Paediatric intraventricular meningiomas. A report of two cases

Sushma Ramraje[1], Sakshi Kulkarni[2] , Barnik Choudhury [2]

1. Associate Professor, 2. Assistant Professor

Department of Pathology, Grant Medical College,Mumbai-400008

CASE REPORT

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Corresponding Author:
Ramraje. Sushma. N
2/15, Dhanwantari, Doctors Quarters,
JJ Hospital Campus, Byculla, Mumbai-400008
Email: sushmaramraje@yahoo.com

Abstract

Meningiomas are rare in children comprising less than 3% of paediatric brain tumours and only 1.5-1.8% of all intracranial neoplasms. Intraventricular meningiomas (IVM) account for 0.5-5% of all meningiomas. They arise in the ventricles from arachnoid cap cells contained within the choroid plexus, the tela choroidea, or the velum interpositum. Paediatric tumours also show an association with neurofibromatosis type 2 and previous radiation exposure.

We present two cases of intraventricular meningiomas, both in children. The age and site of the lesion in the two cases are uncommon. Excision of the lesions caused a total subsidence of the symptoms with no reported recurrences.

Key Words: Intraventricular meningioma (IVM), children

Background

Meningiomas are central nervous system neoplasms derived from arachnoid cap cells. They are the second most common brain tumours after gliomas, are rare in children[1] and account for 15–25% of primary intracranial tumours in adults but less than 3% of brain tumours in children. Primary meningiomas in the ventricular system without dural attachments are an extremely rare but a well-described entity originating within the ventricles in the brain. They arise from the arachnoid cells embedded in the choroid plexus. They are often located in lateral ventricles and occasionally in the third and fourth ventricles. Because the choroid plexus is more bulky in the lateral ventricles, incidence of lateral ventricle meningiomas is higher as compared to those in the third or fourth ventricles.[2] Their peak incidence is in the third and fourth decades.

We present the clinical, radiological and pathological features of intraventricular meningioma in two patients, aged 13 years and 8 years. Neither patient showed evidence of association with neurofibromatosis or previous radiation exposure.

Case details

Case 1: A 13-year-old female presented with headache, vomiting and ataxia for a duration of two months. Magnetic resonance imaging (MRI) scan (Figure 1) showed a well-defined, midline mass lesion in the posterior fossa, predominantly hypointense on T_1 and heterogeneously hyperintense on T_2-weighted images with intense enhancement on post contrast study (Figure 2). It measured 4.3x3.5 cms in size, compressing the fourth ventricle leading to mild dilatation of lateral, third and fourth ventricles. The mass appeared to spread along the ventricular surface of pons and in the interpeduncular cistern and was diagnosed clinically and radiologically as medulloblastoma. Posterior craniotomy with complete enucleation of the lesion was performed along with an intra-operative squash cytology. A diagnosis of medulloblastoma was entertained. We received gray-white tissue bits measuring 5x4x3cm. Histologically, the tumour comprised of cells arranged in lobulated clusters with areas of whorling. Individual cells were monomorphic, round to polygonal with moderate amount of pale eosinophilic to clear cytoplasm, ovoid nuclei with fine granular chromatin and conspicuous nucleoli. Mitotic activity was low (Figure 3). On immunohistochemistry, the cells showed strong positivity for epithelial membrane antigen. The final diagnosis was meningothelial meningioma.

Histologically, medulloblastomas are composed of densely packed cells with hyperchromatic nuclei, scant cytoplasm, high mitotic index, occasional Homer-Wright pseudorosettes and frequent apoptosis which were not seen in our case

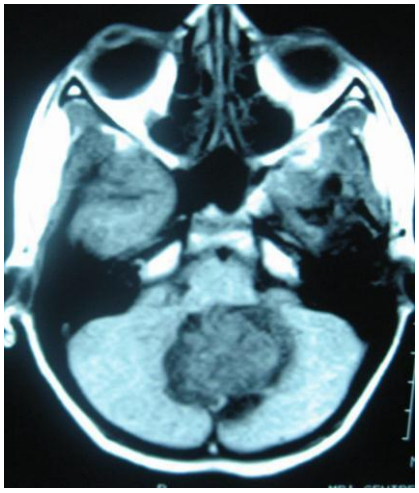

Figure 1: MR imaging brain, T1 weighted axial image: A well defined midline posterior fossa mass lesion

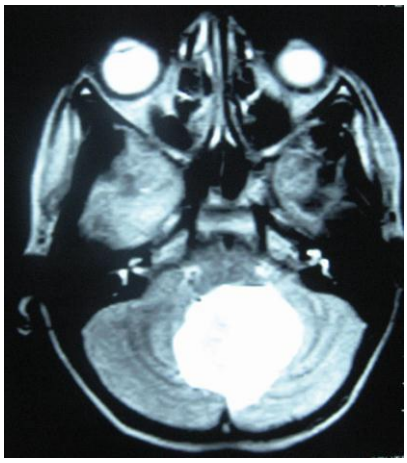

Figure 2: MR imaging brain, T1 weighted post-contrast image: Intensely enhancing mass lesion in the midline in posterior fossa.

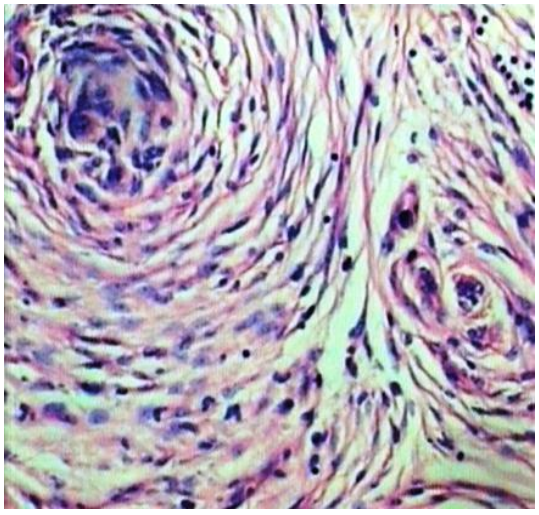

Figure 3: Cells arranged in clusters with areas of whorling (H&E 400X).

Case 2: An 8-year-old female presented with headache, vomiting and ataxia for six months and reduced vision for one month. MRI scan showed a midline heterogeneously enhancing mass lesion in the posterior fossa in the region of the fourth ventricle (Figures 4, 5). It measured 7.6x4.1x4 cms and showed intratumoral cystic areas. Anteriorly it was compressing the brainstem, superiorly reaching up to the root of the fourth ventricle, extending inferiorly into cistern magna and through the foramen magnum into the cervical spinal canal up to the lower margin of C_2 vertebral body and was diagnosed clinically and radiologically as medulloblastoma. Posterior craniotomy with complete enucleation and an intra-operative squash cytology was performed. A diagnosis of high grade tumour suggestive of medulloblastoma was offered. We received fragmented gray white bits measuring 7.5x4x4 cms. Histologically, fascicles of tumour cells with tapering cytoplasmic processes and ovoid bland nuclei were seen. Mitotic activity was low (Figure 6). On immunohistochemistry cells were strongly positive for epithelial membrane antigen. The tumour was diagnosed as fibroblastic meningioma.

Both children showed complete recovery with no recurrence to date.

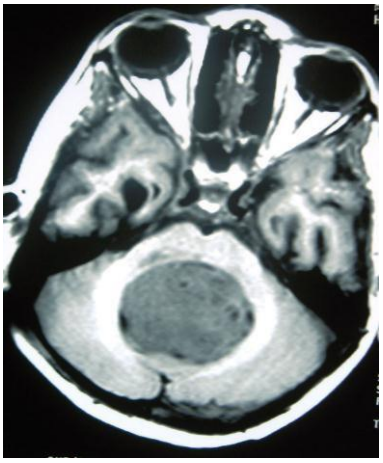

Figure 4: MR imaging brain, T1 weighted axial image : A well defined posterior fossa mass lesion in the region of fourth ventricle

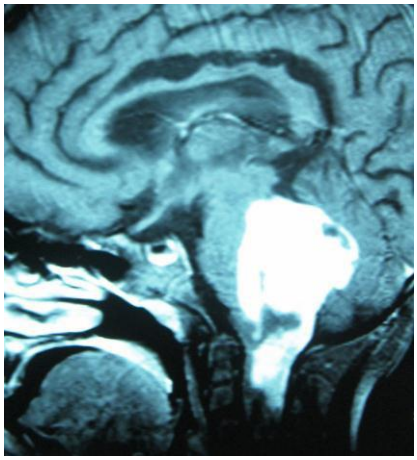

Figure 5: MR imaging brain, T1 weighted sagittal post-contrast image: Intensely enhancing posterior fossa mass lesion in the region of fourth ventricle

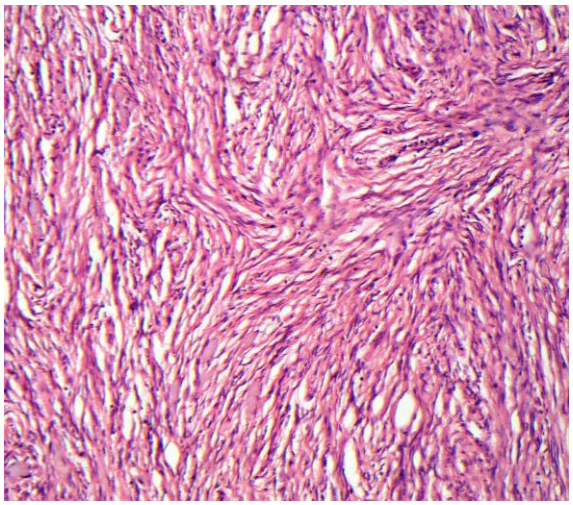

Figure 6: Fascicles of tumor cells with tapering cytoplasmic processes and bland nuclei. (H&E, 200X).

Discussion

Meningiomas arise from arachnoid cap cells, the specialised cells in arachnoid granulations. Similarly, intraventricular meningiomas arise from arachnoid cells present in the choroid plexus[2]. Third ventricle tumours arise from the tela of the velum interpositum, which is the space between the two layers of tela in the roof of the third ventricle that contains the posterior medial choroidal arteries and internal cerebral veins.[3] Fourth ventricle meningiomas arise from choroids or interior tela choroidea.[4]

The incidence of lateral ventricle meningiomas in adults is between 0.5-5% of all intracranial meningiomas[5, 6] and IVMs are rare in the third and fourth ventricles.[7] In children younger than 20 years of age, they account for less than 3% of all childhood tumours of the central nervous system.[8]

Paediatric meningiomas differ from their adult counterparts by their male preponderance, atypicality of location, higher rates of malignant change, recurrence and association with neurofibromatosis.[9] They are frequently cystic as seen in one of our cases.

IVMs are slow-growing tumours that become large prior to detection. Ventricles of the brain provide space for tumour expansion, and until the cerebrospinal fluid pathways are mechanically occluded, manifestations are mild and non-specific. Cushing and Eisenhardt[10] described five clinical features of lateral ventricle tumors: 1) pressure symptoms with ipsilateral headache; 2) contralateral macula splitting homonymous hemianopia; 3) contralateral sensorimotor paresis, more marked sensory involvement, and numbness over trigeminal distribution; 4) cerebellar involvement in more than half of patients; and 5) paralexia, worsened by surgical intervention in left-sided tumours.

The clinical symptoms are due to increased intracranial pressure, whatever their location. Headache, temporal lobe seizures and visual field deficits are commonly seen. [5,11] Cerebellar signs and hypothalamic features with or without endocrinopathy may be seen in fourth and third ventricle tumours respectively.

Neuroimaging is necessary for diagnosis. IVMs may be densely calcified and visible on plain skull radiographs.[12] Computed tomography (CT) scans may show calcification, obstructive hydrocephalus, and uniform contrast enhancement, but MR imaging is needed for diagnosis. The tumour is hypo- or isointense on T1-weighted MR images and enhances densely and uniformly after the addition of gadolinium. Fibroblastic meningiomas, commonly seen in the lateral ventricles, are hypointense on T2-weighted images.[13] Tumours that occur in cases of neurofibromatosis 2 are well delineated on MRI. CT and MR images show intense uniform enhancement after injection of contrast media due to their lack of blood-brain barrier.

The histopathology of IVMs can be of any type (predominantly fibrous, fibroblastic, meningothelial, or psammomatous) as defined by the World Health Organization's classification of meningiomas. Generally, they are diagnosed based on morphological features alone. They are positive for vimentin and epithelial membrane antigen.

In conclusion, IVMs are rare tumours in the paediatric population presenting with features of raised intracranial pressure. However, they can be removed intact without damage to surrounding areas with meticulous planned surgery and show no recurrence.

References

1. Maranhão-Filho P, João Campos CS, Lima MA. Intracranial meningiomas in children: Ten-year experience. Pediatr Neurol. 2008;39: 415-417.

2. Bhatoe HS, Singh P, Dutta V. Intraventricular meningiomas: A clinicopathological study and review. Neurosurgical Focus.2006; 20(3):E9.

3. Tung H, Apuzzo MLJ: Meningiomas of the third ventricle, in Al-Mefty O (ed): Meningiomas. New York: Raven Press 1991, pp 583-591.

4. Guidetti B, Delfini R. Lateral and fourth ventricle meningiomas, in Al-Mefty O (ed): Meningiomas. New York: Raven Press, 1991, pp 569-587.

5. Erman T, Göcer AI, Erdogan E. Intraventricular meningiomas: a review and report of eight cases. Neurosurg Quarterly.2004; 14:154-160.

6. Lapras C, Deruty R, Bret PH. Tumors of the lateral ventricles. Adv Tech Stand Neurosurg.1984; 11:103-167.

7. Nakamura M, Roser F, Bundschuh O, Vorkapic P, Samii M. Intraventricular meningiomas: a review of 16 cases with reference to the literature. Surg Neurol.2003; 59:491-504.

8. Rushing EJ, Olsen C, Mena H, Rueda ME, Lee YS, Keating RF, Packer RJ, Santi M. Central nervous system meningiomas in the first two decades of life: a clinicopathological analysis of 87 patients. J Neurosurg. 2005;103:489–495.

9. Arivazhagan A, Devi BI, Kolluri SV, Abraham RG, Sampath S, Chandramouli BA. Pediatric intracranial meningiomas--do they differ from their counterparts in adults? Pediatric Neurosurg. 2008; 44:43-48.

10. Cushing H, Eisenhardt L. Meningiomas. Their classification, regional behavior, life history and surgical end results. Springfield, IL: Charles C Thomas, 1938, pp 139-149.

11. Tew JM, Larson JJ Jr: Intraventricular meningiomas, in Kaye AH, Black PML (eds): Operative Neurosurgery. London: Churchill Livingstone, 2000, pp 575-585.

12. Bhatoe HS, Gill HS, Sreeram MN. Densely calcified lateral ventricular meningioma. Ind J Radiol Imaging. 1994; 4:117-118.

13.McDermott MW. Intraventricular meningiomas. Neurosurg Clin N Am. 2003; 14:559- 569.

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PEER REVIEW

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CONFLICTS OF INTEREST

The authors declare that they have no competing interests