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Case Report

Primary hemangiopericytoma in parietal bone: literature review and case report

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Abstract: Background: Hemangiopericytoma (HPC) is a tumor originating from pericytes surrounding the capillary walls. Most HPCs grow slowly, but some display aggressive growth. Treatment for HPC is total resection or resection plus adjuvant radiation. Case presentation: A 14-year-old girl presented with a blood vessel-rich tumor with a smooth surface located on the left side of the parietal bone, attached to the inner surface of the skull. The dura was completely intact. The final diagnosis was HPC based on the histopathologic and immunohistochemical findings. The patient underwent total resection of the tumor and cranioplasty. The dura was not incised because of the lack of invasive growth. Within the 2-year follow-up, the patient showed an excellent prognosis without any local recurrence or positive radiologic features. Conclusion: The patient presented with common intracranial hypertension symptoms. A 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 an excellent prognosis during 2 years of follow-up.

Keywords: Hemangiopericytoma, skull

Introduction

Hemangiopericytoma (HPC), a rare mesenchymal tumor originating from pericytes surrounding the capillary walls, was first delineated 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 with slow growth, though fewer than approximately 20% of HPCs display aggressive behavior, including borderline or frank malignancy. No standard treatment for intracranial HPC has been established by international guidelines. The commonly adopted treatment for HPC is surgery alone or surgical resection plus adjuvant radiation, both of which show promising outcomes. Factors associated with inferior overall survival (OS) include age, World Health Organization (WHO) grade, multifocal disease, disseminated disease, and chemotherapy [2].

Case description

A 14-year-old girl presented with progressive headaches 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 complaints, and her past medical history was unremarkable. Physical examination revealed no sensory or motor abnormalities in her extremities.

Brain computed tomography (CT) revealed a roundish lesion with mixed density (**Figure 1C**, **1D**). The tumor was approximately 37×24 mm in size and located on the left side of the parietal bone protruding toward the cranial cavity (**Figure 1E**, **1F**). Moreover, the tumor had eroded the inner table of the skull. The T1-weighted images from magnetic resonance imaging (MRI) demonstrated a heterogeneous mass with intact dura (**Figure 1A**, **1B**).

During surgical resection, we observed a normal appearance of the skull. We removed the left side of the parietal bone encompassing the entire tumor with a 5-mm margin. We found that the blood vessel-rich tumor with a smooth surface was attached to the inner surface of

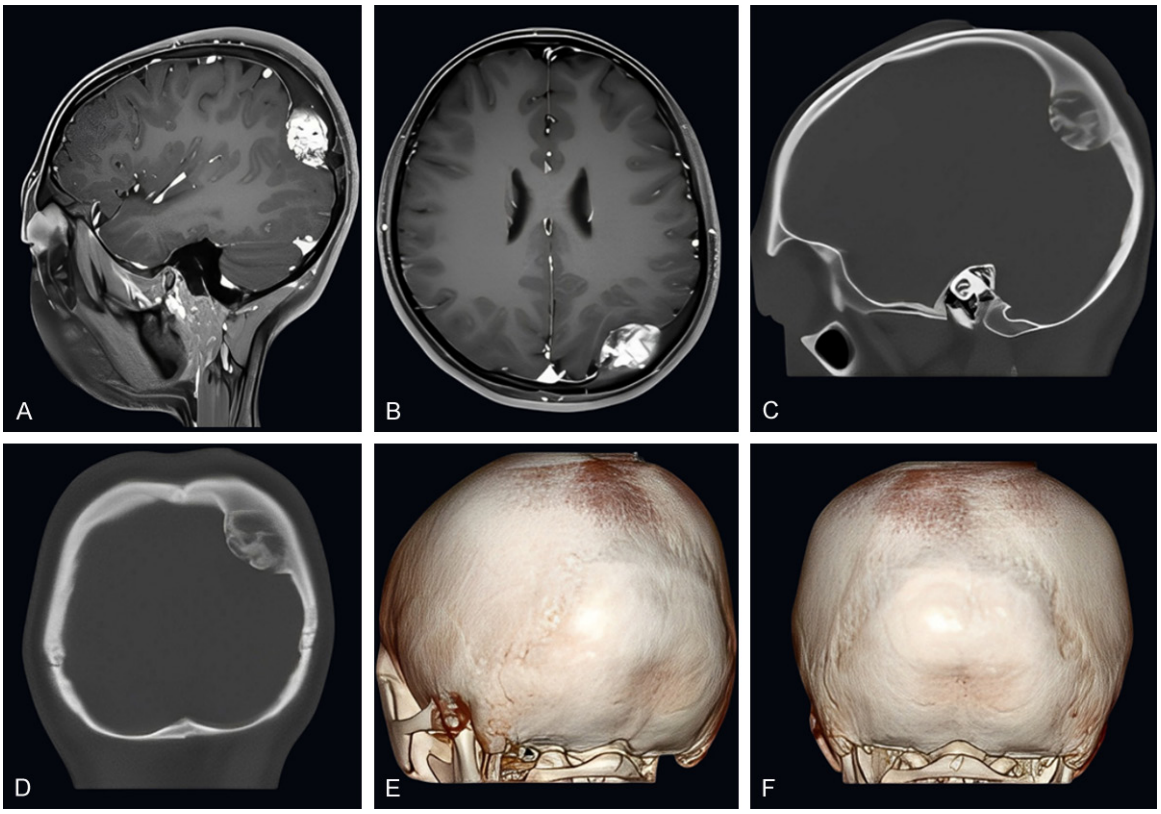

Figure 1. Preoperative radiologic images of the patient. A. Sagittal T1-weighted MRI view of head. B. Axial T1-weighted MRI view of head. C, D. CT scan showing the brain neoplasm. E, F. Exterior line of skull.

the skull; however, the dura was completely intact. Therefore, we did not incise the dura but performed cranioplasty using a customized titanium mesh. Radiotherapy was not arranged after the surgery.

Microscopic examination of hematoxylin-eosin-stained tissue sections showed that the blood vessels were surrounded by a mass of spindle or round-shaped cells. Small mitotic figures were also observed. Immunohistochemical study showed positive staining of smooth muscle actin, signal transducers and activators of transcription 6 (STAT-6), Oligo-2, CD99, cyclin D1, and beta-catenin and negative reactions to CD31, CD34, desmin, epithelial membrane antigen (EMA), factor VIII, and S-100 protein. The positive rate of Ki-67/MIB-1 was 5% (**Figure 2**).

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

Discussion

Hemangiopericytomas (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 pathologic diagnosis. The adult type is more common than the infantile type; it occurs most frequently in deep soft tissues and rarely in the lower extremities, pelvis, retroperitoneum, and intracranial cavities [6]. More than 75% of HPCs are benign and have slow growth, but the remainder demonstrate aggressive behavior, such as local recurrence, extraneural metastases, and neural axis metastases. The infantile type of HPC often occurs before 1 year of age. Before 1993, HPC was considered to be a meningioma due to similar patient symptoms and incomplete investigations. With the development of

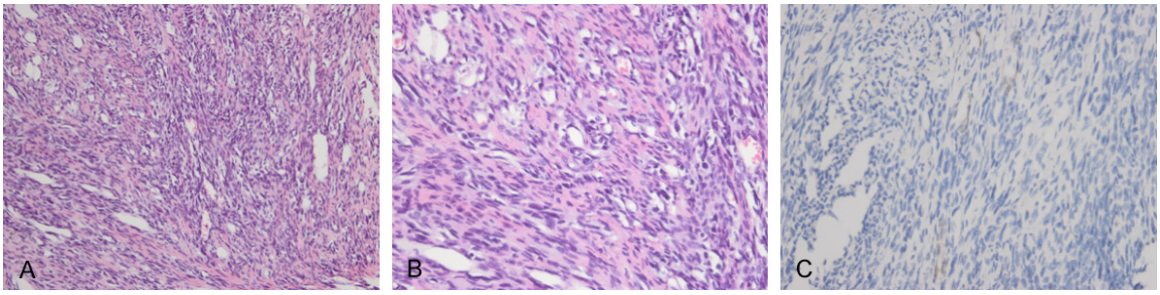

Figure 2. Immunohistochemical study showing H&E (A: 200×, B: 400×) and SMA (C: 400×) staining results.

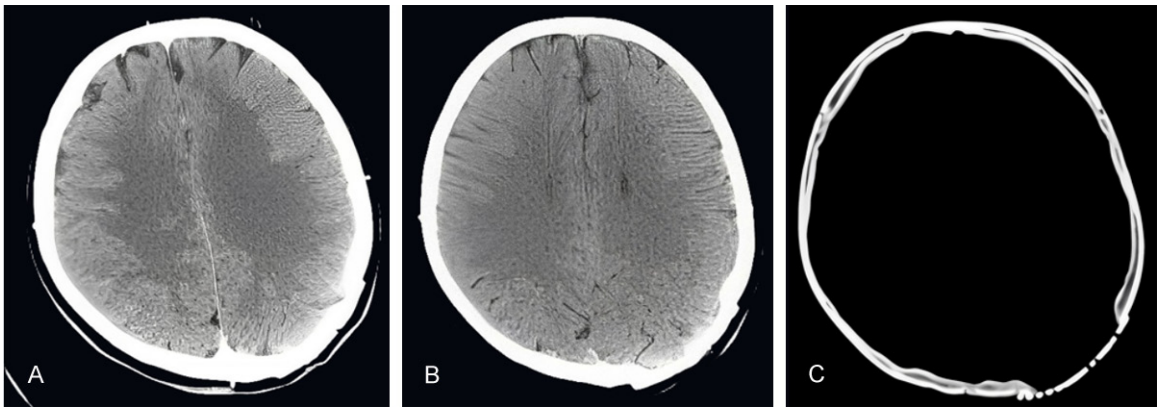

Figure 3. Follow-up radiologic survey. A-C. CT scans of the fourth month, fifteenth month, and 2nd year after surgery, respectively.

Table 1. Review of hemangiopericytoma of the skull in the past few years

Author	*Gender & Age*	*Location*	*Size (cm)*	*Treatment*	*Follow-up*
Sipal et al., 2009	M, 56	Parietal, R	3.4×3.7	Resection	9 M, no metastasis
Cross & Mixon, 1996	M, 62	Temporal, L	1.0×1.0	Resection	2 Y, no metastasis
Vilendecic, Grahovac, Lambasa, Jelec, & Topic, 2012	F, 47	Occipital, L and Neck	6.1×4.5×5.1	Resection, Radiotherapy	Not mentioned
Chin, Rabb, Hinton, & Apuzzo, 1993	F, 35	Temporal, R	Not mentioned	Embolization, Resection	9 M, no metastasis
Birzgalis, Ramsden, Lye, & Richardson, 1990 (3 cases)	F, 50	Temporal, R	Not mentioned	External beam radiotherapy	Reccurrence after 4 years
	F, 55	Temporal, L	Not mentioned	Embolization, Subtotal resection	18 M, no metastasis
	M, 18	Temporal, L	Not mentioned	Embolization, almost totally resection, radiotherapy	Not mention
Current case	F, 14	Parietal, L	3.7×2.4	Resection	2 Y, no metastasis

basic research and radiography, HPC has been reclassified as a distinct pathologic entity of solitary fibrous tumor according to the 2013 WHO classification of soft tissue tumors [7] and further categorized into WHO Grade III anaplastic HPC and WHO Grade II HPC [8].

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

Since 1990, only seven skull HPC cases have been reported (**Table 1**); of these cases, five were temporal HPCs, one was from the occipital bone, and one was parietal HPC. All seven

cases had similar common presentations, including headache, dizziness, and vomiting. Temporal cases may present with 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, reported by Birzgalis et al., was treated with radiotherapy and recurred after 4 years.

The radiologic features of HPCs are not discriminatory compared to those of meningioma and other intracranial tumors. Using MRI images, HPCs are more likely to be round or oval in shape and have a mixed iso-low signal or uneven iso signal in T1WI. Some aggressive HPCs may appear as a mass with mixed iso-high signals in irregular or lobulated shapes in T1WI. Edema may surround the mass in T2WI. A final diagnosis is established based on the pathologic and immunohistochemical findings.

Histopathological findings provide the most important evidence for diagnosis of HPCs, especially when no particular clinical or radiologic differences exist between HPC and other intracranial tumors. Typical pathologic findings from microscopic examination show the tumor vessels surrounded by a proliferation of fusiform or round tumor cells. Positive immunohistochemical markers include STAT-6 and CD99, and negative markers include desmin, CD34, EMA, and S-100.

Distinguishing HPCs from other central nervous system (CNS) tumors is challenging. Patients with HPC have a shorter disease course compared with those with meningioma. Moreover, MRI often shows a hybrid intense mass in HPC, whereas meningioma always features a homogeneous enhanced mass. The immunohistochemical method can most accurately confirm whether the tumor is HPC or meningioma as the EMA and S-100 markers are negative for HPC but positive for meningioma. Our patient's presentation resembled that of benign tumors of the skull, such as skull base osteoma, skull base ossifying fibroma, and giant cell tumor of the skull. However, erosion of the inner table is more distinctive for primary HPC than for benign skull tumors.

No standard treatment is established for intracranial HPC. In some previously reported cases, a presurgical biopsy was performed for diagnosis. However, profuse bleeding always occurred during the biopsy due to abundant feeding vessels involved, leading to hypovolemic shock, especially for infantile patients. To address this outcome, embolization of the feeding vessels during angiography before surgery has been tested [13, 14]. Additionally, adjuvant therapies for primary intracranial HPC, such as postoperative external beam radiotherapy, may reduce the risk of local recurrence and metastasis to the CNS. 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 period of 54.6 months, which is lower than the 76.1-month mean survival for patients with a negative surgical margin. Therefore, a clear surgical margin is important for HPC prognosis [9].

Whether radiotherapy could improve OS (Overall Survival) for HPC patients is still in dispute [2, 4]. The average time for recurrence was 10.3 years among irradiated patients compared to 5.3 years for 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 originated from and was limited to the parietal skull, so we did not arrange for postoperative radiation therapy. Higher risks of local recurrence are observed in the first 5 years after initial diagnosis; therefore, brain CT scans every 6 months to 1 year are especially important for follow-up. Given that patients may suffer from distant metastasis in the follow-up period, imaging areas in follow-up examinations should be broadened.

Conclusion

HPC is a rare mesenchymal tumor that originates from pericytes surrounding the capillary walls. Intracranial HPCs make up fewer than 1% of CNS tumors. Primary HPC in the parietal skull, as reported in this case, has rarely been diagnosed. Our patient presented with common intracranial hypertension symptoms. A 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 an excellent prognosis during the 2 years of follow-up. In future follow-ups, we will remain alert to the potential of local recurrence and distant metastases.

Disclosure of conflict of interest

None.

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.

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