Case Report

Anaplastic cerebral hemangiopericytoma: Rare variant of a rare disease

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ABSTRACT

Intracranial hemangiopericytoma (HPC) is a rare tumor of central nervous system, anaplastic type (grade 3) being the rarest. HPC closely mimics meningiomas in clinical and radiological features, thus, its diagnosis and treatment is a challenge. We report a rare case of histopathologically diagnosed anaplastic HPC of frontal lobe of the brain. A lady in her 60's presented with neurological signs of impaired memory, headache, decreased vision and slurring of speech that gradually progressed to aphasia over a period of 2 months. A space occupying lesion was identified on magnetic resonance imaging in the left frontal region. Left frontal craniotomy and Simpson grade 1 excision of the tumor was done. Postoperative radiotherapy was administered in view of positive margins on histopathological specimen. We describe the clinical, radiological, and histological features of this tumor, its outcome on completion of treatment and on subsequent follow-up along with a review of the literature.

Key words: Anaplastic hemangiopericytoma, aphasia, follow-up, radiotherapy

INTRODUCTION

Hemangiopericytoma (HPC) is an uncommon neoplasm arising from pericytes around capillaries. Meningeal HPC's constitute 2.4% of all meningeal and 1% of central nervous system neoplasms.[1,2] They occur most often in the deep soft tissues. HPC mimics meningioma in clinical and radiological features. A detailed histopathological review is the only means of its accurate diagnosis. The prognosis and appropriate treatment of these uncommon tumors are not well understood. In literature, very few cases of meningeal HPCs have been reported.[2] No definite treatment guidelines have thus been possible. Anaplastic variety is the rarest of all HPC and reports of this variant are still rarer. Only few papers have reported results of postoperative radiotherapy for unresectable or incompletely excised anaplastic meningeal HPC. We present a case of anaplastic (grade 3) HPC of brain, treated with surgery and postoperative radiotherapy.

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CASE REPORT

A 63-year-old lady presented to us with complaints of slurring of speech for 2 months which gradually progressed to aphasia by the time she presented to the hospital, impaired memory for 1½ months, decreased vision, headache and vomiting for 15 days. On clinical examination, she was found to be fully conscious, but aphasic. Visual acuity was impaired (left > right). Papilledema was present bilaterally.

Contrast-enhanced magnetic resonance imaging (MRI) of the brain revealed a heterogeneous lesion in the left frontal lobe, measuring 6 cm × 4 cm × 4 cm, which showed T2 and fluid attenuated inversion recovery hyperintensity, surrounded by thick rim of isointensity [Figure 1]. The lesion had caused effacement of the adjacent sulci, the adjacent part of rostrum and genu of corpus callosum. Mass effect was seen along ipsilateral frontal horn of lateral ventricle and left basal ganglia, with perilesional edema, hemorrhage, calcification and areas of necrosis within mass. There was midline shift of 9 mm toward right with subfalcine herniation. Ipsilateral ventricular system was compressed while contralateral ventricles were dilated.

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She underwent frontal craniotomy and Simpson grade 1 excision of the tumor. The vascular tumor (with some necrotic areas) was found fixed to the dura. Gross total excision was done. Following surgery, speech and vision returned to normal.

Histopathology revealed anaplastic HPC, grade 3 (WHO, 2007) with many slit-like and stag-horn (HPC) blood vessels lined by a single layer endothelial cells. Tumor cells were oval to elongate, arranged in sheets and short fascicles. Tumor cells showed moderate nuclear pleomorphism. Some areas showed brisk mitosis (12–14/10 high-power fields [HPF]). No normal brain parenchyma was included in the material. No dura was identified [Figure 2].

Immunohistochemistry was done for CD34, bcl-2 and epithelial membrane antigen (EMA). The specimen showed expression of CD34 and bcl-2 while EMA was negative.

Postoperative MRI brain showed areas of gliosis left frontal lobe with the extraaxial subacute hematoma with dural enhancement (postoperative changes).

Following postoperative recovery, patient was subjected to adjuvant whole brain radiotherapy (WBRT), in view of high-grade disease and positive margins on detailed histopathology. WBRT was delivered using Co-60 and field borders defined on conventional simulator. Dose of 54 Gy in 30 fractions, 180 cGy/fraction/day was delivered. Subsequently, the patient was put on the follow-up since 6 months and asymptomatic.

DISCUSSION

The mere existence of HPC as a distinct pathological entity is still ignored by most clinicians and pathologists. Formerly called angioblastic meningioma, these have only recently been included as a group by itself (WHO classification 1993) due to their distinct clinical and pathological characteristics.[3,4]

Hemangiopericytoma occurs in both sexes with 60% in men, mainly in fourth to fifth decade of life.[2] These tumors usually progress to a favorable outcome, but 20–30% of cases behave in a malignant fashion. It needs to be considered that malignant cases may be very aggressive with a high rate of local recurrence approaching 91% and a propensity to metastasize.[5]

Usually, single mass is seen attached to the meninges of the brain or spinal cord (resembling meningioma radiographically). It may also be confused with fibrous histocytoma, solitary fibrous tumor, metastasis or mesenchymal osteochondroma. These can also present with hypoglycemia due to the secretion of IGF-I and IGF-II.

A dense reticulin network typically investing individual cells is one of the most characteristic features of the tumor.[6] IHC is of limited help in direct recognition of HPC. There is no definite immunohistochemical marker for this tumor; however, lesional cells may be reactive for vimentin, actin, factor XIII a, laminin and CD34. EMA and S100 are generally not expressed. Pathological findings are confirmatory and allow for the correct diagnosis.[7,8]

Grade 2 tumors are the most common. Criteria for grade 3 include >5 mitoses/10 HPF with moderate to high nuclear atypia, moderate to high cellularity, presence of necrosis, vascular invasion and pleomorphism.[9]

In cases of benign HPC, complete surgical resection is sufficient. On the other hand, in cases of malignant HPC, the addition of radiation therapy or chemotherapy may be indicated, especially in high-grade lesions, large tumors, +ve surgical margins. Unresectable tumors require radiotherapy.

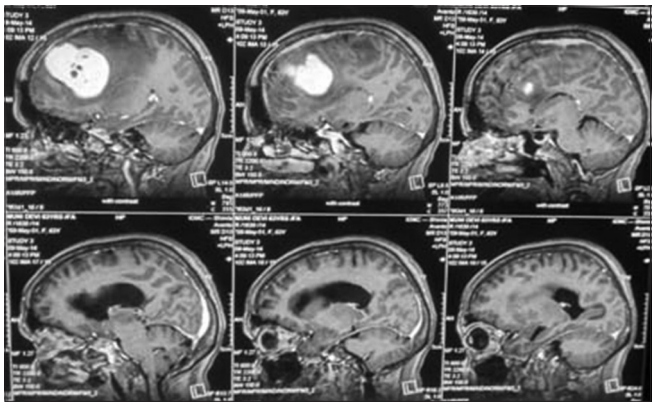

Figure 1: T1-weighted image showing the postcontrast enhancement of an extraaxial mass in fronto-parietal lobe: Saggital section

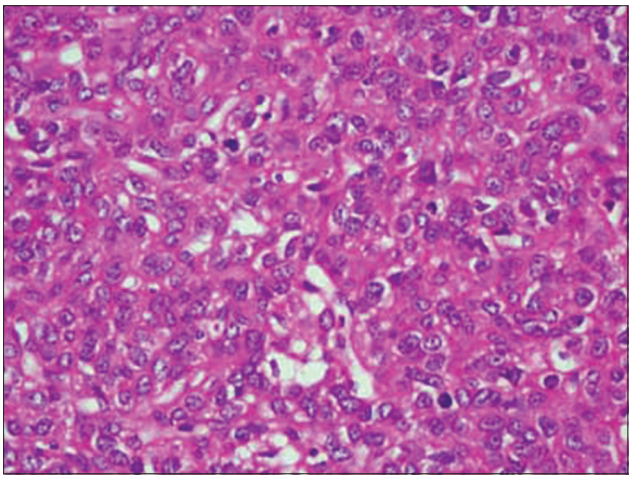

Figure 2: Tumor is composed of oval to spindle-shaped cells with frequent mitotic activity (H and E, ×40)

The drugs actinomycin-D, adriamycin, cyclophosphamide, methotrexate and vincristine can be used for chemotherapy of unresectable or metastatic disease.

Ours is a rare case of grade 3 tumor. Melone *et al*.[10] in a retrospective review of 43 patients treated at Rome university found surgery and adjuvant radiotherapy to hinder tumor progression. Anaplastic variant was associated with reduced overall survival and reduced recurrence interval. This underlines the importance of this rare variant of this disease.

CONCLUSION

Anaplastic HPCs, as a rare variant, as well as intracranial HPC need to be reported. This would definitely aid in improving treatment strategies for this rare tumor. The role of chemotherapy in this aggressive variant also is not clear and needs to be explored.

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