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Case Report

Hemangiopericytoma of the pineal region: a rare case report and literature review

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Abstract: Hemangiopericytomas (HPC) are rare, aggressive vascular tumors arising from mesenchymal cells with pericytic differentiation. They account for less than 1% of intracranial tumors, and those occurring in the pineal region is extremely rare, we report such a rare of hemangiopericytoma of the pineal region. A 38-year-old female presented with headache of two-month duration. The radiological features and histological findings in this case are discussed in the study.

Keywords: Pineal region, hemangiopericytoma, meningioma, magnetic resonance imaging, pathology

Introduction

Intracranial hemangiopericytoma (HPC) is a rare, highly vascularized mesenchymal tumor that develops from malignant transformation of pericytes, which are contractile spindle cells surrounding the walls of capillariesand post-capillaryvenules [1]. HPC has been estimated to represent 0.4% of all primary central nervous system tumors [2], and it is considered a World Health Organization (WHO) grade II neoplasm, with anaplastic variants classified as WHO grade III [3]. Primary intracranial HPC in the pineal region has occurred rarely and reports are also seldom encountered. In this study, we report a patient with HPC of the pineal region, and to the best of our knowledge, only seven cases of pineal region HPCs have been reported so far in the English-language literature [4-10].

Case report

A 38-year-old female presented with a two-month history of headaches. Her neurological examination indicated no focal deficits. Magnetic resonance imaging (MRI) brain showed a well-defined lession in pineal region, isointense on axial T1-weighted image (**Figure 1A**) and iso- to hypointense on axial T2-weighted image (**Figure 1B**). On axial, coronal and sagittal postcontrast T1-weighted MR images, contrast enhancement of the mass is avid (**Figure 1C-E**), causing obstructive hydrocephalus. The mass was found to be attached to the tentorium and was highly vascular (**Figure 1F**, **1G**). A provisional diagnosis of pineal meningioma was considered.

Histopathology revealed that the tumour cells were round to spindle-shaped, and branching vessels inside the tumour tissue showed the dilated 'staghorn' appearance (**Figure 2A**). Tumour cells exhibited strong immunoreactivity for Vimentin and S-100 protein, positive for bcl-2 (**Figure 1B-D**), while being negative for CD34, epithelial membrane antigen (EMA) and glial fibrillary protein (GFAP). Based on the aforementioned features, a diagnosis of HPC was made.

Literature search

We performed a PubMed search for all cases of HPC of the pineal region up to May 2016. Cases were analyzed for basic demographic

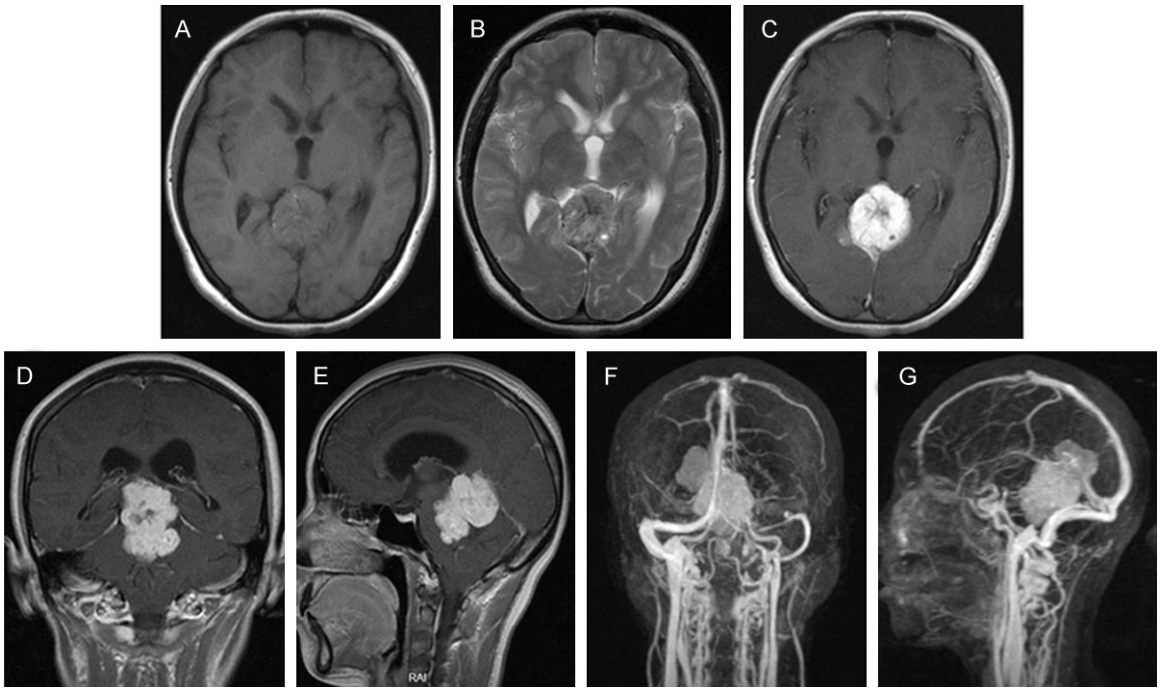

Figure 1. Magnetic resonance (MR) images. MRI brain show a well-defined lession in the pineal region, isointense on axial T1-weighted image (A) and iso- to hypointense on axial T2-weighted image (B). On axial, coronal and sagittal postcontrast T1-weighted MR images, homogeneous contrast enhancement of the mass is noted (C-E), and the mass was highly vascular (F, G).

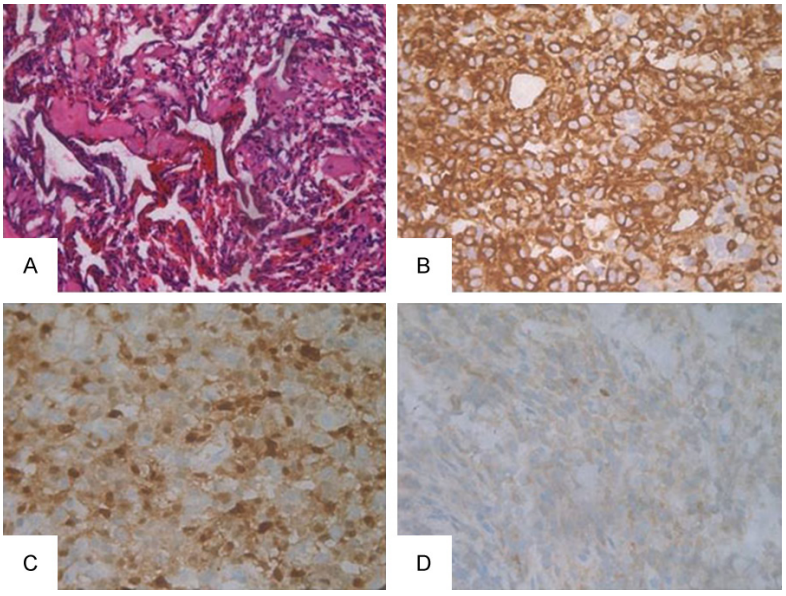

Figure 2. Histological features of tumour samples. Histopathology shows cellular tumor with characteristic staghorn vessels intersecting the tumor (A) (hematoxylin and eosin, original magnification, ×400), and tumor cells strongly expressing Vimentin (B), and S-100 protein (C), positive for bcl-2 (D) (immunohistochemistry, original magnification, ×400).

features including age, sex, clinical manifestation, treatment, and clinical outcome (**Table 1**).

Discussion

HPCs are rare vascular tumors originating from the pericytes of the capillary wall, and they may occur any part of the body, most commonly seen in the lower extremities, pelvis, and the head and neck areas [11]. HPCs represent less than 1% of all primary central nervous system tumors, pineal region location is rare with very few reported cases in English-language literature. HPC was initially considered to be one of the variants of meningioma and was interchangeably called as angioblastic meningioma (hemangiopericytic type). Due to its different histomorphology, biological behavior and immunophenotype, WHO in the current 2007 classification laid down HPC as a subtype of "mesenchymal, non-meningo-

Table 1. Summary of previously reported cases of Hemangiopericytoma of the pineal region

Study	Age/gender	Chief complaint	Duration of symptoms (mo)	Treatment	Clinical outcome
Olson JR et al, [4] 1969	28/Male	Severe frontal headaches	1	Surgery	Dead at 12 hours
Stone JL et al, [5] 1983	27/Female	Headaches and papilledema	3	Surgery + postop radiation	Clinically stable at 12 months
Lesoin F et al, [6] 1984	33/Male	Headache and vomiting	15 day	Surgery	Recurrence and dead at 8 months
Sell JJ et al, [7] 1996	31/Female	Headaches	Several	Surgery	Not reported
Jian BJ et al, [8] 2010	56/Female	Headaches and neurocognitive deterioration	Not reported	Surgery + postop radiation	Disease free at 48 months
Hasturk AE et al, [9] 2011	37/Female	Headache, memory loss and vomiting	2 days	Surgery + postop radiation	Not reported
Maiti TK et al, [10] 2014	24/Male	Headache and vomiting	1	Surgery + postop radiation	Disease free at 12 months
Current case	38/Female	Headaches	2	Surgery + postop radiation	Recurrence at 56 months

thelial tumors" [3]. Because HPCs are remarkably similar to meningioma in clinical and radiographic presentation, histological confirmation is the only definitive means of distinguishing the diagnosis. To distinguish the two tumors, a number of immunohistochemical has been developed. The HPCs generally dose not produce Epithelial Membrane Antigen (EMA), so EMA is weak in HPCs, but strong and diffuse in meningioma. HPCs should be kept in mind in the differential diagnosis of pineal region tumors.

In conclusion, HPCs are rare intracranial tumours arising from pericytes. We have reported a rare case of HPC that arose in the pineal region. Correct diagnosis of HPCs can only be made through immunohistochemical analysis because of the clinical and radiological similarity between the HPCs and the more frequent meningiomas. In particular, the unusual location of HPC often makes it difficult to diagnose via radiological study alone, due to the radiological features are not pathognomic. Biologically, HPCs are more invasive than meningiomas, and for this reason, it is very important to make a preoperative correct diagnosis.

Disclosure of conflict of interest

None.

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