

Epidermoid cyst in Anterior, Middle & Extension of Posterior cranial fossa: rare Imaging with review of literature

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Abstract: Epidermoid cysts are benign slow growing more often extra-axial tumors that insinuate between brain structures, we present the clinical, imaging, and pathological findings in 35 years old female patients with atypical epidermoid cysts which was situated anterior, middle & posterior cranial fossa. NCCT head revealed hypodense lesion over right temporal and perisylvian region with extension in prepontine cistern with mass effect & midline shift and MRI findings revealed a non-enhancing heterogeneous signal intensity cystic lesion in right frontal & temporal region extending into prepontine cistern with restricted diffusion. Patient was deteriorated in night of same day of admission, emergency Fronto-temporal craniotomy with anterior peterousectomy and subtotal resection was done. The histological examination confirms the epidermoid cyst. The timing of ectodermal tissue sequestration during fetal development may account for the occurrence of atypical epidermoid cysts.

Key words: Epidermoid cyst, anterior, middle, posterior cranial fossa, prepontine cistern

Introduction

Epidermoid cysts are benign, slow growing extra-axial tumors that account for 1% of all intracranial tumors (10). Embryologically, they are derived from ectodermal inclusions during neural tube closure from the third to the fifth weeks of embryogenesis (10, 8). They frequently occur at the Cerebellopontine angles and parasellar regions, insinuating between brain structures. Conversely, epidermoid cysts in intraparenchymal or intradiploic locations are very rare, accounting for less than 5% of all intracranial epidermoid

cysts (1). Here, we reported rare cases of 35 years old female demonstrated an extra-axial epidermoid cyst which was situated in anterior, middle as well as posterior cranial fossa.

Case Report

We reported a clinical case concerning a 35 years old Female who presented convulsions during 5 years, headache & ataxia during 5 months, slurring of speech during 3 months & diminished of vision in both eyes during 2 months. The clinical examination found lower motor neuron type grade IV Facial nerve palsy

on right sided with ataxia & vision of bilateral eyes is only perception of light with right sided oculomotor nerve & lateral rectus palsy. The NCCT Head (Figure 1) show hypodense lesion over right temporal and perisylvian region with extension in prepontine cistern with mass effect & midline shift. MRI brain (Figure 2) with contrast showed heterogeneous signal intensity cystic lesion in right frontal & temporal region extending into the prepontine cistern with mass effect and midline shift. The lesion appears hypointense on T1W and

hyperintense on T2W sequence and showing restriction on diffusion weighted images. Patient was deteriorated in night of same day of admission so underwent emergency surgery via fronto-temporal approach with anterior petrousectomy and subtotal resection. A solid part of the tumor and the capsule were deliberately left due to adhesions to the Neurovascular structure. It was a pearly tumor. The histological examination (Figure 2) confirms the epidermoid cyst.

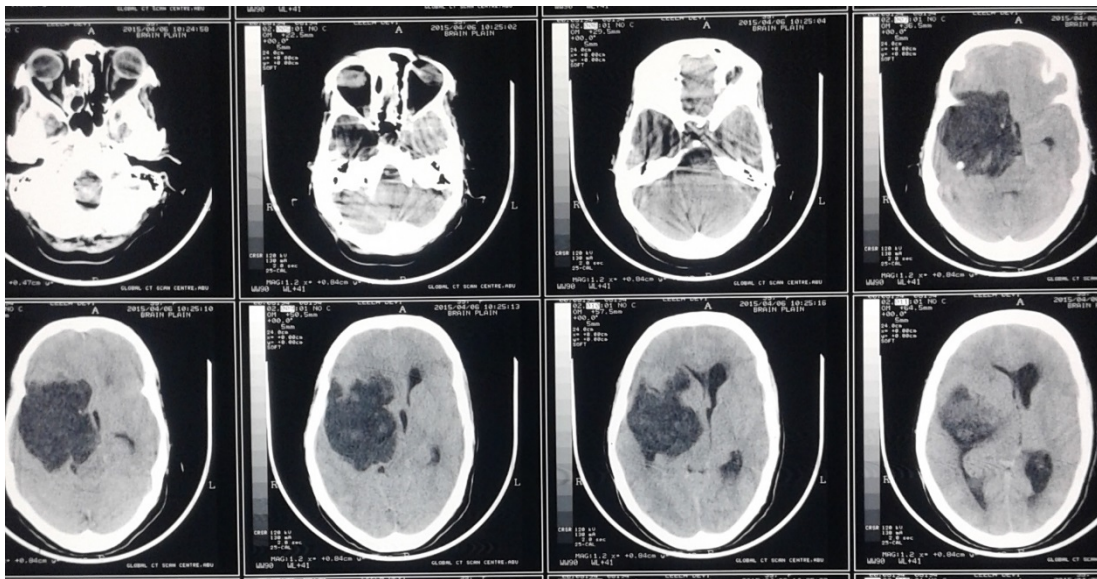


Figure 1 - NCCT Head showed right temporal lobe and perisylvian region hypodense lesion with extension in prepontine cistern, mass effect & midline shift

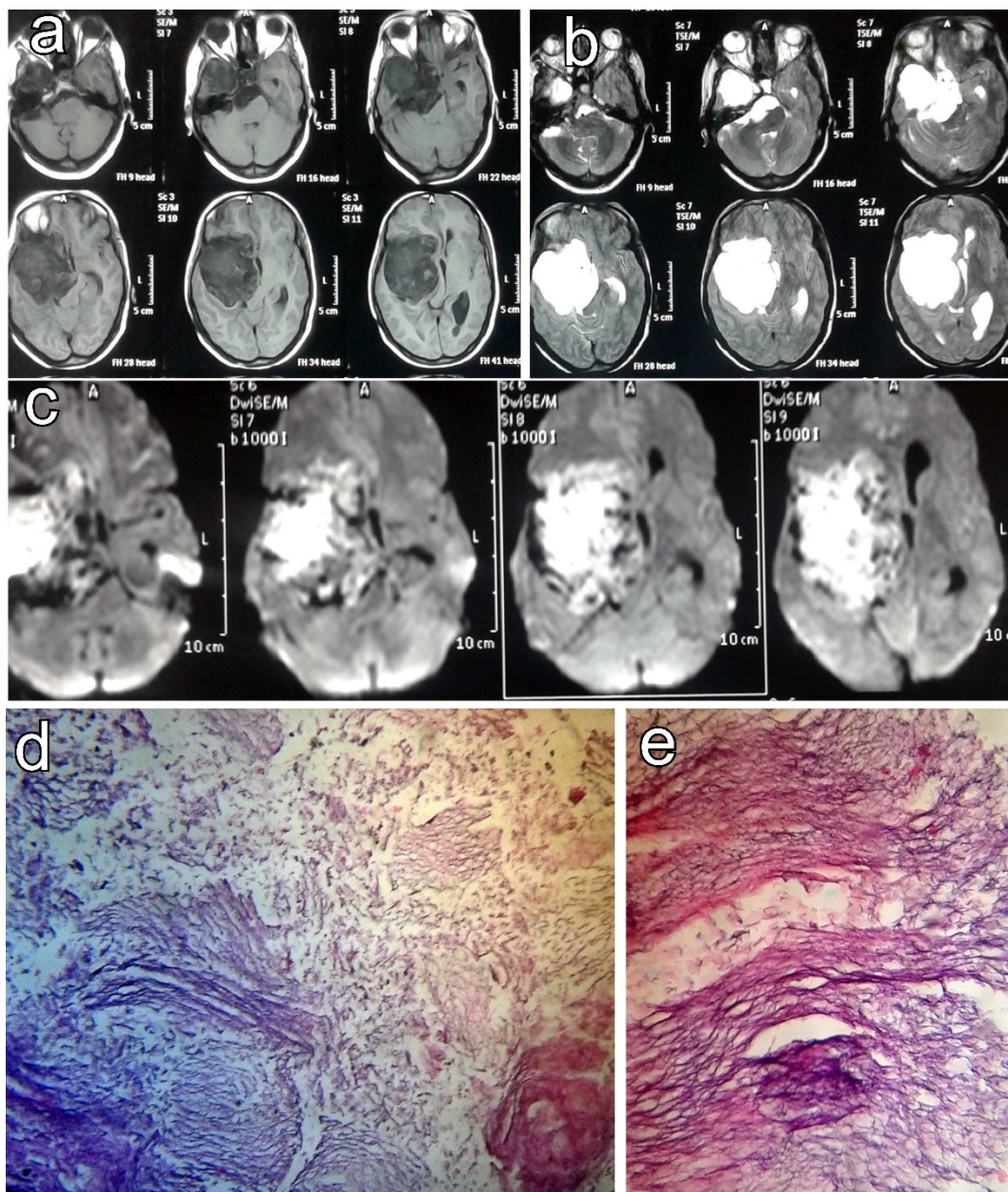


Figure 2 a, b, c - MRI brain with contrast showed heterogeneous signal intensity cystic lesion in right frontal & temporal region extending into the prepontine cistern with mass effect and midline shift. The lesion appears hypointense on T1W and hyperintense on T2W sequence and showing restriction on diffusion weighted.
d, e - H & E 40 & 10 Magnification - Histopathological finding shows an epidermoid cyst lined by maturing and keratinizing stratified squamous epithelium and squames, which is formed from degenerative keratinocyte

Discussion

Epidermoid cysts can occur throughout the neuroaxis, most commonly in the Cerebellopontine angles (40–50%) and the parasellar region (10). Conversely, atypical Epidermoid cysts are rare, with intra-axial epidermoid cysts accounting for less than 1.5% of all intracranial epidermoid cysts (1) and intradiploic epidermoid cysts (including congenital cholesteatomas) accounting for approximately 3% (6, 7). In our case is 1st reported case in world's literature, in which epidermoid cysts involving anterior, middle as well as post fossa. Occasionally the Epidermoid found in pineal gland (9) or the brainstem (11). The proposed embryological pathogenesis of the typical epidermoid cyst involves trapped ectodermal components travelling along the otic vesicles during neural tube closure, thus accounting for the propensity for its location at the Cerebellopontine angles (2). Cell entrapment from the mesectodermal origin of the neural crest within the primitive cerebral hemisphere may lead to the formation of such rare intracerebral lesions (5). Along this line of reasoning, intraparenchymal epidermoid are thought to arise when the ectodermal inclusion occurs before the third week of embryogenesis (when the primary cerebral vesicle is being formed), while intradiploic epidermoid cysts originate from aberrant ectodermal remnants that become trapped after neural tube closure (1, 6, 2). The radiological differential diagnoses of an epidermoid cyst include an arachnoid cyst, dermoid cyst, abscesses, metastasis, or slow

growing brain tumors. FLAIR and DWI sequences (4) have been proposed as useful discriminator in differentiating an arachnoid cyst from the typical epidermoid cyst, in which the signal intensity of the arachnoid cyst follows CSF signal without restricted diffusion. A dermoid cyst is usually at a more central location with foci of calcifications which could be differentiated from epidermoid cyst (12). Abscesses show typical rim of the contrast enhancement, metastasis is usually multiple with known primary tumor and heterogeneous contrast enhancement, and cystic neoplasm usually shows solid component with enhancement. Current debate is ongoing regarding the optimal treatment for an epidermoid cyst. On the one hand, gross total resection of epidermoid cysts offers a definite treatment with prevention of its occurrence or aseptic meningitis (7). Conversely, epidermoid is often located in close proximity to neurovasculature and vital brain parenchyma, and thus a conservative resection can be considered given the slow growing nature of this tumor, with a linear growth rate similar to normal epithelial cells (one generation per month)(3).

Conclusion

Epidermoid presented in anterior, middle and posterior cranial fossa is extremely rare case with unusual location. This is a first case reported in world's literature. Accurate preoperative diagnosis can be very difficult due to the radiological similarities to other common intracerebral cysts (e.g., astrocytomas or gliomas). Magnetic resonance imaging (MRI) studies, especially with

diffusion-weighted images, allow greater accuracy in the preoperative differential diagnosis. Radical surgical removal should be attempted, but a less aggressive surgical strategy should be considered if there is strong adherence between the tumor capsule and the Neurovascular structure of brain tissue, particularly in eloquent areas.

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