

## Primary Hemangiopericytoma in Parietal Bone: Literature Review and Case Report

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### Keywords:

Hemangiopericytoma; Skull

### Abbreviations:

CT: computed tomography; EMA: epithelial membrane antigen; HPC: hemangiopericytoma; SMA: smooth muscle actin; STAT: signal transducers and activators of tranion; MRI: magnetic resonance imaging

## 1. Abstract

**Introductions:** Hemangiopericytoma (HPC) is a tumor from pericytes surrounding capillary walls. Most HPCs grow slowly, but others display aggressive growth. Treatment for HPC is total resection or resection plus adjuvant radiation.

**Patient concerns:** A 14-year-old girl presented with a tumor located on the left side of the parietal bone. The blood vessel-rich tumor with a smooth surface was attached to the inner surface of the skull. The dura was completely intact.

**Diagnosis:** Based on the histopathological and immunohistochemical findings, the final diagnosis was HPC.

**Interventions:** The patient was subjected to total resection of the tumor and cranioplasty. The dura was not opened because of lack of invasive growth in it.

**Outcomes:** Within the two-year follow-up, the patient showed excellent prognosis without any local recurrence or any positive radiological features.

**Conclusions:** The patient presented common intracranial hypertension symptoms. The blood vessel-rich tumor with a smooth surface was attached to the inner surface of the skull with the dura completely intact. Simple surgical resection without radiotherapy offered excellent prognosis during the two years of follow-up study.

## 2. Introduction

Hemangiopericytoma (HPC), a rare mesenchymal tumor originated from pericytes surrounding capillary walls, was delineated first by Stout and Murray in 1942 [1]. HPCs consist of a mass of fusiform and round tumor cells [1-4]. Intracranial HPCs are often benign and have slow growth, whereas less than 20% of HPCs display aggressive behavior, including borderline or frank malignancy. No standard treatment for intracranial HPC has been established in national or international guidelines. The commonly adopted treatment for HPC is surgery alone or surgical resection plus adjuvant radiation, which show promising prognosis. Factors associated with inferior overall survival (OS) include age, WHO grade, multifocal disease, disseminated disease, and chemotherapy [2].

## 3. Case Description

A 14-year-old girl presented with progressive headache and dizziness for 5 months. She developed nausea and vomiting in the following days and became frail with low spirits. She did not report a history of trauma or any systemic complaint and had no remarkable past medical history. Physical examinations revealed no sensory or motor abnormalities in her extremities.

Brain computed tomography (CT) revealed a roundish lesion with mixed density. The tumor was on the left side of the parietal bone, about 37 mm x 24 mm in size, protruding toward the cranial

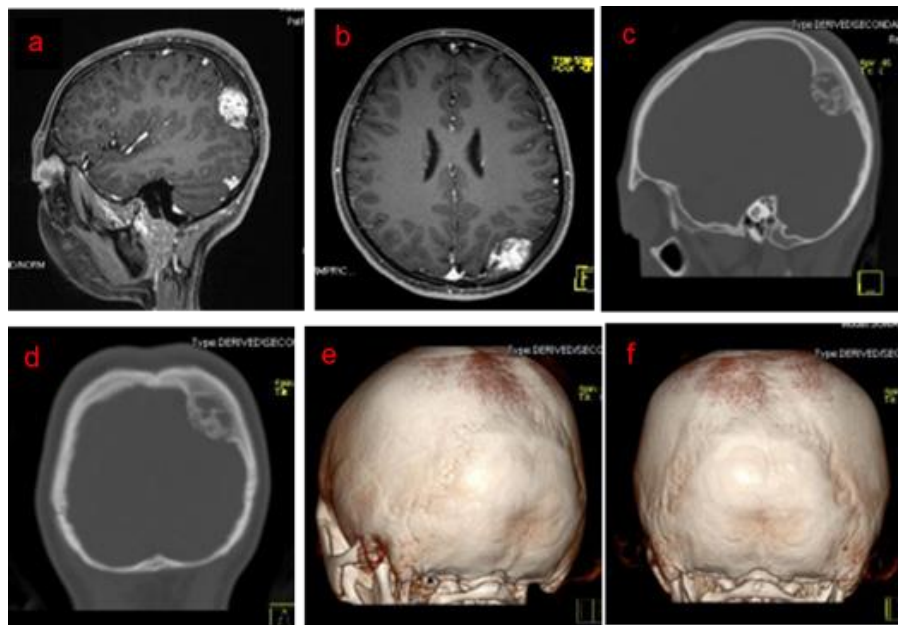
cavity. Moreover, the tumor eroded the inner table of the skull. The T1-weighted images of magnetic resonance imaging (MRI) demonstrated a mass with heterogeneous components and intact dura (Figure 1a & b).

During surgical resection, we observed normal appearance of the skull. We removed the left side of the parietal bone encompassing the entire tumor with an additional 5 mm margin. We found that the blood vessel-rich tumor with a smooth surface was attached to the inner surface of the skull. However, the dura was completely intact. Therefore, we did not make an incision on the dura but performed cranioplasty using a customized titanium mesh. Radiotherapy was not arranged after the surgery.

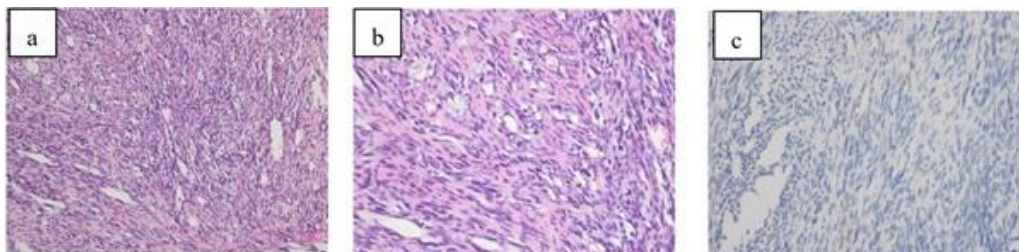
Optical microscopy examination of hematoxylin-eosin (HE)-stained tissue sections showed that the blood vessels were be-

sieged with a mass of proliferous spindle or round-shaped cells. Little mitotic figures were observed. Immunohistochemical study showed positive staining of SMA, STAT-6, Oligo-2, CD99, CyclinD1, and beta-catenins but negative reaction to CD31, CD34, desmin, EMA, Factor-VIII, and S-100. The positive rate of Ki-67(MIB1) was 5% (Figure 2).

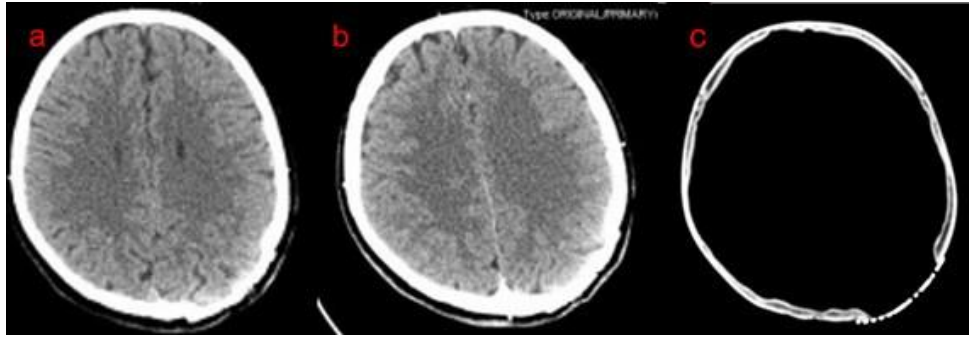
The surgical resection was performed 2 years ago. Approximately every 6 months, the patient underwent a brain CT scan for prognosis evaluation. The CT images taken at the 4th, 15th, and 24th months after the resection showed excellent prognosis without any local recurrence or any positive radiological features. Clinical follow-up examination also did not find any abnormal focal neurological symptoms or functions (Figure 3).



**Figure 1:** Preoperative radiological images of the patient head. a, b. Sagittal T1-weighted MRI view showing demarcated, white and large mass in the parietal Bone. c, d. CT scan showing circular hybrid dense shadow. e, f. Exterior line of skull showing the intact outline of the skull.



**Figure 2:** Representative microscopic images showing proliferative tumor cells. HE (a: 200x, b: 400x) and SMA (c: 400x) staining results.



**Figure 3:** Follow-up radiological survey identified lack of evidence of recurrence. a–c. The CT scans of the fourth month, fifteenth month, and second year after surgery, respectively.

#### 4. Discussion

HPCs consist of a mass of fusiform and round tumor cells, forming a “staghorn pattern” around the dilated vasculature. Enzinger and Smith [5] classified HPCs into adult- and infantile types according to onset time, disparate presentation, and pathological diagnosis. The more common form of HPC is the adult type, which occurs more frequently in deep soft tissues and rarely in lower extremities, pelvis, retroperitoneum, and intracranial cavities [6]. More than 75% of HPCs are benign and have slow growth, but the rest demonstrate aggressive behavior, such as local recurrence, extraneural metastases, and neural axis metastases. The infantile type often occurs before 1 year old of age. Before 1993, HPC was considered as meningioma due to similar patient symptoms and incomplete investigations. With the development of basic research and radiography, HPC has been reconsidered as one pathological entity within solitary fibrous tumors according to the 2013 WHO classification of soft tissue tumors [7] and categorized into WHO Grade III anaplastic hemangiopericytoma (AHPC) and WHO Grade II HPC [8].

In this report, the case of primary HPC in the parietal bone is particularly rare compared with previously reported cases. The present case differs from common brain HPC, which frequently involves eroded dura, and from skull tumors, which have broken inter table. The patient presented only with intracranial hypertension symptoms such as headache, dizziness, nausea, and vomiting, similar to patients with other intracranial tumors.

Since 1990, only seven skull HPC cases have been reported (Table 1). Among them, five cases were temporal HPCs, one case was from occipital bone, and one was parietal HPC. All the cases had similar common presentations such headache, dizziness, and vomiting. Temporal cases could have some aural or vestibular symptoms, including otalgia, deafness, and vertigo. Most patients underwent total resection without metastases in the follow-up years. Only one case, which was reported by Birzgalis, A. R. et al., was treated with radiotherapy and recurred after 4 years.

The radiological features of HPCs are not discriminative compared with those of meningioma and other intracranial tumors. In MRI images, HPCs are more likely to be in round or oval shapes and

have a mixed iso-low signal or uneven iso-signal in T1WI. Some aggressive HPCs are shown to be a mass with mixed iso-high signals in unregular or lobulated shapes in T1WI. Edema might surround the mass in T2WI. The final diagnosis is established based on the findings of pathological and immunohistochemical methods.

Histopathological findings are the most important for diagnosis of HPCs, especially when no particular differences exist between HPC and other intracranial tumors clinically or radiologically. Typical findings are that the tumor vessels are besieged with a mass of fusiform or round-shaped proliferous tumor cells under the microscope in the pathological examination. The positive immunohistochemical markers include STAT-6 and CD99, while the negative markers include desmin, CD34, EMA, and S-100.

Distinguishing HPCs from other CNS tumors is a challenge. Patients with HPC have a shorter course of the disease compared with those with meningioma. Moreover, MRI often shows a hybrid intense mass in HPC, whereas meningioma always features a homogenous enhanced mass. The most accurate way to confirm whether the tumor is HPC or meningioma is to use immunohistochemical method. The markers EMA and S-100 are negative for HPC but positive for meningioma. The patient in the present case resembled more with the diagnosis of benign tumors of the skull, such as skull base osteoma, skull base ossifying fibroma, and giant cell tumor of the skull. However, the erosion on the inner table is more distinctive for the primary HPC compared with that of benign skull tumors.

No standard treatment for intracranial HPC has been established. In some of the reported cases, presurgical biopsy was performed for diagnosis. However, profuse bleeding during the biopsy always occurred due to abundant feeding vessels involved, leading to hypovolemic shock especially for infantile patients. Therefore, embolization of the feeding vessels during the angiography before the surgery has been tested [13,14]. Additionally, adjuvant therapies, such as postoperative external beam radiotherapy, for primary intracranial hemangiopericytoma could reduce the risk of local recurrence and metastasis to central nervous system. No study has reported the relationship between the excision pattern (local or extended) and the outcome, but a negative surgical margin should

be achieved. All patients with positive surgical margins were dead within the mean survival of 54.6 months, which is lower than the 76.1 months of mean survival in patients with a negative surgical margin. Therefore, a clear surgical margin is important for HPC prognosis [9].

Whether radiotherapy could benefit the overall survival is still in dispute [2,4]. The average time for recurrence among irradiated patients was 10.3 years compared with 5.3 years in non-irradiated patients [9], particularly when the tumor is deep in the brain,

where it is difficult to obtain surgical access. In this case, the tumor was originated from and limited in the parietal skull, hence we did not arrange for the post-operative radiation therapy. For the first 5 years after initial diagnosis, higher risks of local recurrence are observed; therefore, brain CT scan every six months or yearly is especially important for follow up. In the follow-up period, patients may suffer from distant metastasis; hence, imaging areas in the follow-up examinations should be broadened.

**Table 1:** The review of hemangiopericytoma in the skull in the past few years.

Author	Gender & Age	Location	Size(cm)	Treatment	Follow-up
Sipal et al., 2009	M, 56	Parietal, R	3.4x3.7	Resection	9M, no metastasis
Cross & Mixon, 1996	M, 62	Temporal, L	1.0x1.0	Resection	2Y, no metastasis
Vilendecic, Grahovac, Lambasa, Jelec, & Topic, 2012	F, 47	Occipital, L and Neck	6.1x4.5x5.1	Resection, Radiotherapy	Not mentioned
Chin, Rabb, Hinton, & Apuzzo, 1993	F, 35	Temporal, R	Not mention	Embolization, Resection	9M, no metastasis
Birzgalis, Ramsden, Lye, & Richardson, 1990 (3 cases)	F, 50	Temporal, R	Not mention	External beam radiotherapy	Reccurred after 4 years
	F, 55	Temporal, L	Not mention	Embolization, Subtotal resection	18M, no metastasis
	M, 18	Temporal, L	Not mention	Embolization, almost totally resection, radiotherapy	Not mention
Current case	F, 14	Parietal, L	3.7x2.4	Resection	2Y, no metastasis

## 5. Conclusion

HPC is a rare mesenchymal tumor that originates from pericytes surrounding capillary walls. Intracranial HPC consists of less than 1% of central nervous system tumors. In this report, the case with primary hemangiopericytoma in the parietal skull has been rarely diagnosed. The patient presented with common intracranial hypertension symptoms. The blood vessel-rich tumor with a smooth surface was attached to the inner surface of the skull with completely intact dura. Simple surgical resection without radiotherapy offered excellent prognosis during the 2 years of follow-up. In future follow-up, we will continuously watch out the potential of local recurrence and distant metastases.

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