Case Report:

Intracranial Haemangiopericytoma: Case Report

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Abstract:
A rare case of an intracranial haemangiopericytoma with extra-cranial extension in a 60-year-old female patient’s is presented. Intracranial haemangiopericytomas are uncommon, dural-based tumors. It is classified as a tumor of uncertain origin in the WHO system. Haemangiopericytomas arise from Pericytes of Zimmerman that surround capillaries and not archetypical meningotheial arachnoid cap cells. Most frequently they arise in soft tissue sites, but rarely present in the CNS. Macroscopically many haemangiopericytomas resemble meningioma but are distinct histologically. These tumors show more aggressive natural history. Vascular nature suggests a different therapeutic approach thus fore warning a surgeon to a potentially more challenging resection.

Key words: Haemangiopericytoma, Meningioma, Magnetic resonance imaging (MRI)

Introduction:
Meningeal hemangiopericytoma is rare and its incidence is <1% of all central nervous system tumors and 2.4% of all meningiomas. Meningeal hemangiopericytomas have a high rate of local recurrence and distant metastasis in contradiction of usual benign meningiomas which rarely metastasize extracranially. The most commonly reported sites of metastasis are bone, liver, lung, central nervous system, and abdominal cavity. It is classified as a tumor of uncertain origin in the WHO system. Haemangiopericytomas arise from Pericytes of Zimmerman that surround capillaries and not archetypical meningotheial arachnoid cap cells. Most frequently they arise in soft tissue sites, but rarely present in the CNS. Angiographically, they can resemble supratentonal hemangioblastomas, glioblastoma multiforme or some metastatic tumors. Unlike glioblastomas and some metastatic lesions, they are insensitive to radiation. Therefore surgical extirpation is the definitive treatment, and long term follow up should be the first hand rule. However, there are difficulties in the histologic diagnosis of hemangiopericytoma because other soft tissue neoplasms may have areas of rich “hemangiopericytomalike” vascularity. These facts make obvious the importance of their recognition by both the radiologist and the pathologist. Their awareness of the entity will avoid unnecessary radiation therapy and guard against delay in recognizing recurrences.

Case report:
A 60 yr. old female patient, presented with a large swelling over the skull in the midline overlying the parietal bone on either side, of 2
years duration. It was small initially but grew rapidly in size in the last 2 months (Fig 1 A and B). On examination a large, firm globular mass with a broad base towards the skull in the midline was noted. Contrast enhanced MRI brain revealed a large 10.1 x 9.5 x 7.9 cms, extra-axial mass in the high parietal region, extending from the midline and overlying the skull vault on either side causing destruction of underlying bone and indenting the cerebral surface. It was predominantly hypointense with few areas of hyperintense signal on T1W and predominantly hyperintense on T2W and FLAIR images (Fig 2 A and B).

The lesion showed no blooming on GRE and was not suppressed on STIR images (Fig 3 A and B). Post contrast images show mild to moderate inhomogeneous enhancement with no early draining veins (Fig 4 A and B). Mild indentation of superior sagittal sinus was seen without any vascular encasement (Fig 5 A and B). Based on these findings the possibility of atypical meningioma / haemangiopericytoma was raised. Patient was operated and the tissue was sent for histo-pathological examination and was diagnosed as haemangiopericytoma.

Photomicrograph shows dilated "staghorn" blood capillaries (black arrow) lined by a single layer of endothelial cell (Figure 6 A). The capillaries are separated by closely packed cells (arising from pericytes) with ill-defined cytoplasm (pentagon). Some of the cells are showing plasmacytoid features and giant cells (black arrow) (Figure 6 B). A typical mytosis can be seen the characteristic appearance of a haemangiopericytoma.

**Discussion:**

Haemangiopericytomas are rare tumors, accounting for less than 1% of all primary CNS neoplasms. Their overall frequency is 2.4% of all meningiomas.\(^{[1,9]}\) The term "haemangiopericytoma" was first described by Schmidt in 1937 and named by Stout in 1942 and co-workers to describe a group of malignant neoplasms that originate from pericyte cells found around the reticular sheath of capillaries and post capillary venules.\(^{[1,10]}\) Angioblastic meningioma was the term used by Bailey et al in 1982 to describe a meningeal tumour observed in 3 cases.\(^{[11]}\) The current 1993 WHO classification has eliminated the term angioblastic meningioma in favor of haemangiopericytoma.

Haemangiopericytoma's are typically iso-intense to grey matter on T1WI and T2WI sequences and are often heterogeneous. Strong but inhomogeneous enhancement occurs following contrast administration. Prominent vascular channels are frequently identified. Main differential diagnosis for haemangiopericytoma's is meningioma. Other dural based masses include lymphoma, Pre-operative identification of this tumor is important because of their aggressive nature, high rate of local recurrence and propensity of late distant metastasis. Macroscopically haemangiopericytoma resemble those of meningiomas, three quarter of all cases are firm, well circumscribed, or encapsulated globular masses that can have either a narrow or broad based dural attachment.\(^{[3,12]}\) Haemangiopericytoma are richly vascularised tumors with numerous penetrating blood vessels. Microscopically these tumors are highly cellular, vascular and have dense pervasive reticulin network lobules of neoplastic cells surround so called stag horn vessels.\(^{[12]}\)
Average age of onset is 42 yrs and has slight male preponderances. Symptomatology is related to tumor location as with meningioma. Due to faster growth rate of these tumors there is shorter interval between symptom onset and diagnosis (as in our case interval is two months in the onset of symptom and diagnosis). Different imaging modalities for diagnosis of haemangiopericytoma are angiography, CT and MRI. Angiography reveals hypervascular lesions that typically have prolonged, dense but heterogeneous tumour stain. Certain angioarchitectural pattern has been shown to be common to haemangiopericytoma and may help to distinguish them from meningioma. The features of this pattern include: Dual supply from internal carotid or vertebral and external carotid arteries is seen with a dominant supply from external carotid, a myriad of cork screw vessels arising from a main feeder within the tumour, dense fluffy long lasting tumour stain rather than the sunburst pattern of meningioma and lack of early draining vein. On CT heterogeneous appearing lesions on both pre and post contrast studies. Low density cystic or necrotic areas are common and enhancement is typically strong but inhomogeneous. Unlike meningiomas there is an absence of tumoral calcification or adjacent hyperostosis. Instead bone erosion is often described. The findings of MRI are variable. Signs of extradural mass are often but not invariably present. Haemangiopericytoma generally have a broad based dural attachment and may have a dural tail. However a narrow dural base has been described in one-third of cases in one series. Metastasis, sarcoidosis, solitary fibrous tumour, gliosarcoma and neurofibroma.

Conclusion:
We have presented a rare case of intra-cranial haemangiopericytoma with extra-cranial extension which is dural based tumour. Its main differential was an atypical meningioma. This was successfully operated upon and histopathology proved it to be a hemangiopericytoma.

Fig 1 : Patient in frontal and lateral views showing a large, globular swelling over the vertex in the midline.
Fig. 2: (A) Plain T1W sagittal image shows large lesion which appears predominantly hypointense with few hyperintense areas in the centre with underlying bone erosion. (B) Plain T2W coronal image lesion appears predominantly hyperintense with few areas of hypointense signal.

Fig 3: (A) GRE image shows no blooming. (B) STIR image shows no fat suppression.

Fig 4: Plain T1W sagittal image T1W sagittal (A) and Post Gadolinium enhanced (B) images shows strong but inhomogenous enhancement with dural enhancement

Fig 5: MR Brain Venogram coronal (A) and sagittal (B) images show mild indentation of superior sagittal sinus however no encasement is seen.
Fig 6: (A) Photomicrograph shows dilated "staghorn" blood capillaries (black arrow) lined by a single layer of endothelial cell. The capillaries are separated by closely packed cells (arising from pericytes) with ill-defined cytoplasm (pentagon). (B) Some of the cells are showing plasmacytoid features and giant cells (black arrow). A typical mitosis can be seen the characteristic appearance of a haemangiopericytoma.

References: