

Interhemispheric Epidermoid Tumor: A Case Report

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DOI: [10.36348/sjpm.2020.v05i12.004](https://doi.org/10.36348/sjpm.2020.v05i12.004)

Received: 29.10.2020 | Accepted: 10.11.2020 | Published: 16.12.2020

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Abstract

Intracranial epidermoid tumors are inclusion tumors of the central nervous system. They are congenital neoplasms that grow through desquamation of keratin, cholesterol and cellular debris. They cause mass effect and gradual development of neurological symptoms. Here we present a case of a 65 year old lady who presented with new onset seizures, CT scan suggesting a low grade cortically based neoplasm and a diagnosis of epidermoid cyst was made from histopathological sections.

Keywords: Epidermoid cyst, congenital, keratin, inter hemispheric.

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INTRODUCTION

Epidermoids are benign, slowly growing congenital lesions and are most commonly located in the cerebellopontine angle, followed by the suprasellar cisterns and other locations include Sylvian fissure, brainstem, intraventricular, pineal regions, intradiploic space of skull and spinal cord. Interhemispheric epidermoids are very rare and only 19 cases have been reported. Clinical presentation is related to gradual mass effect and depends largely on the location of the tumour. Patients usually present with focal onset seizures with or without secondary generalisation and are aged between 17 and 45 years. Some patients present with frontal and temporal lobe dysfunction, raised intra cranial tension, hemiparesis and neurological deficits.

CASE REPORT

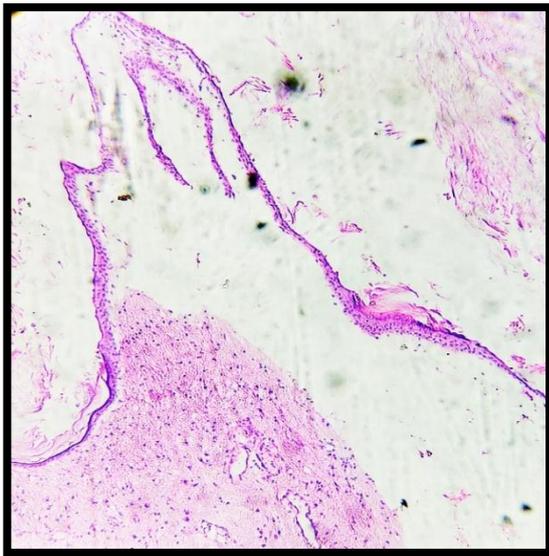
A 65 year old lady presented with two episodes of seizure associated with loss of consciousness in the last one month period. She had no history of fever, trauma or any other significant illness in the past. CT scan Head revealed a well-defined hyper intense, non-enhancing lesion in the left antero-superior frontal convexity and anterior interhemispheric fissure measuring 4.7x5x3.3 cm with calcification and perilesional edema. Lesion is crossing the midline to right side. A possibility of a low grade cortically based neoplasm with differentials, Oligodendroglioma and Dysembryoplastic Neuroepithelial tumor was made



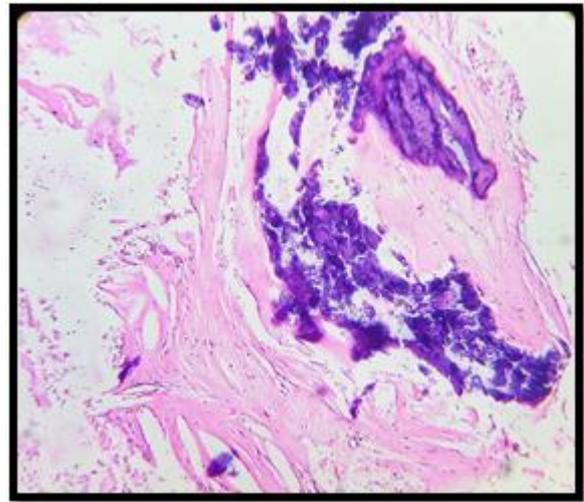
Cyst wall lined by squamous epithelium with keratin flakes and wall showing brain tissue

Intra operatively it was an extra axial easily suckable, friable lesion in the left hemisphere, crossing to the opposite side with calcified areas. We received the specimen piecemeal with calcified tissue bits. H&E sections showed fragments of a cyst wall, lined by stratified squamous epithelium with lumen showing keratin flakes and glial tissue in the wall. Areas showing calcification, ossification and cholesterol cleft formation were also seen. No hair or hair fragments

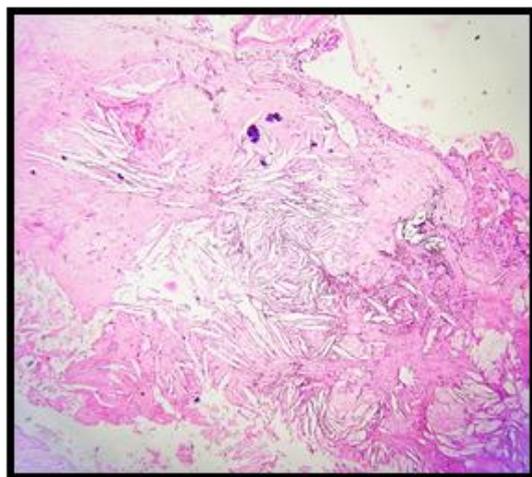
seen. A diagnosis of interhemispheric congenital epidermoid cyst was made.



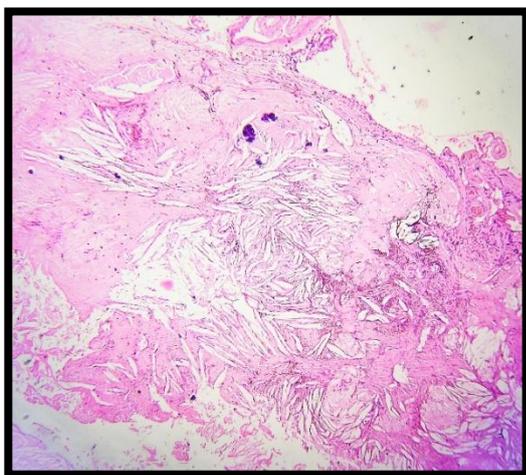
Cyst wall lined by squamous epithelium with keratin flakes and wall showing brain tissue



Section showing areas of ossification



Sections showing areas of calcification



Section showing cholesterol cleft formation.

DISCUSSION

Epidermoid cysts are benign congenital lesions and account for 0.5% to 1.8% of all intracranial brain tumors. During the third to fifth week of embryonic life, the ectoderm on the mid dorsal region forms a neural plate, which then infolds to form the neural tube. During infolding, if the surrounding ectoderm does not separate from the neural ectoderm completely, nests of these cells may be entrapped along with neural ectoderm. These nests of cells later grow within the central nervous system forming a spectrum of lesions, viz: epidermoids, dermoids and dermal sinuses. Epidermoids grow slowly and become symptomatic during the third to fifth decades of life, unlike dermoids, which are fast growing and which become symptomatic in the second decade. The dermoid cysts are known for its midline location, but epidermoids are common in the lateral location. The lateral preference of most of the epidermoids are due to proliferation of multipotential embryonic cell rests carried laterally with the developing neurovasculature. Leak of epidermoid tissue from the capsule may result in inflammatory reaction with formation of thickened hyalinised and fibrous arachnoid membrane. This may lead to dense adhesions. If tumor capsule is adherent to neurovascular structures, it is preferable to leave a small tag of cyst rather than damaging vascular and neural structure. Rupture or spillage of epidermoid tissue during excision may lead to severe chemical meningitis. Complete isolation of the epidermoid tumor from the surrounding tissue with cotton, removal of the content followed by capsular dissection, gentle and complete removal of content without spillage, irrigation with diluted hydrocortisone fluid and perioperative intravenous steroid reduce the chance of chemical meningitis.

Goldman and Gandy has documented a squamous cell carcinoma developed in the bed of the lesion 33 years after surgical resection of a benign right lateral intraventricular epidermoid tumor.

CONCLUSION

Interhemispheric epidermoids are rare congenital tumors. The rarity of these tumors has often led to delayed diagnosis. Achieving safe complete excision without spillage is the surgical goal to prevent chemical meningitis and recurrence.

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