University Journal of Medicine and Medical Specialities

ISSN 2455- 2852 2019, Vol. 5(7)

Medulloblastoma -Case report

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Abstract : Medulloblastoma is an infratentorial, mid-line tumour. Incidence of Medulloblastoma is 1.5- 2 cases per 1,00,000 population(1). It presents as 2-3 months of chronic headache, vomiting, ataxia and visual disturbances. Here is a case of one and half year old child who was developmentally normal brought with altered sensorium and seizures with no history suggestive of increased intracranial tension. The blood reports were all normal. The presentation was so acute that acute metabolic encephalopathy and poisoning were the working diagnosis. CT brain sprang a surprise as medulloblastoma. This case is presented in view of the rarity of presentation.

Keyword :medulloblastoma ,seizures,radiotherapy

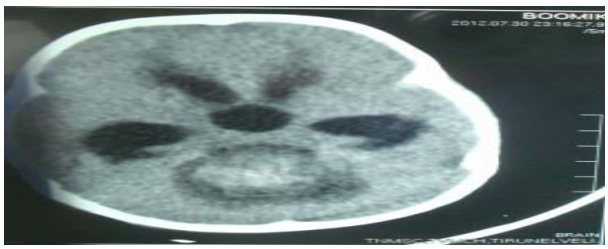
INTRODUCTION

Medulloblastoma is the 2nd most common tumour between 1-10 years of age. It has got male preponderance. It is a contrast enhancing tumour which infiltrates into the fourth ventricle and causes hydrocephalus2. It belongs to the group of embroyonal tumours.

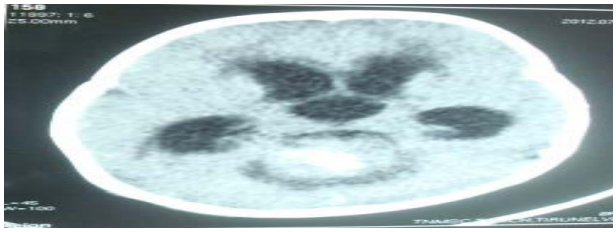
CASE REPORT

A one and half year female child was brought with complaints of sudden onset of irritable cry followed by tonic posturing of all four limbs lasting for more than one hour. The child was apparently normal till 10 pm, had taken dinner and slept in the cradle. At 11pm child woke up with a shrill cry with vomiting and tonic posturing. There was no history of fever ,refusal of feeds or altered sensorium prior to onset of the event, no history of cough , loose stools or bleeding manifestation.

There was no nystagmus ,head rolling , projectile vomiting, truncal ataxia,trauma or ear discharge. There was no history of suggestive of cranial nerve palsy or history of contact with pesticides or tuberculosis. There was no family history of seizures. The child was developmentally normal child with an uneventful antenatal, perinatal and neonatal history. On examination, the child was received in a state of altered sensorium with nystagmus, upward rolling of eye balls and pin point pupils not reacting to light. Fundus showed no evidence of papilledema. The child had excessive salivation ,tachycardia and palpable peripheral pulses. The gastric aspirate had no abnormal smell .CBG was 89 mg/dl. The child was given two doses of diazepam followed with phenytoin. The Child was on Jacson Rees circuit with 100% FIO2. After 45 minutes, the child developed irregular re spiration and was started on mechanical ventilation ,had recurrent episodes of seizures which were managed with injection phenobarbitone followed by midazolam infusion. At this point, the differential diagnosis was acute encephalophaty-?metabolic, status epileticus, poisoning or space occupying lesion .Base line lab investigation did not yield any clue. Complete blood count,renal parameters ,serum electrolytes, peripheral smear and blood cultures were non-contributory.organophosphorus poisononig was considered in view of excessive salivation and pin point pupils. Serum cholinesterase was sent,which was within normal limits. CT brain revealed a space occupying lesion in the midline infiltrating the fourth ventricle with dilatation of all other ventricles with hyper intense signal in the mass suggestive of bleeding. The possibilities of cerebellar astrocytoma and medulloblastoma were entertained . The location of tumour was more in favour of medulloblastoma. Astrocytoma is infratentorial but it is not a mid line tumour. The child was started on 3% Nacl and injection dexamethasone. A guarded lumbar puncture was done and a clear CSF was obtained which was not under tension. CSF cytology revealed malignant cells. The child was planned for surgery and followed by radiotherapy. The child became vitally unstable despite mechanical ventilation and succumbed.



CT BRAIN SHOWING THE MASS IN THE FOURTH VENTRICLE WITH GROSS HYDROCEPHALUS



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DISCUSSION

Medulloblastoma is the 2nd most common brain tumour between 1-10 years of age. It has Male preponderance2. It is a mid line, Infratentorial tumour. It belongs to the group of embroyonal tumours (primitive neuroectoderm). Primitive neuroectoderm consist of medulloblastoma, supratentorial PNET, ependymoblastoma, medulloepithelioblastoma, atypical teratoid. Histologically medulloblastoma composed of mono-morphic sheet of undifferentiated small ,blue ,rounded cells3 with the characteristic homer wright rosettes with neuronal differentiation. The tumour is immunoreactive for synaptophysin. Chang staging4 is a surgical staging based on the size of tumour, extension in to the aqueduct of sylvius, foramen lushka, brain stem and foramen of magnum and presence or absence of metastasis.Risk stratification; average risk includes age greater than 3 years, residual tumour after resection is less than 1.5 cm2 and stage m0. High risk comprises of age less than 3 years, residual tumour size greater than 1.5 cm2 and leptomeningeal seeding. Worst prognosis is seen in age <3 yrs, disseminated disease, CSF analysis positive for malignant cells , amplification of mycc gene and tyrosine receptor 2 positivity. This child belonged to high risk group in view of age below 3 years , large tumour size and leptomeningeal seeding

MANAGEMENT

Protocols are Surgery- Total or near total resection5 followed by irradiation with or without chemotherapy.The feasibility of complete resection is associated with histology of tumour as patients with desmoplastic medulloblastoma has more chance of complete resection than patients with classical medulloblastoma11. The best survival results for children with medulloblastoma have been obtained when radiation therapy is initiated within 4 to 6 weeks postsurgery.6,7,8.The radiation dose for average risk patients is 54 to 55 Gy to the tumour bed and 23.4Gy to the entire neuraxis while in case of high risk patients the spinal radiation is given at 36 Gy units. Preradiation chemotherapy has been related to a poorer rate of survival9,10 Combination of cis platin, lomustine and vincristine or cisplastin,cyclophosamide and vincristine are used. Post radiation chemotherapy has become standard protocol. Stem cell rescue can be tried but has limited success. However in children less than 3 years radiotherapy has its own long term sequalae.

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