International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2025: 7.089

Diagnostic Dilemma: In a Case of Retro Clival-Prepontine and Anterior Cerebellar SOL and Review of Literature

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Abstract: ***Introduction/ Background: Preoperative identification of lesion pathology is crucial for surgical planning and management. However, overlapping radiological features may lead to diagnostic uncertainty. We present a rare case of retro clival–prepontine and anterior cerebellar space-occupying lesions (SOLs), which mimicked two distinct entities radiologically but were proven histologically to represent a single pathology—an epidermoid cyst in different stages of evolution. Case Presentation: A 40-year-old woman presented with a three-month history of progressive headache, intermittent diplopia, facial numbness, and mild gait imbalance. Examination revealed left sixth nerve palsy, trigeminal hypoesthesia, and mild ataxia. Imaging showed two lesions: a hyperdense, non-enhancing midline prepontine mass without diffusion restriction, suggestive of a neuro-enteric cyst, and a left cerebellar lesion with restricted diffusion, consistent with an epidermoid cyst. Surgical excision via a left retro sigmoid suboccipital approach revealed both lesions interconnected by a thin arachnoid membrane. Histopathology confirmed keratinizing squamous epithelium with keratin flakes, diagnostic of an epidermoid cyst. Discussion: Intracranial epidermoid cysts are rare congenital lesions (0.2–1.8% of intracranial tumours) arising from ectodermal remnants during neural tube closure. They commonly appear hypodense on CT and show diffusion restriction on MRI. However, atypical "white" epidermoid may be hyperdense on CT and hyperintense on T1 due to high protein, lipid, or haemorrhagic content. The coexistence of typical and atypical features in this case led to a diagnostic dilemma, initially suggesting two separate pathologies. Conclusion: This case underscores the radiological variability of epidermoid cysts and their potential to mimic other cystic lesions such as neuro-enteric or dermoid cysts. Awareness of atypical imaging patterns and consideration of evolving cyst content can help avoid misdiagnosis and guide appropriate surgical management.***

Keywords: Intracranial Epidermoid, Radiological dilemma, cerebellopontine angle

1. Case Report

Introduction

Preoperative identification of the nature of a lesion has implications for optimal management. It helps the surgeon in taking decision regarding surgical excision of the lesion. Occasionally, there is significant overlap between the radiologic features. We report a case in which two lesion that appeared to be two different pathologies on pre-operative imaging was shown to be single pathology in different stages of pathogenesis.

Case Report

A 40yrs old female without any comorbidity presented to us with history of gradually progressive headache and intermittent diplopia for 3months, also associated with mild facial numbness and imbalance while walking. There was no history of seizures or hearing loss. On examination there was partial sixth nerve palsy on left side, mild hypoesthesia in trigeminal V1-V2 distribution on left side and mild gait ataxia. Sensory / motor and systemic examination was unremarkable.

She underwent non-contrast computed tomography (NCCT) scan at a local hospital, which revealed a well-defined hyperdense lesion in prepontine and pre-medullary cisterns compressing adjacent brainstem and a hypodense lesion in the anterolateral part of cerebellum on left side. There was no calcification or hemorrhage. (fig. 1)

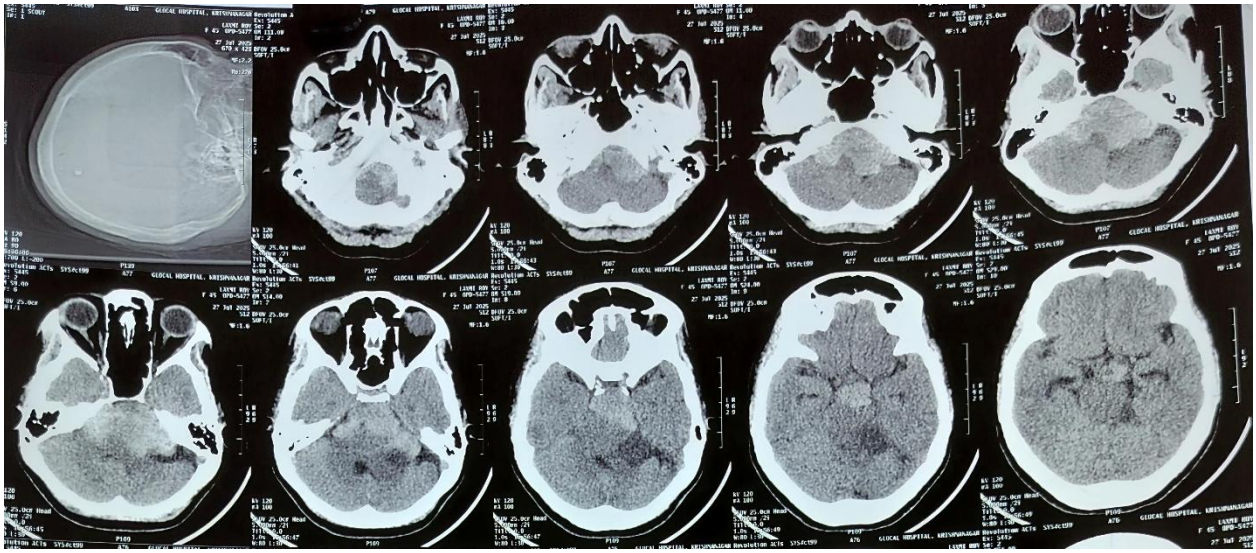

Figure 1

Further evaluation with brain magnetic resonance imaging (MRI), showed an extra axial lesion predominantly hyperintense on T1 and hypointense lesion on T2, T2 FLAIR sequence in prepontine cistern extending up to the cerebellopontine angle cisterns on both sides (Lt>Rt). DWI and ADC map reveals no restriction of diffusion in this lesion. Post contrast T1 WI does not reveal any enhancement of the lesion. The lesion is surrounded by mild oedema and yields mass effect with compression of 4th ventricle leading to mild dilatation of 3rd and lateral ventricles. The lesion variably encases, splays and compresses adjacent vessels e.g. basilar and vertebral arteries. Radiologically possibilities of neuro-enteric cyst considered for this lesion.

On the other-hand the lesion in the anterolateral part of the cerebellum on left side was hypointense on T1 and hyperintense on T2 with restriction of diffusion on DWI and ADC map and no contrast enhancement, strongly indicating an epidermoid cyst. (fig. 2, 3, 4, 5)

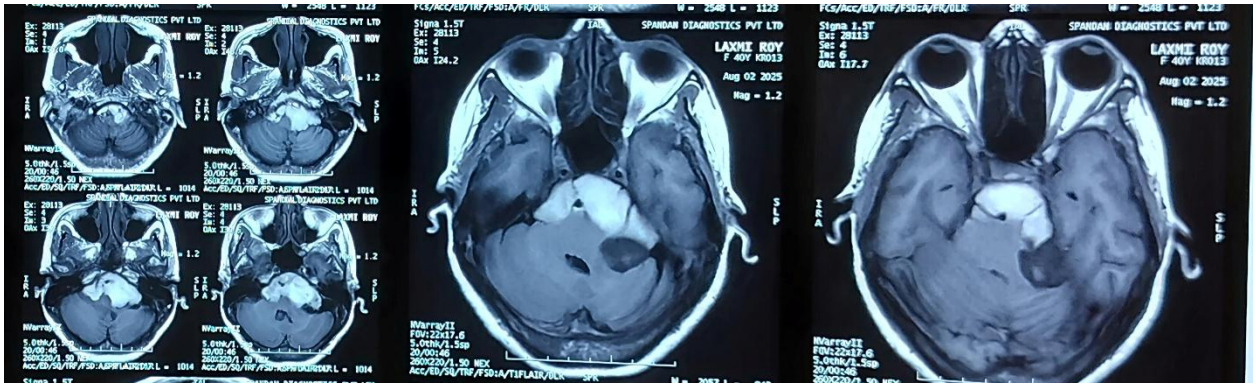

Figure 2

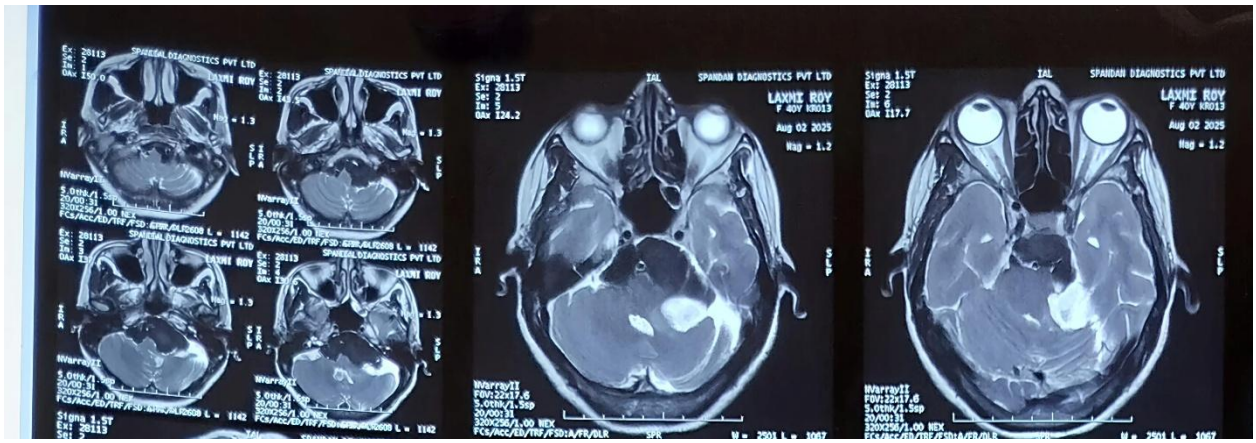

Figure 3

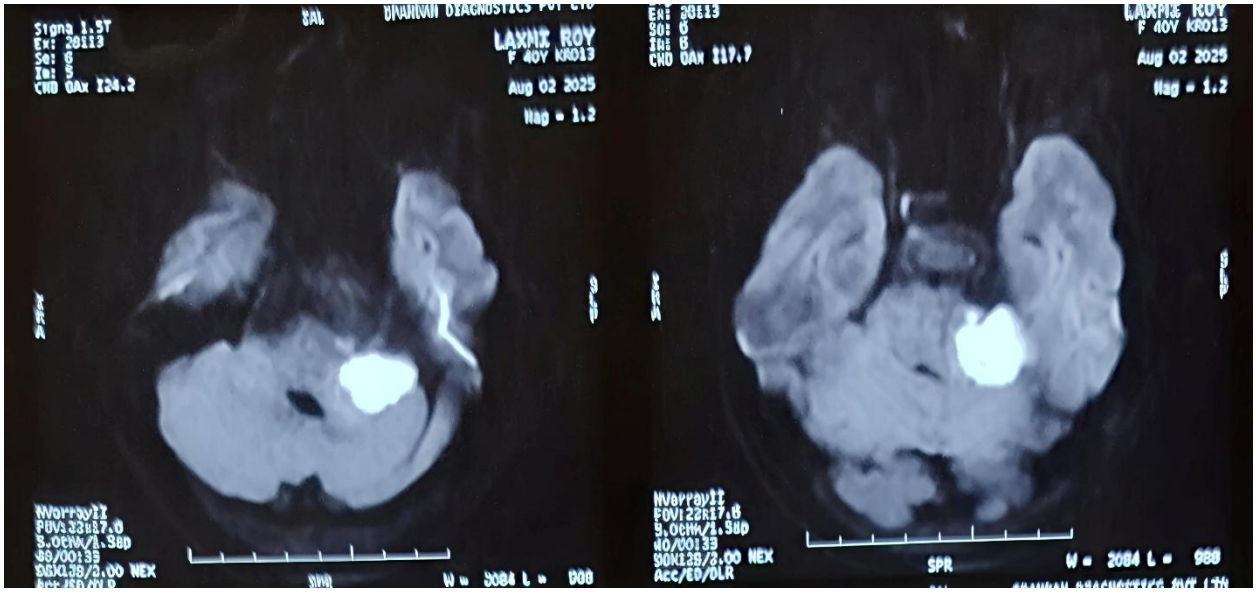

Figure 4

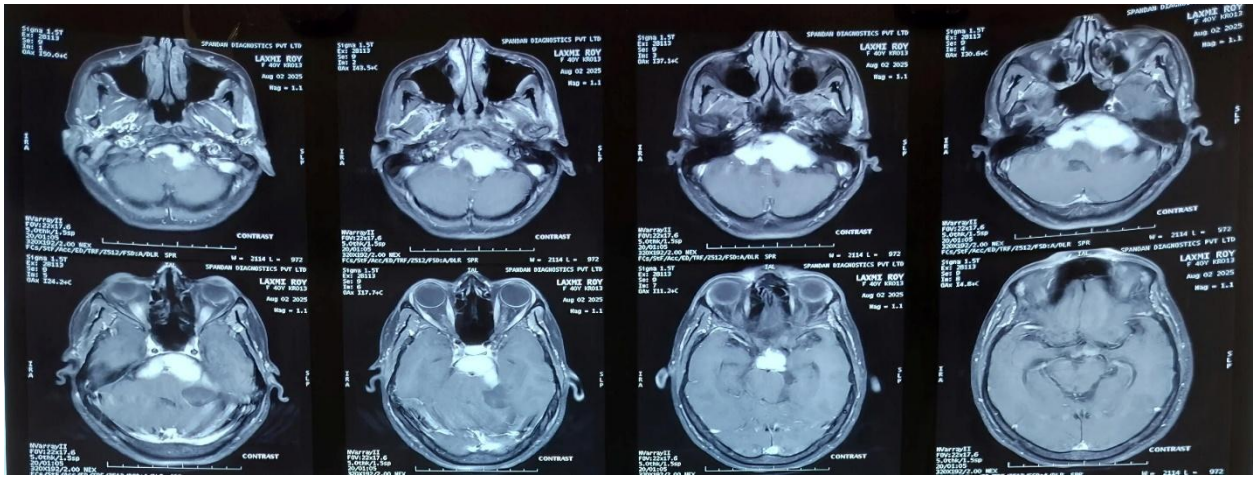

Figure 5: Patient underwent gross total excision of the midline SOL in prepontine cistern and cystic SOL from left cerebellar hemisphere through left retro sigmoid suboccipital craniotomy. Intraoperatively there was dense whitish fluid filling the retro clival cistern found. Whereas a pearly white, flaky material was found on the other lesion. Both the lesions were found to be interconnected with a thin arachnoid covering.

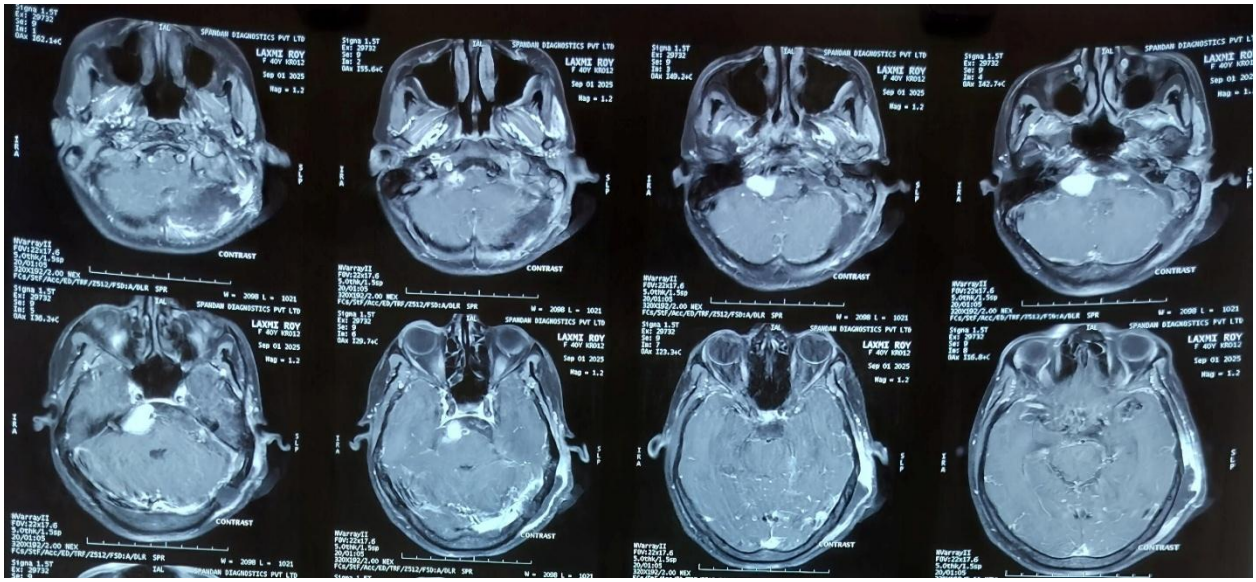

Figure 6: On histopathology both the lesions showed fibrous wall lined by keratinizing squamous epithelium containing degenerated squamous and flaky keratin suggestive of epidermoid cyst.

2. Discussion/ Review of literature

Intracranial epidermoid cysts are rare, benign congenital lesions and account for 0.2%–1.8% of all intracranial tumors (1). Epidermoid cysts are thought to arise from abnormal entrapment of ectodermal elements during neural tube closure, though other hypotheses have been suggested, including traumatic or iatrogenic etiologies (2). Epidermoid cysts can be intradural in location, with cisternal predilection constituting up to 90% of all intracranial epidermoid cysts. The remainder of intracranial epidermoid cysts are extradural, mostly presenting within the skull, and are called "intra-diploic epidermoid cysts" (3). Epidermoid cysts can be asymptomatic, particularly when small, or can have a variety of clinical presentations ranging from acute to chronic symptoms related to increased intracranial pressure and compression of adjacent neurovascular structures (4). Benign epidermoid cystic lesions characteristically demonstrate slow linear growth with a turnover rate similar to normal human skin (2). Therefore, any rapid change in symptoms or imaging findings can indicate hemorrhage into the cyst or malignant transformation.

On CT, epidermoid cysts appear similar in density to cerebrospinal fluid (CSF), typically registering around 0 Hounsfield Units (HU) due to a mix of cellular debris and high cholesterol content. This similarity can cause them to resemble arachnoid cysts. Calcification occurs in a small percentage of cases (10–25%), and on rare occasions, epidermoid cysts may appear hyperdense due to factors like hemorrhage, saponification, or high protein levels (5).

On MRI, epidermoid cysts usually appear hypointense on T1-weighted images and hyperintense on T2-weighted images with restricted diffusion, and they do not show post-contrast enhancement on gadolinium-enhanced images. Epidermoid cysts show a bright signal on diffusion-weighted imaging (DWI) and have apparent diffusion coefficient (ADC) values similar to the surrounding brain tissue (6). Rarely they can be hyperintense on T1, hypointense on T2 without diffusion restriction and are known as "white" epidermoid (7).

Benign epidermoid cysts generally have a favorable prognosis with resection, although they have a high recurrence rate (> 20%) (8). Hasegawa et al. analyzed 63 patients with intracranial ECs and found that up to 25% of patients required multiple resections over a mean follow-up of about 89 months (about 7.5 years) (9). It is important to note, though, that postsurgical adhesions and disorganized disease-brain interfaces in recurrent epidermoid cysts render optimal resection challenging. Radiation therapy use has been shown to decrease recurrence but only in small series. In a study performed by Morshed et al., 8 patients with recurrent epidermoid cysts were treated with radiation therapy and had no disease recurrence at the end of a short median follow-up period of 2.9 years (10). Nonetheless, while not always feasible, neurosurgeons attempt to utilize every available resource to ensure the most extensive yet safe resection during the initial surgery (11).

Differential Diagnosis
Neuro enteric cyst

- On CT, they vary in density ranging from fluid-like density similar to CSF, to iso-dense to brain parenchyma to hyperdense
- MRI is the modality of choice, and appearances depend on the variable protein content.

T1: variable, ranging from isointense to CSF to very high T1 signal (white)
T2: variable, ranging from isointense to CSF to very low T2 signal (black)
FLAIR: T2 high signal lesions do not suppress on FLAIR
DWI/ADC: ranging from mild diffusion restriction to facilitated diffusion (12).
Dermoid cyst

- On CT typically they appear as well-defined low attenuating (fat density) lobulated masses. Very rarely they demonstrate hyper density, thought to be due to a combination of saponification, microcalcification and blood products.
- On MRI,

T1: typically, hyperintense (due to cholesterol components)
T2: variable, signal ranging from hypo- to hyperintense
T1 C+ (Gd): generally, do not enhance (13).

Radiological dilemma
Due to the retro clival location and MRI findings radiologically the midline SOL was thought to be a neuro-enteric cyst or dermoid cyst whereas the lesion anterolateral to left cerebellum was thought to be an epidermoid cyst.
But intraoperative and histological findings confirmed the diagnosis of epidermoid cyst for both the lesions and proved it to be a single lesion.

3. Conclusion

This case highlights the diagnostic dilemma posed by an unusual retro clival epidermoid indicating, single epidermoid cysts may appear partly

- Hyperdense on CT due to factors like saponification, high protein levels or hemorrhage (5). Which can be hyperintense on T1 due to high protein or lipid content (mixed triglycerides and no cholesterol) and in rare instances of hemorrhage into the cyst (7). [white epidermoid]
- Hypodense on CT, hypointense on T1 and hyperintense on T2 with diffusion restriction (6).
  [Typical black epidermoid]
  Due to its different stages of pathogenesis.

List of Abbreviations
1) MRI: Magnetic Resonance Imaging
2) HOD: Head of the Department
3) NRSMCH: Nil Ratan Sircar Medical college & Hospital
4) Dept: Department
5) OT: Operation Theatre
6) ITU: Intensive Therapy Unit
7) CT: Computed Tomography scan
8) P + C: Plain + Contrast images
9) Fig: Figure
10) SOL: Space Occupying Lesion
11) HPE: Histopathological Examination
12) WHO: World Health Organisation

13) CSF: Cerebro Spinal Fluid
14) MRS: Magnetic Resonance Spectroscopy
15) CECT: Contrast Enhanced Computed Tomography scan
16) ADC: apparent diffusion coefficient
17) DWI: diffusion-weighted imaging

Declarations

1) Ethics approval and consent to participate: This is a case report of a patient already admitted under us for her treatment (implied consent). However, every part of the treatment, including all imaging and of course surgery were done after proper counselling and written informed risk consent from the patient's relatives.
2) Consent for publication: Due written informed consent was taken from patient's relatives regarding case report publication.
3) Availability of data and material: All necessary data related to the patient and her treatment has been procured from her case files and her MRI Brain images. The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.
4) Conflict of Interests: None. The authors declare that they have no competing interests.
5) Funding: No separate funding was necessary as the patient underwent his treatment in a government hospital where the treatment is free of cost.
6) Authors' contribution:
   - AG was the principal investigator and was involved in Data collection, literature review, analysis and interpretation of the clinical course of the patient and writing the manuscript.
   - DS was involved in the patient treatment and Data collection.
   - KS provided Treatment guidance and also contributed to literature review.
   - KR was the Treating neurosurgeon under whom the patient was admitted. He planned and executed the surgery and provided insight regarding the rarity of the clinical presentation.
   - All authors read and approved the final manuscript.
7) Acknowledgements:
   - Prof. Dr. Kaushik Roy, HOD, Dept of Neurosurgery, NRSMCH
   - Spandan MRI Centre and Dept of Radiology, NRSMCH
   - Sisters of ward, Neuro OT and Neuro ITU.

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