Rare meningeal-derived malignant hemangiopericytoma/solitary fibrous tumor grade II-III presenting as a subcutaneous mass on the scalp

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INTRODUCTION

Hemangiopericytoma, also known as a solitary fibrous tumor, is a rare, highly vascular, soft tissue tumor derived from pericytes, which are mesenchymal cells that line capillaries.[1] It was first described in the literature by Stout and Murray[2] in 1942 as a complex neoplasm composed of capillaries and perivascular cells, but lacking organoid features. Telomerase reverse-transcriptase promoter mutation is associated with clinically aggressive types of hemangiopericytoma. Hemangioperiocytomas (HPC) are locally destructive tumors with various malignant potential. Metastasis can occur by hematogenous spread to lung, lymph nodes, or bone.[3] Diagnosis is often controversial because of nonspecific clinical findings, lack of diagnostic immunohistochemical stains, and limited guidelines on classification and prognosis.[4,5] We present a case of a rare, meningeal-derived, solitary, fibrous tumor arising on the scalp of an elderly man.

Abbreviations used:

HPC: hemangioperiocytomas
STAT6: signal transducer and activator of transcription 6

CASE REPORT

A 68-year-old white man with significant medical history including malignant melanoma of the skin presented to the dermatology clinic with a slowly enlarging mass on the frontal aspect of the scalp for 5 months. On physical examination, a 5-cm fluctuant subcutaneous mass was present on the midline frontal aspect of the scalp, clinically consistent with a hematoma (Fig 1). Because of clinical suspicion, magnetic resonance imaging without contrast was performed, which revealed a soft tissue mass in the frontoparietal junction that caused displacement of the sagittal sinus. The patient was referred to plastic surgery for surgical removal.

During removal of the tumor by the plastic surgeon, the mass appeared to penetrate the skull and complete removal was not possible. Gross examination revealed a well-circumscribed, 6.0 × 4.0 × 3.5-cm, gray-white, firm mass. Microscopic evaluation at low power demonstrated a relatively uniform, densely packed, hypercellular proliferation of hyperchromatic blue cells with a high nuclear to cytoplasmic ratio, admixed with areas demonstrating slight cytoplasmic pallor and small foci of hemorrhage. The architecture was generally patternless, and telangiectatic, thin-walled, "staghorn,"

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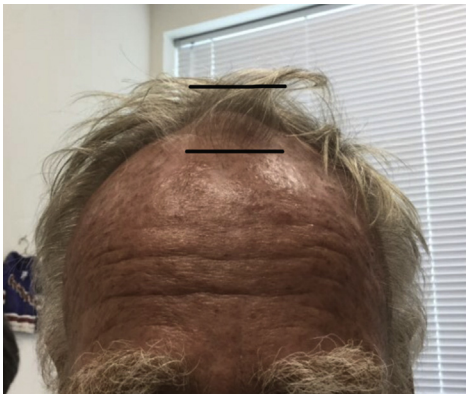

Fig 1. A 5-cm large subcutaneous nodule on the mid scalp.

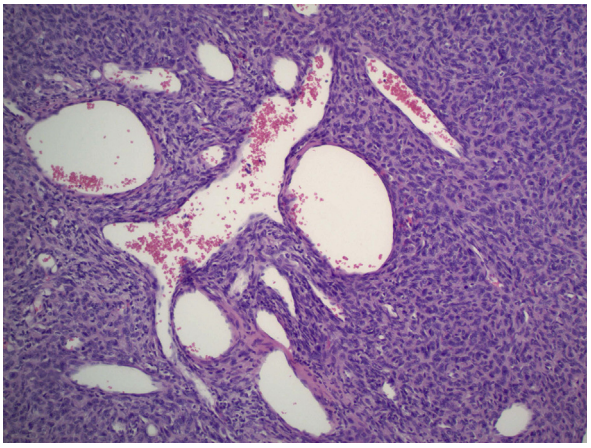

Fig 2. Telangiectatic thin-walled "staghorn" blood vessels in a hypercellular patternless spindle and epithelioid blue cell tumor, Hematoxylin and eosin stain. (Original magnification: ×200.)

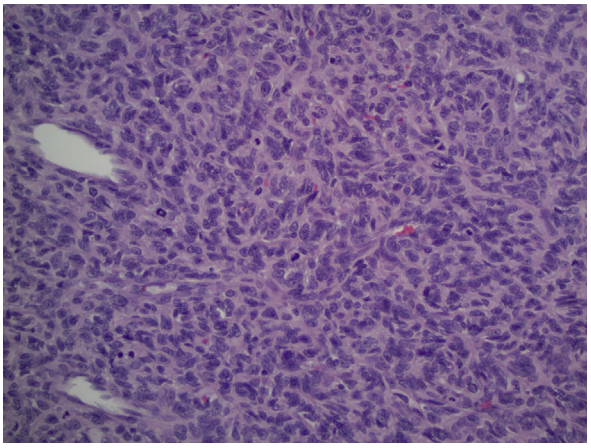

Fig 3. Patternless hypercellularity with variable nuclear pleomorphism and frequent mitoses including single atypical "ring" mitosis, Hematoxylin and eosin stain. (Original magnification: ×100.)

branching blood vessels were noted, particularly toward the tumor periphery (Fig 2). The neoplasm consisted of a mixture of hyperchromatic, atypical, spindled and epithelioid cells with focal areas showing significant cellular pleomorphism and scattered foci of necrosis. Areas with high mitotic activity including the presence of atypical mitoses were readily identifiable (Fig 3). Tumor cells were positive for signal transducer and activator of transcription 6 (STAT6), *B-cell lymphoma 2*, *cluster of differentiation 99*, and vimentin and were negative for pancytokeratin AE1/AE3, keratin 34 beta E-12, cytokeratin 5,2, epithelial membrane antigen, P40, S-100 protein, melanoma antigen recognized by T cells, human melanoma black, desmin, and progesterone receptor. Thus, an initial diagnosis of hemangiopericytoma was made.

Neurosurgery was consulted and further evaluated the tumor with magnetic resonance imaging with and without contrast, which showed a 4.4 × 3.7-cm, midline, posterior, frontal, calvarial-enhancing, hypercellular tumor with subgaleal and extradural extension along with localized superior sagittal sinus thrombosis. Neurosurgery performed a bifrontal craniectomy with overlying graft and cranioplasty for gross total removal of the mass. Pathology reconfirmed the tumor as a STAT6—positive malignant hemangiopericytoma/solitary fibrous tumor, World Health Organization grade II to III. Postoperative radiation therapy was completed in accordance with recommendations from oncology.

DISCUSSION

HPCs and solitary fibrous tumors are exceedingly rare and represent approximately 1% of all angiogenic tumors.[6] They most commonly occur on the lower extremities and trunk, followed by the head and neck region.[3,5] Only a handful of malignant, transcalvarial, meningeal-derived, solitary fibrous tumors have been reported. Although the etiology of HPCs is unknown, numerous factors have been associated with an increased incidence, such as trauma, steroid use, and hormone imbalances.[7]

In 2016, the World Health Organization reclassified solitary fibrous tumor and hemangiopericytoma as 1 entity and created a classification system with grades 1 to 3. Grade 1 is classified as a spindle cell tumor with low cellularity. Grade 2 corresponds to a spindle cell tumor with more cellularity, less collagen thickening, and staghorn vessels. Grade 3 is classified as a spindle cell lesion with the presence of 5 or more mitotic figures.[8]

HPCs can present in any age group but most commonly occur in adults between the fifth and

seventh decades of life. Two different clinical variants are described: infantile hemangiopericytoma, which presents in children younger than 1 year, and the more common variant, adult hemangiopericytoma. Infantile hemangiopericytoma most commonly occurs in the oral cavity and is associated with response to chemotherapy and overall better prognosis than the adult hemangiopericytoma variant.[4]

The majority of malignant HPCs occur as encapsulated soft tissue tumors with gradual growth, with various presentation based on location site.[4] Masses are usually asymptomatic but may cause pain once they become locally destructive and displace adjacent structures. Intracranial and central nervous system-derived HPCs, as observed in our case, typically arise from the dura.[9] Radiologic findings are nonspecific, demonstrating a well-circumscribed soft tissue mass that may appear lobulated and which enhances with contrast.

Although imaging can assist in the evaluation for malignant hemangiopericytoma, diagnosis requires histologic examination. Histology of classic HPCs consists of tightly arranged spindle cells around a central lumen lined by endothelial cells. Blood vessels exhibit a classic staghorn pattern of branching.[7] The presence of mitotic figures, cellular atypia, and necrosis indicates a diagnosis of malignant hemangiopericytoma.[3] Immunohistochemistry can aid in the diagnosis of HPCs, but stains are inconsistently expressed and therefore may create difficulty in the diagnosis of atypical cases. Typical immunohistochemical markers of HPCs include expression of CD34, B-cell lymphoma 2, cluster of differentiation 99, and vimentin. More specific and sensitive markers for HPCs include STAT6, which is classically confined to the nucleus of hemangiopericytoma cells, and GRIA2, a novel diagnostic marker for hemangiopericytoma found through gene-expression profiling.[9] In a study by Bertero et al,[10] 93.3% of HPCs stained positively for STAT6. HPCs are typically negative for the expression of actin, desmin, S100, MDM2, CDK4, and epithelial markers epithelial membrane antigen and low-molecular-weight cytokeratins.[10]

Although some HPCs are benign, with a good prognosis and few recurrences, the prognosis for malignant HPCs is variable. Treatment options for hemangiopericytomas include surgery, radiation, and chemotherapy. For localized tumors, surgery with wide local excision is the first-line treatment.[4] Radiation therapy is recommended on a case-by-case basis for recurrent, incompletely resected, or central nervous system-derived HPCs. Like radiation therapy, the role of chemotherapy in the treatment of HPCs is controversial mainly because of the lack of studies available on this rare tumor.

Our case demonstrated malignant hemangiopericytoma arising after head trauma. Because malignant HPCs can have unusual and nonspecific presentations, as observed in our case, the dermatologist must be aware of this entity. Physicians must have a high clinical suspicion to provide a reasonable time to diagnosis. Because of the varying nature of malignant HPCs and solitary fibrous tumors, long-term follow-up of patients with this tumor is crucial.

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