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Squamous Carcinoma Arising in a Cerebellopontine Angle Epidermoid: CT and MR Findings

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Intracranial epidermoid tumors are rare lesions, which reportedly account for less than 1% of intracranial tumors that arise as a result of the inclusion of squamous epithelium during embryogenesis [1]. This report describes the CT and MR findings of the rarer occurrence of a squamous cell carcinoma arising in a cerebellopontine angle epidermoid and extending into the cerebellum.

Case Report

A 74-year-old man presented at another outside institution with vertigo, nausea, and vomiting. A CT examination of the head was reported to show a low-density, roughly spherical mass in the region of the left cerebellopontine angle. There was some peripheral high density, suggestive of an irregular, calcified component. MR confirmed the extraaxial location of the lesion and it was thought to be consistent with an epidermoid. On angiography, the lesion was avascular. A left suboccipital craniotomy revealed a pearly white lesion, typical of a benign epidermoid adherent to the arachnoid of the left cerebellar hemisphere. The aspirated fluid was yellowish and oily. No intraaxial extension was present. Pathologically, a benign epidermoid cyst with pseudostratified squamous epithelium was diagnosed.

The patient did well until 13 months later, when he experienced headaches, ataxia, confusion, nausea, and vomiting. Repeat CT examination revealed a 3-cm recurrent hypodense lesion with an irregular rim of enhancement. There was resultant obstructive hydrocephalus. A ventriculostomy was performed, at which time an extraaxial cystic lesion extending inferiorly to the left cerebellar tonsil was noted. Because of the adherent nature of the cyst wall, it could not be totally removed.

Pathologic inspection revealed a moderately differentiated squamous cell carcinoma consisting of cells with large hyperchromatic nuclei, nuclear folding, rare mitoses, and occasional squamous pearls (Fig. 1A). In some areas a transition between dysplastic squamous epithelium lining a cyst wall and overt carcinoma was apparent.

Two months later, the patient was admitted to our institution with ataxia, nausea, and vomiting. Contrast-enhanced CT scans (GE 9800 Quick, Milwaukee) revealed an enhancing lesion with irregular borders and postoperative changes (Figs. 1B and 1C). The mass displaced the fourth ventricle to the right. The center of the mass did not

enhance. T1-weighted MR examination (600/20/1) (Signa, 1.5 T, General Electric, Milwaukee) revealed evidence of prior surgery and a hypointense mass in the posterior fossa. The T2-weighted (2250/80/1) images showed extensive edema and a cystic component. Gadopentetate-dimeglumine-enhanced T1-weighted (600/20/1) images outlined a heterogeneously enhancing mass with irregular, crumpled margins (Figs. 1D–1F).

A third debulking was performed. Pathologic examination of the tissue removed again showed poorly differentiated squamous carcinoma with markedly anaplastic cells and many mitotic figures (Fig. 1G). Radiation therapy was begun; however, the patient died 7 weeks after admission. A postmortem examination, limited to the posterior fossa, revealed an extensive tumor infiltrating the left cerebellum and extending across the midline.

Discussion

This case represents the unusual occurrence of malignant transformation in an epidermoid [2]. Such changes have been reported at postmortem examinations [3]. In 1960 Davidson and Small [4] described a case in which malignancy was diagnosed in a living patient. Nasaki et al. [5] described such a tumor visualized by CT. In our case the lesion was identified on both CT and MR.

Of the cases previously reported, 13 were male patients and seven were female, a similar male predominance to that seen in benign epidermoid [6]. The age peak for malignant transformation is also similar to that for epidermoid tumor occurrence [7]. The 13-month period of apparent malignant transformation in our patient was rather short: the longest reported period between epidermoid resection and detection of malignancy is 33 years [2]. Malignant tumor location is most common in the parapontine areas (five cases), parapituitary/basal (four cases), frontal (four cases), cerebellopontine angle (four cases), fourth ventricle (two cases), sella turcica (one case), and parietooccipital region (one case) [7]. Because of the central location of the tumor, obstructive hydrocephalus is a frequent problem. The complex pattern of tumor growth and the insinuation in and around neurovascular structures make total resection difficult [8].

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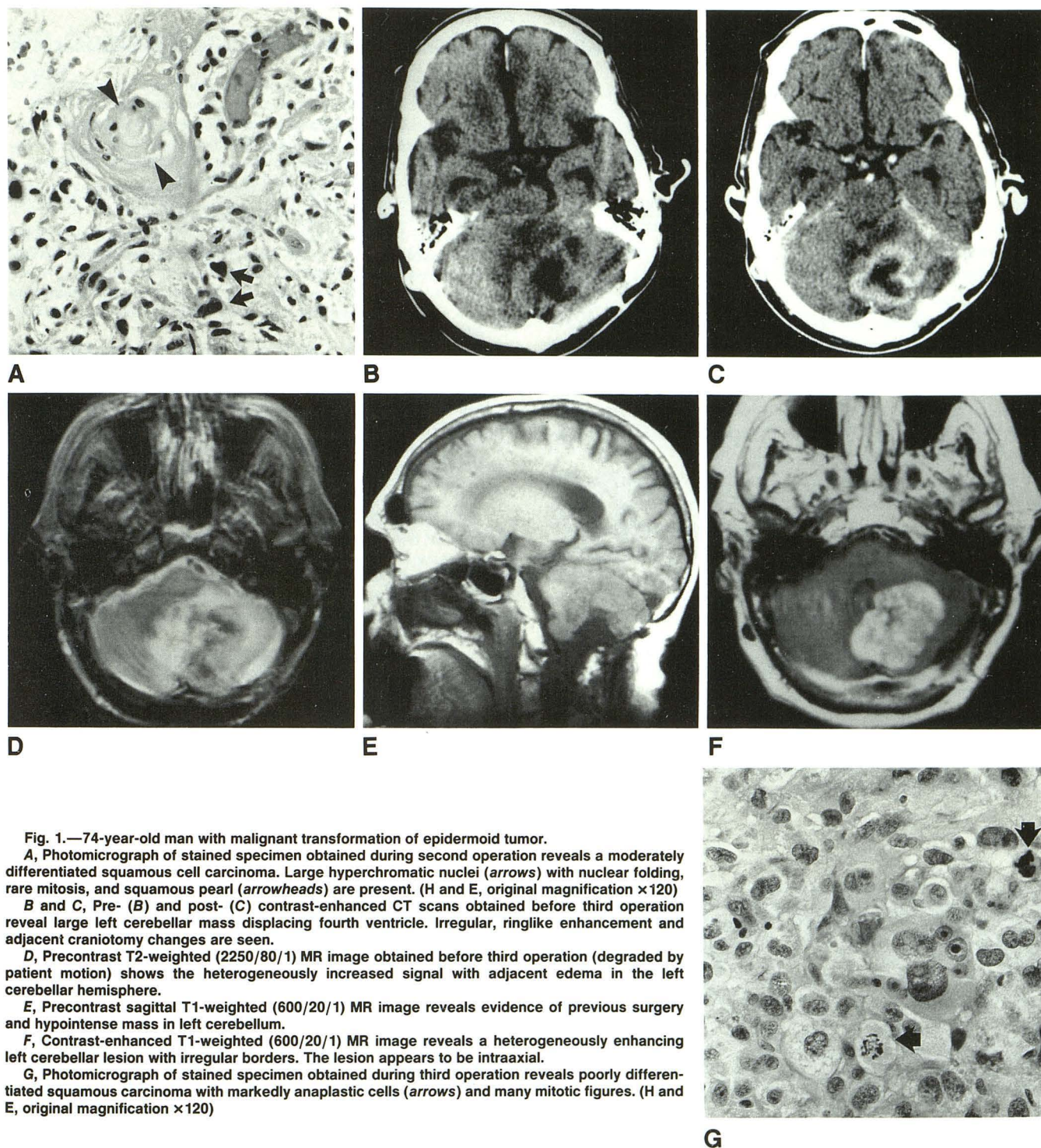


Fig. 1.—74-year-old man with malignant transformation of epidermoid tumor.

A, Photomicrograph of stained specimen obtained during second operation reveals a moderately differentiated squamous cell carcinoma. Large hyperchromatic nuclei (arrows) with nuclear folding, rare mitosis, and squamous pearl (arrowheads) are present. (H and E, original magnification $\times 120$)
 B and C, Pre- (B) and post- (C) contrast-enhanced CT scans obtained before third operation reveal large left cerebellar mass displacing fourth ventricle. Irregular, ringlike enhancement and adjacent craniotomy changes are seen.

D, Precontrast T2-weighted (2250/80/1) MR image obtained before third operation (degraded by patient motion) shows the heterogeneously increased signal with adjacent edema in the left cerebellar hemisphere.

E, Precontrast sagittal T1-weighted (600/20/1) MR image reveals evidence of previous surgery and hypointense mass in left cerebellum.

F, Contrast-enhanced T1-weighted (600/20/1) MR image reveals a heterogeneously enhancing left cerebellar lesion with irregular borders. The lesion appears to be intraaxial.

G, Photomicrograph of stained specimen obtained during third operation reveals poorly differentiated squamous carcinoma with markedly anaplastic cells (arrows) and many mitotic figures. (H and E, original magnification $\times 120$)

The clinical outcome in patients with intracranial squamous carcinoma degeneration is bleaker than that in cases of benign epidermoids. Only one patient has been reported to have lived 3 years after documentation of malignancy [2]. This differs from the generally good prognosis for benign epidermoids

reported by Yamakawa et al. [6]. Although 24% of their patients had recurrence, the average interval was almost nine years. If a second occurrence was discovered, the average interval was another 12½ years. The 20-year survival rate was approximately 93%.

Postcontrast CT examination just prior to the discovery of carcinoma in our case displayed ring enhancement, a rare occurrence in benign epidermoids [9]. Enhancement in pathologically benign tumors has been attributed to peritumoral granulation [10]. However, Lewis et al. [7] correlated more irregular CT enhancement with malignancy. In our case, the irregular nodular rim enhancement represented an area of malignancy.

MR appearance of benign epidermoids has been well documented [9, 11, 12]. The generally accepted pattern of low signal intensity on T1-weighted images and high intensity on T2-weighted images was investigated by Horowitz et al. [9], who described two subtypes of epidermoids: black and white, with low intensity on T1-weighted scans and high intensity on T2-weighted scans, respectively. Black epidermoids were found to have lower lipid content. The enhancement seen on MR images is due to the breakdown in the blood-brain barrier like that seen on CT with iodinated contrast infusion [13]. Finally, because of its multiplanar capability, MR is better able than CT to define tumor extent [14].

Epidermoid tumors are uncommon intracranial lesions and secondary carcinomatous transformation is unusual. Malignant change must be considered when a recurrent lesion shows intense enhancement, either on postcontrast CT or MR examination.

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