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Case Report

An intracranial space occupying lesion in a 4-year-old child: a case report

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ABSTRACT

Primary central nervous system tumours are a heterogenous group of diseases that are collectively, the second most common malignancy in childhood and adolescence. The overall mortality among this group approaches 30%. Despite of high mortality rates, outcomes of these malignancies has improved with neurosurgery, radiation therapy, and chemotherapy as a therapeutic modality. Here authors have reported a 4-year female child who was diagnosed with brainstem glioma in present institute. The child presented with the typical clinical presentations of brainstem glioma.

Keywords: Brainstem, Hemiparesis, Tumour

INTRODUCTION

The etiology of pediatric brain tumours is not well defined. A male predominance is noted in the incidence of medulloblastoma and ependymoma. Familial and hereditary syndromes associated with an increased incidence of brain tumours.[1] The incidence of central nervous system (CNS) tumours in India ranges from 5 to 10 per 100,000 population with an increasing trend and accounts for 2% of malignancies.

The most common tumor was astrocytoma (34.7%) followed by medulloblastoma and supratentorial Primary neuroectodermal tumour (22.4%) and craniopharyngioma. Most of the astrocytic tumors were reported to be low grade commonly pilocytic astrocytoma and subependymal giant cell astrocytoma.[2]

CASE REPORT

A 4-year-old girl was brought to the OPD with a history of aggressive behaviour since 2 months, followed by intermittent fever, vomiting and a progressive gradual onset of left sided hemiparesis, drooling of saliva from left side and excessive tears from the left eye. There was no history of seizures, trauma, ear infections, jaundice, contact with tuberculosis. There was a history of admission for nutritional rehabilitation at 1.5 years of age.

On examination patient was conscious and oriented to time, person and place. Authors noted left sided hemiparesis with increased tone and clasp knife type spasticity. There was left abducent nerve paralysis and left facial nerve LMN type paralysis. Autonomic, cerebellar and sensory systems were intact. The child had a circumduction gait with wide base stance. There were no meningeal signs of irritation.

Blood investigations revealed normal haemogram pattern with normal leucocytes and platelet count. Serum sodium, potassium, calcium, uric acid was within normal limits. Serum lipid profile was borderline elevated. Neuroimaging (MRI T1 weighted image brain) depicted brainstem space occupying lesion (Glioma) mostly involving pons and medulla and right half of midbrain (Figure 1). Also noted was compression of fourth

ventricle with moderate obstructive hydrocephalus (Figure 2).

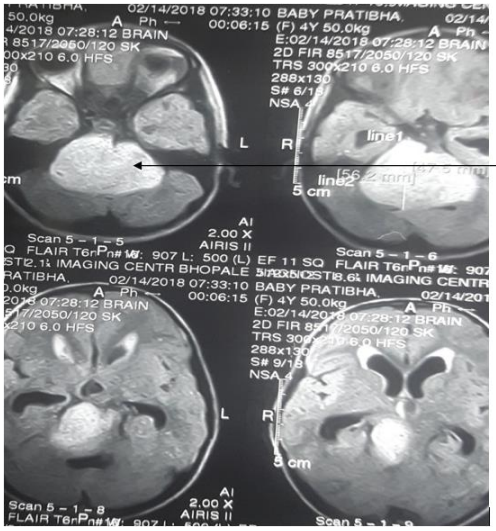

Figure 1: MRI T1 weighted image showing a mass lesion in the brainstem with its extensions over midbrain.

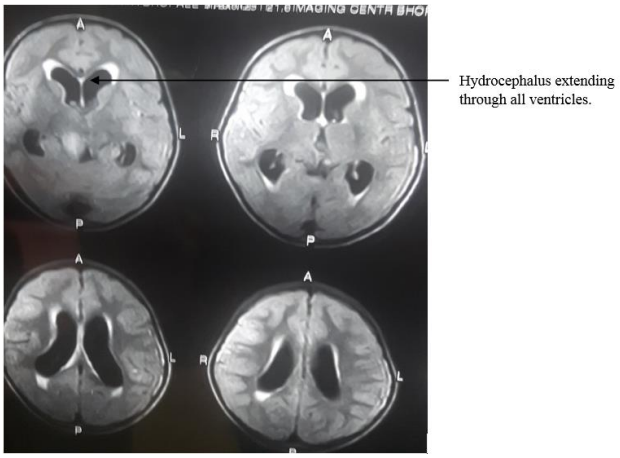

Figure 2: T1 weighted MRI brain showing moderate hydrocephalus.

The patient was then referred for radiation therapy. Due to monetary constraints they did not undertake the treatment and left against medical advice.

DISCUSSION

The recent literatures report a slight predominance of infratentorial tumour location (43.2%) followed by the supratentorial regions (40.9%), spinal cord (4.9%) and multiple sites (11%). There are age related differences in primary location of tumours. During the 1st year of life supratentorial tumours predominate and include most commonly choroid plexus complex tumours and teratoma. In children, aged 1-10 years, infratentorial tumours predominate with high incidence of medulloblastoma. The infratentorial tumours most commonly present with classic triad of headache, nausea and vomiting.[3-5]

Primary CNS tumors are more common in the first decade of life. The reported incidence of tumors is higher in whites. The overall incidence is highest among those between 0 and 4 years of age. The above-mentioned child falls in this group. The etiology of these tumors relate to genetic factors, environmental factors, maternal exposure to radiation and immunosuppression therapy.[6]

Gliomas are tumors of the central nervous system that arise from astrocytic or oligodendroglial lineages. The gliomas are classified into low grade and high-grade gliomas. High grade gliomas are relatively rare in children and comprise approximately 4% of all brain tumors in the age group of 0 – 14. These are generally slow growing tumors and the clinical presentation depends on the site of tumor. Generalized increased ICP can result in headache, vomiting, irritability. Cerebral tumors present with gait changes, ataxia, and nystagmus. Tumors within the optic pathway cause changes in visual fields, hormonal abnormalities and/ behavioural anomalies. Lesions in brainstem produce cranial nerve palsies and upper motor neuron signs such as contralateral weakness and ipsilateral ataxia. Supratentorial tumors present with hemiparesis or seizures.[7]

As discussed above the child in the case is clinically suspected to have a glioma in the brain stem. The grade of severity can be assessed with histopathological or neuroimaging correlation. With the advent of newer chemotherapy regimens such as TPDCV (thioguanine, procarbazine, dibromodulcitol, lomustine, vincristine), the survival rate in low grade gliomas have far increased.

Perkins et al has described that chemotherapy is equally effective in such case of CNS tumors and cranial radiotherapy is to be avoided to the maximum possible extent.[8]

Chris Johns et al highlights the therapeutic implications of targeting epigenetic lesions induced by histone mutations.[9] However, the impact of K27M and G34R/V on the global chromatin landscape is still largely unknown. In these articles they have tried to study the effect of various combinations of chemotherapy and radiotherapy in treating gliomas. Despite all these studies the therapeutic success of gliomas is incomplete.

CONCLUSION

To conclude pilocytic low grade gliomas encompass several histologic variants of tumors. The overall prognosis of these tumors is poor. Recent literatures on chemotherapy has quoted an increase in the survival rate of these tumors based on the type of chemotherapy received and the complete health status of the patient. Current management also includes surgical resection as

an option in combination with chemotherapy. Survival in these tumors still requires multiple therapeutic studies to try to improve the survival.

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