RESEARCH PAPER

A Neuro-Oncologic Surprise: Malignant Transformation of a Cerebellopontine Angle Epidermoid Cyst—Case Report and Management Review

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ABSTRACT

Background

Intracranial epidermoid cysts are rare intracranial tumors. Malignant transformation to squamous cell carcinoma (SCC) is well documented but uncommon. We present a case of recurrent cerebellopontine angle (CPA) epidermoid undergoing malignant transformation after two prior resections.

Case Report

A 42 year old male presented in 2014 with the history of headache with occasional vomiting for 1 year, imbalance while walking for 2 months. On examination, right sided cerebellar signs were present. Fundus examination showed features of early papilledema. Magnetic Resonance Imaging (MRI) Brain (plain and contrast) showed features suggestive of epidermoid tumour with moderate hydrocephalus. Right paramedian, suboccipital craniotomy radical excision of epidermoid was done. On histopathological examination (HPE), a keratinous cyst of epidermal type was seen. He made a steady recovery. In 2024, he presented with right side occipital headache with vertigo for 1 month. On examination, right sided cerebellar signs were present. MRI brain (plain with contrast) showed recurrence of right CP angle lesion. Re-craniotomy and excision of solid cystic lesion. HPE showed features of epidermoid cyst. He got symptomatically better and was discharged. He was on regular followup. In 2025, he presented with multiple episodes of non-projectile vomiting, mild headache and generalised tiredness. MRI brain revealed a multilobulated T2/FLAIR hyperintense lesion with new solid enhancement in the Cerebello-pontine angle, just deep to the transverse-sigmoid junction. PET CT whole body scan was done which showed no other lesions in the body. The patient was planned for re-exploration and excision of lesion after Institutional Tumour board discussion. Intraoperatively - the tumour was on the transverse sigmoid sinus, endoscopic sub-total excision of lesion was done. Pearly keratinous debris was noted in the cavity with yellowish cystic fluid, reddish coloured firm, moderately vascular lesion over the transverse sigmoid junction. The frozen section suggested malignant etiology. Histopathology-H&E showed features of squamous cell carcinoma. The patient was on follow-up in the OPD and had an unremarkable course. He is scheduled for medical and radiation therapy.

Conclusion

Malignant transformation should be suspected when recurrent epidermoid cysts develop new enhancement or rapid growth. Early detection, maximal safe resection, and adjuvant radiotherapy improve outcomes.

Keywords: Epidermoid cyst, Squamous cell carcinoma, Malignant transformation, Cerebellopontine angle, Neurosurgery

How to cite this article: Gupta J, Soogoor DV, Naidu PB, Balasubramanian A, D'Cruze L. A Neuro-Oncologic Surprise: Malignant Transformation of a Cerebellopontine Angle Epidermoid Cyst—Case Report and Management Review. Int J Drug Deliv Technol. 2026;16(23s): 328-331. DOI: 10.25258/ijddt.16.23s.32

Source of support: Nil.

Conflict of interest: None

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Introduction

Epidermoid cysts are benign congenital inclusion cysts representing about 1% of intracranial tumors [1]. Malignant transformation into squamous cell carcinoma (SCC) is extremely rare, with fewer than 100 cases reported since 1912 [2]. The cerebellopontine angle (CPA) is the most frequent intracranial location. Transformation should be suspected when a previously non-enhancing lesion develops new nodular enhancement or rapid growth. We report a recurrent CPA epidermoid cyst transforming into SCC after two resections spanning 11 years which shows how important radio-neuro - pathological discussions are for diagnosing and managing such a case.

Case Report

A 53-year-old male presented with headache, multiple episodes of vomiting and imbalance while walking for 1 month. At 42 years of age, he had history of headache with occasional vomiting for 1 year and imbalance while walking for 2 months. He was well built and nourished, normal motor and sensory examination, right side dysmetria, dysdiadokinesia and swaying to right side while walking. Fundus examination showed features of early papilledema. Magnetic Resonance Imaging (MR) Brain (plain and contrast) showed a T1 hypo- intense, T2 hyper- intense, DWI uniform restriction, no T1 contrast enhancement, ADC no drop, FLAIR - hypointense- features suggestive of epidermoid tumor (Figure 1- A, B, C, D , E, F) with moderate hydrocephalus.

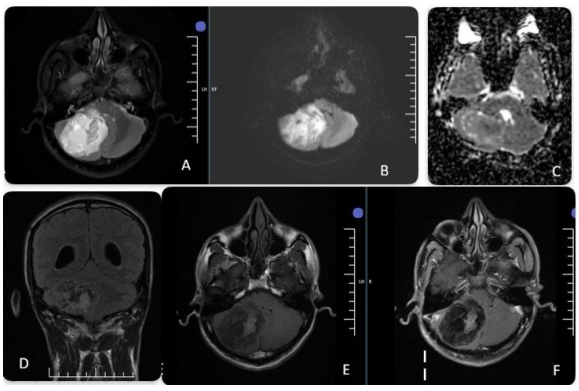

Figure- 1 - MRI brain(plain with contrast) from 2014 showing presence of epidermoid cyst in right CP angle

A- T2 weighted MRI showing heterogenous hyper-intense lesion in CPA

B- Diffusion weighted imaging showing intense restriction

C- ADC map showing hypo- intensity

D- T2w FLAIR MRI coronal showing predominantly hypo-intense lesion

E- T1w MRI showing hypointense lesion

F- T1w post contrast MRI showing no contrast uptake in lesion

He underwent right paramedian suboccipital craniotomy and radical excision of lesion. On histopathology, a keratinous cyst of epidermal type was noted which was friable, grey- white in colour and soft in consistency (Figure 2- A).

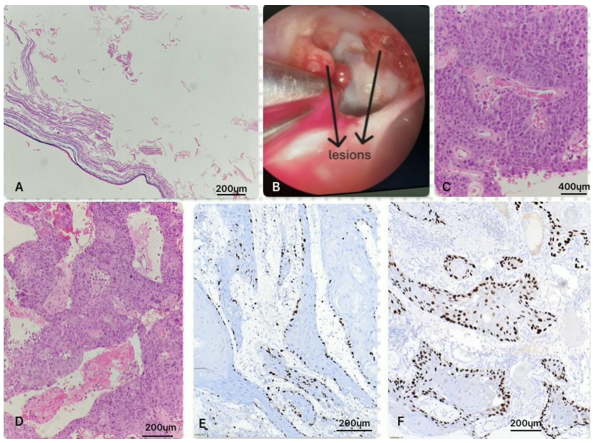

Figure 2 -

A- H&E of 2014 biopsy at 200x : cyst wall lined by flattened squamous epithelium containing flakes of keratin material

B- Intraoperative endoscopic image showing the tumour- 2024

C-H&E of 2025 biopsy at 400x: showing nests of atypical squamous cells with brisk mitosis

D- H&E of 2025 biopsy at 200x : Nests of tumour cells with presence of keratin pearls and focal necrosis surrounding blood vessels

E- IHC for Ki-67 shows high proliferative index

F- IHC for p40 shows nuclear positivity in the tumour cells confirming squamous origin

He made a steady recovery. In 2024, he presented with right sub- occipital headache with vertigo for 1 month. On examination, he had swaying to the right, right sided nystagmus and right dysmetria, MRI Brain and CT brain - right cerebellar multicystic lesion with small solid component with mass effect. Patient underwent re-exploration and excision of lesion. HPE showed features of epidermoid cyst. He got symptomatically better and was discharged. He was on regular followup in OPD, had no new complaints. In 2025, he presented to OPD with multiple episodes of non- projectile vomiting, mild headache and generalised tiredness. MRI revealed a multilobulated T2/FLAIR hyperintense lesion with new solid enhancement in the

Cerbello-pontine angle, just deep to the transverse-sigmoid junction (Figure 3-A,B,C,D,E,F).

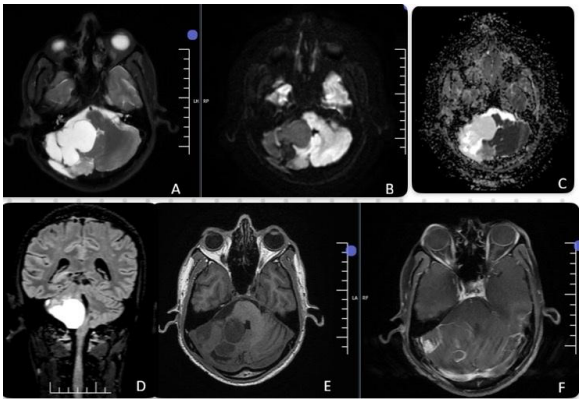

Figure 3- MRI brain(plain with contrast)- 2025 showing enhancing nodule in CP angle.
A-T2 weighted MRI showing cystic hyper-intense lesion in CPA
B- Dwi MRI showing no restriction
C- ADC showing hyper-intense lesion
D- T2w FLAIR coronal MRI showing Hyper-intense lesion
E- T1w MRI showing hypo-intense lesion
F- T1w Post - contrast MRI showing intensely enhancing lesion in the CPA

PET CT whole body scan was done which showed no other lesions in the body. The case was discussed in the Institutional Tumour board and consensus was given to go ahead with surgery. The patient was planned for re-exploration and excision of lesion. Intraoperatively, tumour was on the transverse sigmoid sinus, endoscopic sub-total excision of lesion was done. Intraoperatively pearly keratinous debris was noted in the cavity with yellowish cystic fluid, reddish coloured firm, moderately vascular lesion over the transverse sigmoid junction (Figure 2- B). The frozen section suggested malignant etiology.

Histopathology

H&E showed atypical squamoid cells with increased nucleo-cytoplasmic ratio, vesicular chromatin with prominent nucleoli, and intercellular bridges. Atypical mitotic features, hyalinised vessel and anucleate keratin material (Figure 2- C, D). Ki- 67 was 8% (Figure 2- E). P-40 is positive (Figure 2- F).

Postoperative Course

The patient's symptoms improved. He was discharged home. On follow-up, his symptoms improved. The case was discussed in tumour board and decision was to plan the patient for adjuvant radiotherapy and chemotherapy.

Patient’s perspective

Patient and attender tell that they were scared after they had heard the diagnosis, especially because they were on regular followup and had been told that the lesion is usually benign but prone to recurrence. They were thankful for our role in the treatment and for explaining everything in clear simple terms.

Discussion

The malignant transformation of an intracranial epidermoid cyst into squamous cell carcinoma (SCC) is an exceptionally rare clinical entity, with an estimated incidence of less than 2% among all epidermoid cysts [3]. In the context of this 53-year-old male, the presentation aligns with Hamlat’s Type 2 classification (Table 1), defined as malignant transformation arising from a remnant cyst following prior partial resection [4].

Type	Description of Origin / Presentation
Type 1	Malignant transformation of an epidermoid cyst
Type 2	Malignant transformation from a remnant epidermoid cyst (e.g., recurrence post-resection)
Type 3	Malignant transformation with leptomeningeal carcinomatosis (spread to the meninges)
Type 4	SCC arising from other benign cysts (e.g., dermoid cysts)
Type 5	Other malignancies (non-SCC) arising in benign cysts

Table 1- Hamlat classification[4]

The ten-year interval between the initial surgery (2014) and the current malignant presentation supports the chronic inflammatory hypothesis, which postulates that repeated surgical interventions and the persistence of cystic remnants induce a metaplastic change in the squamous epithelium, eventually leading to dysplasia and carcinoma [3]. Radiologically, the hallmark of this transformation is the appearance of contrast enhancement. While benign epidermoid cysts typically appear as non-enhancing, diffusion-restricting lesions that mimic cerebrospinal fluid, the development of a solid, enhancing component-as observed in this patient

deep to the transverse-sigmoid junction-is a critical red flag for malignancy [5]. This rapid radiological progression, coupled with the worsening clinical deficits (facial nerve palsy and hearing loss) shortly after the 2024 resection, reflects the aggressive biological behavior of SCC compared to the linear, indolent growth of benign epidermoids.Intraoperatively, the case highlighted the distinct macroscopic picture often described in the literature: the coexistence of "pearly keratinous debris" (typical of the benign precursor) with a "reddish, firm, vascular" mass (representing the malignant clone) [6]. The location of the tumor, deep to the transverse-sigmoid junction, posed a significant surgical challenge. The use of an endoscope to achieve a sub-total excision was a novel technical adaptation, allowing visualization in a corridor inaccessible to the standard microscope. This aligns with modern skull base approaches where endoscopic assistance is utilized to minimize retraction on critical neurovascular structures while addressing "blind spots" in the CPA. Given the sub-total excision and the confirmed malignant etiology, the prognosis is guarded. The literature indicates that adjuvant radiotherapy is the cornerstone of postoperative management for these patients, as it significantly extends median survival compared to surgery alone. While stereotactic radiosurgery (SRS) or fractionated stereotactic radiotherapy (FSRT) are standard recommendations, the overall survival rate remains poor compared to benign lesions, necessitating rigorous surveillance with serial MRI to monitor the residual vascular component [7].

Conclusion

Malignant transformation should be suspected when recurrent epidermoid cysts develop new enhancement or rapid growth. Regular followup, early detection, maximal safe resection, and adjuvant therapy improve outcomes.

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