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Case Report

Disseminated glioneuronal tumor: a rare presentation in children

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ABSTRACT

Gliomas, though most common pediatric central nervous system tumor, can manifest as disseminated glio-neuronal tumor, a rare variant in children. Clinical presentation depends on its location, type and age of child. We are presenting 8 years old male child with fever, projectile vomiting and severe headache which woke him up from deep sleep for 1 month. He had positive meningeal signs and raised intracranial tension with cerebrospinal fluid picture suggestive of partially treated meningitis. There was no improvement even on adequate duration of intravenous antibiotics and had appearance of new onset false localizing signs, MRI brain showed features of cryptococcal meningitis for which India ink staining was negative. As clinical picture was unlike of meningitis, repeat 3 tesla MRI brain was done. Expert neuro-radiologist's opinion was in favor of disseminated glio-neuronal tumor which was confirmed on histopathological examination. Child underwent laminectomy in TATA memorial hospital and advised palliative care. Child succumbed at home within 6 months of illness.

Keywords: Pediatric CNS tumor, Glioma, Disseminated Glioneuronal Tumor

INTRODUCTION

Gliomas are most common pediatric CNS tumors.[1] It is a leading cause of morbidity and mortality in children worldwide. Disseminated glio-neuronal tumors in pediatric population are rare. Clinical presentation depends on its location, type, age of child and obstruction to cerebrospinal fluid pathway. MRI is the standard neuroimaging modality, diagnosis is confirmed by histopathological analysis. It has multimodal therapeutic approach.[1]

CASE REPORT

An 8 years old male child born out of non-consanguineous marriage was with fever, projectile vomiting and severe headache which woke him up from his deep sleep for 1 month. He was treated previously for meningitis however he had recurrence of the symptoms. There was no past history of tuberculosis or contact of tuberculosis. Child was developmentally appropriate for age. On general examination vitals were stable. Child was frail as weight (17 kg: below 3rd centiles) and height (122 cm: 3rd and 50th centiles). On central nervous system examination child had signs of meningeal irritation with positive Kernig's sign and neck rigidity. Fundus examination was normal. On investigations, complete blood count was suggestive of moderate anaemia Hb- 8.4 g% (11.5-14.5 g%), total leukocyte count-7800/mm^3 (4000-12000/mm^3), platelets- 3,20,000/mm^3 (150-400x10^3/mm^3), blood culture: no growth. Cerebrospinal fluid analysis showed evidence of partially treated meningitis (proteins-267 mg/dl, sugar- 30 mg/dl and cells-60, all lymphocytes) for which child was started on injection ceftriaxone (100 mg/kg/day) and meropenem (40 mg/kg/dose) for 14 days. Child gradually developed signs of raised intracranial pressure including papilledema, 6th nerve palsy and hypertension and was treated appropriately. The work up for tuberculosis and sero-status was negative. MRI brain (Figure 1 and 2) showed cystic lesions in bilateral thalamus and diffuse intracranial and intraspinal leptomeningeal enhancement with obstructive hydrocephalus. Differential

diagnosis were tubercular meningitis with raised intracranial pressure, cryptococcal meningitis or space occupying lesions. On reviewing MRI, radiologist's opinion was in favour of cryptococcal meningitis for which India ink staining for cryptococcal was negative. As clinical picture was unlike of partially treated meningitis or tubercular meningitis, a repeat 3 tesla MRI was repeated. Senior radiologist's opinion was in favour of a leptomeningeal glio-neuronal tumour. Biopsy from leptomeningeal region and PET scan were suggestive of high-grade Astrocytoma consistent with drop metastasis from thalamic space occupying lesion.

Immunohistochemistry was positive for p53 protein and ATRX protein. Child was referred to Tata memorial hospital for further management. Child underwent laminectomy. Child was bedridden and expired at home within 6 months of illness.

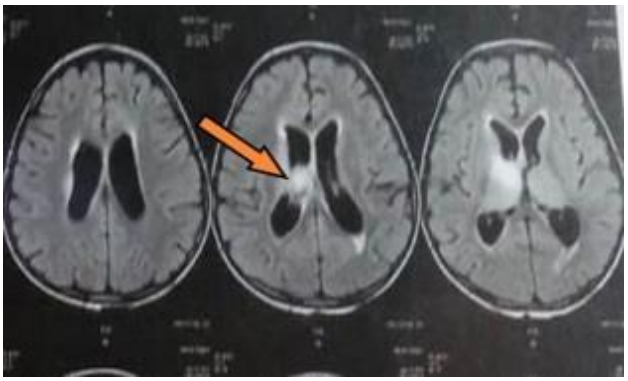

Figure 1: Axial T1 view showing diffuse infiltrative lesion involving thalamus and bilateral dilated ventricle.

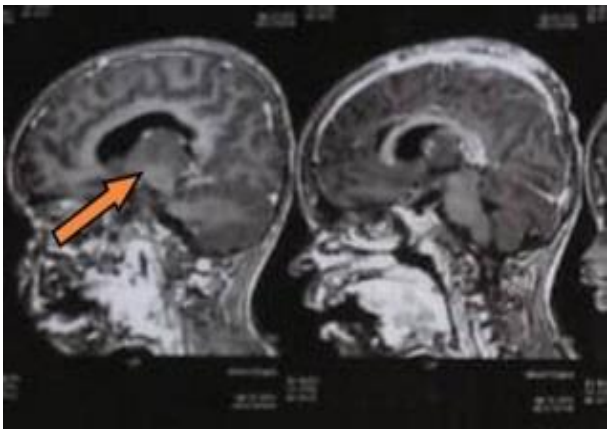

Figure 2: Sagittal T1 view showing involvement of thalamus, septum pellucidum, fornices, brainstem and cerebellar vermis.

DISCUSSION

A diffuse lepto-meningeal glio-neuronal tumor represents 5-10% of brain tumors in pediatric population. As per recent WHO 2016 classification system gliomas are classified into two types-non-diffuse (low grade) and diffuse gliomas (high grade). Diffuse gliomas include astrocytomas, oligodendrogliomas and rare mixed oligodendroglial-astrocytic of WHO grade-2 (low grade), grade-3 (anaplastic) or grade 4 (glioblastoma).[2]

Radio-logically these tumors are characterized by either widespread leptomeningeal enhancement or with discrete intraparenchymal lesions unlike low grade gliomas. Usual sites involve thalamus (13%), brainstem, basal cisterns, spinal cord (3%).[3,4] On histological examination they appear as rounded cells with background of desmoplasia.[5-8] High grade gliomas develop symptoms in a very short course in the form of headache, vomiting, altered sensorium, seizures and focal neurological deficits (as per specific area involved).

Though having a benign appearance these tumors often behave aggressively and are resistant to standard chemotherapy. Genetically these tumors are found to express BRAF rearrangement /duplication, loss of chromosome 1p and 1p19q co-deletions.[5-10] Currently high-grade gliomas is a tissue diagnosis, therefore, confirmatory diagnosis is made after surgical/biopsy specimen analysis by trained neuropathologist. Treatment relies on supportive management, chemotherapy and radiotherapy.

These tumors usually have bad prognosis with 5 years survival rate of around 10%, with recurrence as high as 27.5%.[11] Palliative care maintains the quality of life.

Thus, though there are enough data about its epidemiology in western population, there are only few reports from developing country like India. This is because of lack of national cancer registry due to which we depend on local hospital-based registries to know their incidence. Thus, awareness among health care professionals is being encouraged to report each and every case of pediatric CNS tumor. This will contribute in developing new resources and interventions, thus promoting quality of life of children.

CONCLUSION

In children diagnosis of CNS tumor can be delayed due to non-specific complaints. It is mandatory to co -relate a report clinically and obtain expert opinion if no response of standard therapy. Complete resection is best treatment option but some tumors are aggressive and recurrence is also common. Prognosis depends on age of onset, location and intrinsic properties of tumor.

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REFERENCES

1. Kliegman RM, Stanton BF, Geme JW, Schor NF. Brain tumors in childhood. In: Kliegman RM, eds. Nelson Textbook of Pediatrics. 20th ed. India: Elsevier; 2016: 2453-2460.
2. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, et al. The 2007 WHO classification of tumours of the central nervous system. Acta Neuropathol. 2007;114(2):97-109.
3. Kramm CM, Butenhoff S, Rausche U, Metz M, Kortmann RD, Pietsch T, et al. Thalamic high-grade gliomas in children: a distinct clinical subset? Neuro Oncol. 2011;13(6):680-9.
4. Wolff B, Ng A, Roth D, Parthey K, Metz M, Eyrich M, et al. Pediatric high grade glioma of the spinal cord: results of the HIT-GBM database. J Neurooncol. 2012;107(1):139-46.
5. Rodriguez FJ, Perry A, Rosenblum MK, Krawitz S, Cohen KJ, Lin D, et al. Disseminated oligodendroglial-like leptomeningeal tumor of childhood: a distinctive clinicopathologic entity. Acta Neuropathol. 2012;124(5):627-41.
6. Gardiman MP, Fassan M, Orvieto E, Avella D, Denaro L, Calderone M, Severino M, et al. Diffuse leptomeningeal glioneuronal tumors: a new entity? Brain Pathol. 2010;20(2):361-6.
7. Agamanolis DP, Katsetos CD, Klonk CJ, Bartkowski HM, Ganapathy S, Staugaitis SM, et al. An unusual form of superficially disseminated glioma in children: report of 3 cases. J Child Neurol. 2012;27(6):727-33.
8. Armao DM, Stone J, Castillo M, Mitchell KM, Bouldin TW, Suzuki K. Diffuse leptomeningeal oligodendrogliomatosis: radiologic/pathologic correlation. AJNR Am J Neuroradiol. 2000;21(6):1122-6.
9. Bourne TD, Mandell JW, Matsumoto JA, Jane JA, Lopes MB. Primary disseminated leptomeningeal oligodendroglioma with 1p deletion. Case report. J Neurosurg. 2006;105(6):465-9.
10. Michotte A, Chaskis C, Sadones J, Veld PI, Neyns B. Primary leptomeningeal anaplastic oligodendroglioma with a 1p36-19q13 deletion: report of a unique case successfully treated with Temozolomide. J Neurol Sci. 2009;287(1):267-70.
11. Broniscer A, Gajjar A. Supratentorial high-grade astrocytoma and diffuse brainstem glioma: two challenges for the pediatric oncologist. Oncologist. 2004;9(2):197-206.

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