Metastatic retroperitoneal hemangiopericytoma

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Hemangiopericytomas are rare mesenchymal lesions arising from pericytes within the walls of capillaries. They often have an unpredictable course. We present a case of a large retroperitoneal hemangiopericytoma in a 65-year-old woman who initially presented with upper gastrointestinal discomfort. Following exptirpative surgery, pathology was consistent with hemangiopericytoma of low malignant potential. Widespread metastasis was discovered on follow up imaging, 17 months following surgery. To our knowledge, this is the first case report demonstrating a primary retroperitoneal hemangioperictoma with confirmed metastases.

Key Words: hemangiopericytoma, retroperitoneal tumors, metastases

Introduction

Hemangiopericytomas are extremely rare tumors originating from pericytes that can be found surrounding small vessels and capillaries.[1] There are fewer than 200 case reports of these vascular tumors and they most commonly arise from soft tissue and bones in the lower extremities. Only about 25% of these cases involve the retroperitoneum or pelvis.[2] Hemangiopericytomas are usually well circumscribed and have been treated successfully with extirpative surgery. Histopathologically, these tumors have been shown to stain positive for CD34 and negative for desmin and actin on histology. Metastasis is reported to occur in 20% to 50% of cases.[2,3] With so few cases reported, chemotherapeutic regimens are not standardized.

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Case Report

A 65-year-old, otherwise healthy female, presented to her primary care physician complaining of indigestion and stomach discomfort. After failing conservative medical therapy, a CT scan was performed, revealing a large left retroperitoneal mass superior to the kidney. She was then referred to an endocrinologist where the work up for a functional adrenal mass was negative. The patient was eventually referred to our tertiary referral center where repeat imaging (MRI) demonstrated a stable lesion without metastasis, Figure 1. The differential diagnosis included renal angiomyolipoma, adrenal myelolipoma and retroperitoneal sarcoma. She was sent to interventional radiology for angiography and potential embolization for a presumed diagnosis of angiomyolipoma. Angiographic imaging revealed a discrete left peri-adrenal tumor separate from the left kidney that was supplied by large branches of the splenic, adrenal, and renal vasculature, Figure 2.

It was recommended that the patient undergo surgical resection. The patient underwent a left laparoscopic adrenalectomy and retroperitoneal mass excision. The mass was not adhered to the renal capsule, although it was intimate in proximity. Preoperative angiography was helpful in our surgical approach to identify unforeseen vessels and minimize intraoperative blood loss. The patient's postoperative recovery was uneventful, though it did involve a postoperative transfusion. The specimen measured 13.0 cm x 9.0 cm x 6.5 cm and weighed 380.43 grams. It consisted primarily of tumor attached to a small portion of uninvolved, residual adrenal gland. Pathological analysis revealed a solitary fibrous tumor consistent with a hemangiopericytoma of low malignant potential on the histologic basis of mitotic activity, Figure 3. The tumor cells were also strongly immunoreactive for CD34, suggesting a diagnosis of hemangiopericytoma.

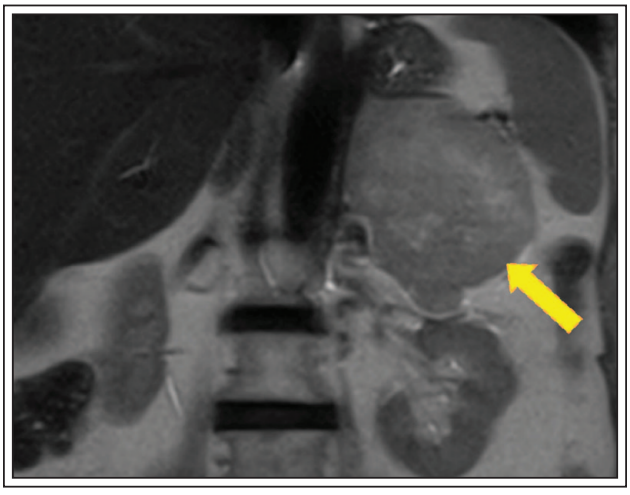

Figure 1. Abdominal MRI (coronal view) revealing left peri-adrenal mass (arrow).

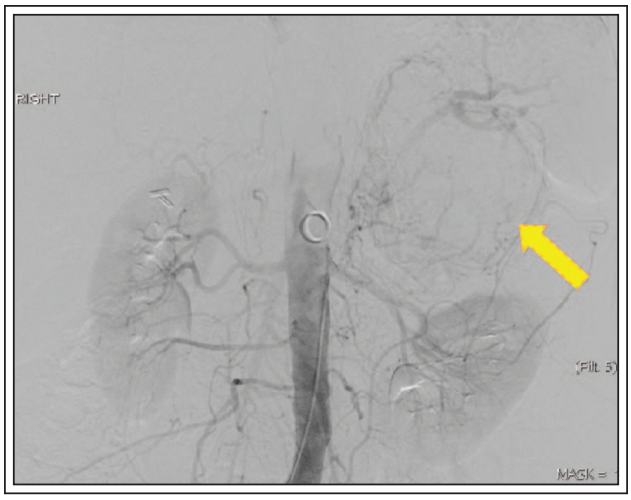

Figure 2. Abdominal angiography revealing peri-adrenal tumor (arrow) supplied by branches of the splenic, adrenal and left main renal arteries.

A consensus tumor board reviewed the pathology and the patient was evaluated 6 months after surgery with abdominal/pelvic MRI followed by yearly imaging thereafter. Despite remaining asymptomatic, the patient was found to have multiple metastatic deposits in the liver, lungs, and vertebrae (at the level of L1, L3, L4 and sacrum) on routine MRI seventeen months after surgery. The patient was referred to medical oncology and was initiated on systemic chemotherapy. Unfortunately she quickly succumbed to rapidly progressing disease.

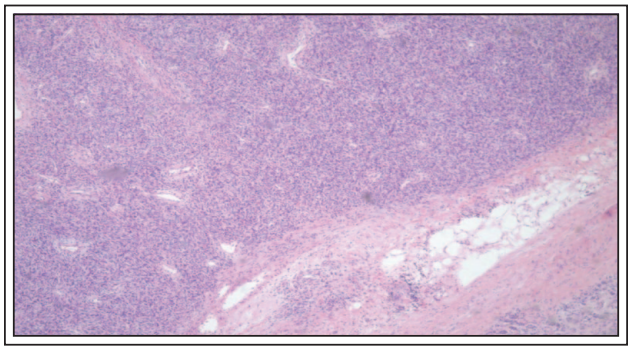

Figure 3. Histology specimen from primary tumor revealing perivascular hyalinization, spindle cells and positive staining for CD34 consistent with hemangiopericytoma.

Discussion

Hemangiopericytomas are rare mesenchymal lesions arising from pericytes within the walls of capillaries. They often have an unpredictable course. They most commonly occur within bony structures, and comprise only about 1% of vascular tumors. In general, hemangiopericytomas have a reported 5 year recurrence rate approaching 50%, with distant metastases most commonly found in the lungs, liver, and brain.[4,5] Although these tumors occasionally are immunoreactive with CD 34, there is a general lack of consensus on the utility of immunohistochemical staining to positively identify tumor cells. Furthermore, assessing the mitotic activity and other histological features that may suggest tumor aggression is misleading. For example, on pathological analysis, our patient's tumor stained positive for CD 34 and negative for HMB-45. However, there was minimal atypia, no necrosis, and no mitotic activity; all features that would suggest a tumor of low malignant potential.

This report describes a case of metastatic retroperitoneal hemangiopericytoma that was considered to have non-aggressive features after extirpative surgery. Seventeen months following laparoscopic excision, frank metastases to the liver, lungs, and vertebrae were discovered on imaging. The patient was subsequently treated with systemic bevacizumab and temozolomide. To our knowledge, this is the first case report demonstrating a primary retroperitoneal hemangioperictoma with confirmed metastases.

Conclusion

Hemangiopericytoma is a rare mesenchymal sarcoma, that when encountered, should be respected for its malignant potential. These tumors present a diagnostic challenge due to their ability to mimic other more commonly benign tumors (both radiographically and histologically). Further studies are necessary to determine the exact role or prognostic value of preoperative biopsy. Following extirpative surgery, even in the setting of a favorable histopathology, these patients should be followed with frequent follow up imaging to monitor local recurrence and distant metastasis. □

References

1. Stout AP, Murray MR. Hemangiopericytoma: a vascular tumor featuring Zimmermann's pericytes. *Ann Surg* 1942;116(1):26-33.
2. Enzinger FM, Smith BH. Hemangiopericytoma. An analysis of 106 cases. *Hum Pathol* 1976;7(1):61-82.
3. Malkowicz SB. Retroperitoneal Tumors: Campbell-Walsh Urology 10th edit. Campbell MF, Wein AJ, Kavoussi LR. Philadelphia, Pa. Saunders Elsevier 2011:1509.
4. Espat NJ, Lewis JJ, Leung D et al. Conventional hemangiopericytoma: modern analysis of outcome. *Cancer* 2002;95(8):1746-1751.
5. Hansen CP, Francis D, Bertelsen S. Primary hemangiopericytoma of the lung. Case report. *Scand J Thorac Cardiovasc Surg* 1990;24(1):89-92.