Intraventricular Hemangiopericytoma: A Case Report and Literature Review

James E. Towner, MD, Department of Neurosurgery; Yan Michael Li, MD, PhD, Department of Neurosurgery; Mahlon D. Johnson, MD, PhD, Department of Pathology and Laboratory Medicine
University of Rochester School of Medicine and Dentistry

Introduction

Hemangiopericytomas (HPC) are vascular mesenchymal tumors, classified by the World Health Organization (WHO) as grade II or, if anaplastic elements are identified, grade III neoplasms. In 1974, the first intracranial HPC was reported by Begg and Garrett, who described a left parietal intraparenchymal HPC. They believed that entities described as angioblastic meningiomas originated from pericytes and should be reclassified as HPC. In 1993, the WHO revised classification of angioblastic meningiomas from meningioma variant to HPC. Exceedingly rare for intracranial location - only 11 cases of intracranial HPC in the literature.

Case Report

History and Physical

- 23-year-old right-handed man with a chief complaint of bifrontal headache for 1 month.
- Described headaches as pulsating, worse in the morning and improved by evening.
- Endorsed occasional photophobia, blurring vision, nausea, emesis, right sided numbness, and intermittent episodes of dizziness with food finding and blurring.
- Found to have a right homonymous hemianopia and a right pronator drift.

Radiographic Evaluation

CT and MRI scan were obtained, revealing a large mass in the left lateral ventricle as seen in Figures 1A,B,C.

Operative Course

- Left superior parietal transcallosal approach to the tumor.
- Intraparenchymal piece removal of the tumor was undertaken, resulting in a radiographic gross total resection (GTR), illustrated in Figure 2.

Pathology

- Histology showed a hypocellular, spindle cell tumor with slit-like blood vessels illustrated in Figure 3A.
- Retinulin stained showed extensive pericellular staining, seen in Figure 3B.
- Tumor cell staining highly positive for vimentin and CD34, patchy for epithelial membrane antigen (EMA), scattered for BCL-2; and rare for CD34.
- Final diagnosis of HPC solitary fibrous tumor (SFT) WHO grade III.

Postoperative Course and Follow-up

- Transient worsening of expressive aphasia, resolved prior to discharge. Rapid resolution of right hand weakness and headache after surgery. No change in right homonymous hemianopia.
- Received adjuvant treatment of 60 Gy of local radiation over 30 fractions.
- No evidence of recurrence or metastasis found at 6-month follow-up scans, which included MRI of the neuraxis and PET CT.

Discussion

Epidemiology

- CNS HPC - 0.4% of all intracranial neoplasms and 2-3% of all primary tumors of the dura.
- 65-76% of all CNS HPC are located supratentorially and average age of diagnosis is 41 years.2
- 91% of intracranial HPC are located in the lateral ventricle and average age of diagnosis is 41 years.
- Intracranial HPC have a high incidence of recurrence, ranging from 34% to 90%, and a trend to metastasize (in order of decreasing frequency, bone, lung, and liver), ranging from 12% to 55%.3
- Mean survival 13 years, with 5- and 10-year survival estimates of 86% and 66%, respectively, and 10-year progression-free survival ranging from 79% to 39%.4
- Average time to recurrence with grade III neoplasms 59 months compared with 95 months in grade II neoplasms.5

Radiographic Evaluation

- Solid heterogeneously hypodense mass on CT, isointense on T2-weighted MRI, mixed intensity on T1 MRI, and heterogeneously enhancing post-gadolinium contrast.5
- Differential diagnosis based on radiographic evaluation includes meningioma, SFT, high-grade glioma, ependymoma, chordoid papilloma, and metastasis, particularly renal cell carcinoma (renal cell carcinoma involved intraventricular spread in 37.5% of all patients who had brain metastasis compared with 8% for all other diseases involving brain metastases).3

Pathology

- Very vascular, with abundant capillaries and pathognomonic clusters of staghorn blood vessels.
- Prominent basilar laminar on reticulin or collagen IV stains help differentiate HPC from meningiomas.
- Immunohistochemically, focal CD34 positivity and negative for EMA; in comparison, meningiomas are generally negative for CD34 and positive for EMA.
- Intraventricular HPC likely originate from pericytes in the tela choidea of the choroid plexus.
- Outside the CNS, HPC is considered to be on same neoplastic continuum as SFT. Although WHO still distinguishes, some feel CNS SFT and HPC are merely different histologic grades of the same neoplasm based on radiographic, pathologic, and prognostic similarities.

Treatment

- Surgical resection gold standard - compared with subtotal resection, GTR associated with increase in overall survival, from 175 to 235 months, and recurrence-free survival, from 54 to 117 months.
- Rates of intracranial HPC GTR range from 38% to 83%. Rate of GTR of intracranial HPC is 91% despite deep location and rarity (likely in part because of lack of venous sinus involvement).5
- 35% patients with intracranial HPC receive adjuvant radiation, but exact role and dose are controversial.2

Conclusion

- Standard treatment involves aggressive surgical resection.
- Role of adjuvant radiation less well defined, but commonly pursued postoperatively.
- Regardless of extent of resection or adjuvant treatment, close follow-up to evaluate for evidence of local recurrence and distant metastasis essential.

References