

Case Report

Malignant squamous cell carcinoma arising in a previously resected cerebellopontine angle epidermoid

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Abstract

Background: Malignant squamous cell differentiation of an epidermoid cyst can carry a significantly poor prognosis and very little is known about this entity.**Case Description:** We present the case of a 35-year-old lady, with primary malignant squamous cell carcinoma (SCC) arising from a previously partially resected cerebellopontine angle epidermoid cyst almost 5 years after initial resection. We also review the relevant literature.**Conclusion:** The imaging findings, histopathology, and management of a malignant SCC arising from a benign epidermoid cyst are discussed with recommendation for increased surveillance and follow-up, even for classically accepted benign lesions.**Key Words:** Cerebellopontine angle tumour, Epidermoid, Squamous cell carcinoma, Malignant transformation**Access this article online****Website:**www.surgicalneurologyint.com**DOI:**

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INTRODUCTION

Epidermoid cysts are benign, slow growing, extra-axial tumors that account for ~1% of all intracranial tumors. Embryologically, they are derived from ectodermal inclusions during neural tube closure.^[6] Intracranial squamous cell carcinoma (SCC) is commonly seen as a metastatic deposit, however, there are case reports of malignant transformation of epidermoid into SCC. We present the case of a 35-year-old female with primary malignant SCC arising in a partially resected cerebellopontine angle (CP angle) epidermoid.

CASE REPORT

The patient originally presented in 2010 with headaches. Neurological exam was unremarkable. Magnetic resonance imaging (MRI) demonstrated a CP angle lesion that was lobulated and predominantly of low

signal intensity in the prepontine and right ambient cistern, with a few small foci of increased signal intensity on diffusion imaging, which was consistent with an epidermoid along with a small left-sided acoustic neuroma. At that time, a right retrosigmoid craniotomy was performed with the intention of decompressing the brainstem and cranial nerves. The right CP angle lesion was partially resected with a small residual left coating the anterior brainstem in

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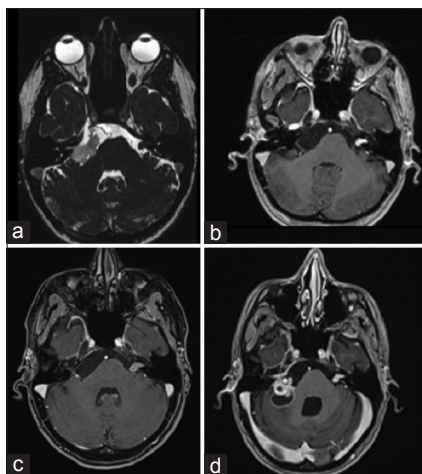


Figure 1: (a) MRI T2 CIS axial demonstrating pre-pontine and ambient cistern lesion in 2010. (b) MRI T1 post gadolinium in 2010 demonstrating no significant enhancement. (c) MRI T1 post gadolinium in 2015 demonstrating no significant enhancement. (d) MRI T1 post gadolinium in 2016 demonstrating lobulated, enhancing mixed cystic/solid lesion

the prepontine cistern. Histology demonstrated an epidermoid tumor with typical histological features of lamellar keratin and benign squamous epithelium. The small residual was monitored with serial MRI scans. Approximately 5 years later, routine follow-up MRI demonstrated marked growth of the residual lesion with development of a multilobulated enhancing cystic component. The patient complained of a 6-month history of right-sided facial numbness. After discussion with the patient, the decision was made to reoperate and resect this lesion. A repeat right retrosigmoid craniotomy was performed. Intraoperatively, the lesion was cystic with an anteromedial solid component adhering to the lateral aspect of the brainstem. There appeared to be a dural attachment which was resected. The patient awoke well initially, however, subsequently developed a House–Brackmann grade 5 facial palsy on day 2 [Figure 1].

Pathological findings

Histology demonstrated SCC with areas of poorly differentiated sarcomatoid elements. The squamous component comprised large epithelial cells with hyperchromatic, pleomorphic nuclei, and abundant, dense eosinophilic cytoplasm. Keratinization and intercellular bridges were present. There were no benign squamous cellular features. An acellular keratin pearl was present. The sarcomatoid component comprised highly atypical spindle cells displaying hyperchromatic, pleomorphic nuclei, and brisk mitotic activity. Areas of necrosis were also present. Immunohistochemistry was performed. The squamous cell component was positive for CK5/6, and had positive nuclear staining for P63. Some of the sarcomatoid spindle cells showed positive staining for cytokeratin markers AE1/3 and CK5/6 and P63. They

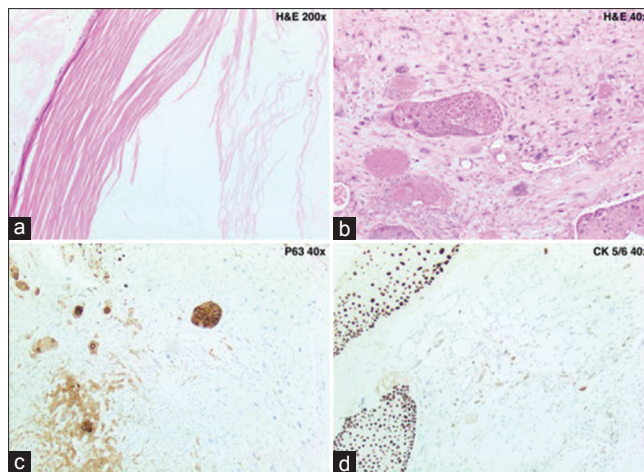


Figure 2: (a) first histology H and E staining demonstrating benign squamous cyst wall and keratin with no malignant features. (b) Second resection histology H and E staining showing metastatic squamous cell carcinoma features. (c) and (d) immunohistochemistry staining demonstrating malignant squamous cell markers of CK 5/6 and positive nuclear staining for P63

were negative for CD34, S100, desmin, GFAP, CD31, and myogenin. The previous histology was reviewed which showed benign squamous cyst wall and keratin. There were no malignant features [Figure 2].

DISCUSSION

Malignant transformation of an epidermoid tumor into SCC is a rare but described phenomenon.^[1,4,5,8-11] By far the most common cause of intracranial SCC is metastasis from elsewhere. True primary SCC is thought to arise from malignant transformation of a dermoid or epidermoid within the central nervous system.^[3,7] Garcia first described this in 1981 and proposed a set of criteria for the diagnosis of primary squamous cell carcinoma; Tumor restricted to the intracranial compartment, no invasion or extension beyond the dura or cranial bones, no communication with the middle ear, air sinuses or sella, no evidence of nasopharyngeal tumor.^[3] Hamlat *et al.* added to this in 2005, proposing that there also be benign squamous epithelium within the main tumor mass and no evidence of a primary tumor elsewhere.^[4] Prognosis has been considered poor across the literature with a median survival of 9 months, although this has been increased to 26 months in some cases with radiotherapy.^[2] Any rapid growth of a lesion in the site of a previously resected epidermoid or dermoid or change in the MRI characteristics of any residual should be considered suspicious and prompt either operative intervention to gain tissue for analysis or a more frequent surveillance schedule.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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