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A CASE OF CEREBELLOPONTINE ANGLE TUMOUR PRESENTING AS SEIZURE DISORDER

General Medicine

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ABSTRACT

Cerebellopontine angle (CP Angle) tumour are most commonly in the form of vestibular schwannoma, meningioma and epidermoid cyst. The CP Angle is a triangular space in the posterior cranial fossathat is bounded by the tentorium superiorly, brainstem posteriomedially, and the petrous part of the temporal bone posterolaterally. It is occupied by the CP Angle cistern in which lies the cranial nerves V, VI, VII and VIII and also the anterior inferior cerebellar artery. The epidermoid cyst which was picked-up on the MRI, also known as cholesteatomas are the third most frequent tumour of the CP Angle. They arise from the normal epithelial cells included during the neural tube closure. Their growth is due to the accumulation of keratin and cholesterol produced by desquamation of squamous epithelium lining the mass.

KEYWORDS

INTRODUCTION:-

Cerebellopontine angle (CP Angle) tumour are most commonly in the form of vestibular schwannoma, meningioma and epidermoid cyst. The CP Angle is a triangular space in the posterior cranial fossathat is bounded by the tentorium superiorly, brainstem posteriomedially, and the petrous part of the temporal bone posterolaterally. It is occupied by the CP Angle cistern in which lies the cranial nerves V, VI, VII and VIII and also the anterior inferior cerebellar artery. The lesions in this regionpresent with non-specific symptoms like sensorineural hearing loss, tinnitus and giddiness. The CP Angle tumours are mostly benign and slow growing. The 2 major types are-

a) Sporadic- Unilateral and mostly present between 4^{th} – 6^{th} decade of life.
b) Associated with Neurofibromatosis type II- most commonly bilateral acoustic neuromas in younger patients.

Schwannomas are the primary lesion of the cranial nerves V, VII, IX, X and XI.

Meningiomas arise from proliferation of arachnoid meningothelial cells.

Epidermoid tumours arise from congenital misplacement of ectodermal cells during closure of neural tube.

Arachnoid cysts are CSF filled cysts that arise from splitting of embryonic arachnoid membranes[1].

Case Report:-

A 16 years old female presented to the emergency of KCGMC, Karnal with presenting complaints of Seizures for the past 20 days. The patient was alright 20 days back when she started having generalized tonic-clonic seizures. In the past 20 days she has had 4 to 5 episodes of seizures which were generalized tonic-clonic in nature.

There was no history of fall/ trauma
There was no history of fever
There was no history of diplopia
There was no history of any loss of sensation
There was no history of any preferential weakness
There was no history of incontinence/ retention of urine/ faeces
There was no history of any cranial nerve involvement
There was no history of any aura

In the past history, there was no history of similar complaints in the past. There was no previous history of any cardiac or valvular lesion. There was no previous history of Atrial Fibrillation. There was no previous history of any tumour. There was nothing significant in the treatment history. There was no previous history of any allergy to any drug.

There was nothing significant in the family history.

In the personal history, the patient was vegetarian, was non-alcoholic and non-smoker, sleep was sound and there was no disturbance of bowel and bladder.

On general physical examination, the patient was moderately built and nourished. She was afebrile, had a pulse of 80/ minute, regular, normal volume and character, no radiofemoral delay, equal on both the sides, all peripheral pulses were well felt. The blood pressure was 124/82 mm Hg in the right arm in the supine position. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy, edema. The JVP was not raised. On CNS examination, the patient was conscious, oriented and all other higher mental functions were normal. The Cranial Nerves examination, motor system examination, reflexes, sensory system examination did not reveal any abnormality.

Other systems examination were normal.

A provisional diagnosis of Seizure Disorder was kept.

The investigations done showed- Hb of 10.5 gm%, MCV 78.5 fl, Platelet count of 3.54 lakh/mm^3 , TLC of 15,100 cells/mm^3 , Viral Markers were negative, COVID-19 RT-PCR was negative. Blood Urea was 22 mg/dl, Serum Creatinine was 0.7 mg%, Sr Sodium was 141 mEq/lt, Sr Potassium was 4.0 mEq/lt, Sr Uric acid was 4.5 mg%. ECG was normal. Sr Bilirubin was total of 0.2 mg%, Sr AST was 39 IU/lt, Sr ALT was 63 IU/lt, Sr Alkaline phosphatase was 120 U/lt. Sr Calcium was 9.3 mg%. Random blood sugar was 116 mg%. The chest X-ray was normal.

MRI Brain done showed a lobulated lesion with MR signal similar to the CSF on all sequences with partial suppression on FLAIR sequences with restricted diffusion on DWI sequence in the right CP angle cistern region leading to compression and displacement of pons and medulla towards left side => possibility of right CP Angle epidermoid is likely.

NCCT Head revealed an illdefined hypodensity noted in the pons on the right lateral side.

So, the final diagnosis made was Seizure Disorder with right Cerebellopontine Angle tumour.

The patient was started on treatment. She was given antiepileptic drugs and other supportive treatment. She was seizure free with the treatment given. She was then referred to neurosurgeon for the treatment of the CP Angle tumour.

DISCUSSION:-

CP Angle tumour constitutes about 5-10% of all intracranial tumours.

Vestibular schwannomas constitute about 75-85% of all the CP Angle tumours.

Meningiomas constitute about 10-15% of all the CP Angle tumours.

Epidermoid constitutes about 7-8% of all the CP Angle tumours.

Other rarer forms include- Cranial nerve schwannomas, arachnoid cysts, lipoma, glomus jugulare, metastatic lesions.

The most common presenting symptoms of CP Angle tumours include

tinnitus, hearing loss, giddiness, vertigo, headaches and gait disturbances. Other symptoms pertaining to the brainstem compression symptoms, cranial nerve palsies and hydrocephalus may be seen with larger tumours compressing these structures. One of the rarer symptoms of presentation is seizures[2].

Diagnosis is made on history, physical examination, audiometric and radiological evaluation. The gold standard for evaluation is the MRI imaging of the brain. HRCT is useful for the assessment of bony involvement.

The treatment options for the CP Angle tumour include observation, radiation therapy or microsurgery.

The epidermoid cyst which was picked-up on the MRI, also known as cholesteatomas are the third most frequent tumour of the CP Angle. They arise from the normal epithelial cells included during the neural tube closure. Their growth is due to the accumulation of keratin and cholesterol produced by desquamation of squamous epithelium lining the mass. These slow growing tumours encase and surround the nerves and arteries in the cisterns rather than displacing them. Epidermoid cyst produces no reaction of the adjacent bone structures.

Acoustic neuromas are managed by either surgery, radiation therapy or observation with regular MRI scanning. Steriotactic radiosurgery is suitable for small and medium sized tumours. Surgical option includes three modalities of excision depending the site of the tumour and location of incision for access to the tumour- rectrosigmoid, translabyrinthine and middle fossa. The overall complication rate following surgery is 20% with CSF leak being the most common[3].

CONCLUSION:-

The present case signifies that seizures may be one of the rarer symptoms of presentation in a case of CP Angle where the tumour may appear to have been picked-up on imaging by chance.

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