International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2024: 7.101

A Rare Case Report of Atypical Meningioma in a 7 Year Old Male Child

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Abstract: ***Background: Meningioma are tumors arising from the meninges. They occur intracranially or within spinal canal. Paediatric meninagiomas are relatively rare and comprise of Atypical and Anaplastic variety. They also show an association with neurofibromatosis type 2 and previous radiation exposure. Clinical Description: We present a case of Atypical Meningioma which was detected in 7 year old male child who presented with Bilateral Progressive painless loss of vision and Headache. Patient also had lost bowel and bladder control, and behavioural abnormalities. On MRI brain child was diagnosed with Atypical Meningioma. Management: Meninagioma management includes Medical and Surgical modalities. Medically stabilization of patient and Reduce the intracranial agents, Chemotherapy and Radiotherapy play roles. While neurosurgery and resection of Tumor remain the most crucial step. Our patient was treated conservatively and referred to a higher Centre for specifc neurosurgery. Conclusion: Meningioma can be diagnosed if history, physical exam and brain imaging is highly suspicious. Histopathological examination remains the gold standard after the tumor resection to diagnose Meaningiomas which reveal whorling pattern.***

Keywords: Atypical meningioma, pediatric brain tumor, neurofibromatosis type 2, vision loss in children, intracranial tumor management

1. Introduction

Meningioma refers to a set of tumors that arise contiguously to the meninges. Meningiomas may occur intracranially or within the spinal canal. They are thought to arise from arachnoidal cap cells, which reside in the arachnoid layer covering the surface of the brain.

[1]https: //emedicine. medscape. com/article/1156552 - overview?src=mbl_msp_android&ref=share
They comprise approximately 35% of all primary intracranial neoplasms in adults. In contrast, they are relatively rare in children, accounting for just 1%–2% of primary brain tumors in persons <21 years of age (Pubmed/ncbi)

[2]https: //www.ncbi. nlm. nih. gov/pmc/articles/PMC8018050/

2. Case History

Presenting Concerns
Patient was brought with complaint of bilateral progressive loss of vision since one month. It was gradual in onset with loss of distant vision initially which progressed to loss of near vision as well. The patient also had complaint of Headache and decreased appetite since 20 days.

History and Clinical Findings
- The patient had history of high grade **Fever** for 4 days 1.5 month ago which was relieved by over the counter medication.
- Then the patient developed **Bilateral loss of vision**. Vision was initially lost for distant vision later progressed to near vision as well.
- The patient also **Lost bowel and bladder control** since last 15 days.
- **Behavioural abnormalities** like Anger, Loss of social inhibition.

The patient went to Local Eye hospital where routine investigations and fundus examination was done. ESR was 80mm after 1 hour (elevated) and was found to have Bilateral Papilledema on Fundus examination. MRI Brain was advised and the patient was referred to our hospital.

On admission the patient denied any history of Trauma, Seizures, Projectile vomiting. There was no history of Blood transfusion, Past Hospitalization, TB contact.

There was no ingestion of Drugs or toxins, no history of similar complaints and CNS infections in past. Patient denied any refractive error of vision in past.

He was born Full term vaginally with birthweight 2.5kg, Cried immediately after birth, APGAR was 8 and 9 with No history of NICU admission or neonatal seizures.

There was no history of Previous Neurological deficit, developmental delay or major medical illness. Family history was negative for seizure disorder, neurofibromatosis or any other tumor.

In the emergency department patient was found to be afebrile with following vitals. PR: 154bpm RR: 30/min BP: 90/64mmHg RBS: 152mg/dl. The child was drowsy but arousable. GCS 15/15 (V5M6E4). Pupils were Round regular medically dilated.
- On CNS Examination the patient had normal cranial nerve examination except for the 2nd Cranial nerve as it was non testable.
- He moved all 4 extremities well with 5/5 Tone and 5/5 Power with symmetrical DeepTendonReflexes in both upper limbs and lower limbs.
- Plantar responses were Flexion bilaterally without ankle clonus.
- There was no signs of Meningeal irritation or Abnormality in Cerebellar Signs.
- Chest, Heart and Abdomen was within normal limits with Tanner stage 1 Pre Adolescent.

- Skin and mucus membrane did not reveal any unusual pigmentation or Rash.
- Ophthalmology consult was taken which revealed **Bilateral Papilledema with Macular Star**. No history of Chronic illnesses in Family and siblings.

Diagnostic Focus, Assessment and Course of Illness
As there was Bilateral Papilledema, Treatment of Raised ICP was started.

Inj Mannitol, Inj. Dexamethasone and Acetazolamide was given.
Routine Examination of blood was done.
Hb=10.4 gm/dl, WBC=9700/mm3, Platelets=3.56Lakhs

As there was Raised ICP, Lumbar Puncture was contraindicated for CSF Examination. Patient's MRI Brain was planned.

MRI Brain Findings included

- Fairly large Multilobulated heterogeneously enhancing **Extra axial** lesion measuring 8.9 x 10 x 6.8 cm in **the Anterior cranial fossa** with probably **broad base of dura** of Anterior falx or olfactory groove region.
- Distortion and dilatation of both lateral ventricles.
- Prominent perioptic CSF with **Empty sella** represents **Raised intracranial tension**.

Overall imaging features favour Possibility of an aggressive neoplastic pathology, like **Atypical meningioma**. Possibility of primary intracranial Esthesio - neuroblastoma seems less likely. Advice Histopathological correlation.

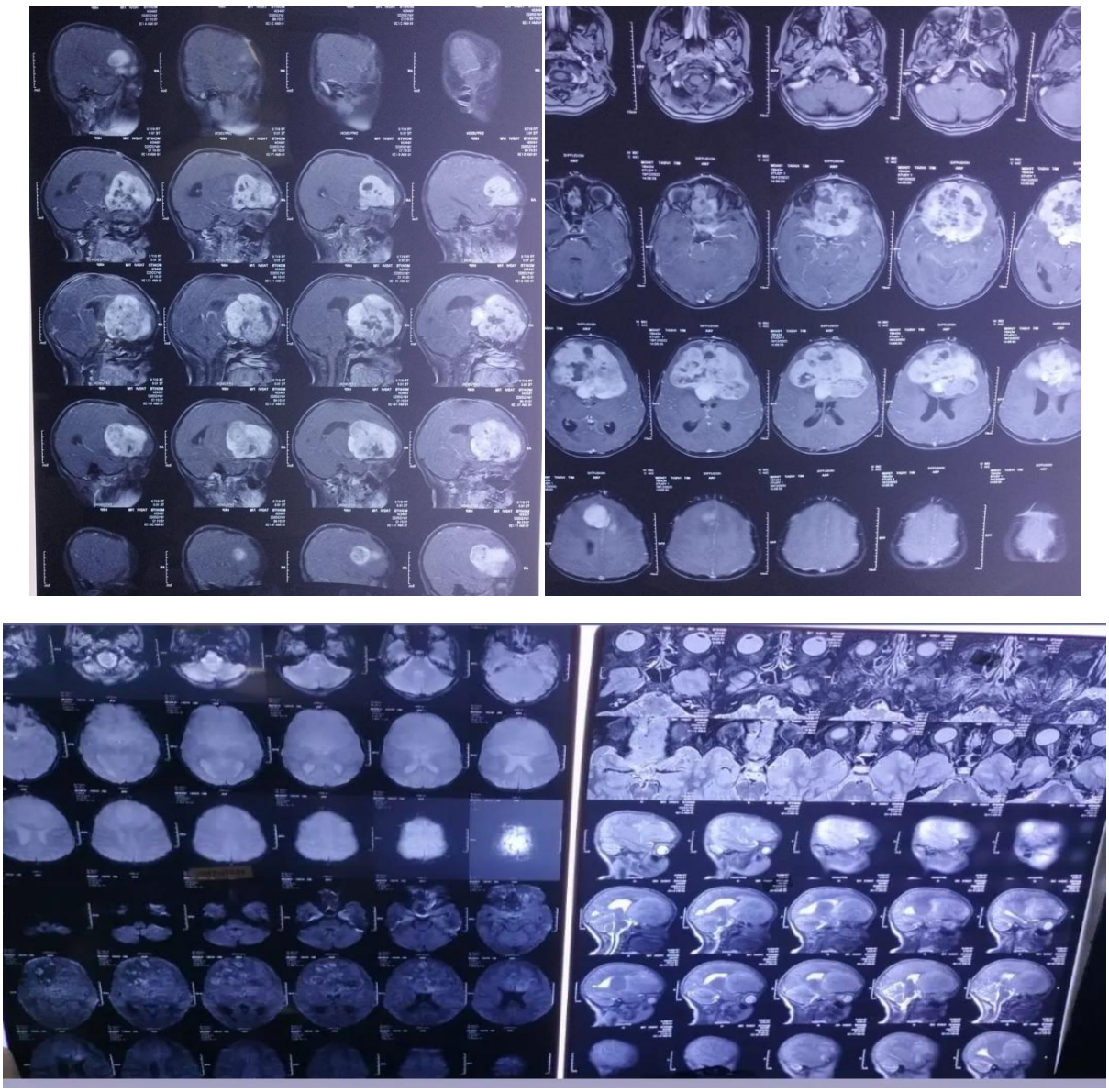

Image 1: MRI Brain showing aggressive nature of Meningioma

3. Outcome

After this patient had been stabilized and diagnosed, the patient was referred to higher centre for further neurosurgical management as our hospital did not had facility for paediatric neurosurgery.

Image 3: Patient After Stabilization and Before Being Referred to HigherCenter. Picture Posted with Consent of Parents

4. Discussion

Introduction

Meningioma, the term coined by Harvey Cushing, refers to a set of tumors that arise contiguously to the meninges. Meningiomas arise from arachnoid cap cells, the specialised cells in arachnoid granulations.

Similarly, intraventricular meningiomas arise from arachnoid cells present in the choroid plexus. Third ventricle tumours arise from the tela of the velum interpositum, which is the space between the two layers of tela in the roof of the third ventricle that contains the posterior medial choroidal arteries and internal cerebral veins. Fourth ventricle meningiomas arise from choroids or interior telachoroidea.

Paediatric meningiomas have unique features compared to their adult counterparts.

- For instance, they are associated with an increased frequency of higher-grade (WHO Grades II and III) tumors, aggressive histological types (eg, papillary, clear cell, etc.), higher mitotic and proliferation indices, more frequent brain invasion, higher recurrence rates and a predilection for atypical sites (particularly the spinal cord)
- They also lack the female sex bias seen in adult tumors, are more often associated with inherited genetic syndromes, mutational profiles.

[https: //www.ncbi. nlm. nih. gov/pmc/articles/PMC8018050/]

Differential diagnosis of progressive painless loss of vision in both eyes in children are vast. It can be attributed to structural or medical causes including toxic, infectious and metabolic.

Epidemiology

The annual incidence of symptomatic meningiomas is approximately 2 cases per 100, 000 individuals in US.

- They are relatively rare in children, accounting for just 1%–2% of primary brain tumors in persons <21 years of age.
- Females are affected nearly 1.5 times more often than males. As they Medscape3 receptors.
- Meningiomas are more prevalent in Africa than in North America or Europe. (Medscape)

https: //emedicine. Medscape. Com/article/1156552 – overview?src=mbl_msp_android&ref=share

Presentation

Meningiomas produce their symptoms by several mechanisms. They may cause symptoms by irritating the underlying cortex, compressing the brain or the cranial nerves.

Localized nonspecific headaches, Monoparesis, Seizures, Apathy, Disinhibited behaviour, Sphincter disturbances, Anosmia with possible ipsilateral optic atrophy and contralateral papilledema (this triad termed Kennedy - Foster syndrome.)

Location	*Symptoms*
Parasagittal	Monoparesis of the contralateral leg
Subfrontal	Change in mentation, apathy or disinhibited behavior, urinary incontinence
Olfactory groove	Anosmia with possible ipsilateral optic atrophy and contralateral papilledema (this triad termed Kennedy - Foster syndrome)
Cavernous sinus	Multiple cranial nerve deficits (II, III, IV, V, VI), leading to decreased vision and diplopia with associated facial numbness
Occipital lobe	Contralateral hemianopsia
Cerebellopontine angle	Decreased hearing with possible facial weakness and facial numbness
Spinal cord	Localized spinal pain, Brown - Sequard (hemispinal cord) syndrome
Optic nerve	Exophthalmos, monocular loss of vision or blindness, ipsilateral dilated pupil that does not react to direct light stimulation but might contract on consensual light stimulation; often, monocular optic nerve swelling with optociliary shunt vessels
Sphenoid wing	Seizures; multiple cranial nerve palsies if the superior orbital fissure involved
Tentorial	May protrude within supratentorial and infratentorial compartments, producing symptoms by compressing specific structures within these 2 compartments
Foramen magnum	Paraparesis, sphincteric troubles, tongue atrophy associated with fasciculation

Vascular: This presentation, although rare, should be considered. Meningiomas of the skull base may narrow and even occlude important cerebral arteries, possibly presenting either as **transient ischemic attack (TIA) -** like episodes or as stroke.

Intraventricular meningiomas may present with **Obstructive hydrocephalus**. Meningiomas in the vicinity of the sellaturcica may produce **Panhypopituitarism**. Meningiomas that compress the visual pathways produce various visual field defects, depending on their location. Rarely, chordoidmeningiomas can present with hematologic disturbances, namely Castleman syndrome.

Pathogenesis

Meningiomas may occur intracranially or within the spinal canal. They are thought to arise from arachnoidal cap cells, which reside in the arachnoid layer covering the surface of the brain.

Genetic association with Neurofibromatosis - Type2, Ghorlin syndrome, Von Hippel Lindau, Cowden, MEN Type 1 is present.

Loss-of-function mutations in NF2 and chromosome 22 losses were common, but pathogenic variants in other genes (SMARCB1, FUBP1, BRAF V600, TERT promoter, CHEK2, SMAD and GATA3) were identified in a minority of cases. [https: //www.ncbi. nlm. nih. gov/pmc/articles/PMC8018050/]

Diagnosis

- Plain skull radiograph may reveal hyperostosis and increased vascular markings of the skull, as well as intracranial calcifications.
- On plain head CT scans, meningiomas are usually dural - based tumors that are isoattenuating to slightly hyperattenuating.
- On T1 - and T2 - weighted MRIs, the tumors have variable signal intensity. If a meningioma is suspected, obtaining an enhanced MRI is imperative. Meningiomas enhance intensely and homogeneously after injection of gadolinium gadopentetate. The edema may be more apparent on MRI than on CT scanning. An enhancing tail involving the dura may be apparent on MRI.

https: //emedicine. medscape. com/article/1156552 - overview?src=mbl_msp_android&ref=share

MRI brain is very useful for diagnosing. With sensitivity of 90% and specificity of 80%.

https: //www.nature. com/articles/s41598 – 022 – 13881 – z

Management

- Medically decrease the ICP by Mannitol, Acetazolamide
- The use of corticosteroids preoperatively and postoperatively has significantly decreased the mortality and morbidity rates associated with surgical resection.
- Antiepileptic drugs should be started preoperatively in supratentorial surgery and continued postoperatively for no less than 3 months.
- Chemotherapy. This modality of treatment is reserved for malignant cases after failure of surgery and radiotherapy to control the disease.
- The main drugs studied include temozolomide, which had no effect against recurrent meningiomas in a phase 2 study, and hydroxyurea (ribonucleotidereductase inhibitor)
- The combination of interferon alpha and 5 - fluorouracil synergistically reduces meningioma cell proliferation in culture and warrants further investigation.
- Molecules to block specific growth factors or enzymes are being developed. Atypical meningioma (WHO grade II) and anaplasticmeningioma (WHO grade III) showed increased fatty acid synthase (FAS) expression. FAS inhibitor (cerulein) decreased meningioma cell survival in vitro. Thus, increased FAS expression in human meningiomas represents a novel therapeutic target for the treatment of unresectable or malignant meningiomas.

Radiotherapy is mainly used as adjuvant therapy for incompletely resected, high - grade and/or recurrent tumors

In general, the ideal treatment of a benign meningioma is surgical resection if possible. Hasegawa et al treated 46 patients with gamma knife radiation (GKR) as the initial treatment modality. GKR may be selected over surgery in patients with significant medical comorbidities.
[https: //emedicine. medscape. com/article/1156552 - overview?src=mbl_msp_android&ref=share]

Prognosis

The relative 5 - year survival rate for atypical and anaplastic meningioma is 63.8% but know that many factors can affect prognosis. This includes the tumor grade and type, traits of the cancer, age and health when diagnosed, and how they respond to treatment. (NIC. gov)

[https: //www.cancer. gov/rare - brain - spine - tumor/tumors/meningioma#: ~: text=The%20relative%205%2Dyear%20survival, how%20they%20respond%20to%20treatment.]

Meningiomas can spread to other areas of the CNS through cerebrospinal fluid (CSF). Grade II meningiomas can invade surrounding tissue, including nearby bone tissue. Grade III meningiomas have irregular cells and are likely to invade the brain or spread to other organs in the body.

- Meningioma grading (I to III) is based on the appearance of the tumor cells under a microscope.
- Grade I (Benign) is the most common type of meningioma.
- Grade II (atypical): Approximately 15 percent to - 20 percent of meningiomas are atypical, which means that the tumor cells do not appear typical or normal.
- Atypical meningiomas are neither malignant nor benign, but may become malignant at some point. Grade II meningiomas also tend to recur and grow faster. Grade III (Anaplastic) is the most aggressive form and is considered malignant.

Good Progonstic factors
Age <60 yrs,
patients presents with seizures,
Frontal Lobe tumors, Low grade tumors,
Less mitotic index,
Presence of MGMT gene promoter hypermethylation,
Karnofsky performance score>70 (Patient is able to take care for himself)

https: //emedicine. medscape. com/article/1156552 - overview?src=mbl_msp_android&ref=share

5. Conclusion

Meningiomas are most common begin tumours which are extra axial in nature. Although they're rare in Paediatric age group most such tumours are aggressive and anaplastic variety. High level of suspicion, thorough clinical examination and genetic/familial history makes diagnosis easy especially when there are signs of raised Intra cranial tension. Early diagnosis and Treatment via complete surgical excision with good margins improves quality of life of patient immensely.

Teaching points

Signs of raised intracranial tension includes projectile vomiting, Seizures, Headache, Papilledema, Poor GC score/Altered sensorium. Hence high index of Suspicion in cases of Raised ICT makes way for diagnosis of Intracranial space occupying lesions.