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An uncommon intracranial malign tumour which was misdiagnosed as Glioblastoma multiforme: Hemangiopericytoma

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ABSTRACT

Background. Hemangiopericytoma (HPC) which is mostly located at the lower extremity and visceral organs was found extremely rare in the central nervous system. Radiological images are not enough to differentiate HPC from other CNS tumours. The case was analyzed to determine presurgical features for diagnosis and the challenges during surgery.

Case. A 65-year-old male patient with headache was diagnosed as Glioblastoma Multiforme (GBM) considering the image findings. However, the intraoperative macroscopic shape and tendency to bleeding were not relevant to the GBM. The mass was reported as Hemangiopericytoma which is a malign tumour, originates from pericapillary bodies of veins, and commonly locates out of the CNS.

Conclusion. Even in advanced age and radiologically considered high-grade glial tumours, HPC should be considered in the differential diagnosis for preoperative preparation.

INTRODUCTION

Hemangiopericytoma (HPC), which is mostly located in the musculoskeletal system and the skin, is rarely seen in the central nervous system. It mostly locates in the lower extremities, retroperitoneum and pelvis 1,2. It accounts for 2% of meningeal tumors, less than 1% of all central nervous system (CNS) tumors 1,3. HPC, first reported by Begg 4, originates from the Zimmerman pericytes around the capillary and postcapillary veins 1. It is a highly malignant tumor with a tendency to local and distant metastases. It is very difficult to distinguish radiologically from the glial and meningeal tumors observed in the central nervous system 5.

Our aim is to present the difficulties encountered during intraoperative treatment of HPC, which is rarely located in the CNS and radiologically mixed with high grade glial tumor.

Keywords
hemangiopericytoma,
diagnosis,
central nervous system,
surgery



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CASE REPORT

A 65-year-old male patient had a headache and dizziness for about 1 month. He was diagnosed with benign positional vertigo and received medical treatment, but did not recover. The patient, who started complaining of weakness and nausea 3 days ago, worsened gradually and started to vomit and prone to sleep for 1 day. He was admitted to the emergency room with unconsciousness and the occipital mass was detected in the CT scan (Figure 1). The patient underwent contrast MR imaging in intensive care unit and a preliminary diagnosis of Glioblastoma Multiforme (GBM) was made radiologically, which was presented with edema, peripheral contrast, necrotic areas in the center (Figure 2). When the mass of the patient was reached during the operation performed under general anesthesia, it was observed that the mass was very smooth and shiny surface, rather fragile, bleeding tendency, but easily dissected in the surrounding tissues. When frozen sample was sent to pathology, it was reported that the mass was extremely malignant, but the diagnosis could not be made before immunohistochemical staining. Mass excision was performed by performing difficult bleeding control. It was observed that the mass was removed close to total by postoperative imaging. HPC was diagnosed after further pathological examination of the removed mass. After histopathological diagnosis, radiotherapy was planned and the patient was discharged in good condition. The patient who presented with the same complaints after 3 months showed that the mass recurred as large as the preoperative volume. The patient was missing while preparing for reoperation.

DISCUSSION

Intracranial HPC is a malignant tumor that makes up less than 1% of all CNS tumors. The location in CNS is extremely rare. It tends to locate in the lower limbs and intra-abdominal visceral organs 3. Unlike other meningiomas, it is more common in young men (38-44 years) and tends to be supratentorial 6.

It is very difficult to differentiate HPC with benign meningioma before surgery 5. Differential diagnosis can be made by observing multilobulate mass and dural connection in CT and MRI imaging. In addition, osteolytic areas can be observed in x-ray and CT imaging. However, in our patient, there was no multilobulate mass and osteolytic area images in

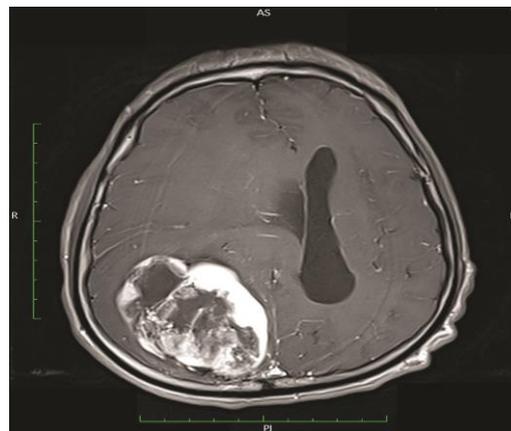
preoperative imaging (Figure 1 and 2). There was an edematous mass image with peripheral contrast, necrotic areas in the center. Considering the current imaging findings and the patient's age, primarily GBM was considered and surgical planning was done accordingly. However, it was understood that the character of the tumor was different due to its intraoperative macroscopic shape and the mass tending to bleed. Since meningeal artery bleeding is common during HPC surgery, preoperative arterial embolization is recommended to prevent bleeding 7. Tumor embolization also reduces the operation-related mortality and morbidity rate 3.

HPC tends to recur, especially in primer surgical localization. Distant organ metastases are frequently observed 8. 5-year life after postoperative adjuvant radiotherapy has been reported as 57% 9. However, despite our patient's radiotherapy, relapse was observed after 3 months and was mortal.



Figure 1. There was no osteolytic bone lesion in CT images.

Figure 2. Due to the peripheral contrast enhancing and the central necrosis, the preliminary diagnosis was Glioblastoma multiforme.



CONCLUSION

HPC is a highly malignant tumor, which is more common in young men and rarely seen in CNS. It is quite difficult to make a preoperative diagnosis. Multilobulence in CT and MRI imaging and osteolytic bone lesions are valuable for preliminary diagnosis. Even in advanced age and radiologically considered high grade glial tumors, HPC should be considered in differential diagnosis for preoperative preparation. Unlike other glial tumors, preoperative tumor embolization should be planned since HPC tends to bleed more. In addition, since the distant metastasis of HPC is common, the whole-body scan must be performed.

DECLARATION OF INTEREST: None

SUBMISSION DECLARATION: We confirm that this study is an original contribution, has not been published before and is not currently being considered for publication elsewhere.

CONSENT FOR PUBLICATION: Since the patient died, the family were given detailed information of the procedure and informed written consent was obtained from them.

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