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Primary Diffuse Leptomeningeal Gliomatosis

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Abstract: Primary Diffuse Leptomeningeal Gliomatosis [PDLG] is a rare high grade central nervous system neoplasm, in which focal or diffuse evidence of gliomatous tissue is identified in subarachnoid space with no evidence of primary central nervous system neoplasm.Limited literature available on this entity defines the aggressive nature of this disease with dismal prognosis.
Keywords: Primary Diffuse leptomeningeal Gliomatosis.

INTRODUCTION

The incidence of all primary central nervous system [CNS] neoplasm in India ranges from 5 to 10 per one lakh population and accounts for about2% of all malignancies [1]. Gliomas account for 26.4% of all primary brain tumours, with malignant gliomas accounting for 19.9% of them [2]. PDLG is a rare high grade CNS neoplasm in which there is focal or diffuse evidence of gliomatous tissue infiltrating the subarachnoid space in the absence of a primary central nervous system neoplasm [3]. This condition was first described by Moore in 1954.Patient usually present with features of raised intracranial tension,cranial neuropathies and seizures. Due to nonspecific nature of clinical presentation, antemortemdiagnosis posea challenge to clinician. We report the case of a 15 year old young male who presented with features of raised intra cranial tension which was eventually diagnosed as PDLG.

CASE DESCRIPTION

This 15 year old boy presented to us with headache, vomitingand rapidly progressing quadriparesisof two weeks duration.Clinical examination revealed intact cranial nerve and sensory function but with grade zero powerin bothupper and lower limb. Magnetic Resonance Imagingrevealed T2 hyperintense diffuse leptomeningeal and ependymal enhancement in cerebral and cerebellar convexity,basal cistern,cerebellopontine cistern extending to sylvian fissure and fourth ventricle[Figure 1].Susceptibility weighted images showed multiple foci of blooming suggestive of high grade nature of the disease.Similar intraduralextramedullary nodular lesion was seen coating the entire spinal cord and cauda with diffuse compressive myelopathy[Figure 2].There was also a suspicious hyperintense lesion of 1.0 x0.5 cm in the left medial temporal lobe [Figure 3]. Cerebrospinal fluidcytology was negative for malignant dissemination or infective meningitis.Radiological diagnosis ofdiffusehigh grade glial neoplasm was made. Patient underwent ventriculoperitoneal shunt placement, excision of gliotic tissue at D1 spinal cord level and excision of left temporal lobe lesion. Histopathological examination of spinal cord lesion was consistent with primary diffuse leptomeningeal gliomatosis (GFAP strong positive, synaptophysinnegative and MIB labelling index very high 25-40%) [Figure 4a,b,c&d].Histology of suspicious temporal lobe lesion reported as reactive gliosis.

In view of the limited literature available onthe standard treatment protocol for this disease, patient was planned for craniospinal radiotherapy. Patient died while was on treatment due to disease progression.

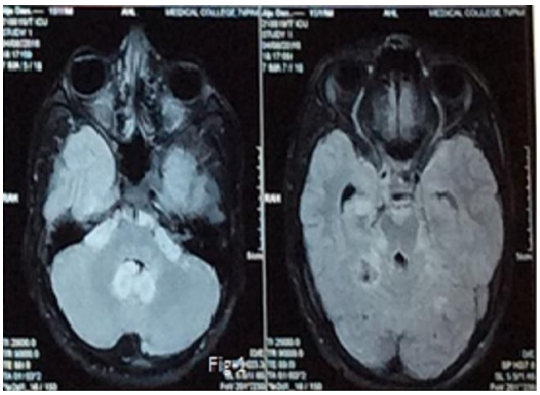

Fig-1: T2hyperintensity in basal and cerebellopontine cistern

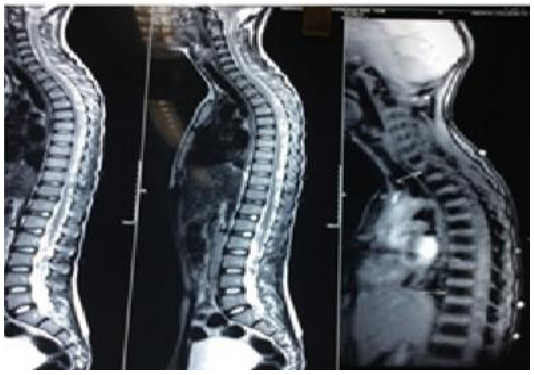

Fig-2: Multiple intraduralextramedullary nodular lesion coating entire spinal cord and cauda

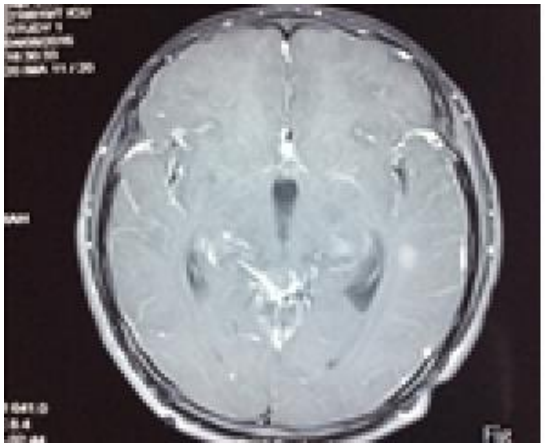

Fig-3: Enhancing lesion in temporal lobe

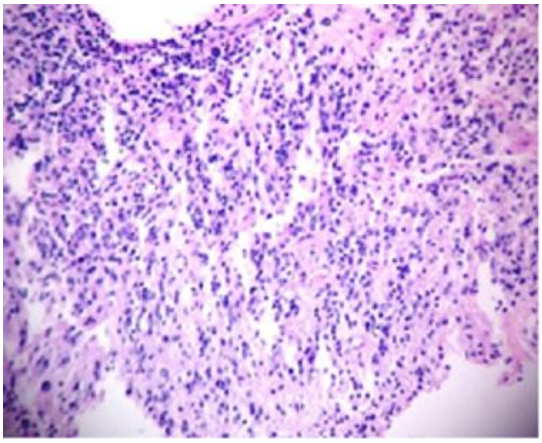

Fig-4a: HPE showing high grade neuroglial neoplasm

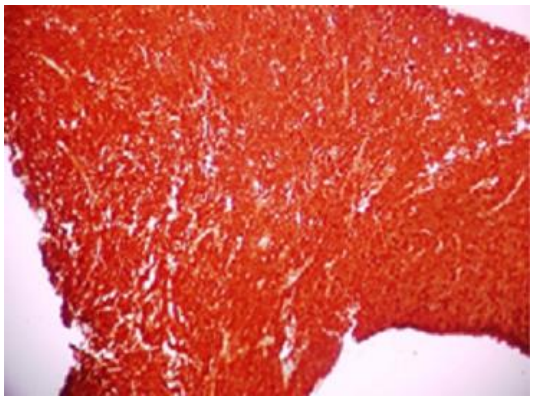

Fig-4b: IHC showing GFAP expression

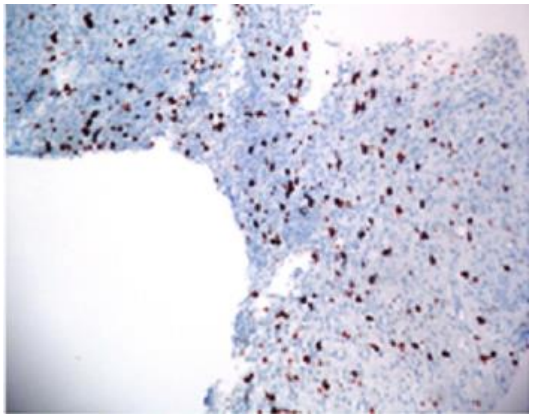

Fig-4c: MIB showing high index (25-40%)

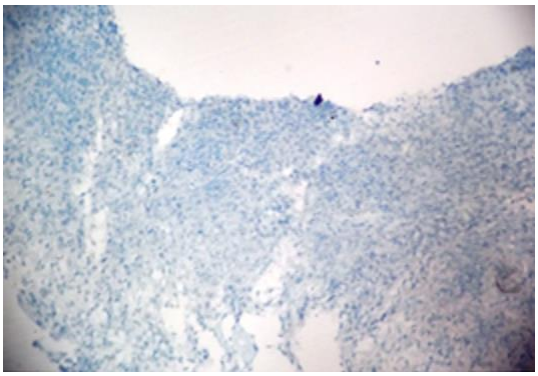

Fig-4d: IHC showing synaptophysin negative

DISCUSSION

PDLG is a rare condition in which there is diffuse infiltration of meninges by neoplastic glial cells without evidence of primary tumour in brain and spinal cord [4]. Primary leptomeningeal glioma can be in the form of a single solitary tumour or a diffuse tumour involving the intracranial or the spinal cord'sleptomeninges [5]. The usual presentation includes symptoms and signs of raised intracranial pressure such as headache, nausea, vomiting, seizures and pappilloedema. Based on a review of similar reported cases, It has been suggested that the presence of raised intracranial pressure with marked leptomeningeal enhancement on imaging should raise the suspicion of PDLG [5]. Debeno *et al*. have proposed the following three diagnostic criteria for PDLG [6] (a) No apparent attachment of extramedullary meningeal tumour to the neural parenchyma.(b)No evidence of primary neoplasia within the neuraxis.(c)The existence of distinct leptomeningeal encapsulation around the tumour.

A common finding in imaging studies is the focal or diffuse leptomeningeal enhancement with contrast .CSF cytology for malignant cells is usually negative as seen with this patient and for this reason it is advised to do GFAP staining in CSF cytology to detect cells of glial origin [7]. There is no definitive curative treatment for this ultimately fatal disease. Treatments are usually directed on symptom relief.

Ventriculoperitoneal shunting is used if there are features of raised intracranial tension.Only a few cases have been reported to have responded to treatment [8, 9] and the longest survival has been reported with the combination of radiotherapy and Temozolamide[9].

CONCLUSION

Primary Diffuse LeptomenigealGliomatosis is a rare entity and this case report reiterates the fact that prognosis of this neoplasm remains dismal and better treatment options have to be defined.

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