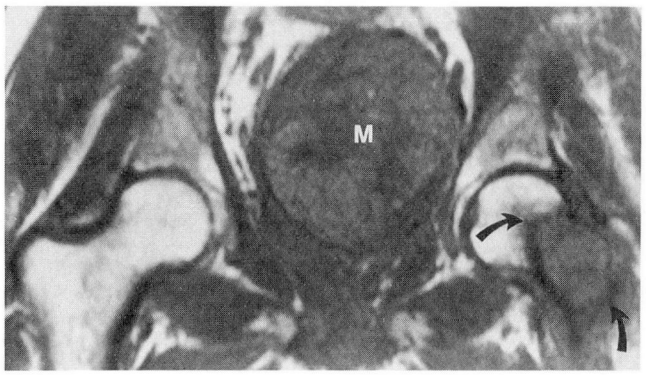

FIGURE 4. Coronal T-1 weighted (TR 615 milliseconds, TE 26 milliseconds) spin-echo MRI of the pelvis. The well-defined pelvic mass (M) is of low signal intensity. The lesion of the left proximal femur is also noted (curved arrows).

vascular mass of the left pelvis. A computed tomography (CT) scan of the abdomen and pelvis with intravenous and oral contrast material was obtained and revealed a large, well-defined left retroperitoneal mass containing a few small calcifications. The mass extended into the pelvis and displaced the bladder, rectosigmoid colon, and rectum (*Figure 3*). Hydronephrosis with marked parenchymal loss of the left kidney was also present, indicating chronic obstruction of the left distal ureter by the mass. Spin-echo magnetic resonance imaging (MRI) of the pelvis demonstrated the mass to have decreased signal on T1-weighted images and very high signal on T2-weighted images; the left femoral neck lesion showed similar signal characteristics (*Figure 4*). Ultrasonography demonstrated the pelvic mass to be of mixed (predominantly low) echogenicity, with a few scattered echogenic foci of calcification. A needle biopsy of the pelvic mass was then performed under CT guidance (see Discussion). Angiography of the pelvis confirmed the mass to be hypervascular and supplied by both internal iliac arteries (*Figure 5*). Preoperative embolization of the anterior divisions of the internal iliac arteries was performed bilaterally.

An open biopsy of the left proximal femur was performed, and the pelvic mass was resected. The pelvic mass was found to be adherent to the posterior wall of the bladder, and there was no evidence of bowel involvement. Despite embolotherapy, considerable blood loss occurred, requiring multiple transfusions intraoperatively.

The resected pelvic specimen weighed 735 g and measured 14 × 12 × 9 cm. It was encapsulated by a white-yellow, slightly glistening fibrous layer. The cut surface was pink-tan and consisted of soft tissue with areas of hemorrhage, necrosis, and fibrosis (*Figure 6*).

Microscopic examination (*Figure 7*) showed a monotonous population of tightly packed anaplastic tumor cells arranged in a vaguely spindled pattern and compressing small and large vascular channels lined with benign endothelium. The nuclei were enlarged, irregular, and contained easily identifiable nucleoli, and nuclear chromatin was stippled. Cytoplasmic borders were indistinct, and the nuclear-cytoplasmic ratio was increased. One to three mitoses per high-power field were seen. Necrotic hemorrhagic tissue was present. The left femoral biopsy was consistent with necrotic metastatic tumor.

DIAGNOSIS: HIGH-GRADE SARCOMA CONSISTENT WITH MALIGNANT HEMANGIOPERICYTOMA

The patient underwent a left total hip replacement. Radiation therapy of 5,500 cGy was administered to the left proximal femur. Chemotherapy with mesna, ifosfamide, DTIC, and adriamycin was given. A metastatic workup revealed no evidence of other sites of involvement. The patient remains free of recurrence or metastases 2 years after surgical treatment.

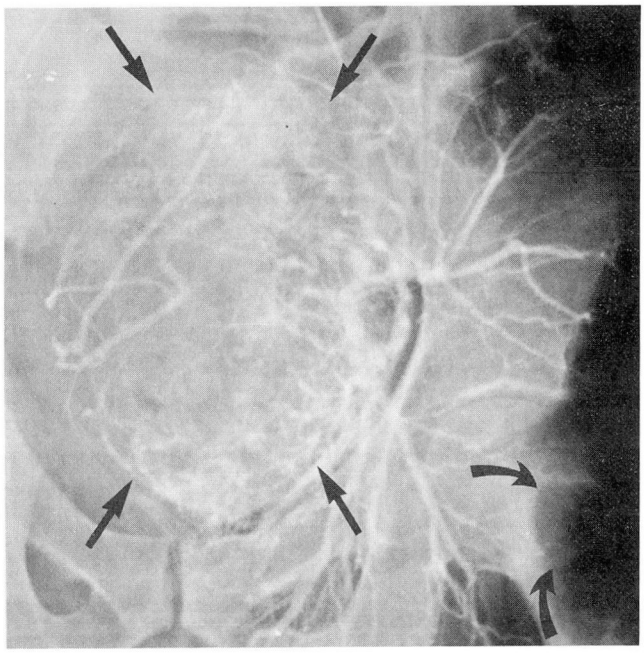

FIGURE 5. Late arterial phase of a left internal iliac arteriogram confirms the hypervascular nature of this pelvic mass (straight arrows). The left proximal femoral lesion is again noted (curved arrows).

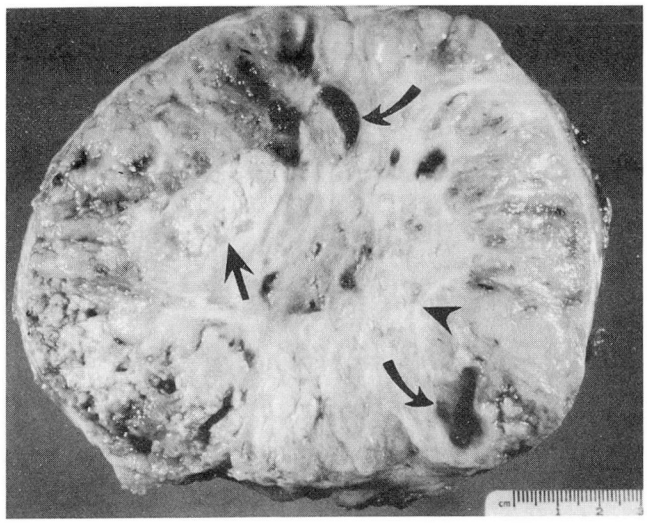

FIGURE 6. The cut surface of the gross specimen shows areas of necrosis (straight arrow), hemorrhage (curved arrow), and fibrosis (arrowhead).

DISCUSSION

The histologic differential diagnosis of this tumor included malignant hemangiopericytoma, monophasic synovial sarcoma, and fibrosarcoma. Despite the use of special stains, immunohistochemistry, and electron microscopy, the exact histologic classification of the resected specimen remained challenging. The histology of the needle biopsy suggested hemangiopericytoma; the histology of the resected specimen suggested high-grade sarcoma consistent with malignant hemangiopericytoma. The tumor was considered a high-grade sarcoma based on mitotic rate, cellular anaplasia, the presence of necrosis, and its aggressive behavior. The clinical, radiologic, and histologic features all suggested a diagnosis of high-grade sarcoma consistent with malignant hemangiopericytoma. The subsequent resection specimen from the left proximal femur consisted almost entirely of necrotic metastatic tumor.

High-grade sarcomas located in the retroperitoneum and the pelvis have been well described. The most common location of malignant hemangiopericytoma is the lower extremity, while other common locations are the pelvic fossa and the retroperitoneum. Patients most commonly present with a painless mass between 20 and 70 years of age, with no male or female preponderance.[1]

Although grading is important for staging and prognosis, there is a lack of agreement upon standard criteria for the grading of sarcomas. Spindle-cell sarcomas are often graded from I to III, with III being the most malignant. The assignment is based on the degree of cellularity, the amount of cellular pleomorphism, and the mitotic activity. Other systems include the degree of necrosis as a component to grading.[2]

Retroperitoneal sarcomas are treated with as complete a surgical excision as possible. Because tumor-free margins are often difficult to achieve, radiation and adjuvant chemotherapy have also been used in trials, but with limited success.[3] Despite treatment, a significant percentage of tumors will recur locally or with metastases (usually pulmonary) within 5 years. Long-term follow-up is recommended for patients with malignant hemangiopericytoma because metastases have been shown to occur beyond 5 years.[4]

When evaluating the patient with a suspected retroperitoneal or pelvic mass, CT has been shown to be highly effective. In the female patient in whom a gynecologic lesion is suspected, pelvic ultrasound may be useful initially. Retroperitoneal tumors often reach a large size before causing symptoms from compressive effects on adjacent structures. Most commonly, a retroperitoneal mass is due to a sarcoma which demonstrates the CT appearance of a soft tissue mass

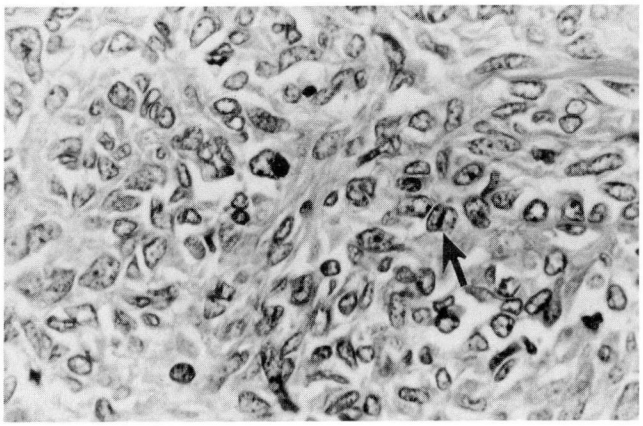

FIGURE 7. Photomicrograph showing hypercellularity, marked nuclear pleomorphism, and indistinct cytoplasmic borders. A cleaved nucleus is visible (arrow), which could represent new mitosis (hematoxylin and eosin, ×550).

causing displacement, compression, or obliteration of normal retroperitoneal structures. The presence or absence of invasion of adjacent structures cannot always be determined with certainty. Occasionally, CT findings may suggest a specific histologic diagnosis, such as well-differentiated liposarcoma, which may contain areas of fat attenuation, or leiomyosarcoma, which often contains areas of tissue necrosis and cystic degeneration.[5] MRI may be helpful for further evaluation of retroperitoneal or pelvic masses. Its multiplanar capability and excellent soft tissue contrast may be useful in evaluating the relationship to adjacent structures and in defining the extent of neoplasm. MRI signal characteristics may also be useful in differentiating benign lipoma from liposarcoma.[6]

The described radiologic features of hemangiopericytoma include a well-defined soft-tissue mass containing multiple areas of low attenuation, occasionally with small calcific foci. Marked contrast enhancement is present, and displacement of the gastrointestinal tract is common. Compression of adjacent viscera with associated hydronephrosis has been described. Angiography consistently demonstrates hypervascularity, and sonography demonstrates well-circumscribed hypoechoic lesions with areas of "through-transmission." MRI shows variable T1-weighted appearances, but T2-weighted images show very high signal intensity. Metastatic disease at the time of diagnosis is common, usually to the lungs and retroperitoneum. Metastases to bone, liver, and regional lymph nodes also occur.[7,8]

In this case, the metastatic lesion to the left proximal femur was mainly cystic due to necrosis with a lytic radiographic appearance (*Figure 1*). The large left retroperitoneal mass extending into the pelvis displaced the urinary bladder and obstructed the left distal ureter resulting in hydronephrosis with marked parenchymal loss of the left kidney, indicating obstruction of a chronic nature. Significant displacement and compression of the rectosigmoid colon and rectum were also present.

As in other reported cases, the well-defined mass, which correlates with encapsulation, was of predominantly soft-tissue attenuation by CT, with areas of low attenuation (*Figure 3*) that correspond to the necrosis and hemorrhage noted on gross examination of the specimen (*Figure 6*). Marked enhancement and a few calcific foci were also present. The sonographic findings of a mainly hypoechoic mass were also similar to that seen in other reported cases. MRI demonstrated decreased signal on the T1-weighted images (*Figure 4*) and very high signal intensity on the T2-weighted images. The early phases of the radionuclide bone scan indicated the hypervascular nature of this mass. Angiography confirmed its hypervascularity (*Figure 5*), and despite embolotherapy, marked hemorrhage was encountered at the time of resection. However, hemorrhage may have been greater without embolotherapy, making resection incomplete or impossible, as has occurred in several previously described cases.[7]

The recognition of hypervascularity in a retroperitoneal or pelvic mass and the determination of its relationship to adjacent structures are important in management and surgical planning. Contrast-enhanced CT and angiography are most effective by demonstrating the tumor's appearance, relationship to adjacent structures, vascularity, and blood supply. Preoperative arterial embolization may be extremely helpful in reducing tumor vascularity, minimizing the risk of intraoperative hemorrhage, and, therefore, allowing complete surgical excision.

ACKNOWLEDGMENTS

The authors wish to thank Charles Biscotti, MD, and Ralph Tuthill, MD, Department of Pathology, The Cleveland Clinic Foundation, for their assistance with the pathology discussion, and Sherry Kappa, Department of Diagnostic Radiology, for her assistance in the preparation of this manuscript.

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