Monocular Blindness as Presentation Manifestation of Neuroblastoma

Sanjeev Sivakumar, MD¹, Janet Poulik, MD²,
and Lalitha Sivaswamy, MD³

Keywords

optic canal, neuroblastoma, metastases

The Neurohospitalist
2016, Vol. 6(1) 41
© The Author(s) 2015
Reprints and permission:
sagepub.com/journalsPermissions.nav
DOI: 10.1177/1941874415585546
nhos.sagepub.com

Figure 1. A, 3.0T Magnetic resonance imaging of the brain and orbits. T1-weighted postcontrast image demonstrating an enhancing mass in the region of the sphenoid bone with involvement of the sphenoid sinuses and posterior ethmoid air cells. The mass involves the bilateral cavernous sinuses, superior orbital fissures (red arrows), and right optic canal with mass effect on both optic nerves (blue arrows). The right internal carotid artery is completely obliterated (absence of flow void, white arrow). B, Microbiopsy, hematoxylin & eosin, magnification $\times 40$. Poorly differentiated neuroblastoma with high mitotic karyorrhectic index consisting of cells with scant cytoplasm and nuclei with salt and pepper chromatin and many cells with one or more prominent nucleoli.

A previously healthy 4-year-old patient was admitted with a 2-week history of extreme irritability with no apparent etiology on initial examination. Coincidentally, the child was noted to have intermittent strabismus involving the right eye. His visual acuity was 20/200 on the affected side with an afferent pupillary defect. On dilated direct ophthalmoscopy, a swollen optic disc was noted. There was no restriction of ocular movements.

Magnetic resonance imaging of the brain and the orbits revealed a large, lobulated, contrast-enhancing mass in the region of the sphenoid bone (Figure 1A). The right optic nerve appeared to be completely surrounded by the mass (accounting for the edema of the optic disc on examination).

Meta-iodobenzylguanidine (MIBG) scan (using iodine-123-MIBG) showed evidence of diffuse metastases in axial and appendicular bones. The primary lesion was identified as a neuroblastoma involving the left adrenal medulla. Biopsy of the adrenal mass revealed round blue cells with scant

cytoplasm and scattered Homer-Wright rosettes suggestive of neuroblastoma (Figure 1B). Cytogenetics revealed amplification of the Neuroblastoma MYC Oncogene (NMYC), which correlates with increased frequency of intracranial or orbital, liver, and lung metastasis.¹

Neuroblastoma—the most common extracranial solid tumor of childhood—consists of cells derived from the neural crest. Patients presenting with blindness often have stage IV disease. At the time of initial diagnosis, up to 18% of children will have metastases to the cranial bones/orbit.¹ Neurologists who encounter optic disc swelling should entertain a broad differential diagnosis and consider early imaging to avoid potentially fatal pitfalls.

Authors' Note

Consent from the appropriate institutional review board was obtained prior to the preparation of the manuscript.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

Reference

1. DuBois SG, Kalika Y, Lukens JN, et al. Metastatic sites in stage IV and IVS neuroblastoma correlate with age, tumor biology, and survival. *J Pediatr Hematol Oncol*. 1999;21(3):181-189.

¹ Department of Neurology, Detroit Medical Center/Wayne State University, Detroit, MI, USA

² Department of Pathology, Children's Hospital of Michigan, Detroit, MI, USA

³ Department of Pediatrics and Neurology, Children's Hospital of Michigan, Detroit, MI, USA

Corresponding Author:

Lalitha Sivaswamy, Department of Pediatrics and Neurology, Wayne State University School of Medicine, Children's Hospital of Michigan, 3901 Beaubien, Detroit, MI 48201, USA.

Email: lsivaswamy@med.wayne.edu