

# Imaging Findings of an Epidermoid Cyst with Malignant Transformation to Squamous Cell Carcinoma

## *Skuamöz Hücreli Karsinomaya Malign Dönüşüm Gösteren Bir Epidermoid Kistin Görüntüleme Özellikleri*

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### ABSTRACT

We report imaging findings of a 64-year-old male patient with a ruptured epidermoid cyst (EC) known to be constant over the 23-year follow-up and showing malignant transformation to squamous cell carcinoma (SCC). Computed tomography (CT) and magnetic resonance imaging (MRI) findings including diffusion weighted imaging (DWI), 1H+MR spectroscopy (MRS), dynamic susceptibility contrast perfusion (DSC) MRI of EC, and its rare complications are presented together with a review of the literature. Fluid-low-attenuated-inversion-recovery (FLAIR) and T1-weighted images with gadolinium are the best sequences together with DWI to show the relationship of the EC, the SCC and the border between. Primary brain SCC enhances mostly ring-like or peripherally, but diffuse enhancement is also possible. To our knowledge, no MRS and DSC findings have been reported in the literature yet.

**KEYWORDS:** Epidermoid cyst, Malignant transformation, Intracranial squamous cell carcinoma, Magnetic resonance imaging

### ÖZ

Makalede 64 yaşında bir erkek hastada, 23 yıldır stabil olarak takip edilen bir epidermoid kistin (EK) rüptürü ve zemininde gelişen skuamöz hücreli karsinomaya (SHK) malign dönüşümün görüntüleme özellikleri tarif edilmektedir. Bu nadir klinik durumun bilgisayarlı tomografi (BT) ve diffüzyon ağırlık görüntüleme (DAG), 1H+ MR spektroskopisi (MRS) ve dinamik manyetik duyarlılık kontrast perfüzyon (DMDKP) MR gibi ileri manyetik rezonans görüntüleme (MRG) bulgularının yanı sıra EK ve onun nadir komplikasyonları da literatür bilgisi dahilinde tarif edilmiştir. FLAIR ve gadolinyumlu kontrastlı T1 ağırlıklı görüntüler, DAG ile beraber birleştirildiğinde EK, SHK ve aralarındaki sınır ve ilişkiyi göstermede en iyi sekanslardır. Primer beyin SHK sıklıkla periferik veya yüzük tarzı kontrastlanma gösterir, ancak diffüz kontrastlanma da mümkündür. Bildiğimiz kadarıyla MRS ve DMDKP bulguları şimdiye kadar henüz literatürde tanımlanmamıştır.

**ANAHTAR SÖZCÜKLER:** Epidermoid kist, Malign dönüşüm, İntrakranial skuamöz hücreli karsinom, Manyetik rezonans görüntüleme

### INTRODUCTION

Epidermoid cysts (EC) are one of the most common benign intracranial cystic lesions-observed in daily practice. Current imaging technology makes it easy to recognize them and the radiologic diagnosis is almost always possible (14). Squamous cell carcinoma (SCC) of the brain is rare and mostly found secondary to metastasis of a primary tumor in the body or direct spread of a head or neck tumor via neural foramina (7,9,12). Reported cases of primary intracranial SCC developing from an EC in the literature are fewer than 60, most of which were written upon clinical and pathologic findings with poor imaging content (12). We report imaging findings of a ruptured EC known to be constant over the 23-year follow-up and showing malignant transformation.

### CASE REPORT

A 64-year-old male patient presented with gradual onset of gait disturbance for the last 3 weeks with no other neurologic complaints. His neurologic examination was normal except the gait disturbance. His history revealed that he was diagnosed with an EC by cranial computed tomography (CT) performed due to a headache in 1990 (Figure 1A). The patient was followed up with control CT scans and received no therapeutic intervention.

Because of his new developing symptom, a new contrast enhanced (CE) CT (Brilliance-64, Philips, Best, Netherlands) examination and magnetic resonance imaging (MRI) (1.5T Achieva, Philips, Best, Netherlands) were obtained. CT showed dilated lateral ventricles and cerebral sulci, due to mild cerebral atrophy. The EC was located in the same area as

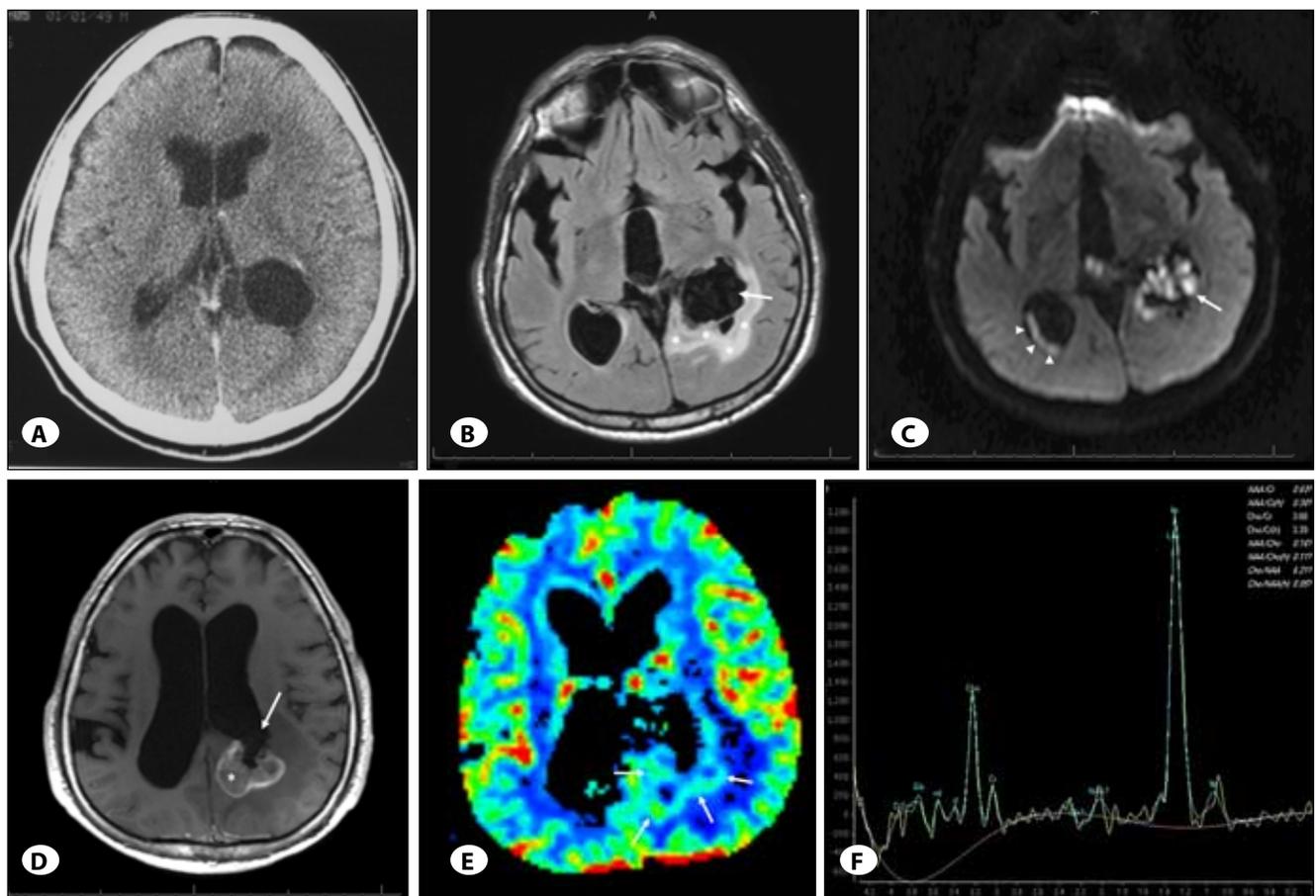
seen on the previous imaging. Its content was isointense with cerebrospinal-fluid (CSF) on T1 (TR:650ms, TE:15ms) and T2-weighted (TR:4000ms, TE:120ms) images, but hyperintense on fluid-attenuated inversion recovery (FLAIR) sequence (TR:6000ms TE:120ms TI:2000ms) (Figure 1B) and the lesion showed diffusion restriction on diffusion weighted imaging (DWI) (single-shot-spin-echo TR:3620ms TE:89ms ETL:89). DWI also showed sediment-like masses with restricted diffusion in the occipital horn of the right lateral ventricle. These suggested that the lesion was EC and ruptured (Figure 1C).

Another solid component with accompanying vasogenic edema in the brain parenchyma was detected just adjacent to the EC. This heterogeneous solid mass was hypointense on T2 and FLAIR and enhanced on post-gadolinium (Gd) T1-weighted images. It also showed diffusion restriction (Figures

1B-D). Figures 1E-F summarizes dynamic susceptibility contrast perfusion (DSC) MRI (Gradient echo TR:633ms TE:30ms) and single voxel  $^1\text{H}^+\text{MR}$  spectroscopy (MRS) (TE:144ms) findings of the solid mass.

Based on the imaging findings, we considered the lesion as a primary intracranial SCC transformed from the EC. For the differential diagnosis, we suggested also high grade primary brain neoplasm or metastasis of unknown origin.

The solid component was surgically removed through the left occipito-parietal approach. On histopathological examination, well-differentiated SCC with moderate atypia and keratinized squamous cells was found. Areas adjacent to the tumor were full of abundant lamellar keratin commonly observed in epidermoid cysts. No benign epithelial fragment was found throughout the whole operation material. The tumor had



**Figure 1:** **A)** Non-enhanced CT scan performed 23 years ago. Hypodense, homogeneous, well-defined cystic lesion is located in the posterior horn of the left lateral ventricle and compressing occipital horn of the left lateral ventricle. MR images were obtained in our institute **B)** In the axial FLAIR, the lateral ventricles are slightly dilated compared to A and there are tiny intensities in the cyst due to the epidermoid cyst content. Also vasogenic edema (\*) is seen in the adjacent brain parenchyma. **C)** DWI shows restriction in the epidermoid cyst (arrow) and some sediment-like masses with diffusion restrictions (arrowheads) were found inside the posterior horn of the right lateral ventricle which suggests rupture of the cyst. **D)** Post Gd T1-weighted image shows enhancement in the lesion adjacent to the left ventricle (\*). The peripheral part of the tumor is thick and enhances well. **E)** DCS perfusion MRI. On the CBV map, the mass (arrows) shows increased CBV compared to normal white matter, and decreased CBV compared to cortical grey matter. **F)** Single voxel  $^1\text{H}^+\text{MR}$  spectroscopy(MRS) showed high lactate and lipid peaks with increased ratio of Cho/NAA(h: 8.85), Cho/Cr(h: 3.4) and decreased NAA/Cr(h:0.38).

an infiltrative feature through the brain parenchyma unlike metastatic carcinomas which had the character of well-defined borders with the adjacent parenchyma (Figures 2A, B). This finding supported the diagnosis of primary intracranial SCC, which was consistent with the imaging findings.

An ear-nose-throat evaluation, a full dermatologic examination and a whole body imaging of PET/CT scan were performed to exclude a SCC metastasis. All were negative regarding any extracranial foci of SCC. The patient was diagnosed as primary intracranial SCC based on the radiologic and pathologic evidences, according to Garcia et al. (7) and Hamlat's et al. (9) criteria (Table I).

First-month follow-up MRI after surgery showed no residual tumor (Figure 3A). Unfortunately, the third-month follow-up MRI showed a recurrent mass at the surgical site (Figure 3B).

### DISCUSSION

ECs are believed to arise from ectodermal squamous epithelial cell remnants. EC walls are lined with keratinizing stratified squamous epithelial cells. Desquamation occurs in time and ECs become enlarged with concentric portions of keratin, water and cholesterol (9,13,14). Usually, they are followed without any intervention. Surgery is needed only if the patient has symptoms due to mass effect (3, 5, 6, 8).

The underlying mechanism causing malignant transformation is not well known, but chronic inflammation with cyst rupture, subtotal resection of the cyst may be potential factors (9). Our case supports the first mechanism. In most of the reported cases, there was a history of previous surgery, ECs were subtotally resected and SCC developed from the residual lesion (3,11,12).

In our case, a heterogeneous solid lesion next to the EC, with strong contrast enhancement, invaded the adjacent parenchyma, causing vasogenic edema, and CBV increased on the DSC MRI. These were supportive features of a high-grade brain tumor with mass effect as well. MRS showed increased lactate peak as a sign of anaerobic metabolism observed in infarct, inflammation or high grade tumors, with increased Cho/NAA ratio as a sign of increased cellular membrane turnover generally seen with neoplasms. Cho/Cr ratio increased to 3.4, although not specific, this rate can also be seen in a neck SCC or in a high grade brain neoplasm (1, 4).

In the literature, the ECs are usually hypodense cyst-like masses and the SCC is usually an enhancing invasive tumor attached to EC. Rough calcification was seen in three of the cases (2,10,15) On MRI, the lesions were mostly heterogeneous, hypointense on T2-weighted images due to high cellularity, and some hyperintensity was observed

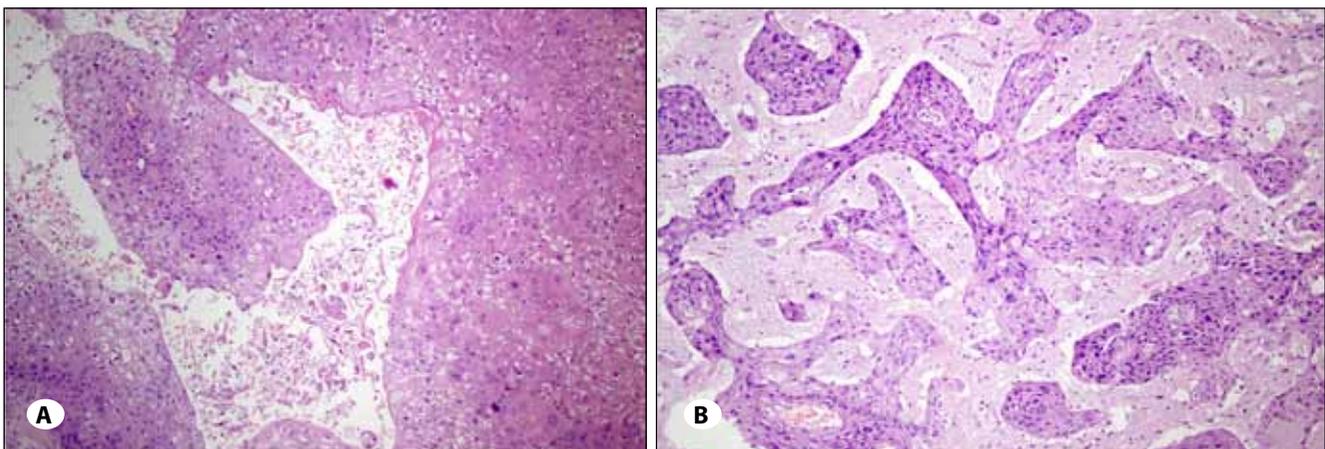
**Table I:** The Criteria for Diagnosing Primary Intracranial Squamous Cell Carcinoma

#### The criteria by Garcia et al. (7)

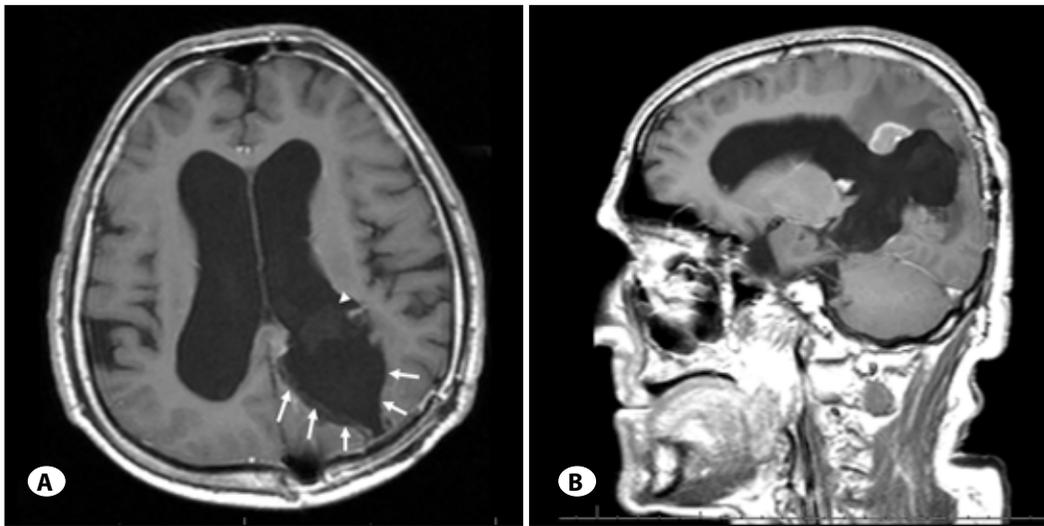
- 1) The tumor must be restricted to the intracranial intradural compartment
- 2) There must be neither invasion nor extension of the tumor beyond the dura or cranial bones through cranial orifices
- 3) The tumor must have no communication with the middle ear, air sinuses or sella turcica
- 4) There must be no evidence of nasopharyngeal tumor

#### Additional criteria by Hamlat et al. (9)

- 1) Benign squamous epithelium must be present within the main tumor mass
- 2) There must be no evidence of a primary tumor elsewhere



**Figure 2:** Microscopic evaluation of the resected specimen. **A)** Large fragments of keratin are seen around the well-differentiated squamous cell carcinoma. That may be compatible with the content of an epidermoid cyst. **B)** The tumor shows a quality of a pattern that is highly infiltrative to the adjacent parenchyma unlike metastatic carcinomas which has the character of well-defined borders with the adjacent parenchyma.



**Figure 3: A)** The first-month follow-up after surgery shows no residual tumor on contrast enhanced (CE) T1-weighted axial image. Arrows show surgical site. The arrowhead points residual epidermoid cyst. **B)** The third-month follow-up MRI shows a recurrent mass in the surgical site on CE T1-weighted sagittal image. Therefore, the patient was sent to radiotherapy.

probably due to necrosis. Providing support to this finding, restricted and increased diffusion areas were seen in the SCC part of the tumor on DWI in our case. FLAIR and T1-weighted images with Gd are the best sequences together with DWI to diagnose EC and SCC and to reveal the relationship between them. Tumors enhance mostly ring-like or peripherally but diffuse enhancement is also possible (2,15). To our knowledge, no MRS and DSC findings have been reported in the literature yet.

### CONCLUSION

Primary brain SCC is rare and may develop from an underlying EC. It can be diagnosed correctly by MRI and DWI with respect to clinical history. MRS and DSC MRI may support the diagnosis.

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