Case Report

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FOURTH VENTRICLE EPIDERMOID TUMOUR: AN INSTITUTIONAL REVIEW

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ABSTRACT

Background: Intracranial epidermoids are uncommon tumours, with an incidence of around 0.5-2 % of all intracranial tumours. Age of presentation is 3rd - 4th decade. Most common sites of involvement are the basal cisterns with Fourth ventricle involvement being rarely reported.

Methods: Over a period of 9 years, a total of 13 cases of epidermoids located in the fourth ventricle were admitted in our Department of Neurosurgery. The history and clinical course was studied. Radiological diagnosis was established with CT and MRI Brain with contrast and was operated upon. Diagnosis was confirmed by histopathology. A retrospective study was carried out and results were reviewed.

Results : A total of 13 cases were studied that included 10 males and 3 females. Age of presentation varied from 16 years to 55 years. Most cases presented with cerebellar signs and symptoms, mostly ataxia and incoordination. Only 2 patients had symptoms of raised intracranial pressure, that too, mild in manifestation. Subtotal resection was done in 11 patients, while total removal was possible in rest 2.Characteristic pearly appearance was found intraoperatively. Except two patients, all other faired well postoperatively. One had recurrence of symptoms later on.

Conclusion: Fourth ventricle epidermoid tumours, although uncommon, have fairly good outcome, even if subtotal removal is done. If neglected, can be life threatening. Subtotal removal may result in recurrence, that can be detected by clinical assessment and MRI. Although total resection should be the goal, as growth is slow, even near total removal may suffice if tumour encases vital structures.

KEYWORD

epidermoid , cistern , ataxia

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INTRODUCTION

Epidermoid cysts commonly known as pearly tumours are benign congenital lesions resulting from ectodermal inclusions persisting during neural tube closure around later half of first embryonic month.[1],[2]They may be intracranial or spinal in location with intracranial epidermoids constituting less than 2% of intracranial masses[1],[3]The majority of such tumours are located in the cerebellopontine angle(around 60%) and the parasellar regions[3],[4]Epidermoids of the fourth ventricle are rare entities constituting around 5-18% of all intracranial epidermoid cysts in most of the studies[5],[6],[7]They have a typical slow growth ,and hence , even though being predominantly congenital , manifests in middle age groups when become sufficiently enlarged to cause neural compression. Clinical features suggest predominant cerebellar involvement but their location, extent and size may produce other symptoms. On neuroimaging, magnetic resonance (MR) depicts inhomogenous hypointense fourth ventricle lesions on T1 weighted images , nonenhancing on contrast and hyperintense on T2 weighted sequence. MRI has replaced CT as the imaging modality of choice , giving better structural details , thus helping optimal surgical planning.

MATERIALS AND METHODS

A total of 13 cases of epidermoid cyst of the fourth ventricle presented to us between March 2008 to March 2017 ,diagnosis being made preoperatively on the basis of CT and MRI Brain plain and contrast. Routine blood investigations were done . All the 13 patients were planned for surgical removal of mass after proper consent and counselling. With patients in prone position , suboccipital craniotomy was done. Dura was opened and pearly white tissue identified. Microscopic assisted surgery was undertaken with an aim to remove most of the lesion, but causing least damage to vital structures.In 6 cases , endoscopic removal of the tumour was done. Surgically removed tissue was subjected to histopathological study. Follow up was done at intervals with proper assessment of the clinical and neuroimaging status. Thorough examination of the patients was done at the time of admission, before operation, after surgery and on follow-ups. Analysis of the outcome was carried out and results were reviewed.

RESULTS

The study group consisted of 13 patients. Total number of patients with brain tumours that got admitted during the same period was 768.Posterior fossa tumours totalled 231 cases. Total number of intracranial epidermoid tumours was 22. Male patients dominated the study with 10 males and 3 females. Age of presentation ranged from 16 years to 55 years with the average being 31.6. Of the 13 cases, 11 were exclusively within the fourth ventricle, while in two cases

epidermoid spilled into the cerebellopontine angle cistern. The tumour reached the foramen magnum in 6 cases. Clinical features depended on location with ataxia being the most common one. Gait ataxia was present in all and appendicular ataxia i.e., incoordination seen in 7cases. Long standing headache was noted in 10 patients .Other important manifestations included vertigo, dysmetria, nystagmus, tremors, dysarthric speech, cranial nerve involvement(seventh cranial nerve in 2 cases) and deranged mentation (in 3 cases).4 cases complained of visual disturbances and on and off vomiting.

CT scan and MRI brain plain and contrast were performed in all patients.MRI revealed masses localised in fourth ventricle and filling and expanding it , and on occasions entering the outlets of the ventricle , the Foramen of Luschka and Magendie. The anteroposterior dimension ranged from 24mm to 49mm, transverse extent from 29 mm to 58mm and cranio caudal extent from 37 mm to 56 mm. CT revealed hypodense lesion compressing the brainstem with minimal hydrocephalous.MR revealed lobulated hypointense T1WI and hyperintense T2WI masses without any enhancement on contrast.

Suboccipital craniectomy was carried out and dura opened in 'Y' shape to reveal whitish fish scale coloured/ pearly white flakes ,that in some cases seeped to adjacent cisterns. Subtotal resection was carried out in 11 cases, surgery being curtailed due to underlying adhesion to brainstem while total removal was possible in 2 patients. Adherent nonseperable capsule was left as such and dura was closed. Postoperative period was uneventful in 8 patients. Two patients developed chemical meningitis on fourth and seventh day respectively that responded to steroids. One patient developed left sided facial palsy and two others had abducens impairment. Post operative MRI was done to asses completeness of removal. Patients were discharged and were followed up at 1 month , 3 months, 6 months and then at yearly intervals, which revealed symptomatic recurrence in one of the cases.

Pathological evaluation showed white capsule with pearly white keratin debris macroscopically while microscopy showed stratified squamous layers with anucleate squamae .

DISCUSSION

Epidermoid cysts or pearly tumours represent 0.2-1.8% of all intracranial tumours.[1],[2] They are considered congenital lesions that arise due to ectopic epithelial remnants that persist after the period of neural tube closure between 3rd and 5th embryonic week[1],[3]These tumours have a predilection for cisterns, most commonly cerebellopontine angle accounting for 60 % cases[2] Epidermoids of the fourth ventricle are second favoured site of occurrence with an incidence of 5-18%.[4],[5],[6]French pathologist Cruveilhier was the first to describe it.[7]

With an outer capsule, epidermoids have a stratified squamous epithelium with variable degree of keratinisation, and keratin debris with solid crystals of cholesterol, that fill up the centre of the cysts, that gives its characteristics pearly appearance.[8] They have a tendency to insinuate within available spaces ,and hence spread to adjacent cisterns and may encase vessels and nerves.

Being a very slow growing mass, clinical symptoms arise in adulthood although the tumour is considered to be congenital. As the squamous epithelium gets keratinised, keratin, cholesterol debris fill up the centre of the cyst and slowly enlarge the cyst and eventually it reaches sizes that start compressing nearby neurovascular entities. Fourth ventricle epidermoids present with gait ataxia, incoordination ,tremors, nystagmus,vertigo and seizures .[2],[6],[9] Long standing headache without other signs of hydrocephalus is commonly seen.

Neuroimaging reveals hypointense CSF like lesions on T1 weighted image with hyperintensity on T2 weighted sequence showing no uptake of contrast material. Although presence of cholesterol should result in hyperintense T1 signals, hypointensity is presumed to be due to heterogeneousity of the cyst and crystalline chemical form of cholesterol.[10],[11] Arachnoid cysts and dermoids constitute the major differentials . Arachnoid cyst follow CSF intensity patterns on MR Pulse sequence.[12]DWI shows restricted diffusion in epidermoids but not in arachnoid cysts and FLAIR shows signals hyperintense relative to CSF. Dermoids are excluded by their preferential midline location and presence of hair follicles, sebaceous and sweat glands. CT reveals nonenhancing extra axial hypodense lesion.

Total resection should be the goal as epidermoids can recur ,albeit slowly. Rapid recurrence with neurological deterioration points to rare instances of malignant transformation. Easily removable capsule with its contents should be removed avoiding any spillage that may predispose to chemical meningitis. But in cases where epidermoid seeps or insinuates into adjoining spaces, attempts at total removal may endanger nerves or vessels. In such cases, subtotal resection is justified. [4] Post surgery outcome is good even after subtotal removal but close monitoring and regular follow-up is a must.

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