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Metastatic cervical paravertebral solitary fibrous tumor detected by fluorodeoxyglucose positron emission tomography-computed tomography

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ABSTRACT

Solitary fibrous tumors/hemangiopericytomas (SFT/HPC) are soft tissue tumors that can arise from the abdomen, pleura, head and neck, or extremities. We report an unusual case of recurrent hemangiopericytoma in a 67-year-old female presenting with a painless and palpable mass within her right posterior neck. Eight years after initial resection of the mass, a follow-up MRI showed multiple enlarging calvarial lesions. A whole body FDG-PET/CT revealed not only hypermetabolic calvarial lesions but also numerous hypermetabolic axillary node and osseous metastases. Though the majority of these soft tissue tumors exhibit benign behavior and carry a favorable prognosis, patients with these slow growing tumors are at risk for local recurrence and distant metastases which demonstrate substantial FDG avidity. Additional studies are needed to clarify the role of whole body FDG-PET/CT in the surveillance of SFT/HPC to detect recurrent or metastatic lesions.

Introduction

Hemangiopericytoma or solitary fibrous tumors (SFT/HPC) are rare mesenchymal tumors that often present as a painless mass most commonly arising from the abdomen and pleura, though they can also be found in the extremities, head and neck or trunk, and central nervous system. Although the majority of these tumors exhibit benign behavior and have a good prognosis, physicians must be aware that a minority of patients with these slow-growing tumors are at risk for local recurrence and distant metastases [1–3]. Here, we present an unusual case of cervical paravertebral SFT/HPC with both local recurrence and widespread metastatic disease detected by fluorodeoxyglucose positron emission tomography-computed tomography (FDG-PET/CT).

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Case report

A healthy 67-year-old female presented to her primary care physician with a painless mass in her right posterior neck. A magnetic resonance imaging (MRI) of the cervical spine showed a paravertebral soft tissue mass (Fig. 1), which was resected in February 2007 and treated with adjuvant radiation. Pathological examination revealed a highly cellular spindle cell neoplasm with frequent staghorn vessels and thick bands of collagen accompanied by scattered multinucleated giant cells, which was diagnosed as a hemangiopericytoma (HPC). Subsequent staging computed tomography (CT) of the chest, abdomen, and pelvis at the time showed no other sites of involvement. Serial surveillance MRI scans of the cervical spine were ordered from 2008 to 2014, which showed minimally enhancing scar tissue and post-treatment changes in the neck, but no new mass-like areas of enhancement or cervical lymphadenopathy suspicious for recurrence or local metastases.

However, an MRI of the cervical spine performed on April 2015 revealed 3 new nodules within the right posterior occipital calvarium measuring up to 9 mm in diameter (Fig. 2) suspicious for local metastases. A review of prior studies revealed that in retrospect, these slow-growing nodules were present on an earlier MRI from February 2013. In June 2015, she underwent a resection of the calvarial lesions and pathology confirmed recurrent HPC. Microscopic examination also showed that the tumor demonstrated increased nuclear pleomorphism and a mitotic index that had increased from 1 mitosis to 5 mitoses per high-powered field, consistent with a Ki-67 index of 8%-10%. Additionally, a STAT6 immunohistochemical stain demonstrated strong nuclear staining, consistent with a diagnosis of recurrent SFT/HPC with increased mitotic activity.

Given the recurrent nature of her malignancy, the patient underwent a staging whole body FDG-PET/CT, which showed

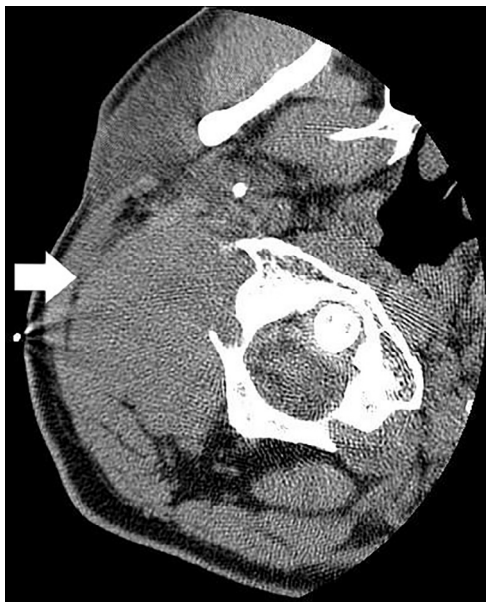

Fig. 1 – Noncontrast enhanced computed tomography demonstrating original appearance of patient's posterior cervical neck mass.

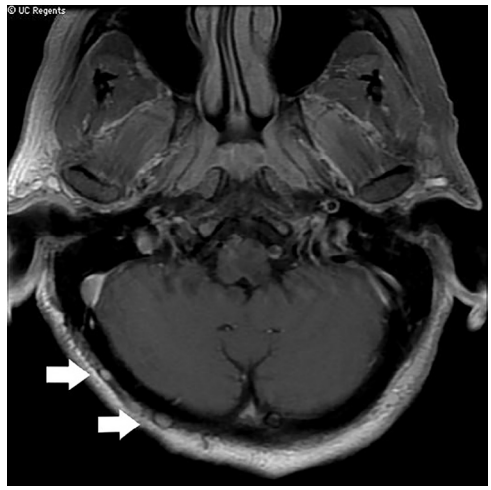

Fig. 2 – Calvarial lesions representing metastatic hemangiopericytoma or solitary fibrous tumors seen on contrast-enhanced T1-weighted brain magnetic resonance imaging.

distant metastases, including numerous hypermetabolic metastases within the right axillary lymph nodes and bony pelvis involving the right acetabulum, right sacrum, and left posterior iliac bone (Fig. 3) as well as multiple hypermetabolic, ovoid, and variably lytic or sclerotic lesions within the calvarium (Fig. 4).

Discussion

Hemangiopericytomas (HPC) have historically represented a difficult pathologic diagnosis given the substantial histologic overlap with solitary fibrous tumors (SFT) as well as synovial sarcomas [4,5]. However, recent studies have shown that HPC and SFT arise from the same inciting mutation, specifically, an inversion at 12q13 with a fusion of the *NAB2* and *STAT6* genes resulting in nuclear expression of *STAT6* that can be detected by immunohistochemistry. These findings confirm that HPC and SFT, previously thought to be unique tumors, are essentially equivalent entities with varying histology and aggressiveness [6,7]. This has resulted in a complicated nomenclature, with soft tissue pathologists favoring the term solitary fibrous tumor while neuropathologists prefer the term hemangiopericytoma when such tumors arise from the central nervous system (CNS) given the clinical implications of the term, including high rates of recurrent disease and risk of distant metastasis many years after initial resection, as seen in this case [8].

Although soft tissue pathologists now exclusively utilize the term SFT, neuropathologists have grouped these two entities together under the term *solitary fibrous tumor/hemangiopericytoma* and created a unique grading system that has been reflected in the 2016 update to the World Health Organization Classification of Tumors of the CNS. A grade I SFT/HPC is characterized by a spindle cell lesion with abundant collagen whereas lesions accompanied by "staghorn" vasculature represent a grade II SFT/HPC and can also be referred to as a hemangiopericytoma. Tumors characterized by high mitotic activity (5 + mitoses/