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Sunil Munakomi, Binod Bhattarai, Iype Cherian

ABSTRACT

Introduction: Epidermoid tumors in the cervico-medullary junction are rare entities. Their presentation with features of acute hydrocephalus is a rare epiphenomenon.
Case Report: Herein we discuss a rare tumor in the posterior fossa presenting with features of hydrocephalus. We discuss the diagnostic modalities, differential diagnosis and management undertaken in the same.
Conclusion: Epidermoids can present with features of acute hydrocephalus. In them, surgical removal of the lesion is the therapeutic target. Specific problems pertaining to them is the insinuating nature of the lesion to surrounding neurovascular structures that may preclude its complete removal. Chemical meningitis, pseudo-meningocele and recurrence may complicate the postoperative period.

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Keywords: Epidermoid tumor, Loss of consciousness, Posterior fossa, Hydrocephalus, Weakness

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INTRODUCTION

Intracranial epidermoid tumors are rare congenital inclusion cysts constituting 0.2–1.8% of primary intracranial neoplasms [1, 2]. First described by a French artist [3], the first full description of this entity was given by the French pathologist Cruveilhier in 1829 [4]. They form between the third and fifth week of embryonic development as a result of displaced epithelial remnants that persists despite neural tube closure [1, 3, 5]. Herein we describe a similar tumor locating in the cervico-medullary region presenting with features of progressive hydrocephalus. Though benign, these tumors insinuate into the surrounding vital structures thereby making their complete excision difficult.

CASE REPORT

A 47-year-old female from Lamjung, Nepal presented to the emergency department of our hospital with a history of on and off headache since last six months and persistent projectile vomiting for last two days. There was no history of trauma, loss of consciousness, aura, weakness of body parts, bladder bowel incontinence or fever with chills and rigor. There was no significant past medical or surgical illnesses. Her Glasgow Coma Scale (GCS) was 15/15. Higher mental function was normal. All cranial nerves were intact. There were no sensory or motor deficits. Cerebellar signs were absent. Fundoscopy revealed bilateral papilledema. Computed tomography (CT) scan of head revealed hypodense and

isodense lesion with anterior nodule pressing upon the fourth ventricle and extending inferiorly up to cervico-medullary junction (Figure 1). There was no contrast enhancement within the lesion. There was also evolving hydrocephalus. Fluid attenuated inverse recovery (FLAIR) and diffusion weighted image (DWI) sequence was performed to differentiate it from the arachnoid cysts. The diagnosis of posterior fossa epidermoid tumor with evolving hydrocephalus was made. Detailed counseling was done and the consent for the surgery was taken. She underwent midline sub-occipital craniectomy with gross excision of tumor. Intraoperatively tumor consisting of large bed of pearls with pseudocapsule adhering to midbrain and surrounding cisterns was seen (Figure 2). The tumor was insinuating basal nerves and extending caudally to C1 arch. Gross removal of the tumor was done (Figure 3). The tumor bed was thoroughly irrigated with normal saline so as to prevent chemical meningitis and recurrence. The dura was repaired and the wound was closed in layers. Postoperatively, she had uneventful recovery. Repeat CT scan showed no contrast enhancement with minimal extra-axial collection and resolving hydrocephalus. The patient was monitored for features of progressive hydrocephalus till her stay at the hospital. The patient was discharged home on the 10th postoperative day on prophylactic antiepileptic (sodium valproate 300 mg per oral three times daily). Patient followed up in the outpatient clinic one month later with a complain of swelling over the surgical site. Examination revealed soft fluctuant swelling suggestive of pseudo-meningocele. Therapeutic tap was done and the patient was started on tab acetazolamide 250 mg per oral three times daily and tapered over three weeks. There was no further recurrence. Patient was advised for six monthly follow-up.

DISCUSSION

These tumors typically occur in the cerebellopontine angle cisterns [1]. Although, few cases are reported in the fourth ventricle, this location is the second most common for an epidermoid tumor in the posterior fossa [6].

Epidermoids histologically consists of an outer capsule surrounding a layer of keratinized stratified squamous epithelium and inner cystic fluid that usually includes debris, keratin, water and cholesterol. As the epithelial layer desquamates, the cells accumulate and form a cholesterol-rich layer that gives the tumor its characteristic bed of pearls appearance. On contrary to the dermoid tumors, these tumors do not contain any dermal appendages. Though benign, these tumors tend

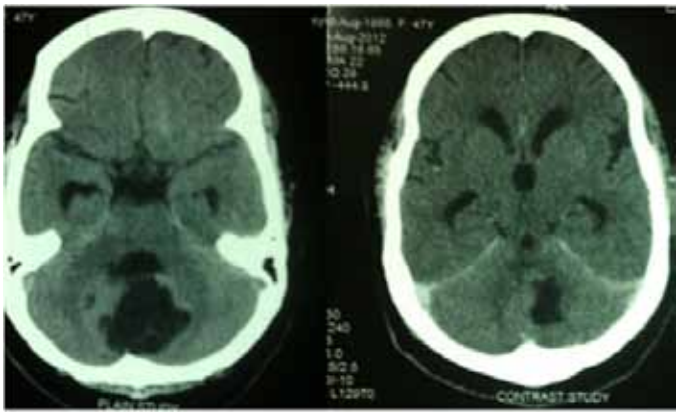

Figure 1: Plain and contrast computed tomography images showing lesion with mixed density in the posterior fossa with evolving hydrocephalus.

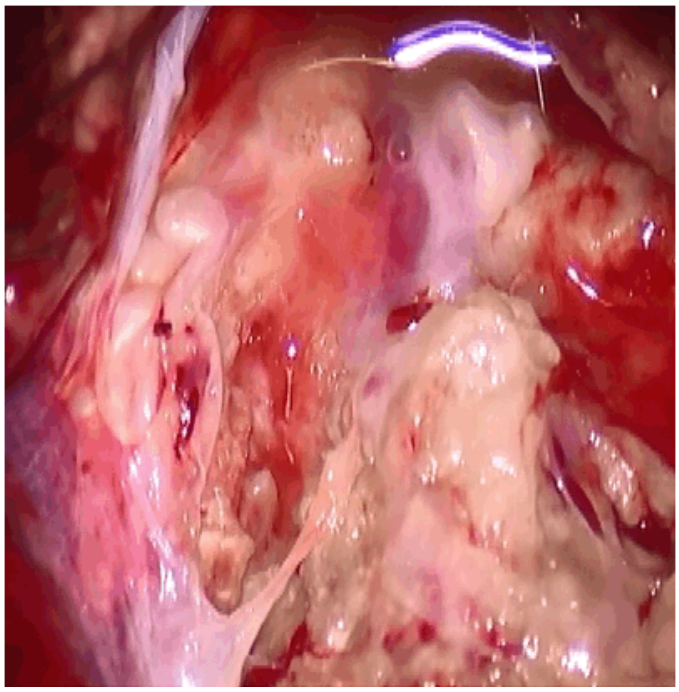

Figure 2: Intraoperative picture showing the bed of pearls appearance of the lesion

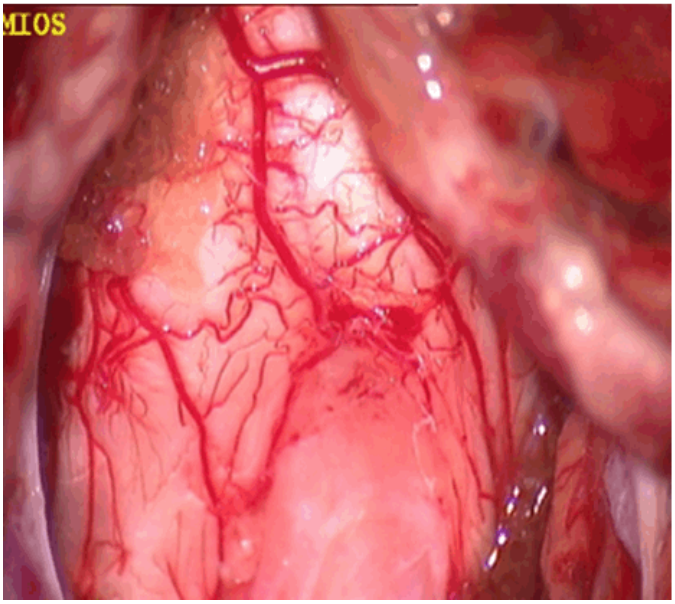

Figure 3: Tumor bed after gross complete removal of the lesion.

to insinuate and encasing adjacent vessels and nerves thereby making its complete removal difficult.

Current armantarium of radio imaging shows cerebrospinal fluid like characteristics but having insinuation to adjacent neurovascular structures [1]. Early distinction from the arachnoid cyst can be easily done by applying the Fluid attenuated inverse recovery (FLAIR) and diffusion weighted image (DWI) sequences. On contrary to arachnoid cysts, it is hyperintense on FLAIR and has restricted diffusion on DW images.

Majority of the patients present with gait problems due to involvement of cerebellar vermis [7]. On contrary to other solid tumors, the incidence of hydrocephalus is rare in such tumors due to egress of the cerebral spinal fluid within the fissures of the tumors to the surrounding foramina [7]. Another presentation may be chemical meningitis due to tumor leakage into the subarachnoid space [6]. The reported rate of recurrence of epidermoid tumors in literature is highly variable. In case of suspected recurrence, main focus should be on the clinical background [6]. While undergoing re-surgery, main attempt should be made removing the areas of abnormal enhancing portion seen on the image studies and sending them for histopathological diagnosis.

CONCLUSION

These tumors though benign in nature, the insinuating nature of the tumor marks the complete removal of the tumor most often cumbersome. Characteristic findings of hyperintensity on FLAIR and restricted diffusion on DWI MR sequences afford easy distinction from arachnoid cyst. Intraoperatively, they have characteristic bed of pearls appearance marking its apparent diagnosis. Postoperative period may be complicated by chemical meningitis, pseudomeningocele and recurrences.

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Author Contributions

Sunil Munakomi – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Binod Bhattarai – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Iype Cherian – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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