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# Recurrence of ruptured intracranial epidermoid cyst – A rare case report and presentation

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## ABSTRACT

**INTRODUCTION:** Intracranial epidermoid cysts are congenital tumors that develop from ectodermal remnants during neuroembryogenesis between the third and fifth weeks of gestation. These tumors are benign and usually present with local mass effect. Here, we present a rare case of ruptured intracranial epidermoid cyst with recurrence.

**PRESENTATION OF CASE:** A 55 years old male patient was brought to emergency with a history of headache and loss of consciousness for 1 h. Radiological imaging showed the features suggestive of ruptured intracranial epidermoid cyst which was operated. Two years later the patient re-presented with headache for 4–5 days where repeat MRI revealed recurrence of the tumor.

**DISCUSSION:** Epidermoid cysts are very slow growing tumor at a linear rate due progressive accumulation of normally dividing epidermal cells. These tumors often reach a large size before the onset of symptoms. At times, the tumor capsule may show infiltration to the brain parenchyma and tight adherence to neurovascular structures which leads to the incomplete removal of the tumor capsule leading to recurrence of tumor.

**CONCLUSION:** Rupture of intracranial epidermoid cyst is a rare phenomenon and recurrence of this tumor in patients is even infrequent.

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## 1. Introduction

Intracranial epidermoid cysts are benign congenital tumors that arise from ectodermal cellular remnants during 3–5 weeks of gestation due to incomplete cleavage of the neural ectoderm from the cutaneous ectoderm. This results in a subsequent inclusion of epiblasts in the neural tube [1]. The incidence of epidermoid cyst is between 1%–2% of all intracranial tumors. This epidermoid tumor was initially described by the French pathologist Cruveilhier as the “most beautiful tumors of all the tumors” based on its pearly nature [2].

Rupture of intracranial epidermoid cysts is a rare phenomenon and recurrence of such tumors after resection is extremely low [2]. We report a rare case of recurrence of ruptured intracranial epidermoid cyst and its presentation as per the SCARE guidelines [3].

## 2. Patient information

### 2.1. Patient presentation

A 55 years old male patient was brought to emergency with a history of headache and loss of consciousness for 1 h. He had complained of headache for the past 6 days. The patient was a known hypertensive, diabetic on medication and regular follow up for the last 6 years. He was non-alcoholic and non-smoker. He did not give any history of food or drugs allergy and psychosocial problems.

### 2.2. Laboratory and radiological findings

His routine lab investigation revealed hemoglobin of 14 gm/dl, total WBC count of 8800/cu mm and random blood sugar of 9.1 mmol/l. His blood differential count and platelets were within normal limit. His HIV, HbsAg and anti HCV tests were non reactive.

Contrast enhanced CT scan revealed a large 4 × 2.9 cm non-enhancing fat density lesion with wall calcifications in left anterior temporal region. The lesion also extended to the left frontal region (Fig. 1) with posterior displacement of the first and second part of the left middle cerebral artery and medial displacement of the anterior cerebral artery. No vascular encasement was seen. CT impression was given as suggestive of epidermoid cyst.

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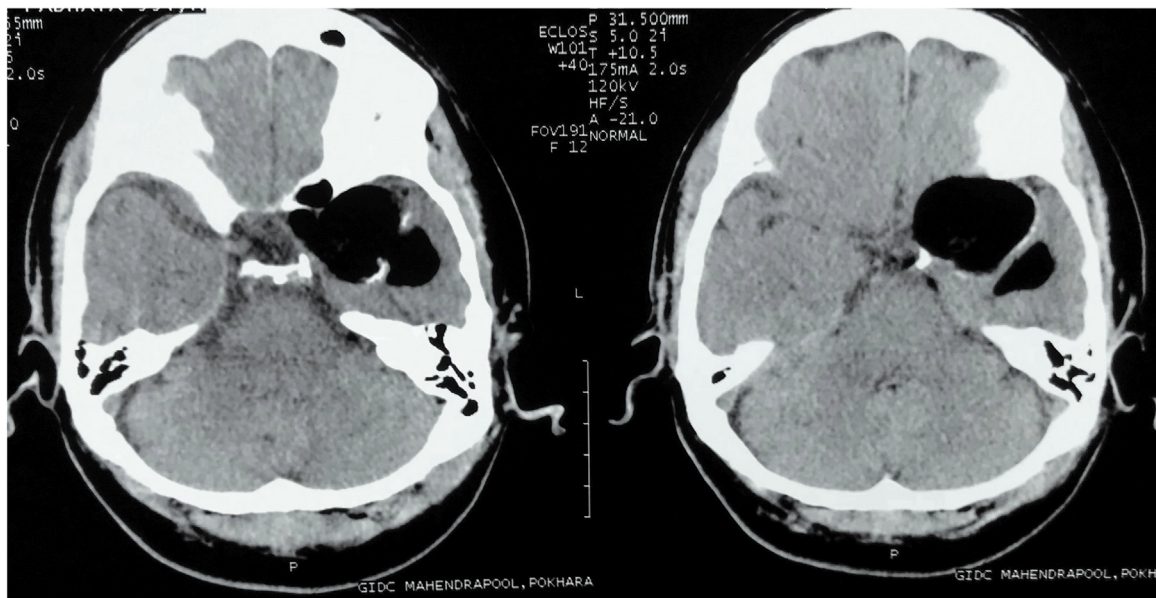


Fig. 1. Axial CT image showing large intracranial fat density lesion with patchy peripheral calcification in left temporal fossa causing mass effect on surrounding structures.

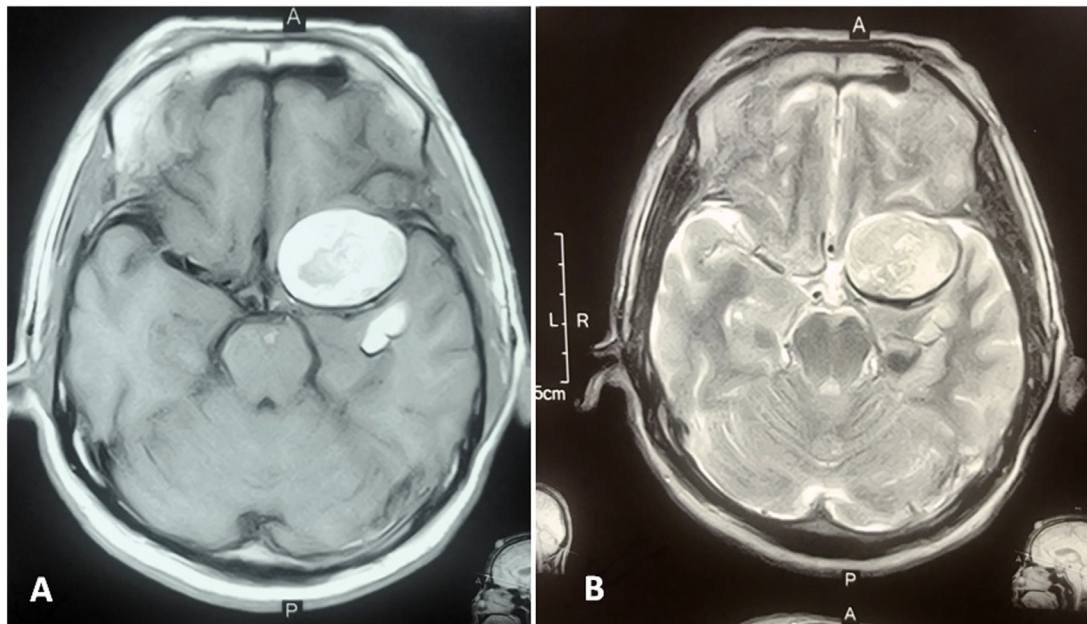


Fig. 2. Axial T1 W image (A) and T2 W image (B) showing hyperintense mass.

Contrast MRI head disclosed heterogeneous, lobulated extra-axial mass lesion in the left middle cranial/ temporal fossa abutting the cavernous sinus. The lesion appeared hyperintense on T1-weighted (Fig. 2A), FLAIR images and heterogeneously hyperintense on T2-weighted images (Fig. 2B). No significant post contrast enhancement was seen. The lesion was heterogeneously hypointense on FAT SAT images (Fig. 3). The lesion measured approximately  $4.1 \times 3.8 \times 5.3$  cm in its largest dimension with patchy peripheral hypointense rim of calcification. The lesion caused mass effect on left temporal lobe, left lentiform nucleus, insular cortex, temporal and frontal horn of left lateral ventricle with posterior displacement of M1 division of left middle cerebral artery.

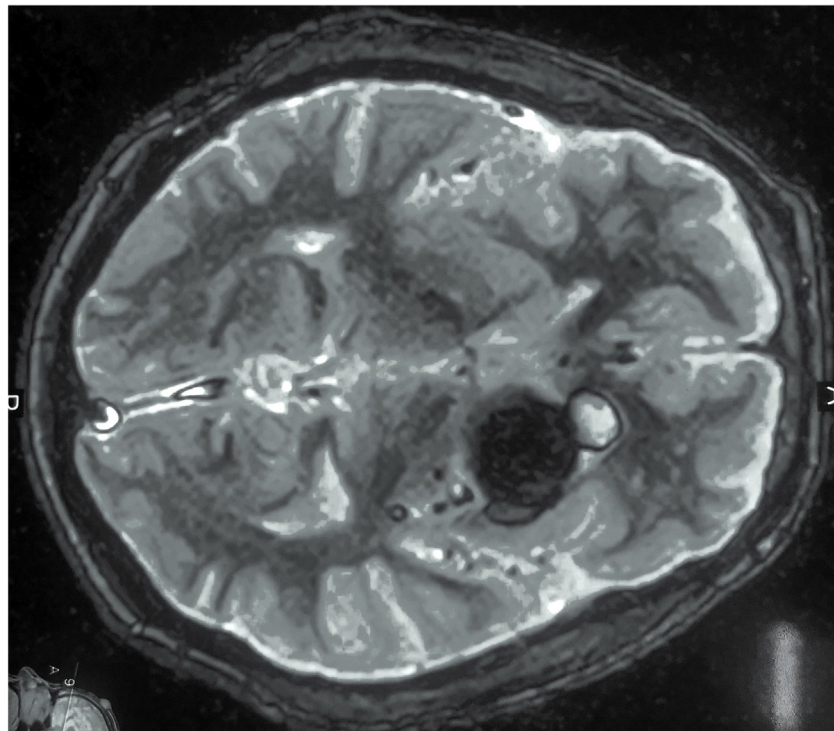
Tiny ill defined T1-weighted hyperintense areas were seen extending posteriorly from the lesion to the external capsule. Similar hyperintense foci were seen in interpeduncular, right ambient

cistern and in left cortical sulci (Fig. 4), suggesting features consistent with mild rupture of the intracranial epidermoid cyst. MRI also disclosed a well defined subcutaneous fat signal intensity lesion in the midline of frontal region.

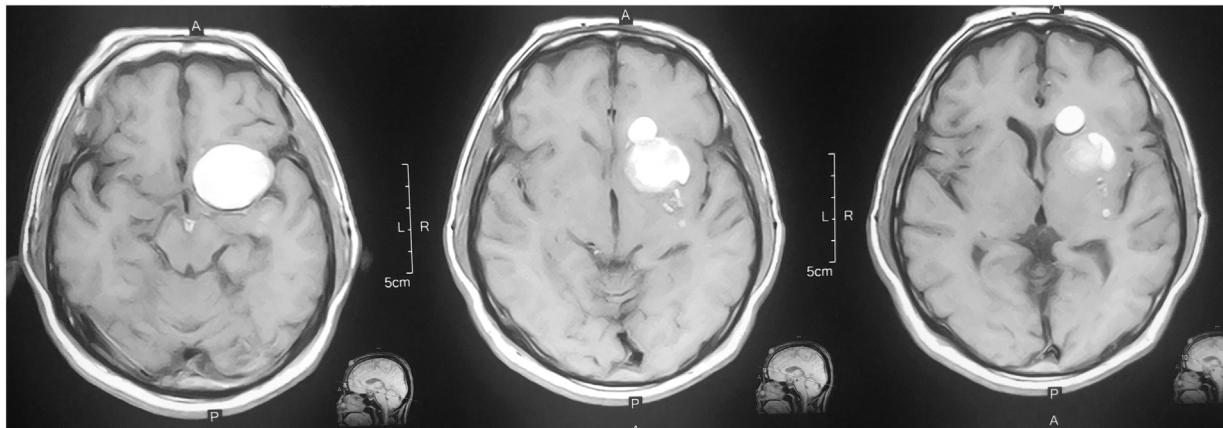
### 2.3. Therapeutic intervention

Patient was taken up for operation by experienced neurosurgical team. A left fronto-temporal craniotomy was done and nearly total gross excision of the lesion was performed. The operation lasted for three hours. Patient was discharged on 7th post-operative day and was kept on tapering dose of steroid for 14 days with no restrictions on any daily activities.

Gross examination of the specimen showed multiple bits of pale white pultaceous material altogether measuring  $8 \times 4$  cm. No brain parenchyma was found grossly. Multiple bits were processed.



**Fig. 3.** Axial Fat suppressed image showing hypointensity within the lesion.



**Fig. 4.** Axial T1W image showing the mass with hyperintense foci in interpeduncular cistern, right ambient cistern along the external capsule suggesting rupture of the cyst.

Microscopic examination showed sheets of keratin flakes with occasional area showing cystically dilated space lined by squamous epithelium along with thick bundles of keratin (Fig. 5). Focal area showed dystrophic calcification (Fig. 6). No brain parenchyma, skin adnexal structures were identified and there was no evidence of atypia. The histopathological diagnosis was given as intracranial epidermoid cyst.

#### 2.4. Follow-up and outcomes

Postoperatively he was on follow up on 2nd, 3rd and 6th months with no signs and symptoms of recurrence of tumor on clinical examination.

Two years later the patient re-presented with headache since 4–5 days. Contrast MRI head revealed non enhancing well defined lobulated mass measuring  $8.4 \times 40 \times 42$  mm in the left anterior temporal region extending superiorly to the frontal horn of the left lateral ventricle causing a mild mass effect on adjacent surfaces. The

lesion was hyperintense on T1-weighted (Fig. 7A), heterogeneously hyperintense on T2-weighted images (Fig. 7B). Well defined cystic area appearing hypointense on T2-weighted (13.7 mm) was seen in superior aspect of the lesion, protruding into the frontal horn of left lateral ventricle, suggesting recurrence of tumor. Post operative changes with gliosis was seen in left temporal and frontal regions. Final MRI impression was of recurrence of tumor with post operative changes and residual lesion in the interpeduncular cistern and external capsule.

#### 3. Discussion

Epidermoid cysts are very slow growing tumor at a linear rate due to progressive accumulation of normally dividing epidermal cells. These tumors can insinuate into the brain parenchyma and often reach a large size before the onset of symptoms [1,2,4]. The most common site for the epidermoid cysts are parasellar region and cerebellopontine angle and are less commonly located in syl-

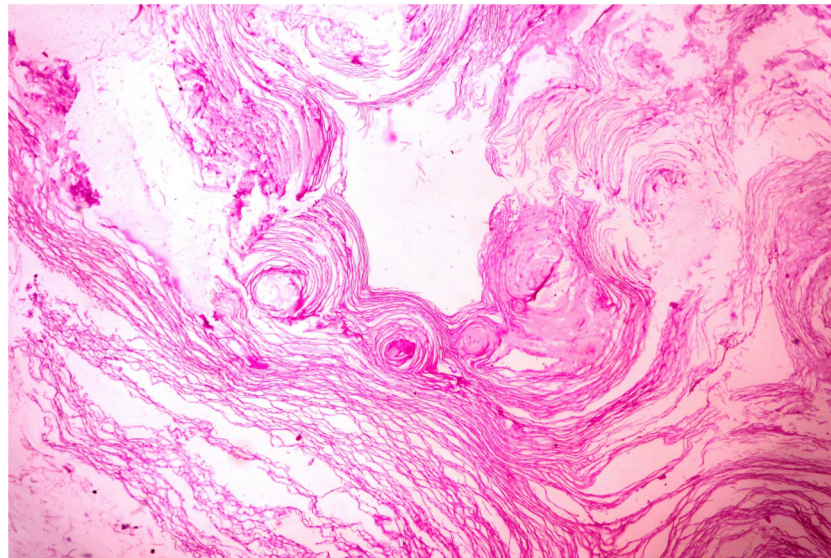


Fig. 5. Microscopic picture showing sheets of keratin flakes with keratin pearls (H&E 40×).

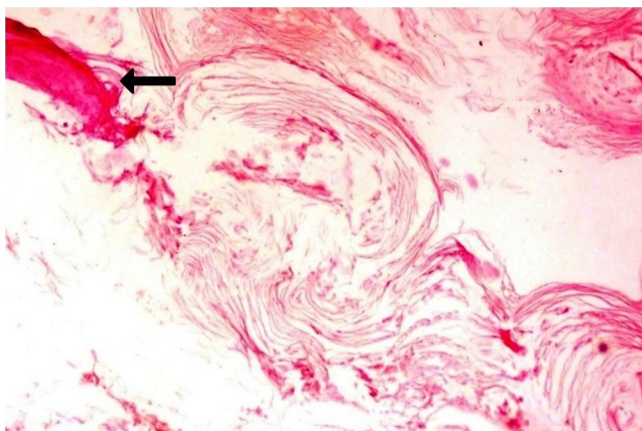


Fig. 6. Microscopic picture showing area of dystrophic calcification (arrow) with surrounding keratin flakes (H&E 40×).

vian fissure, suprasellar region, cerebellar hemispheres, lateral and fourth ventricles [2].

Intracranial epidermoid tumors show a cyst lined by thick layer of stratified squamous epithelium [1]. The epidermoid cyst in some cases show cystic fluid within the lumen [5–7]. The tumor enlarges slowly over a period of time due to accumulation of desquamated epithelial cells which leads to keratin and cholesterol accumulation

giving it a pearly appearance [8,9]. Epidermoid cysts are commonly symptomatic during the adulthood and usually present during the 4th decade of life [1,2]. In our case, the patient presented with the tumor at the age of 63 years. Very few cases present during childhood due to their slow growing nature of the tumor [2]. Ahmed I et al. in their study found the mean age of the paediatric age group patients to be 10.6 years and interestingly all were female patients [1].

Rupture of intracranial epidermoid cyst is a rare phenomenon and is usually spontaneous [10]. Patients with spontaneous ruptured intracranial epidermoid cysts present with various neurologic symptoms such as headaches, seizures, nausea, visual loss, meningeal signs, and altered consciousness [5]. Our patient presented with headache and loss of consciousness. These symptoms depend on the location of the cyst and sometimes the condition may worsen due to hydrocephalus and progressive visual loss from optic chiasm compression [5,11].

On CT scans, dermoid cysts can have mixed densities. The fatty portion of the tumor has negative HU values and areas of calcification appear hyperdense with high HU values. Fat droplets in the subarachnoid space or ventricles appear hypodense. Hydrocephalus may be present if there is rupture into the ventricular system and a fat–fluid level may be present [5,12]. In MRI, the tumors appear hyperintense on both T1 and T2 weighted images. Disseminated lipid droplets in the subarachnoid cisterns or ventricles appear hyperintense on T1-weighted images and are sensitive

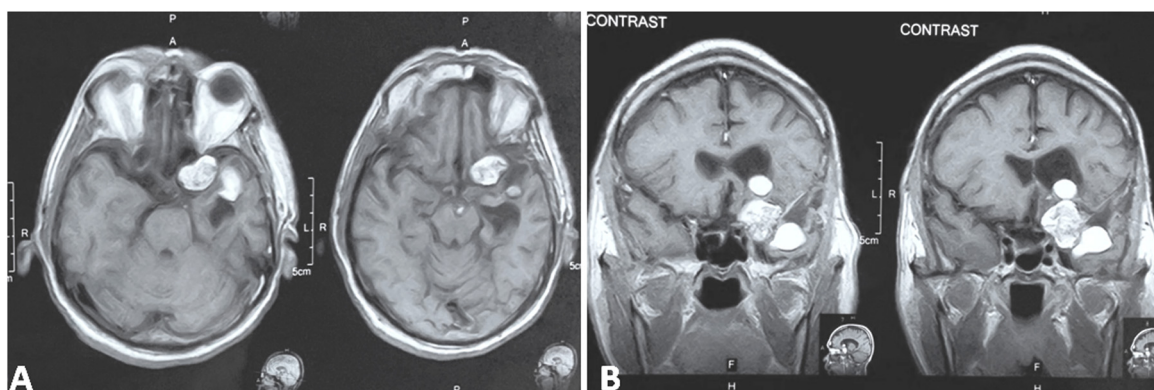


Fig. 7. Post operative follow up T1 W contrast axial (A) and coronal (B) images showing recurrence of tumor.

for the diagnosis of a ruptured cyst [5,13,14]. In cases of these ruptured intracranial cysts into the ventricles, fat-fluid level and movable oil is demonstrated [5,15,16], MRI is the best modality to evaluate epidermoid extension where the solid portion is hypointense on T1-weighted, inhomogeneously hyperintense on T2-weighted images and hyperintense on diffusion weighted images with faint enhancement of the wall [9,17].

The goal of treatment in patients with epidermoid cysts is the complete surgical removal of the primary tumor capsule and intracystic contents without causing damage to the surrounding neurovascular structures [2,5,17,18]. However, at times, intracranial epidermoid tumor capsule may show infiltration to the brain parenchyma and tight adherence to neurovascular structures which leads to the incomplete removal of the tumor capsule [2,6]. Complete removal of the tumor is possible in only 50–80% of intracranial epidermoid cysts [19]. The recurrence rate of the epidermoid cysts is between 1% and 54% in the published studies [6,8,20]. Reoperation in such cases of recurrence should be performed when the patient becomes symptomatic again [2]. In our case, the patient was not operated for the recurrence of the tumor but was given symptomatic treatment.

#### 4. Conclusion

Intracranial epidermoid tumors are rare slow growing tumor which usually ruptures spontaneously but are not fatal. Incomplete removal of the tumor during surgery for preserving the vital neurovascular structures may lead these tumors to recur which may take few years.

#### Declaration of Competing Interest

Authors do not have conflicts of interest.

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#### Ethical approval

Ethical approval for the case report study is not required in Manipal College of Medical Sciences, Pokhara, Nepal.

#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Author contribution

1. Dilasma Ghartimagar – Study design, data collection and paper writing.
2. Manish Kiran Shrestha – Data analysis, interpretation, editing paper writing.
3. Arnab Ghosh – Data interpretation and editing paper.

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#### Registration of research studies

Not applicable.

#### Guarantor

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