CASE REPORT PEER REVIEWED | OPEN ACCESS

Recurrent hemangiopericytoma of fourth ventricle in a 40-year-old women

Yusra Shafique, Ruqaiya Shahid, Shaheen Sharafat

ABSTRACT

Introduction: We report a recurrent case of hemangiopericytoma/solitary fibrous tumor of fourth ventricle that was misdiagnosed as atypical meningioma. Case Report: We report a case of hemangiopericytoma in a 40-year-old female. Hemangiopericytoma can occur anywhere where capillaries present, it is a rare tumor with uncommon location in the Central nervous system. It has only recently been included in 'mesenchymal, non-meningiothelial tumors' in revised WHO classification of 2016. It should be differentiated from meningioma as the hemangiopericytoma is more aggressive and metastasizes and this patient was misdiagnosed as the case of atypical meningioma on magnetic resonance imaging scan. Conclusion: Intracranial hemangiopericytoma/ solitary fibrous tumor should be distinguished from meningioma as the former is locally aggressive and can metastasize.

Keywords: Fourth ventricle, Hemangiopericytoma, Meningioma, Solitary fibrous tumor, WHO classification of CNS tumors

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INTRODUCTION

Hemangiopericytoma, an uncommon, vascular tumor that was first described by Schmidt in 1937 and named by Stout in 1942, accounts for only 2–4% of meningeal tumors and for less than 1% of all intracranial tumors. Hemangiopericytoma (HPC) arises from pericapillary cells or pericytes of Zimmerman and can occur anywhere where capillaries are present. It most commonly occurs in musculoskeletal system (lower extremities), skin, retroperitoneum and pelvis while intracranial localization along with larynx, bone, spleen and thorax is rare [1]. The previous World Health Organization (WHO) grading system of soft tissue (1993) distinguished HPC as separate entity and classified it into group of 'mesenchymal, non-meningiothelial tumors'. Recent WHO classification (2016) has placed hemangiopericytoma within the spectrum of solitary fibrous tumors. As both of these tumors have STAT 6 nuclear expression on immunohistochemistry so their features are regarded as overlapping if not identical. Hemangiopericytoma is considered in WHO as grade II neoplasm, with anaplastic variant regarded as grade III [2, 3].

Hemangiopericytoma cells have prominent intracytoplasmic intermediate filaments similar to smooth muscle cells and fibroblast cells that makeup vascular and perivascular tissues so they stain positive for desmin, actin,

alpha-actinin and vimentin on immunohistochemistry [3, 4].

The biological behavior of hemangiopericytoma is sometimes malignant, with wide range of variations. Local tumor recurrence and widespread extracranial metastasis can occur.

CASE REPORT

A 40-year-old female was presented with a history of headache since three months. Brain magnetic resonance imaging (MRI) scan showed a large lobulated, mushroom shaped, extra- axial, highly vascular mass in para-sagittal region of about 10×6×5 cm.Tumor was crossing the midline and showed a central necrotic component. The mass demonstrated a heterogeneous low signal on T2-weighted images and an iso-low signal on T1-weighted images with signal voids suspected to be a vascular structure. The mass was observed to be compressing the 4th ventricle and became strongly contrast enhanced on T1-weighted images (Figure 1). The case was radiologically reported as atypical meningioma.

After surgical removal of mass the specimen was sent to our histopathology laboratory for evaluation.

Morphology

Histological examination revealed multiple fragments of lobulated, moderately cellular neoplasm, comprising sheets and lobules of spindle cells. Tumor cells were homogenous with moderate amount of eosinophilic cytoplasm and moderately pleomorphic rounded to oval nuclei. Mitotic count of 10–14/HPF was present. Intra-tumoral staghorn vessels with focal sinusoid like vessels were present in tumor. Tumor necrosis was not present. A focus of vascular invasion at the periphery of the tumor was identified. Special stain Reticulin highlighted rich, nested pattern of tumor cell arrangement (Figure 2). Immunohistochemical stains were performed with the Dako kits (Dako North America, Inc.,CA, USA).

Tumor cells revealed positivity with CD 34, vimentin, Bcl2, CD99 and negative results for epithelial membrane antigen (EMA) and alkaline phosphatase (Figure 3). The findings and immunohistochemistry (IHC) confirmed the diagnosis of hemangiopericytoma/solitary fibrous tumor, WHO grade II.

DISCUSSION

Hemangiopericytoma is a rare neoplasm and constitutes about 0.4% of central nervous system tumors [5, 6]. Hemangiopericytoma arises from Zimmermans pericytes which are contractile spindle cells surrounding the capillaries and post capillary venules [7].

According to literature, the mean age at the time of diagnosis of HPC ranges from 38–50 years with male preponderance [8]. Headache is the most common symptom, other symptoms include intracranial hypertension, seizures and motor and sensory deficit.

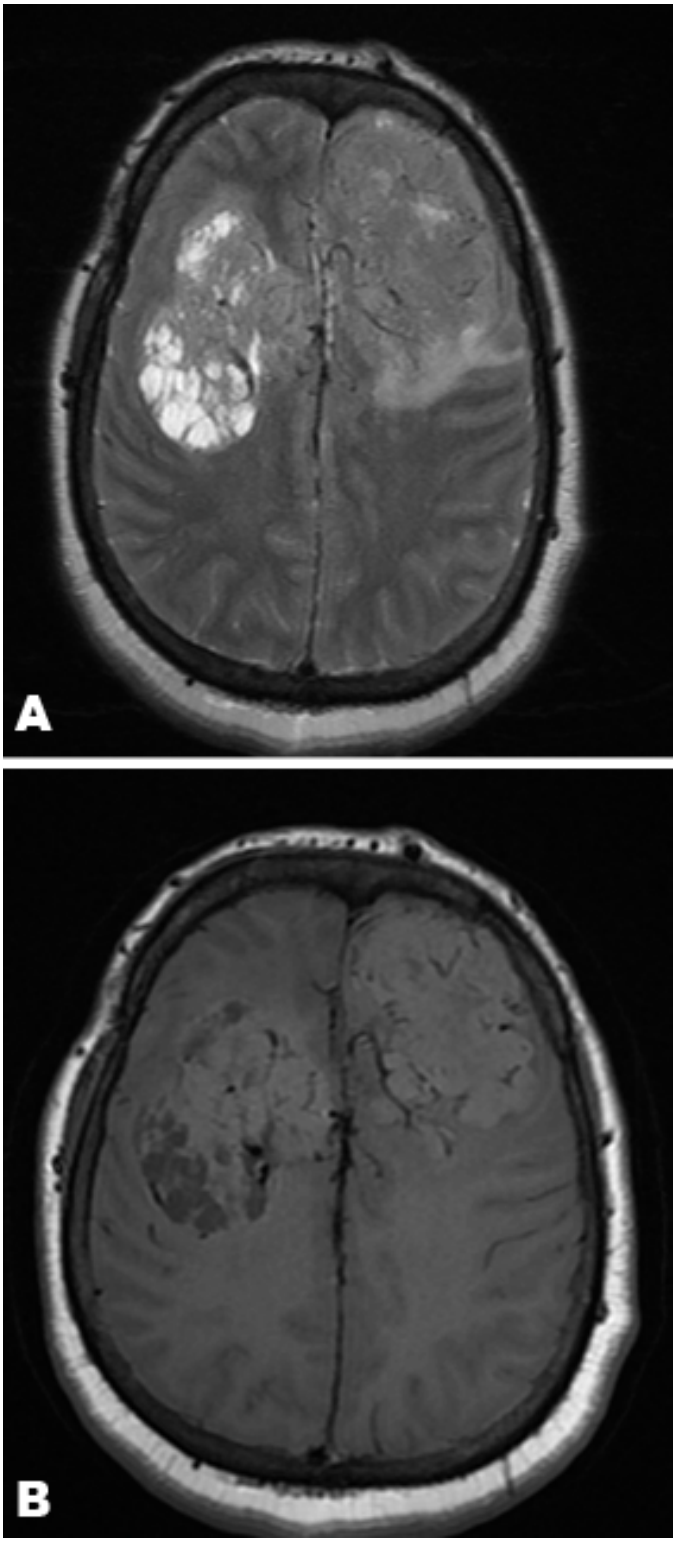

Figure 1: (A) Axial T1-weighted and (B) Axial T2-weighted magnetic resonance imaging scan revealing a lobulated mass with a central necrotic component showing heterogeneous low signal on T2-weighted images and an iso-low signal on T1-weighted images with signal voids suspected to be a vascular structure.

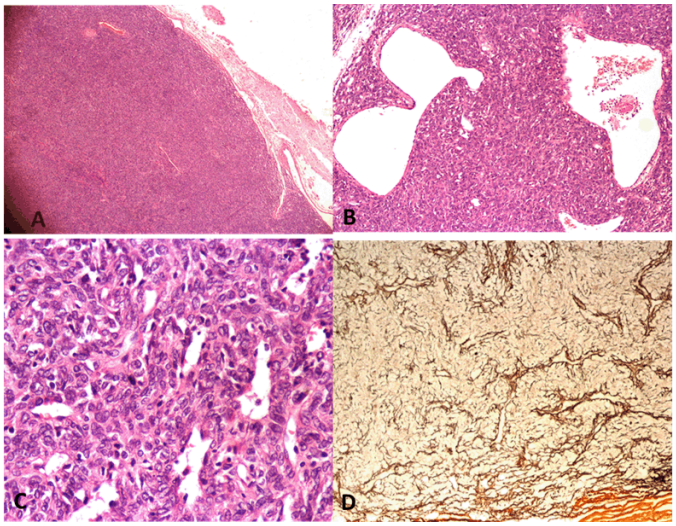

Figure 2: Photomicrographs of H&E sections of brain at (A) x100 (B) x200 (C) x400x magnification showing sheets and lobules of spindle cells with moderate eosinophilic cytoplasm and rounded to oval nucleoli and intratumoral staghorn vessels. (D) Special stain Reticulin highlighting rich nested pattern.

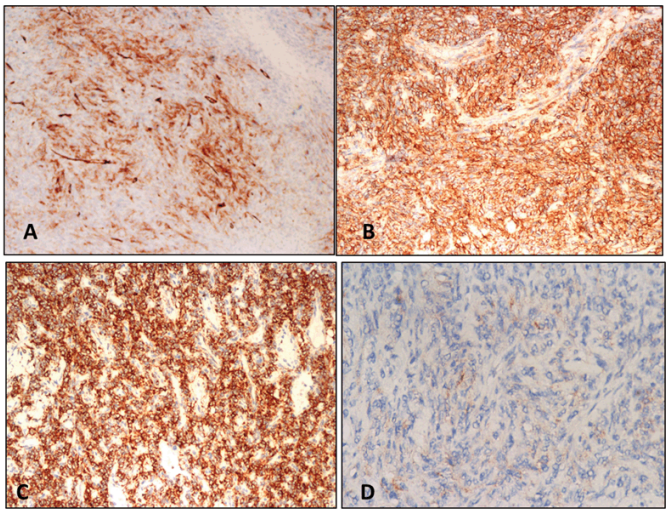

Figure 3: Photomicrograph of immunohistochemical stains at 400x magnification strong positivity against (A) CD34, (B) Bcl2 (C) Vimentin and (D) Negativity against EMA.

On imaging, most intracranial HPCs are supratentorial in distribution; the most common location is the parasagittal area with dural attachment. Other sites are sellar, suprasellar, pineal gland and third ventricle [9]. On imaging HPC have poly lobulated or irregular borders, show parenchymal invasion (mushrooming), bone erosion and contrast enhancement. Hemangiopericytomas do not show dystrophic calcifications [10, 11]. The main differential diagnosis of HPC includes angiomatous/ anaplastic meningioma. Distinguishing HPCs from benign meningioma before surgery can be difficult, but is very important because of the aggressiveness of HPCs, their high rates of local recurrence as high as 91% [12] and distant metastasis [13]. Microscopically, HPC shows high vascularity, a staghorn vascular pattern of spindle cells around a Reticulin network [5, 7]. At times, the histopathologic features of a HPC and meningioma can overlap. Immunohistochemistry can be helpful in these cases. Immunohistochemistry staining for HPC shows an intense reactivity to vimentin but not to EMA, unlike meningioma that is positive for vimentin and EMA. CD34 appear focal or weakly staining in the HPC. Bcl-2 seems to help differentiate these two entities. CD99 seems to be a good marker for HPC with about 84% specificity. Hemangiopericytoma also shows a negative reaction to S-100 protein, factor VIII, CD31 as well as progesterone receptor (PR) [6, 7].

The treatment of choice in hemangiopericytoma is microneurosurgery and adjuvant radiotherapy. Complete resection of tumor is mandatory. Post-surgical radiotherapy is treatment of choice even after complete excision of tumor [3].

CONCLUSION

To conclude intracranial hemangiopericytoma exhibits particular characteristics of WHO grade II or III tumors, which are similar to those of meningioma. However, certain features may aid in differentiating intracranial HPC from meningioma like the dural attachment, growth pattern and immunohistochemical features. Imaging along with pathological examination is required for proper diagnosis.

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Author Contributions

Yusra Shafique – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ruqaiya Shahid – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Shaheen Sharafat – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

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None

Conflict of Interest

Authors declare no conflict of interest.