Unusual Presentations of Posterior Fossa Tumors in Children

The Indian Journal of Pediatrics

ISSN 0019-5456
Volume 77
Number 9

Indian J Pediatr (2010)
77:1039-1040
DOI 10.1007/
s12098-010-0146-6

Springer

Indian J Pediatr (2010) 77:1039–1040
DOI 10.1007/s12098-010-0146-6

SCIENTIFIC LETTERS TO THE EDITOR

Unusual Presentations of Posterior Fossa Tumors in Children

Sweta Mohanty · Ira Shah · Sudha Rao · Neelu Desai

Received: 29 July 2009 / Accepted: 21 May 2010 / Published online: 24 August 2010

Most pediatric posterior fossa tumors (PFTs) present with involvement of brainstem structures like cranial nerves or pyramidal tract, or with signs of raised Intracranial Pressure (ICP) if the passage of CSF is obstructed. Cerebellar signs occur late, as also involvement of vital brainstem centers and herniation. We describe 3 cases of PFTs with unusual presentations.

Case 1 An 8 years old boy came with 8 days' history of vertigo which exacerbated on lying down, frequent falls and bitemporal headache. Temperature, blood pressure, tone and power were normal, but limb reflexes were brisk. Cerebellar signs of dysdiadochokinesia, past pointing and tandem walking were positive. No signs of raised intracranial pressure (ICP) were present. MRI brain revealed diffuse pontine glioma.

Case 2 An 8 years old boy came with complaints of neck pain since 1 year, intermittent frontoparietal headache and vomiting since 3 days. Blood pressure was high. Hypertonia of limbs and brisk reflexes with ill-sustained clonus were present. Neck stiffness and crack-pot sign were positive. Fundoscopy revealed optic disc atrophy. MRI brain showed diffuse pontine glioma.

Case 3 An 8 years old girl came with left eye squint since 12 days. She had normal blood pressure, tone and reflexes, no focal neurological deficit and no signs of raised ICP. However, horizontal nystagmus, dysdiadochokinesia and past pointing suggested cerebellar involvement. MRI brain revealed a mass in cerebellar vermis suggestive of medulloblastoma.

S. Mohanty · I. Shah (✉) · S. Rao · N. Desai
Department of Pediatric Medicine,
B.J.Wadia Hospital for Mumbai,
240 D Walkeshwar Road,
Mumbai 400006, India
e-mail: irashah@pediatriconcall.com

A syndrome typical of brainstem tumors is the combination of multiple bilateral deficits of cranial nerves with pyramidal tract signs and truncal ataxia [1], in the absence of ICP symptoms [2], as they do not obstruct CSF passages early. Spinal pain with headache may be seen with metastatic leptomeningeal involvement. However, Case 2 had neck pain since 1 year, an unusual presentation of PFTs caused by irritation of posterior roots of the cervical cord. Case 1 too had an unusual presentation with positional vertigo. Brisk reflexes in both, and cerebellar signs in Case 1, aided localization. Vomiting, like in other brainstem tumors, was most likely caused due to direct infiltration of the medullary vomiting centre, rather than raised ICP [3].

Cerebellar tumors often attain a surprisingly large size while leaving the passage of CSF intact, and produce localizing symptoms only when the tumor attains considerable volume. Since, at that time, CSF blockage also gives rise to obstructive hydrocephalus, cerebellar signs are obscured by symptoms of increased ICP, as well as remote signs of cranial nerves [3]. Thus, in Case 3 with cerebellar medulloblastoma, it was surprising to find squint due to left abducent paresis with cerebellar signs, in the absence of raised ICP. Such local involvement of cranial nerves is more characteristic of brainstem rather than cerebellar tumors. Also, nystagmus is conspicuously absent or mild in medulloblastomas [1].

In summary, brainstem gliomas have varied presentations. Increased ICP at presentation, although unusual, may be seen, and neck pain may be seen due to nerve root irritation. Positional vertigo is a rare presentation of aggressive PFTs [4].

Eye squint warrants search for cerebellar signs, being an unusual presentation of cerebellar tumors.

References

1. Cohen D. Tumors of the brain and spinal cord. In: Swaiman, editor. Pediatric neurology. 4th ed. Philadelphia: Elsevier; 2006. 1709–18.
2. Maria BL, Menkes JH. Tumors of the nervous system. In: Menkes J, Sarnat H, editors. Child neurology. 6th ed. Philadelphia: Lippincott Williams and Wilkins; 2000. 787–858.
3. Koos WT, Miller MH. Infratentorial tumors- general discussion. In: Koos WT, Miller MH, editors. Intracranial tumors of infants and children. 1st ed. Stuttgart: CV Mosby Company; 1971. 254–304.
4. Chan Y. Posterior fossa lymphoma with initial vertigo presentation. Eur Arch of Otorhinolaryngol. 2009;266:495–500.