Multidirectional Nystagmus as the Presenting Sign of Brain Tumor with Hydrocephalus

Tommaso Bellini1  Laura Siri2  Elisa De Grandis2,3  Clelia Formigoni3  Domenico Tortora4  Emanuela Piccotti1

1 Pediatric Emergency Room and Emergency Medicine, IRCCS Istituto Giannina Gaslini, Genoa, Italy
2 Neuropsychiatry Unit, Gaslini Children Hospital, IRCCS Istituto Giannina Gaslini, Genoa, Italy
3 Department of Neuroscience, Rehabilitation, Ophthalmology, Genetics, Maternal and Child Health (DINOGMI), University of Genoa, Italy
4 Neuroradiology Unit, Gaslini Children Hospital, IRCCS Istituto Giannina Gaslini, Genoa, Italy

Address for correspondence Tommaso Bellini, MD, Pediatric Emergency Room and Emergency Medicine Unit, IRCCS Istituto Giannina Gaslini, Via G. Gaslini 5, 16147 Genoa, Italy (e-mail: tommasobellini@gaslini.org).

A previously healthy 11-year-old boy was admitted to our Emergency Room for a 1-month history of intermittent diplopia. The main clinical signs were right-beating horizontal nystagmus with a vertical component in the upward gaze (Video 1) and bilateral papilledema; he did not suffer headache, vomit, bradycardia, or any additional cerebellar signs such as ataxia, dysmetria, dysdiadochokinesis, or tremor. Brain magnetic resonance showed a large, disseminated, bulky mass involving the fourth ventricle and the mesencephalic aqueduct with cerebrospinal fluid dissemination along the lateral ventricles and the spinal cord, causing decompensating hydrocephalus and intracranial hypertension (Fig. 1). The subsequent biopsy confirmed the histological diagnosis of medulloblastoma and its inoperability due to dissemination.

Video 1


Medulloblastoma is the most common malignant brain tumor in childhood and occurs primarily in the cerebellum; patients affected present with signs and symptoms of cerebellar dysfunction and increased intracranial pressure that may cause cranial nerve deficits, and the diagnosis is often delayed by many months after the first symptoms appear.1 Interestingly, our patient reported only intermittent diplopia and the physical examination showed only horizontal/vertical nystagmus with no other neurological signs, emphasizing the importance of maintaining high clinical suspicion and the need for a detailed and prompt evaluation for the earliest possible diagnosis, even if the patient presents only subtle symptoms as diplopia.2,3

Funding
None.

Conflict of Interest
None declared.

References
Fig. 1  Magnetic resonance (MR) images of an 11-year-old boy with medulloblastoma. Axial (A) diffusion-weighted imaging (DWI) and (B,C) T2-weighted images, (D) sagittal T2-weighted and (E) T1-postcontrast images, and (F) spinal sagittal T2-DRIVE images show a bulky mass lesion in the fourth ventricle and mesencephalic aqueduct with nodular dissemination in lateral ventricles and in the spinal cord (arrows). Lesions determined supratentorial hydrocephalus.