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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

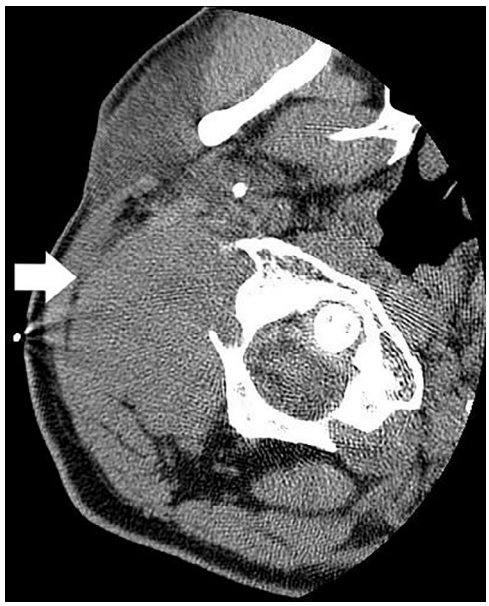

Fig. 1 – Noncontrast enhanced computed tomography demonstrating original appearance of patient's posterior cervical neck mass.

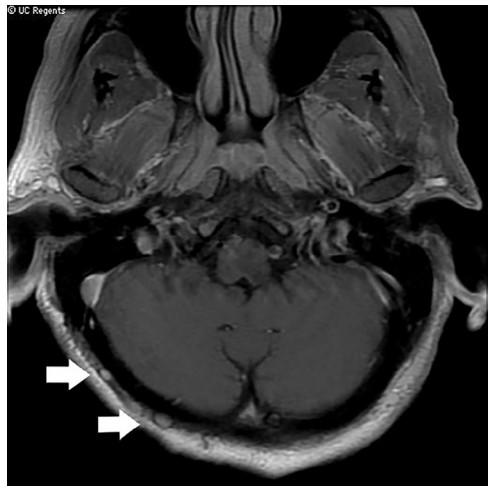

Fig. 2 – Calvarial lesions representing metastatic hemangiopericytoma or solitary fibrous tumors seen on contrast-enhanced T1-weighted brain magnetic resonance imaging.

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/

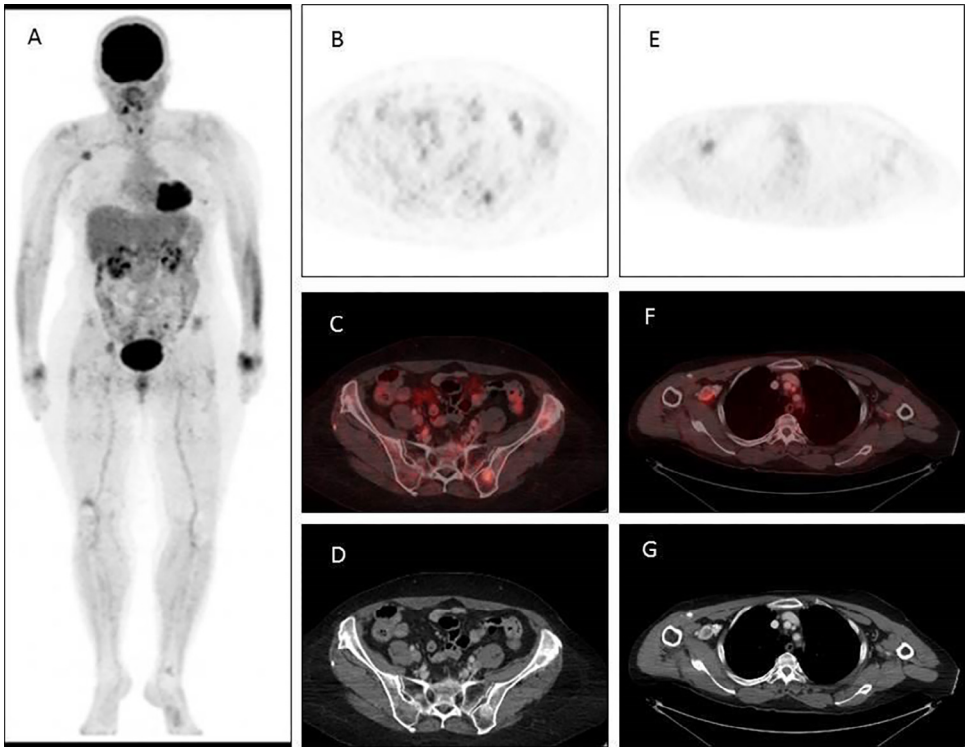

Fig. 3 – Fluorodeoxyglucose positron emission tomography-computed tomographyimages. (A) MIP (maximum Intensity Projection) whole body FDG image demonstrating hypermetabolic osseous and soft tissue metastases. (B) FDG, (C) hybrid, and (D) computed tomography (CT) images demonstrating a hypermetabolic sclerotic left iliac bone metastasis. (E) FDG, (F) hybrid, and (G) CT images demonstrating an enlarged hypermetabolic right axillary lymph node suspicious for metastases.

high-powered field) meet criteria for grade III SFT/HPC also known as an anaplastic hemangiopericytoma [8].

In this particular case, though diagnosed as an HPC at initial presentation, the tumor appears to be confined to the paravertebral soft tissue without CNS involvement and would likely be labeled an SFT under the current terminology. Following recurrence of her SFT/HPC many years later, the histology of her tumor had changed significantly, with greater than 5 mitoses per high powered field and involvement of the calvarium. Though increased mitotic activity is associated with more aggressive clinical behavior [1–3], even low-grade tumors with bland histology have malignant potential and require surveillance for distant metastases [9].

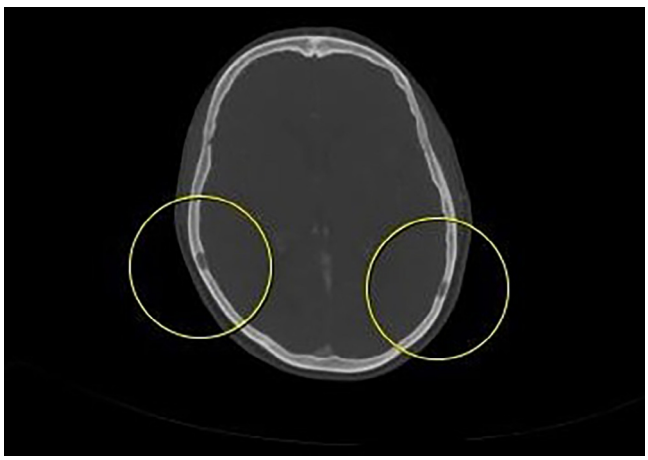

Fig. 4 – Head computed tomography showing lytic and sclerotic lesions within the calvarium with associated hypermetabolism representing osseous metastases.

Treatment for these tumors primarily consists of surgical resection. They often carry a relatively favorable prognosis, with 5- and 10-year disease-specific survival rates of 89% and 73% respectively and metastasis-free survival rates at 5 and 10 years estimated at 74% and 55%, respectively [1]. More recent population-based estimates for 5-, 10-, and 20-year disease-specific survival for SFT/HPCs arising specifically from the head and neck are similar, 84.0%, 79.4%, and 69.4% respectively. Adjuvant chemotherapy, although not uncommon, remains controversial as studies have failed to demonstrate a survival benefit. However, as this case and prior studies have illustrated, 10%-20% of these tumors demonstrate aggressive behavior and develop local recurrence as well as distant metastases years after initial diagnosis [1,10].

Though SFT/HPCs are uncommon, this case demonstrates the need for physicians to be aware of the malignant and recurrent potential of these tumors. The slow-growing nature of these lesions can make local recurrence difficult to detect on CT or MRI, especially when set within a background of post-surgical changes. Other case reports have also shown that SFT/HPCs with recurrent/metastatic lesions can demonstrate substantial FDG avidity [11,12]. One study with limited sample

size has even suggested that the level of FDG avidity is correlated with the malignant potential of pleural SFTs [13]. However, larger-scale studies are needed to determine how frequently SFT/HPCs demonstrate FDG avidity and whether those findings correlate with pathologic grade as well as clinical outcomes such as rates of metastases and survival.

In conclusion, though soft-tissue SFT/HPCs are slow-growing mesenchymal tumors that tend to have a favorable prognosis, their risk of local recurrence and distant metastasis is non-negligible. We illustrate in this case that these tumors can metastasize silently to soft tissues and bones many years after initial treatment which can then be detected by FDG-PET/CT, though they can be missed on other common imaging modalities due to their indolent nature. This case highlights the need for additional research that examines the role of nuclear medicine in the surveillance and follow-up of these SFT/HPCs given the known long-term risk of distant metastases regardless of tumor histology.

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