Meningeal hemangiopericytomas are rare vascular tumors that have a propensity for recurrence and metastasis. Intracranial hemangiopericytomas are rare vascular tumors. They account for 0.5% of primary central nervous system tumors and 2% of meningiomas. Unlike usual benign meningiomas, which rarely metastasize extracranially, meningeal hemangiopericytoma has a high rate of local recurrence and distant metastasis. The treatment paradigms for hemangiopericytomas and meningiomas differ based on their biological behaviors. Hemangiopericytomas have higher rates of recurrence and metastasis compared with meningiomas. Intracranial meningeal hemangiopericytoma is characterized by clinically repeated local recurrences at the primary site. Bone, liver, lung, central nervous system, and abdominal cavity are the most commonly reported sites of metastasis in hemangiopericytomas.

This article describes a case of bone metastasis with extensive involvement of the scapula from intracranial hemangiopericytoma. Bone metastasis can be seen in a relatively late phase of the disease, with metastasis to other organs. Although radiation therapy is effective in controlling pain from bone metastases in unresectable disease and those with extensive visceral metastases, aggressive local surgical control of a solitary bone metastasis may be an option for patients with limited distant disease. The diagnosis may be initially confused with clear cell meningioma and benign meningiomas. The management of bone metastasis is not well reported in the orthopedic literature.

Figure: Anteroposterior radiograph showing proximal humerus and total scapula replacement with a constrained endoprosthesis (Stryker Orthopaedics, Mahwah, New Jersey).

HERRICK J. SIEGEL, MD; ROBERT LOPEZ-BEN, MD; J. HUNTER SUTTON, BA; GENE P. SIEGAL, MD, PHD
Intracranial hemangiopericytomas are rare vascular tumors. They account for 0.5% of primary central nervous system tumors and 2% of all meningiomas. Unlike usual benign meningiomas, which rarely metastasize extracranially, meningeal hemangiopericytomas have a high rate of local recurrence and distant metastasis. The treatment paradigms for hemangiopericytomas and meningiomas are based on their different biological behaviors. Hemangiopericytomas have much higher rates of recurrence and metastasis compared with meningiomas. Bone, liver, lung, central nervous system, and abdominal cavity are the most commonly reported sites of metastasis. This article describes a case of bone metastasis with extensive involvement of the scapula from intracranial hemangiopericytomas.

CASE REPORT

A 51-year-old woman with several years of recently worsened left shoulder pain presented to our institution following referral by her primary care physician. Pain was rated 4/10, and she had limited motion due to pain. Her medical history was remarkable for resection of a posterior fossa clear cell variant meningioma at age 38 and gamma knife treatment for recurrence 5 years prior to presentation to our institution.

On physical examination, a palpable, nontender mass was noted diffusely about the shoulder and dorsal scapula along with a nonremarkable neurovascular examination and with no lymphadenopathy. Radiographs of her left shoulder showed an expansile, lytic lesion involving the lateral portion of the scapula, which appeared to be eroding into the glenoid cavity (Figure 1). The lesion was noted to have a well-demarcated sclerotic border medially and inferiorly. T1-weighted magnetic resonance imaging (MRI) showed a lobulated mass apparently originating in the scapula and exhibiting an exophytic growth pattern into the soft tissues. The mass had an intermediate T1-weighted signal with punctate foci of high T1-weighted signal. The T2-weighted MRI also showed an increased T2-weighted signal with prominent septations throughout the lobulated mass and strandy linear areas of decreased T2-weighted signal.

Fine needle aspiration was performed to rule out a sarcoma or lymphoma. A spindle cell and epitheliod clear cell lesion was identified, and a lymphoma was excluded. An open incisional biopsy was then performed, and microscopically prominent features were those of polygonal cells with indistinct borders, vascular spaces, and collagen deposition in a myxoid background (Figure 2). An increased mitotic rate was appreciated, but abnormal mitoses and significant necrosis were not seen. The immunophenotype revealed the following: VIM (+), EMA (−), PR (−), CD31 (−), LCA (−), CK (−), CD99 (+), and F VIII Rag (+).

After a multi-institutional review of the previously resected brain mass slides and current shoulder specimen, it was concluded that the best diagnosis for the scapula lesion was meningeal anaplastic hemangiopericytoma. A wide resection was performed with clear margins (Figure 3), and endoprosthetic reconstruction with a total scapula and proximal humeral replacement was performed to reconstruct the shoulder girdle (Figures 4, 5). A latissimus dorsi rotational flap was performed as part of the closure to provide adequate soft tissue coverage over the implant and improve cosmesis (Figure 6).

At 7-year follow-up, the patient was pain free, had maintained functional use of the extremity, and was able to perform all activities of daily living, including working as a full-time college professor. The patient was pleased with the cosmetic results and could abduct and forward flex her shoulder to 80°. No evidence of disease existed in the region of the shoulder. However, the brain lesion recurred 2 years postoperatively from the shoulder reconstruction and was subsequently treated with stereotactic radiation. The brain lesion was finally treated with Gamma knife.
Hemangiopericytoma is a rare tumor that was first described and named by Stout and Murray in 1942. This tumor arises from pericapillary cells or pericytes of Zimmerman and can occur anywhere capillaries are found. However, hemangiopericytoma develops mainly in the lower extremities, retroperitoneum, or pelvis and rarely occurs in the larynx, spleen, bone, meninges, or thorax. The peak incidences of hemangiopericytoma are in the fourth and fifth decades of life. However, it is sometimes a locally aggressive and malignant tumor that produces late local recurrences and distant extraneural metastases.

Meningeal hemangiopericytoma is rare, and its incidence is <1% of all central nervous system tumors and 2.4% of meningiomas. The diagnosis of hemangiopericytoma rests on recognition of the architectural pattern and on immunohistochemistry. Hemangiopericytoma is typically cellular and composed of small oval cells, with numerous branching thin-walled vessels of varying calibers, giving a characteristic "staghorn" vascular pattern. Reticulin stain may be helpful in showing individual cell enmeshing. Lack of epithelial membrane antigen and S-100 in the presence of positive staining for vimentin, factor XIIIa and Leu-7 has been considered sufficient to distinguish meningeal hemangiopericytoma from other histological mimics, including fibrous meningiomas and clear cell meningioma.

Meningeal hemangiopericytomas are locally aggressive tumors with a high rate of local recurrence and distant metastasis. In their review of 94 cases, Mena et al reported a 70% recurrence rate and a 27% metastasis rate. Metastases commonly occur in the bone, lung, and liver, but reports of metastases at other sites have been published. Although metastases typically occur 5 to 8 years after diagnosis, they can occasionally be delayed until 20 years after diagnosis.

Bone metastases manifest as osteolytic lesions on plain radiographs. The role of a radionuclide bone scan is limited because metastases may not uptake the isotope, as in 1 of the lesions in this case. Local recurrence tends to predate the onset of distant metastasis in the majority of cases.

Surgery is currently regarded as the main modality of treatment for meningeal hemangiopericytoma, and preoperative tumor embolization has been reported to be useful in reducing the vascularity. In their series of 44 patients treated with postoperative radiotherapy, Guthrie et al reported a significantly longer recurrence-free interval and longer survival. Conservative surgery and postoperative radiotherapy has been suggested, especially for tumors in unfavorable locations. A dose of 50 to 60 Gy is thought to be adequate to reduce the rate of local recurrence.

Chemotherapy has generally been disappointing in the management of metastatic disease. In the series reported by Galanis et al, 1 patient of 7 showed a partial response to doxorubicin. However, isolated case reports of tumor control using a combination of ifosfamide and epirubicin have been published. From current evidence, meningeal hemangiopericytoma is best treated by surgical excision.

Postoperative radiotherapy is recommended to improve local control, particularly in patients for whom complete surgical clearance of the tumor is not possible due to its unfavorable location. Patients need prolonged follow-up because distant metastases are generally a late phenomenon. These tumors are sometimes misdiagnosed as atypical meningiomas, but an accurate histological diagnosis of meningeal hemangiopericytoma is important at the time of initial presentation because these tumors have a different natural history to atypical meningiomas and should be managed accordingly.
The presenting bone metastases were commonly detected by the clinical report of pain. The pain is often insidious in onset and misdiagnosed as inflammatory pain or muscle injury. It is common to find additional metastasis to other organs, such as the liver, lung, and pancreas, at the time of bone metastasis presentation. On plain radiographs, bone lesions are generally well circumscribed, with cortical thinning and expansion. Calcification, which is commonly seen in meningiomas with bone involvement, has not been reported with hemangiopericytoma. On bone scintigraphy, cold-in-hot features have been described. These combined findings of the radiographic features are specific for the diagnosis of bone metastasis from this tumor.15,20

Galanis et al14 and Guthrie et al2 emphasized the importance of complete removal at the first operation to prolong the time to recurrence and extend survival. They also insisted that aggressive surgical management is important for the successful treatment of patients with recurrent hemangiopericytoma. Despite the tumor’s natural tendency to recur and spread outside the central nervous system, it is possible to ensure a long survival time. Palliative radiation of painful bone lesions has been reported to alleviate pain successfully at doses of 30 Gy/10 fractions.57,16 The long-term benefit of wide resection of a solitary metastatic bone lesion is unknown. Palliative therapy with irradiation for bone metastases could provide good local control and maintain the patients’ quality of life during the remainder of their lives.

Intracranial meningeal hemangiopericytoma is characterized by clinically repeated local recurrences at the primary site. Bone metastasis can be seen in a relatively late phase of the disease, with metastasis to other organs. Although radiation therapy is effective in controlling pain from bone metastases in unresectable disease and those with extensive visceral metastases, aggressive local surgical control of a solitary bone metastasis may be an acceptable option for patients with limited distant disease.21

REFERENCES