RESEARCH
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INCIDENTAL FINDING OF EPENDYMOMA IN A CHILD PRESENTING WITH CERVICAL STIFFNESS

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Abstract

Introduction: Tumors derived from ependymal lining of ventricular system are known as Ependymoma. Incidence is 0.4 per 1,00,000 population. Most common amongst childhood brain tumors. Mean age of presentation is 6 years. Clinical presentation depends upon anatomical location. Generally, non invasive, but may extend into ventricular lumen and lead to obstructive hydrocephalus. Surgery is the primary treatment modality. **Case report**: A 6 years female presented with headache , neck pain and abnormal body movements. On examination DTR were exaggerated and signs of meningeal irritation were present.Firstly thought to be meningitis, child was managed conservatively with IV antibiotics and Mannitol. Routine investigations were within normal limits. MRI Brain with contrast study was done which was s/o lesion measuring 1.5 x 1.4 cm in 4th ventricle and 1.4 x 1.1 cm in lateral aspect of medulla oblongata (neoplastic etiology- Ependymoma). **Conclusion**: Ependymoma, although a rare entity, differential of brain tumors should be considered in patients presenting with features of raised ICT and an attempt should be made to ascertain exact etiology. It would help in effective and timely management of these cases.

INTRODUCTION

Tumors derived from ependymal lining of ventricular system are known as Ependymoma. They may also arise from central canal of spinal cord, or cortical rests. Incidence is 0.4 per 1,00,000 population. (1) They account for 1.6% of all CNS tumors. Males are more commonly affected (1.3:1).

Location of tumor also depends upon age, most of them are intracranial in children. Most common of childhood brain tumors, 70% found in posterior fossa. Leptomeningeal spread is seen in 10% cases. (2)

Compared with intracranial ependymomas, spinal ependymomas are less frequent and exhibit better prognosis. Spinal Ependymoma usually present in adults.

Ependymoma is classified into 3 grades (3) –

1. Subependymoma (Grade 1)
2. Ependymoma (Grade 2)
3. Anaplastic (Grade 3)

Grade 1 – Usually asymptomatic and is discovered incidentally on neuro-imaging, has good prognosis. Genetic association is also present.

Grade 2 and 3 are found more commonly in adults and are aggressive tumors. They have more tendency to disseminate.

Mean age of presentation is 6 years. Clinical presentation depends upon anatomical location. One of the most common presenting features is pain for both intracranial and spinal ependymomas. Other symptoms include – change in personality, mood. Sometimes child may directly present with seizures. Tumors can't be diagnosed via history itself; detailed physical evaluation and imaging are required. (4) Supratentorial Ependymoma presents with features of raised intracranial tension (headache, nausea and vomiting). Posterior fossa ependymoma: Tumor may extend into 4th ventricular lumen and lead to obstructive hydrocephalus. Cranial nerve 6-10 may also be involved. Spinal ependymoma has a history of ascending / descending nerve tract involvement with progressive neurological deficit. Bowel bladder may be involved in myxopapillary ependymoma along with saddle anesthesia and lower limb dysfunction. Symptoms vary according to anatomical location of tumor. On physical examination – hydrocephalus, papilledema, ataxia, nystagmus is present in patients with posterior fossa tumors. Upper / lower limb weakness, radicular pain, loss of sensation are typical for spinal tumor. Complications include hemorrhage, infection, worsening of neurological deficit. (4)

Neuroimaging (MRI with contrast) is the investigation of choice. Subependymomas show calcifications, so may be detected on CT scan as well. On MRI, heterogeneous appearance with circumscribed mass lesions is seen. On contrast study, enhancement is seen. Other tumors like astrocytomas, medulloblastoma can be differentiated on DW-MRI. Hemosiderin cap is suggestive of ependymoma, along with associated syringomyelia. Management includes surgery and radiotherapy. Total resection is the mainstay of treatment. (2)

Case Report:

A 6 years female presented with headache x 25 days, neck and back pain x 15 days and 1 episode of abnormal body movements. The headache was increasing in intensity that the child was bed ridden x 10 days, complained of neck and back pain even on slightest movement. On examination DTR were exaggerated in all 4 limbs and signs of meningeal irritation (headache, nausea and vomiting) were present. Kerning Sign and Brudzinski sign could also be elicited. Initially thought to be some infectious / ICSOL etiology, child was started on IV antibiotics and Mannitol. Differential of Pott Spine with Tuberculoma was also kept. Contrary to etiologies patient had no history of fever / photophobia /bowel bladder incontinence / TB contact or history of trauma could not be elicited. Routine investigations were within normal limits. MRI Brain with contrast study was done which was s/o heterogenous enhancing mass lesion with altered signal intensity with central hypointense areas in 4th ventricle. The lesion was causing mass effect on 4th ventricle leading to proximal dilation of 3rd ventricle. The lesion showed lateral extension up to lateral aspect of medulla oblongata. Lesion measuring 1.5 x 1.4 cm in 4th ventricle and 1.4 x 1.1 cm in lateral aspect of medulla oblongata (neoplastic etiology- Ependymoma).

DISCUSSION

Clinically when a child presents with raised ICT features, usually the differential of meningitis and ICSOL is kept. Astrocytomas, medulloblastomas and choroid plexus tumors are other differentials which need to be considered for tumors in posterior

fossa. This can be challenging for the attending physician because of similar clinical features. (5) Around 90% of juvenile ependymomas occur inside the skull, with 70% found below the tentorium (infratentorial) and 30% above it (supratentorial). Supratentorial ependymomas, which range from grade II to grade III tumors, typically develop within the brain tissue, possibly due to ependymal stem cell entrapment during embryonic development.

These tumors often have clear boundaries with neighboring structures, making them well-defined. The prognosis depends on factors like the tumor's grade, type, location, spread, genetic characteristics, patient's age, and the amount of tumor left after surgery (if surgery is an option). (6) The five-year survival rate for ependymoma is around 88.2 percent, yet several variables can influence the outlook. These factors encompass the tumor's grade, molecular subtype, the individual's age and overall health at diagnosis, as well as their response to treatment. (3) The initial approach to treating an ependymoma involves surgery, if feasible. The objective of surgery is twofold: to gather tissue for identifying the tumor type and to excise as much of the tumor as feasible while minimizing additional symptoms. Following surgery, many individuals may not require further treatment. However, for those who do, additional therapies could encompass radiation, chemotherapy, or participation in clinical trials. Clinical trials assess the efficacy of new chemotherapy, targeted therapy, or immunotherapy medications. The selection of treatments is determined by taking into account factors such as the patient's age, residual tumor post-surgery, tumor type, and its location. (7)

CONCLUSION

Ependymoma, although a rare entity, differential of brain tumors should be considered in patients presenting with features of raised ICT and an attempt should be made to ascertain exact etiology. Though it is challenging for the treating physician, when diagnosed well in time it would help in effective and timely management of these cases. This case highlights the importance of timely and precise diagnosis followed by surgery to achieve a favorable outcome and establish a suitable plan for subsequent treatment and follow-up care.

Images:

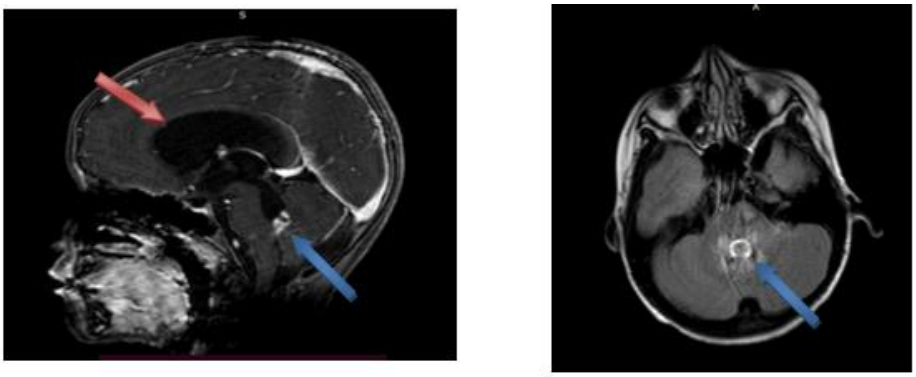

Fig 1 and 2: show location of the tumor – 4th ventricle (blue arrows). Fig 1 also shows dilatation of 3rd ventricle (red arrow).

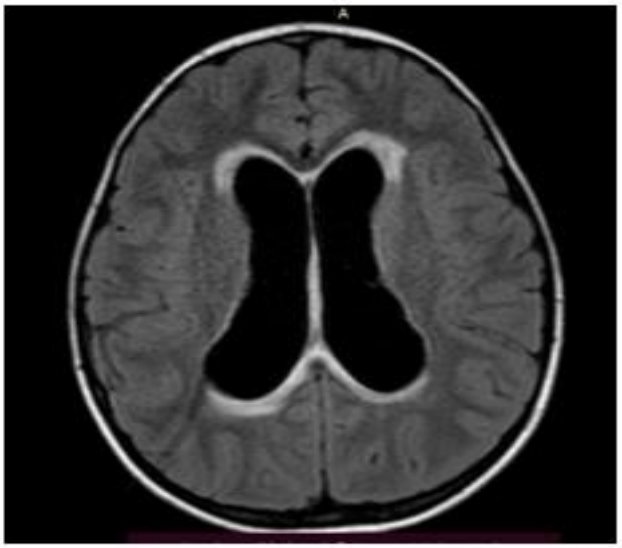

Fig 3: shows presence of ventriculomegaly and peri-ventricular ooze.

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