Case Report

A case series of extremely rare and unusual cases of intracranial hemangiopericytoma: An institutional experience

Abhishek Purkayastha, Neelam Sharma, Virender Suhag, Tejas Pandya

Department of Radiation Oncology, Army Hospital Research and Referral, New Delhi, India

ABSTRACT

We report a case series of three extremely rare and unusual cases of intracranial hemangiopericytoma diagnosed and treated in our institute, scarcely reported in world literature until date. These are highly aggressive and uncommon tumors accounting for <1% of all intracranial neoplasms, arising from Zimmerman's pericytes around capillaries and postcapillary venules. First two patients presented in 2011 and 2012 and were diagnosed as hemangiopericytoma Grade II with vimentin and CD34 positivity on immunohistochemistry. In 2014, a third patient was diagnosed as an anaplastic Grade III tumor, positive for vimentin and negative for epithelial membrane antigen. All three patients were treated with adjuvant radiotherapy to the brain and were kept on follow-up. However, the first patient recurred after 5 years for which he underwent redo-surgery. Other two patients are on follow-up with no evidence of recurrence or distant metastasis.

Key words: Anaplastic, hemangiopericytoma, intracranial, radiotherapy

INTRODUCTION

Hemangiopericytoma is an extremely rare and unusual tumor with an aggressive natural history, accounting for <1% of all intracranial tumors.[1-3] These are highly vascular dural-based neoplasms[2] with a propensity for local recurrence and distant metastasis.[3] They arise predominantly in soft tissues and very rarely in central nervous system (CNS).[2] With radiographic resemblance to meningioma and histological similarity to soft tissue tumors, it poses a diagnostic dilemma and a therapeutic challenge. Radiotherapy (RT) remains the cornerstone of adjuvant therapy due to high probability of local recurrence after surgery[3] while chemotherapy as salvage therapy has shown inconsequential results.[4]

Address for correspondence: Dr. Abhishek Purkayastha, Department of Radiation Oncology, Army Hospital Research and Referral, Dhaula Kuan, Delhi Cantonment - 110 010, New Delhi, India. E-mail: abhi5296@gmail.com

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CASE REPORTS

Case 1

A 28-year-old male presented with headache and vomiting in March 2011. Magnetic resonance imaging (MRI) showed 8 cm × 5.6 cm × 5.5 cm well-circumscribed mass lesion left parieto-occipital region.

Case 2

A 35-year-old male presented with headache in July 2012. MRI brain revealed an extra-axial globular mass lesion 7.0 cm × 6.0 cm × 5.8 cm right temporoparietal region [Figure 1]. Post gross total resection (GTR) histopathological report (HPR) of both patients revealed hemangiopericytoma WHO Grade II, vimentin and CD34 positivity [Figure 2], Ki-67 <10% and <8%, respectively, on immunohistochemistry (IHC). Both patients were treated with adjuvant conformal RT to brain to a dose of 54 Gy in 27 fractions.

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Case 3

A 33-year-old male presented with headache in November 2014. MRI brain showed a well-defined extra-axial mass lesion measuring 7.2 cm × 5.2 cm × 5.1 cm in midline anterior frontal region, compressing and displacing both frontal lobes causing erosion of overlying inner skull table. Magnetic resonance venography showed highly vascularized mass infiltrating anterior one-third of superior sagittal sinus. Metastatic work-up with positron emission tomography scan was negative. He underwent GTR with excision of the tumor. Postsurgery HPR revealed anaplastic hemangiopericytoma WHO Grade III with staghorn pattern of cells [Figure 3]. IHC was immunopositive for vimentin [Figure 4], negative for epithelial membrane antigen (EMA) with Ki-67 > 18%. He received adjuvant conformal RT to brain to a dose of 60 Gy in 30 fractions.

All patients tolerated treatment well and were kept on regular follow-up. The first patient presented with headache and progressive weakness of right upper limb in August 2015. MRI brain revealed 5.7 cm × 5.3 cm × 4.8 cm lesion at the primary site suggestive of recurrence for which he underwent redo-surgery. Postoperative period was uneventful. The other two patients are also on follow-up without any recurrence or distant metastasis.

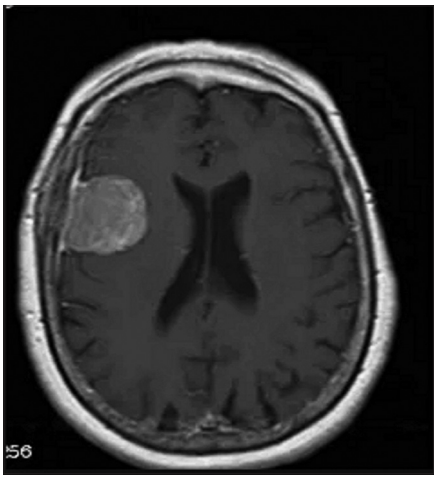

Figure 1: Magnetic resonance imaging brain showing an extra-axial globular mass lesion 7.0 cm × 6.0 cm × 5.8 cm right temporoparietal region in case 2

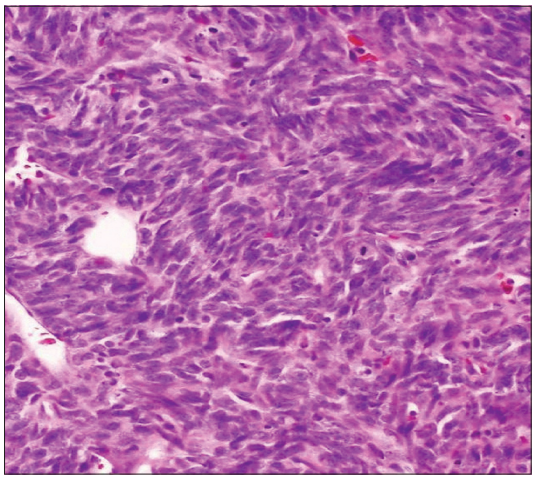

Figure 3: Histopathology report of case 3 showing staghorn pattern in highly cellular tumor cells with scant cytoplasm interspersed with numerous vascular channels and mild nuclear atypia (H and E, ×400)

DISCUSSION

HPC is an extremely rare and unusual neoplasm that accounts for <1% of CNS tumors[1-3] and 0.8% of all CNS tumors diagnosed and treated in our institute from 2008 to 2015. HPCs represent 3–4% of meningeal tumors, 3–5% of all soft tissue sarcomas, and 1% of vascular tumors.[2] About 15–30% of all HPCs occur in head and neck region, and 5% occur in the sinonasal area.[2] Historically, the term hemangiopericytoma was first described by Schmidt in 1937 and named by Stout in 1942. Guthrie *et al*. gave the term angioblastic meningioma; however, WHO in 1993 changed the term to hemangiopericytoma and classified it as a distinct pathologic entity mesenchymal nonmeningothelial

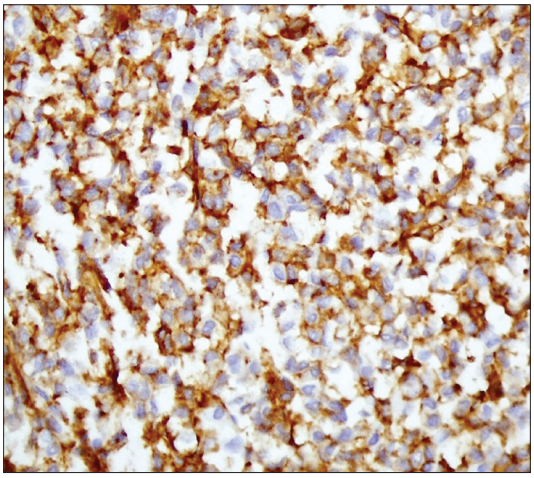

Figure 2: Immunohistochemistry showing CD34 positivity (×40)

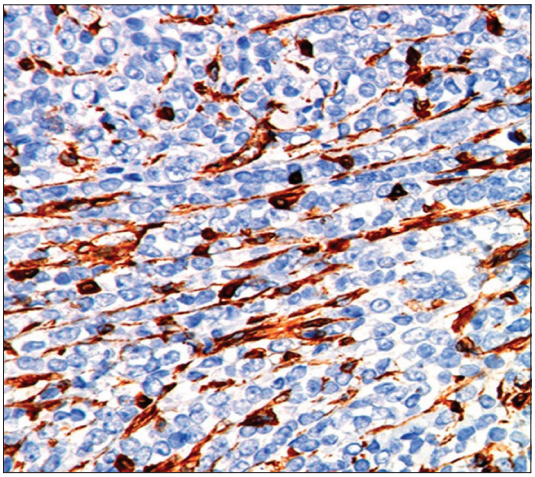

Figure 4: Immunohistochemistry showing tumor cells positive for vimentin (×40)

tumor in 2000. HPC occurs in both sexes with equal frequency and is found mainly in adults[2] at a median age of 42 years.[5]

Most common location of intracranial HPCs is frontal, middle, and posterior fossa.[6] They are highly vascular, occur at an earlier age than other meningeal tumors, recur locoregionally with higher frequency at a mean time of 75 months,[2] and metastasize to bone, lung, liver, kidney, pancreas, and adrenals.[2] HPCs have an indolent course but are notable for high rates of recurrence and distant metastasis many years after treatment,[3] like our first case recurred after 4 years. Studies suggest that high mitotic rates and proliferative indices may signify malignant behavior.

Neoplasms mimicking intracranial HPC are meningiomas and dural-based lymphomas. Clinically and radiographically intracranial HPCs resemble meningiomas[2] which frequently show hyperostosis of bone and calcifications within the tumor on MRI or computed tomography while HPCs show bone erosion and lack of both calcification and hyperostosis.[5,7] HPCs are composed of tightly packed pericytes around thin-walled endothelial lined vascular channels[2] and show a classical staghorn vascular pattern of spindle cells. Since histopathologic features of HPC and meningioma can mimic each other, IHC plays an important role in differentiating these two entities. HPC shows positivity for CD34 while meningioma is EMA positive.[8]

There is a role of RT as definitive modality in case of surgically unresectable and inaccessible tumor or as adjuvant therapy, since this tumor tends to recur even after complete excision.[2,9] Postoperative RT has been associated with decreased local recurrence and increased overall survival (OS)[10] with effect of radiation seen as long as 5 years after treatment. Regarding the role of chemotherapy in the treatment of HPCs, no impact on locoregional disease control, disease-free survival, or OS has been seen. Chamberlain and Glantz[4] reported a case series of 15 patients using chemotherapeutic agents vincristine, adriamycin, and cyclophosphamide followed by alfa-interferon and subsequently by ifosfamide, carboplatin, and etoposide regimen. However, it failed to demonstrate any significant benefit as a salvage therapy in recurrent and refractory cases. Though GTR is the established definite treatment, conformal RT remains the mainstay of adjuvant management as it is reduces local recurrence, increases OS, improves the quality of life and overall prognosis.[3]

CONCLUSION

Intracranial HPC is an extremely rare CNS neoplasm. We reported this case series to highlight its extreme rarity, its clinicopathological findings and define the role of RT as the primary adjuvant treatment affecting the final prognosis. We recommend that the diagnosis of HPC should always be considered in a young adult presenting with focal motor deficit and dural based well-circumscribed vascular mass lesion in view of its higher rates of local recurrence and systemic metastasis as compared to other CNS tumors.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Someya M, Sakata KI, Oouchi A, Nagakura H, Satoh M, Hareyama M. Four cases of meningeal hemangiopericytoma treated with surgery and radiotherapy. Jpn J Clin Oncol 2001;31:548-52.
2. Perez CA, Thorstad WL. Perez and Brady's Principles and Practice of Radiation Oncology. 6th ed. Philadelphia: Lippincott Williams and Wilkins, Walters Kluwer Business; 2013. p. 875-7, 986.
3. Alén JF, Lobato RD, Gómez PA, Boto GR, Lagares A, Ramos A, *et al.* Intracranial hemangiopericytoma: Study of 12 cases. Acta Neurochir (Wien) 2001;143:575-86.
4. Chamberlain MC, Glantz MJ. Sequential salvage chemotherapy for recurrent intracranial hemangiopericytoma. Neurosurgery 2008;63:720-6.
5. Guthrie BL, Ebersold MJ, Scheithauer BW, Shaw EG. Meningeal hemangiopericytoma: Histopathological features, treatment, and long-term follow-up of 44 cases. Neurosurgery 1989;25:514-22.
6. Pitkethly DT, Hardman JM, Kempe LG, Earle KM. Angioblastic meningiomas; clinicopathologic study of 81 cases. J Neurosurg 1970;32:539-44.
7. Chiechi MV, Smirniotopoulos JG, Mena H. Intracranial hemangiopericytomas: MR and CT features. AJNR Am J Neuroradiol 1996;17:1365-71.
8. Rajaram V, Brat DJ, Perry A. Anaplastic meningioma versus meningeal hemangiopericytoma: Immunohistochemical and genetic markers. Hum Pathol 2004;35:1413-8.
9. Soyuer S, Chang EL, Selek U, McCutcheon IE, Maor MH. Intracranial meningeal hemangiopericytoma: The role of radiotherapy: Report of 29 cases and review of the literature. Cancer 2004;100:1491-7.
10. Uemura S, Kuratsu J, Hamada J, Yoshioka S, Kochi M, Ushio Y, *et al.* Effect of radiation therapy against intracranial hemangiopericytoma. Neurol Med Chir (Tokyo) 1992;32:328-32.