Desmoplastic infantile ganglioglioma - A case report


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Discussion

CJD affects individuals between 50-70 years of age. Psychiatric symptoms in the form of cognitive impairment, delusions and hallucinations, depression, euphoria and aggression are occasionally described in the literature and may form prodromal symptoms of CJD in 18-39% of cases.1,2,3,4 A diagnosis of probable CJD of sporadic form was considered in the present case on the basis of clinical features. In this case, dementia appears to have been caused by diffuse carcinomatous meningitis and multiple cortical tumor nodules altering the CSF dynamics and deranged cortical activity.5 The topographic distribution of the lesions in the frontal cortex, hippocampus, striatum account for the clinical features of dementia, ataxia and extrapyramidal symptoms. The diffuse background theta activity on EEG, could be due to multifocal disruption of the synaptic connectivity in the cortex by the tumor deposits. The absence of abnormal prion protein in the brain excluded the diagnosis of CJD. Only one other such report of metastatic disease presenting like CJD is recorded in the literature.5 Though in cases of rapidly progressive dementia with myoclonus and EEG features, CJD forms an important clinical diagnosis, it is imperative to make efforts to exclude other treatable causes mimicking CJD, as happened in this case. This case report highlights that despite advances in clinical diagnosis, confirmation by tissue diagnosis remains a prerequisite for confirmation of CJD.

References

Desmoplastic infantile ganglioglioma is a very rare supratentorial tumor occurring in the first two years of life. A five-month-old female infant presented with recurrent seizures, large head and loss of acquired milestones. Computerized Tomographic Scan of brain showed a large subarachnoid cyst with a solid intensely contrast enhancing tumor in the right temporoparietal region with severe degree of mass effect. Craniotomy and total excision of the tumor followed subsequently by subduro-peritoneal shunt for the extracerebral fluid collection was done. The child made good recovery. Histopathology revealed features of desmoplastic infantile ganglioglioma, viz., marked desmoplastic component with glial and neuronal elements. Immunohistochemistry showed positive staining for glial fibrillary acidic protein (GFAP) with areas of synaptophysin and chromogranin positivity. Desmoplastic infantile ganglioglioma is a rare tumor of infancy, which has excellent prognosis after total excision. No adjuvant therapy is required. This is the first Indian report of desmoplastic infantile ganglioglioma out of less than fifty cases reported worldwide.

Key Words: Desmoplastic infantile ganglioglioma, supratentorial neuroepithelial tumors of infancy, gangliogliomas

Introduction

Desmoplastic infantile ganglioglioma was first described by Vanden Berg et al in 1987. Together with superficial astrocitoma, they are also grouped as desmoplastic supratentorial neuroepithelial tumors of infancy. Desmoplastic infantile gangliogliomas are extremely rare and less than 50 cases have been reported in the literature. A 5-month-old infant with right temporoparietal desmoplastic infantile ganglioglioma which was successfully managed is presented in this report and the relevant literature has been reviewed.

Case Report

A 5-month-old Bengali female child presented with enlargement of head, repeated generalized tonic-clonic seizures and loss of acquired milestones of one-month duration. At the time of admission, the child was irritable. The head circumference was 51 cm. And the anterior fontanelle was wide and tense. There was no neurological deficit. Computerized Tomographic scan (CT scan) of brain showed a large cystic tumor in the right frontotemporoparietal regions, with a moderate-sized, isodense and intensely enhancing surface mass (Figures 1 and 2).

A right frontotemporoparietal craniotomy was performed. The subarachnoid cyst contained xanthochromic fluid. The solid tumor was approximated to the dura. It was firm and relatively avascular and measured about 6 cm x 5 cm x 3 cm. The tumor was excised completely. The postoperative course was uneventful.

Histopathology showed the tumor with marked desmoplastic component with neoplastic astrocytes and a few scattered neuronal elements (Figure 3). Immunohistochemistry showed strong reaction with Glial Fibrillary Acidic Protein (GFAP) with areas showing posi-
Total external ophthalmoplegia induced by phenytoin: A case report and review of literature

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A 28-year-old male developed total external ophthalmoplegia following oral administration of phenytoin. The case is reported and its significance is discussed.

Key Words: External ophthalmoplegia, Phenytoin induced ophthalmoplegia, phenytoin toxicity.

Introduction

There have been innumerable reports concerning phenytoin toxicity, but few have mentioned its effects on eye movements other than nystagmus.1 Ophthalmoplegia has been reported with administration of large doses of Phenytoin,2,3 Phenobar-