Intracranial Epidermoid Cysts with Malignant Transformation: Two Case Reports

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Introduction

- Intracranial epidermoid cysts (IECs) originate from remnants of ectodermal cells during neural tube closure in 3rd – 5th week of embryogenesis.
- Incidence is 0.5–1.8 % of all primary intracranial tumors, common sites are cerebellopontine angle, 4thventricle, and the sellar/parasellar regions.
- Primary mode of management is surgical excision.
- Malignant transformation to squamous cell carcinoma (SCC) is very rare with a single case report of IEC transformation to glioblastoma.

Result (Case 1)

- 48 yr male, presented to **emergency** with history of **altered sensorium** for 1 day, on and off headache & vomiting for 10 days.
- 1 episode of bilateral tonic clonic seizures (**TCS**) 5 days back.
- **CT head (Fig 1a)** showed multiple conglomerate ring enhancing lesions in right temporoparietal lobe with significant perilesional edema and midline shift.
- HPE revealed IEC with full thickness dysplasia of lining epithelium with invasive cell nests of squamous cells & atypical mitotic figures **(Fig 1b-d).**

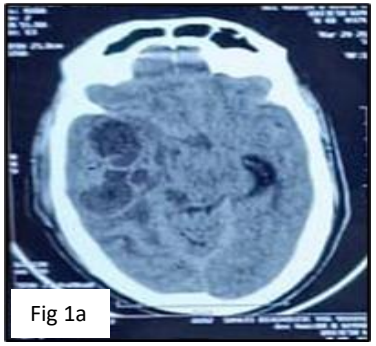


Fig 1a

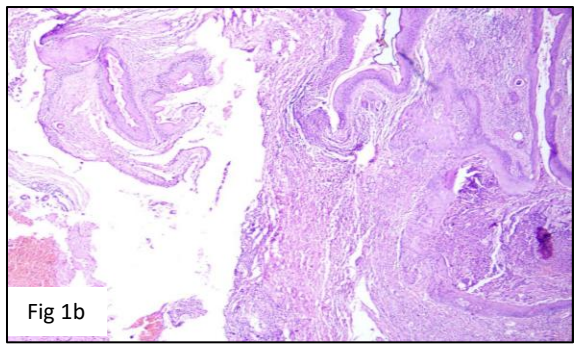


Fig 1b

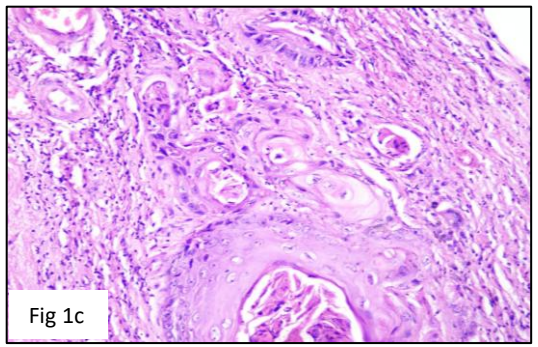


Fig 1c

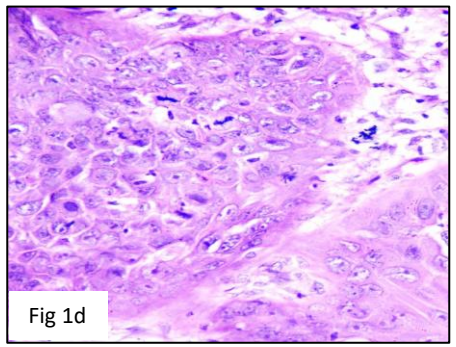
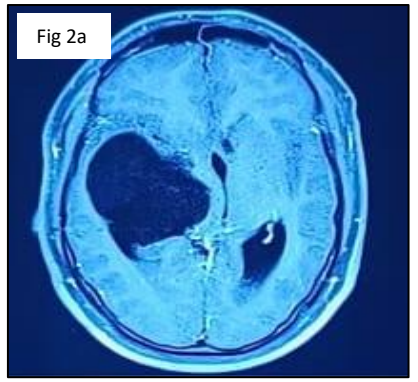


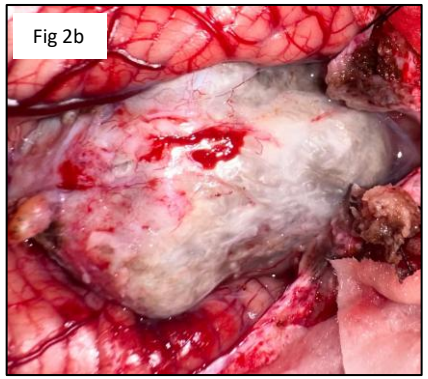
Fig 1d

Fig1 (a) CT head **(b)** HPE, H&E; 100X **(c,d)** HPE, H&E; 400X

Result (Case 2)

- 23 years old male presented with generalised TCS and headache for 5 years.
- MRI **(Fig 2a)** revealed 5.6x5.5x5 cm non enhancing lobulated space occupying lesion (SOL) in right temporal region .
- Intraoperative mass was whitish, friable, mildly vascular and suckable **(Fig2b)**
- Histopathological examination (HPE) was suggestive of epidermoid cyst **(Fig 2c).**





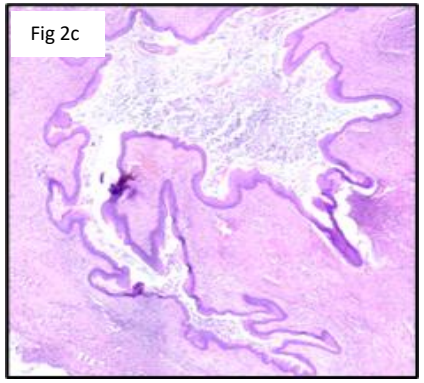
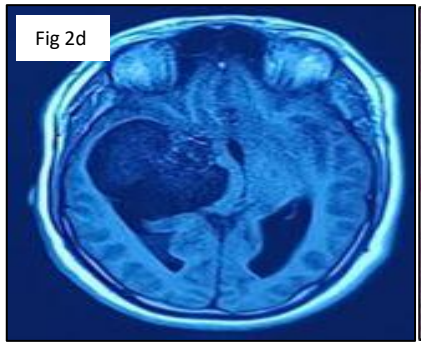
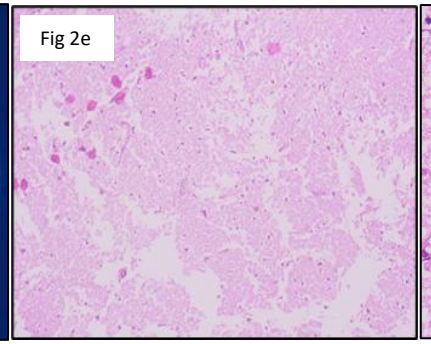


Fig2 (a) MRI brain **(b)** Intraoperative image **(c)** HPE,H&E; 200X).

- **<u>After 7 months</u>,** patient presented with headache and bilateral diminished vision for 20 days.
- MRI **(fig 2d**), right temporal lobe showed a heterogenous enhancing large solid cystic SOL, suspicious of GBM.
- HPE **(fig2 e,f)** revealed low grade glioma with mild cellularity (H&E, 200X, 400X).
- Immunohistochemistry done showed tumor cells positive for IDH1, ATRX retained, weak focal p53 positive.
- Final diagnosis: Oligodendroglioma, CNS WHO grade 2
- FISH for 1p/19q codeletion was advised.





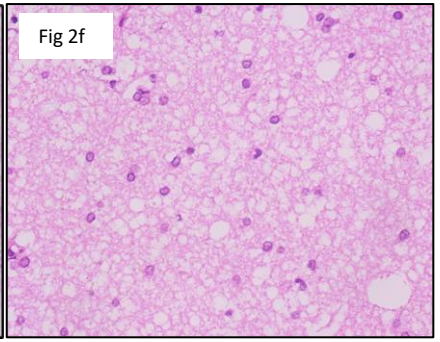
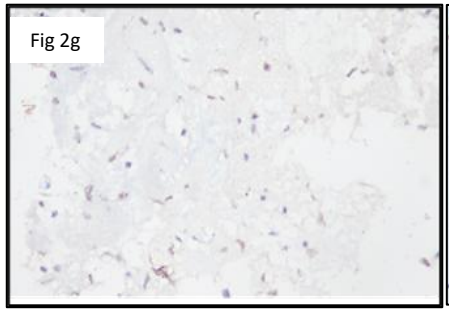
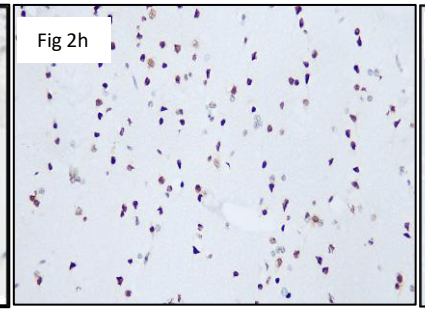


Fig2 (d) MRI brain **(e,f)** HPE, H&E, 200X, 400X.





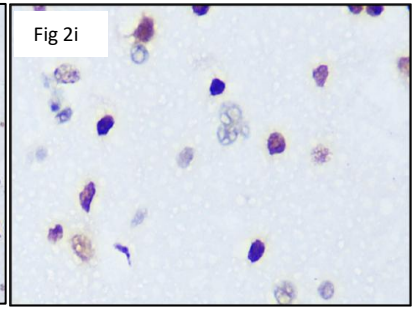


Fig2 (g) IDH1,400X **(h)** ATRX,400X **(i)** p53, OIF

Discussion & Conclusion

- Malignant transformation in IECs is **associated with** rapid onset of symptoms, recurrence, leptomeningeal carcinomatosis & enhancement in CT/ MRI.

- Epidermoid carcinogenesis→**chronic inflammation** from cyst rupture or intraoperative deposition of foreign material.

- **Mantovani et al**. propose model of **converging cancer and inflammatory pathways** in which **NF-κB, HIF1α and STAT3** collectively produce cytokines in stromal, tumor and inflammatory cells.

- **Giannoni et al.** showed cancer-associated fibroblasts utilize these transcription factors causing release of ROS leading to **epithelial–mesenchymal shift** with subsequent stem cell formation.

- Non-SCC transformation to oligodendroglioma is **hitherto unreported.**

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