Malignant Solitary Fibrous Tumor

Solitary fibrous tumor/hemangiopericytoma (SFT/HPC) is a combined entity for which a soft-tissue–type grading system, ranging from grades I to III, was introduced in the 2016 WHO classification of tumors of the CNS.

Hemangiopericytomas were first described in 1942 and initially thought to be a vascular neoplasm related to smooth muscle perivascular cells known as pericytes [1]. However, the diagnosis was largely descriptive, based upon a nonspecific, albeit characteristic, staghorn vascular pattern, and the term became a “wastebasket” diagnosis, which included a variety of unrelated benign and malignant entities. With the advent of diagnostic immunohistochemistry (IHC) and cytogenetic analysis, pathologists were able to exclude histologic mimics, and hemangiopericytoma became more accepted as a distinct entity, although largely a diagnosis of exclusion, until the recognition of phenotypic and behavioral overlap with solitary fibrous tumor (SFT) led pathologists to consider the two as one tumor type.

The discovery of a shared, recurrent, and thus far unique gene fusion in SFT and tumors histologically identified as hemangiopericytoma has confirmed the identical nature of these tumors. At present, the term SFT is preferred and use of the term “hemangiopericytoma” is discouraged in clinical practice. However, some neuropathologists still prefer the term hemangiopericytoma in reference to meningeal tumors to emphasize their aggressive behavior compared with other SFT.

see Spinal hemangiopericytoma.

Solitary fibrous tumor (SFT), a rare mesenchymal neoplasm affecting mainly the visceral pleura, was first described as a primary spindle-cell tumor of the pleura by Klemperer and Rabin in 1931.

After increased recognition of this lesion outside of the thorax, it has also been described in a number of head and neck locations, including the orbit, nasal cavities, paranasal sinuses, thyroid glands, parotid glands, and buccal and parapharyngeal spaces.

see Primary Intracranial Solitary Fibrous Tumor

Hemangiopericytoma (HPC) is a term formerly used to describe a continuum of mesenchymal tumours with elevated cellularity found throughout the body in soft tissue and bone. As the growth pattern of HPC is shared by numerous unrelated benign and malignant tumours, the term as a group of these tumour is now avoided.

This is still a controversial subject among pathologists.

Gengler and Guillou proposed the relocation of haemangiopericytoma tumours in 3:

non-HPC tumours that occasionally display HPC-like features:

synovial sarcoma

tumours with a clear evidence of myoid or pericytic differentiation and corresponding to true haemangiopericytomas:

HPC of spleen

dural haemangiopericytomas

myopericytoma

infantile myofibromatosis

sinonasal HPCs.

solitary fibrous tumors

History and etymology

The term was first used by the American pathologists Arthur Purdy Stout and Margaret Ransone Murray in 1942 to describe a soft tissue tumour presumably of pericytic origin, with a monomorphic population of compact polygonal or fusiform cells and a branching stromal vascular pattern with a “staghorn” form.

Intracranial solitary fibrous tumor/hemangiopericytoma

Intracranial solitary fibrous tumor/hemangiopericytoma.

Case reports

male, age 15, who had migraines for 2 months before the diagnosis of an intracranial tumor. Computed tomography and magnetic resonance imaging showed a mass located in the right parietooccipital region with surrounding edema and a compressed right lateral ventricle. Neurosurgery was utilized to successfully remove the mass, and a single intracranial fibrous tumor (grade I) was identified by postoperative pathological analysis. During an 8-month follow-up period, the patient did not experience any recurrences.

Lessons: SFT is often misdiagnosed as meningioma because of their similar imaging characteristics. However, identifying the distinctive features of SFT on magnetic resonance imaging can distinguish it from meningioma and help to select the appropriate treatment. The complete preoperative imaging data for this case indicated a highly vascularized tumor. Preoperative vascular embolization treatment reduced any difficulties during the subsequent tumor resection and minimized intraoperative bleeding [2)].

An 86-year-old man sustained progressive motor weakness in the left lower extremity for 1 month. Cranial computed tomography (CT) revealed an isodense mass in the right parietal lobe, with a smooth-contoured focal erosion in the adjacent parietal bone. The extra-axial tumor appeared isointense on T1- and hyperintense on T2-weighted magnetic resonance imaging with intense enhancement. On three-dimensional CT angiography, the ectatic left occipital artery coursed into the right parietal foramina and connected with a dilated meningeal vessel supplying the tumor. The focal erosion formed in the inner parietal bone was located adjacent to the feeding vessel. A total tumor

resection was achieved. The microscopic findings of the resected specimen were consistent with a World Health Organization grade III hemangiopericytoma (HPC). Bone erosion and peripheral feeding vessels may be characteristic findings of intracranial solitary fibrous tumor (SFT)/HPC. Careful interpretation of neuroimages could help detect clues for distinguishing an SFT/HPC masquerading as a meningioma from a true meningioma [3)]

A 43-year-old woman with a tumour within the left lateral ventricle with the typical appearances of meningioma on MRI. (1)H MR spectroscopy demonstrated an increased choline peak, suggesting a malignant form of meningioma. Histologically a haemangiopericytoma was found, an exceptionally rare tumour at this site [4)].

1)
Stout AP, Murray MR. HEMANGIOPERICYTOMA: A VASCULAR TUMOR FEATURING ZIMMERMANN'S PERICYTES. Ann Surg. 1942 Jul;116(1):26-33. PubMed PMID: 17858068; PubMed Central PMCID: PMC1543753.
2)
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3)
Sugiyama H, Tsutsumi S, Hashizume A, Inaba T, Ishii H. Are bone erosion and peripheral feeding vessels hallmarks of intracranial solitary fibrous tumor/hemangiopericytoma? Radiol Case Rep. 2022 Jun 1;17(8):2702-2707. doi: 10.1016/j.radcr.2022.04.050. PMID: 35669225; PMCID: PMC9162938.
4)
Hattingen E, Pilatus U, Good C, Franz K, Lanfermann H, Zanella FE. An unusual intraventricular haemangiopericytoma: MRI and spectroscopy. Neuroradiology. 2003 Jun;45(6):386-9. Epub 2003 Apr 18. PubMed PMID: 12707742.