

Lateral Ventricle Epidermoid Cyst Presenting with Right Hemiparesis and Blindness: A Case Report and Review of Literature

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SUMMARY

Epidermoid cyst of the CNS is rare, slow growing, benign neoplasm derived from the remnant of neural tube. It accounts for 0.2% of all intracranial neoplasms. Intraparenchymal and lateral ventricles are the rarest sites within the CNS. The lesion is asymptomatic at the early stage but later may present with mass effects and cranial neuropathies. We present our index case of a 23-year-old male who presented late with a rare lateral ventricle epidermoid cyst associated with calcification resulting in left hemiparesis and blindness. The excised tumour mass aggregate was 11cm and weighed 15g. This is an index case in our region. Lateral ventricle epidermoid cyst is a rare neoplasm that may mimic neurocysticercosis, choroid plexus papilloma and dermoid cyst at both clinical and imaging levels. The treatment of choice is surgical excision; however, there is recurrence rate of 33% with increased chances of chemical meningitis. Nevertheless, the tumour rarely transforms to squamous cell carcinoma.

Keywords: Epidermoid cyst, cerebral lateral ventricles, blindness, rare and index case.

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Introduction

Epidermoid cyst is one of the uncommon slow growing congenital tumours¹. It develops from ectodermal remnants in the neural tube when it separates from the ecto-

-derm between 3rd and 5th weeks of gestation during embryonic life.^{2,3} The neoplasm accounts for only 0.1 to 1.8% of all intracranial tumours² and most of them are asymptomatic but they may present with severe headache due to raised intracranial pressure and aseptic meningitis.⁴ Late manifestations include mass effects, cranial neuropathy, seizures and hydrocephalus.² About 60% of the cases occurs in paediatric age group, however, patients harboring the disease become symptomatic between the second and forth decades of life.¹ The commonest location of the tumour is cerebellopontine angle accounting for 40 to 50% of cases, this is followed by fourth ventricle (17%).³ Other locations include parasellar/sellar and cerebral hemispheres accounting for 10% and 1.5% respectively.³ Location in intra parenchymal and lateral ventricles are the most rare sites.² Sahoo et al reported calcifications in only 10-25% of cases.³ The

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tumours generally are classified based on their anatomical location in the brain which also assists in planning for surgical intervention. The rarity of lateral ventricle epidermoid cyst and this happened to be our index case in our region.

Case report:

A 23-year-old male patient was referred by neurology unit to neurosurgery unit of our hospital with complains of headaches of two months and blindness of one-month duration, however, his problems started one year ago with on and off symptoms before the current complain. The headaches were said to be recurrent, throbbing and globally progressive in intensity which was severe enough and prevented him from doing his daily activities. He occasionally had early morning vomiting that relieved the headaches. He was also noticed to be somnolent with associated irrational talks while awake but no history of seizures or enuresis. He was however, noticed to have developed weakness on the right side of his body but was able to walk with difficulty. The patient complained of progressive visual impairment to total blindness over a period of one month prior to presentation.

He had no prior history of trauma to the head or exposure to radiation or history suggestive of tuberculosis. The patient is not a known hypertensive or diabetic and has no known drug allergy. He was taken to different hospitals before he was referred to our hospital due to worsening of his condition. Examination revealed a young man conscious with stable vital signs, Glasgow coma scale score of 15/15. Confrontation test showed

patient was unable to see objects with no perception of light and the pupils were moderately dilated nonreactive to light bilaterally while, fundoscopy showed blurred optic disc with poorly defined margins in both eyes. Other systemic examinations were normal except the musculoskeletal system where he had right hemiparesis which was more in the upper limb with hyper reflexia in both knees. Clinical diagnosis of space occupying lesion with total blindness due to raised intracranial pressure was made.

His Computer tomography (CT scan) and Magnetic resonance imaging (MRI) of the Brain (figure 1) showed a huge mass originating from left ventricular medial wall of the lateral ventricle expanding into the ventricle on the same side, well encapsulated with some areas of calcifications, has cystic and solid components with features of hydrocephalus.

Patient was counseled, written consent obtained and had craniotomy with tumour excision.

Intra operatively the tumour was cheesy and was completely removed easily by just scooping and the sampled were taken for histopathological for final diagnosis (figure 2).

He did well post operatively and was later discharged home however, patient could not afford to do postoperative magnetic resonance imaging due to financial constraints. He had one follow up visit at our outpatient unit he was however, still blind but symptoms of raised intracranial pressure have completely subsided.



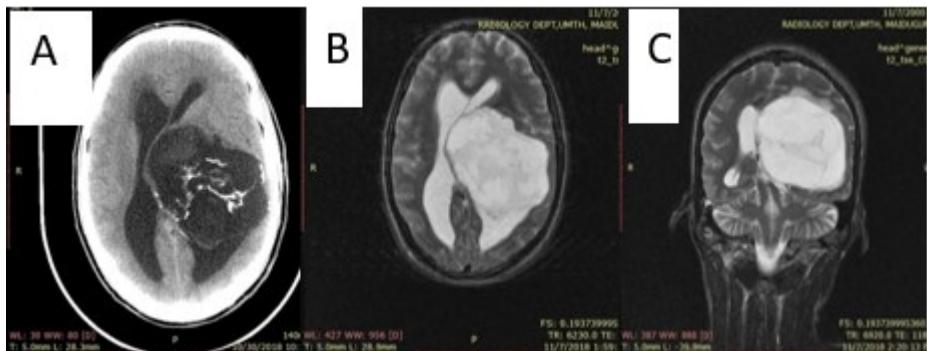


Figure 1: A-C CT and MRI of the Brain A, B Showed well encapsulated mass with cystic and solid component and focal areas of calcifications C, showed a huge mass originating from left ventricular wall of the lateral ventricle expanding into the ventricle.

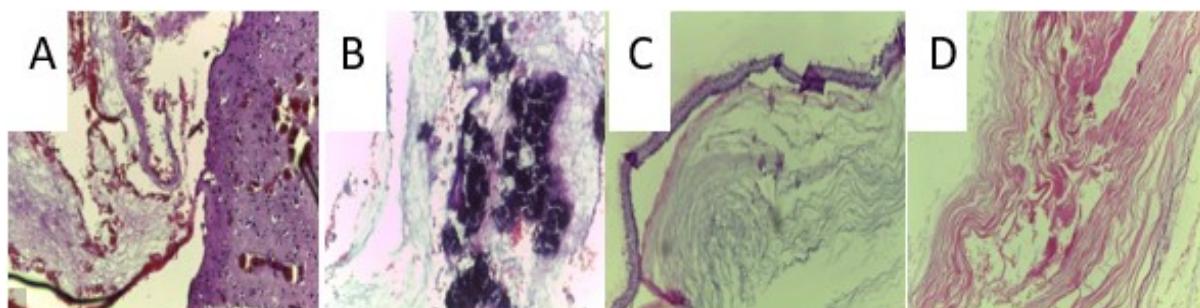


Figure 2: Photomicrograph; A-D show cyst lined by squamous epithelium containing desquamated laminated keratin A, C, D. Areas showing dystrophic calcification B and adjacent normal brain tissue A.

Discussion

Epidermoid tumour was first described in 1683 by Duverney,⁴ the first diagnosed case was reported by a French Pathologist Cruveilhier in 1829 and since then the disease was named as Cruveilhier tumour.¹ The tumour grossly appears bright white and for that reason Remark first suggested the epidermal origin of the lesion.⁴ Intraventricular epidermoid cyst was described to develop when neural tube closes and divides from cutaneous ectoderm where cutaneous cells are left behind within the neural tube.^{1,2} Intradural epidermoid cysts account for 90% of cases while extradural type constitute 10% of cases.³ An acquired

type due to head injury has been reported.² In our case the patient presented with mass effects with features of raised intracranial pressure in the third decade of life and there was no history of trauma, suggesting that it is more likely to be congenital in origin. Intracranial epidermoid tumours are rare and account for only 0.2 to 1.8% of all intracranial tumours.² In the English literature only 25 cases were reported from discovery of the tumour to 2017.¹ The tumours are exceptionally slow growing and usually present from the second to fourth decades of life with equal sex predilection.^{5,6} In the index case the patient is 23 years old male who



presented with one year history of right hemiparesis and blindness with one month history of persistent headache. The locations of the neoplasm form the bases for its classification into diploic region of the calveria type, ventricular, cerebello-pontine angle, optic chiasma and parapituitary region types. Others include pineal area and collicular types.⁴ However, the intrinsic brain stem epidermoid cysts are further classified into pure intrinsic/intra axial brain stem tumour, intra axial component with slight extension to the surrounding cisternal space, predominantly extra axial cisternal component with slight intra axial brain stem infiltration and multiple intracranial epidermoid cysts involving the brain stem.¹ The pathogenesis of the tumour is uncertain^{8,12}, however, the proliferation of pleuri-potent embryonic remnants along the otic and optic cerebral vesicles¹ is one of the hypotheses. Other hypotheses suggested that the tumour grows in cleavage plane between nerve fibers and extends along vessels into the subarachnoid space. The tumour then takes the least resistant path and displaces other brain structures.¹

Early clinical symptoms include severe headache, aseptic meningitis, vestibular symptoms, trigeminal neuralgia and cerebellar sign.^{2,4,5} In our case the patient presented with persistent headache and occasional early morning vomiting. Late complications had been right hemiparesis and blindness due to raised intracranial pressure.

Most epidermoid cysts are well defined hypo-attenuated masses that resemble CSF on CT-scan (figure 1) and they do not enhance.² It may be iso-intense or slightly hyper-intense to CSF on both T1 and T2 weighted MR-

Image.^{2,10,11} Intra-parenchymal tumours most of the time are difficult to diagnose pre-operatively because they resemble other lesions that are considered as mimics of epidermoid tumours.⁷ The mimics include; neurocysticercosis, dermoid cyst and choroid plexus papilloma.² These are all considered as differential diagnosis in the index case before definitive histological confirmation.

Morphologically, intracranial epidermoid cyst has similar features with the cutaneoustype.¹² The cyst cavity is filled with soft, waxy or flaky keratohyalin material that results from the progressive desquamation of the cyst wall.¹² The cyst contents are mucoid constituting cellular debris, keratin and cholesterol which can easily be aspirated or scooped out¹² as it was done surgically to the index patient, (figure 1). The cyst wall is lined by stratified squamous epithelium supported by an outer layer of collagenous stroma.² Cases having epithelial atrophy of the cyst lining, acanthosis, hyperplasia and cones formation have been described.² Although histological variation also exist between genders; females showed more hyperplasia of the epithelial lining while males showed more calcification.² Calcification generally, is seen in 10-25% of cases as also seen in our case (figure 2) and rarely the tumour undergoes malignant transformation due to chronic inflammatory stimulation as a result of repeated cyst rupture.^{1,2,12}

The treatment of choice for epidermoid cyst is surgical,^{1,2} however, radical excision of the cyst wall is difficult because of its firm adherence to neurovascular structures.^{1,9} Recurrence rate of 33% has been reported following partial or incomplete resection,² and it rarely transforms to squamous cell carcinoma.^{5,7,8}



Conclusion: Lateral ventricle epidermoid cyst is rare benign tumour with uncertain pathogenesis. The tumour has equal sex predilection and indolent clinical course and most patients present with late complications due to tumour mass effect which our index case presenting with blindness and remain so despite surgical excision of the tumour. The tumour is best diagnosed with MRI, even though it can be confused with some mimics, therefore definitive diagnosis requires histology. The treatment of choice for the tumour is surgical excision having excellent prognosis when adequate excision. Nevertheless, the tumour rarely transforms to squamous cell carcinoma and has high chances of recurrence.

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