An infant intracranial tumor with nystagmus

Li-Li Ge, Cai-Ping Shi

Department of Ophthalmology, The Children's Hospital, School of Medicine, Zhejiang University, Hangzhou, Zhejiang 310000, China.

To the Editor: A 4-month-old girl was brought to the Department of Ophthalmology in the Children's Hospital, School of Medicine, Zhejiang University, due to "sudden eyeball shaking for a week." The patient was a full-term gravida 2 para 2 infant without any incidence of trauma/surgery; further, the family's medical history did not reveal any risk factors. Ophthalmological examination revealed bilateral horizontal tremor of the eyes, and both eyes showed light-chasing response. The anterior segment examination was normal. After mydriasis, the wide-angle retinal imaging system examination revealed that the binocular optic discs were clear with normal color and flat retina; no obvious abnormal exudation or position occupying was seen, but the left eye showed obvious retinal vasodilation with slight distortion [Figure 1A and 1B]. Skull magnetic resonance imaging suggested the presence of a large tumor in the sellar region with extensive skull/brain transplantation in the skull base and spinal canal, as well as supratentorial hydrocephalus [Figure 1C]. The patient was then admitted to the Neurosurgery Department of the Children's Hospital, Zhejiang University for intracranial tumor resection. The post-operative pathological diagnosis suggested pilomyxoid astrocytoma (PMA; the tumor cells were loose and star-shaped and grew around the blood vessels; immunohistochemistry: Global fibrillary acific protein (GFAP)+, oligodendrocyte transcription factor-2 (Olig2)+, CD34−, neuron specific enolase (NSE)+, epithelial membrane antigen (EMA)±, synaptophysin (SYN)+, P53 tumor suppressor protein (P53)+, H3K27M±, Ki-67 approximately 15%+) [Figure 1D–F].

Pediatric PMA is a rare central nervous system tumor. PMA occurs mainly in the sellar region of infants and young children with a mean age of 18 months.[1] The onset age of PMA in this study was earlier than that reported in the above literature; however, the site of PMA observed in our case was the most common site of the disease according to the above study. A study evaluated the presenting signs and symptoms of children with central nervous system tumors, and found that in addition to symptoms of increased intracranial pressure, papilledema was present in 13% of the patients, strabismus in 7%, cranial nerve palsies in 7%, and abnormal eye movements in 6%.[2] Nystagmus refers to repeated, reciprocating, and involuntary eye movements initiated by the slow movement of eyes away from the visual target. Nystagmus is grouped into infantile nystagmus that usually appears in the first 3 to 6 months of life, and acquired nystagmus (AN) that appears later. Many forms of AN can be attributed to disturbances in the three mechanisms that normally ensure steady gaze, namely: visual fixation, the vestibulo-ocular reflex, and the mechanism that makes it possible to hold the eyes at an eccentric eye position. See-saw nystagmus is linked to both parasellar lesions of the optic chiasma (eg, pituitary tumors) and achiasma.[3] In this study; however, the patient exhibited nystagmus because the occurrence of tumor was in the saddle area, and the conduction pathway was impeded. Lavery *et al*[4] have reported that the occurrence of nystagmus in chiasmatic gliomas was age-dependent; it was seen mainly in younger children. Ehrt *et al*[5] have described the following signs and symptoms of AN: onset after the age of 4 months, oscillopsia, dissociated (asymmetric) nystagmus, preserved optokinetic nystagmus, afferent pupillary defect, papilloedema, and neurological symptoms such as vertigo and nausea. Pediatricians and ophthalmologists must look for signs of AN and refer those patients for neurological and imaging work-up. In our case, the patient developed nystagmus 4 months later; blood vessel expansion at the fundus occurred, and hence it was suspected to be AN caused by intracranial lesions. This case report reminds us that in addition to the routine anterior segment examination, detailed fundus examination should also be performed in infants and young children at the outpatient clinic. If necessary, special eye examinations and imaging evaluations are required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient's or guardian's consent forms. In the form, the patient's guardians have given their consent for her images

Access this article online

Quick Response Code:

Website: www.cmj.org

DOI: 10.1097/CM9.0000000000000597

Correspondence to: Dr. Li-Li Ge, Department of Ophthalmology, The Children's Hospital, School of Medicine, Zhejiang University, Hangzhou, Zhejiang 310000, China
E-Mail: 6512055@zju.edu.cn

Chinese Medical Journal 2020;133(1)

Received: 03-09-2019 **Edited by:** Ning-Ning Wang

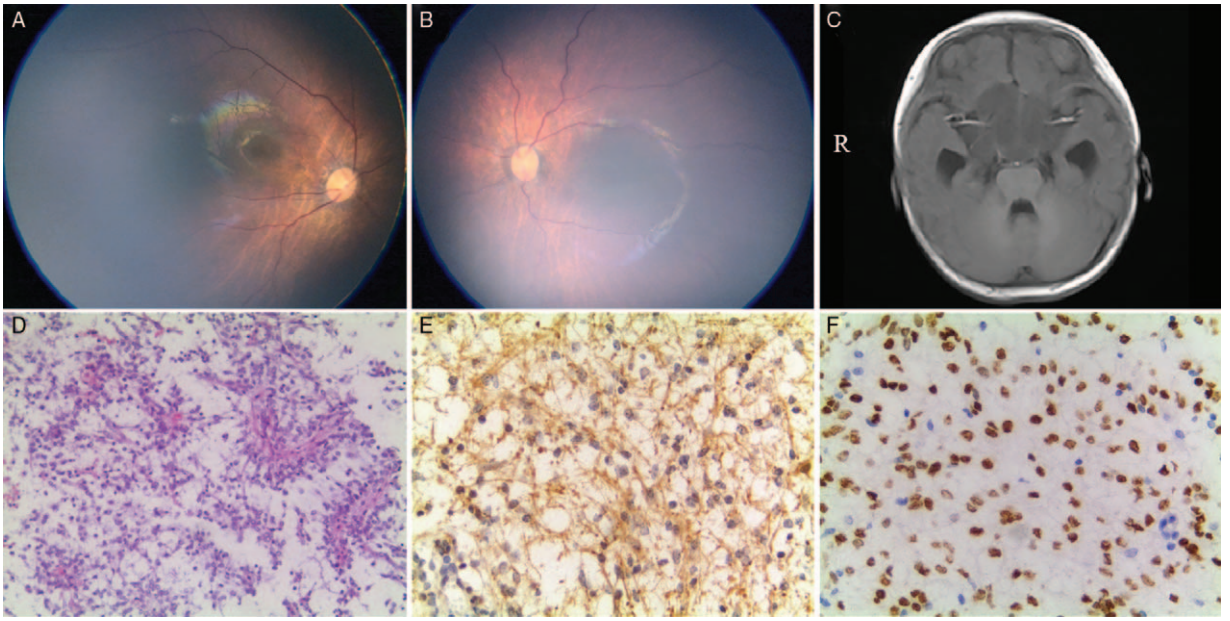

Figure 1: OD fundus examination: the optic disc is clear, with normal color and flat retina; no obvious abnormal exudation or position occupying is seen (A). OS fundus examination: the optic is clear with normal color and flat retina; obvious retinal vasodilation with slight distortion; the retinal blood vessels in the right eye shows no obvious distortion or thickening (B). Head MRI scan shows a mass in the sellar region, the tumor compresses the optic chiasm, which is involved in the bilateral cavernous sinus (C). The tumor cells are loose and star-shaped and grow around the blood vessels (D). Immunohistochemistry: GFAP+ (original magnification ×200) (E). Immunohistochemistry: Olig2+ (original magnification ×200) (F). GFAP: Glial fibrillary acidic protein; MRI: Magnetic resonance imaging; OD: Right eye; Olig: Oligodendrocyte; OS: Left eye.

and other clinical information to be reported in the journal. The patient's guardians understand that her name and initial will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Conflicts of interest

None.

References

1. Komotar RJ, Burger PC, Carson BS, Brem H, Olivi A, Goldthwaite PT, *et al*. Pilocytic and pilomyxoid hypothalamic/chiasmatic astrocytomas. Neurosurgery 2004;54:72–80. doi: 10.1227/01.neu.0000097266. 89676.25.
2. Bresson D, Herman P, Polivka M, Froelich S. Sellar lesions/pathology. Otolaryng Clin N Am 2016;49:63–93. doi: 10.1016/j.otc.2015.09.004.
3. Abadi RV. Mechanisms underlying nystagmus. J R Soc Med 2002;95:231–234. doi: 10.1258/jrsm.95.5.231.
4. Lavery MA, O'Neill JF, Chu FC, Martyn LJ. Acquired nystagmus in early childhood: a presenting sign of intracranial tumor. Ophthalmology 1984;91:425–453. doi: 10.1016/s0161-6420(84)34269-5.
5. Ehrt O. Infantile and acquired nystagmus in childhood. Eur J Paediatr Neurol 2012;16:567–572. doi: 10.1016/j.ejpn.2012.02.010.

How to cite this article: Ge LL, Shi CP. An infant intracranial tumor with nystagmus. Chin Med J 2020;133:99–100. doi: 10.1097/CM9.0000000000000597