INTRODUCTION

Intracranial epidermoid cysts represent approximately 1% of all brain tumors and 7% of cerebellopontine angle (CPA) tumors, and generally regarded as slow growing brain tumors. Intracranial squamous cell carcinoma is a rare complication of epidermoid cysts. The clinical and radiological course of malignant transformation is more aggressive than epidermoid cysts, and their prognosis remains quite poor. Treatment of such malignancies can be problematic, particularly if there is extensive brainstem involvement. We report a case of the CPA epidermoid tumor that transformed into a squamous cell carcinoma. The imaging and clinical features of this case focus attention on the efficacy of radiosurgery in the management of this unusual malignancy.

CASE REPORT

A 43-year-old man visited our clinics in February of 2009 due to the weakness of the right facial expression (House-Brackmann grade III). T2-weighted magnetic resonance imaging (MRI) showed a high signal intensity (SI) in the CPA (Fig. 1A). T1-weighted gadolinium enhanced MRI showed a nonenhancing tumor in the right CPA, sized 58×38 mm (Fig. 1B). Diffusion-weighted (DW) MRI revealed a high signal intensity lesion in the right CPA (Fig. 1C). A subtotal resection was performed via right retrosigmoid suboccipital approach. Histopathological findings were consistent with an epidermoid tumor. Five months later, the patient underwent gamma knife radiosurgery due to highly probable recurrent epidermoid tumor. Two years after, the patient’s neurological deficit had been newly developed, and follow-up magnetic resonance imaging demonstrated a large contrast-enhancing tumor in the left cerebellopontine angle, which compressed the brainstem. After resection of the tumor, histopathological examinations revealed a squamous cell carcinoma probably arising from an underlying epidermoid cyst. We report a case of an epidermoid tumor in the cerebellopontine angle that transformed into a squamous cell carcinoma.

Key Words: Epidermoid cyst · Malignancy.
Malignant Transformation of an Epidermoid Cyst

KH Chon, et al.

Epidermoid cysts are benign and slow growing lesions representing about 1% of all intracranial tumors. The CPA is the most common site of occurrence of intracranial epidermoid tumors, accounting for 7% of tumors in this region. Grossly, epidermoid tumors are typically well defined lesions with an irregular nodular outer surface and a shiny “mother of pearl” appearance. Microscopically, the wall of the epidermoid cyst consists of a layer of stratified squamous epithelium without vascularity. The cyst content is derived from desquamated epithelial cells composed mainly of keratin in concentric layers and cholesterol in a solid inclusion of ectodermal epithelial elements.

An epidermoid tumor is a congenital lesion that arises from...
crystalline state\textsuperscript{6}. Five months later, follow-up brain MRI showed focal enhanced mass on the right cerebellar hemisphere, there is outside operation field in our case. This finding was atypical. The small enhanced tumor was highly suggested malignant transformation. We performed a retrospective review of the initial pathological specimen did not uncover any evidence of squamous cell carcinoma. We recommended reoperation, but patient refused. Kida et al.\textsuperscript{10} reported that GKRS has also been used to treat recurrent benign epidermoids with reported shrinkage of the tumor and resolution of the patient’s symptoms. Patient underwent GKRS. In cases of previously resected epidermoids when new contrast-enhancing lesions are seen, surgical exploration is necessary to document malignant transformation before adjuvant therapy is considered.

Their complication by squamous cell carcinoma is an unusual occurrence. Primary intracranial squamous cell carcinoma shares a rare but well-described association with benign intracranial epidermoid cysts\textsuperscript{7,9}. These mechanisms of such transformation remain unclear. Malignant change in the epithelium-lining cyst is well documented. An ‘\textit{in situ}’ carcinoma has been observed in an epidermoid cyst\textsuperscript{11,16}. Concerning the mechanisms of malignant transformation of benign epidermoid cysts, some authors have suggested that introduction of a foreign material intraoperatively in combination with the inflammation-inducing contents of the epidermoid cyst, may be a cause of cellular atypia leading to neoplasia\textsuperscript{1,13}. Malignant transformation of intracranial epithelial cysts has been classified into five groups. These five groups include an initial malignant transformation of an epidermoid cyst, malignant transformation from a remnant epidermoid cyst, malignant transformation with leptomeningeal carcinomatosis, squamous cell carcinoma from other benign cysts, and other malignancies arising from the benign cysts\textsuperscript{8}. According to Hamlat’s classification, our case can be classified as malignant transformation from a remnant epidermoid cyst.

On MRI, epidermoid tumors typically have low signal intensity on T1-weighted images, high signal intensity on T2-weighted images that is similar to that of cerebrospinal fluid, and no enhancement on gadolinium-enhanced images\textsuperscript{6,13}. In echo-planar DW imaging, all epidermoid tumors appeared sharply hyperintense relative to the brain and CSF\textsuperscript{13}. DW imaging could differentiate epidermoid tumors from other cystic lesions, in that the apparent diffusion coefficient (ADC) of epidermoid tumors were similar to that of brain tissue whereas the ADCs of cystic lesions were similar to that of CSF\textsuperscript{13}. Fast fluid-attenuated inversion recovery (fast-FLAIR) imaging of epidermoid tumors is featured by the increased signal intensity of surrounding CSF in the subarachnoid cisterns and ventricles\textsuperscript{8}. In previous reports, characteristic MRI findings of malignant transformation of epidermoid cysts include a focal enhancing part within the mass and leptomeningeal metastasis\textsuperscript{7,9}. The diagnosis of malignant transformation is still remained controversial, because of atypical features\textsuperscript{4,5,8}. In the present case, MRI can show an enhanced part with associated vasogenic edema in the cerebellum and pons adjacent to the enhancing nodule. These findings were suggestive of malignant transformation of epidermoid cyst. Therefore, ongoing clinical and neuroimaging follow-up at yearly intervals is necessary for previous subtotal resection of the epidermoid cysts.

The treatment options are controversial. Only short-term follow up was available but the results were encouraging. Patients with subtotal resection should be followed up for the identification of recurrence and malignant transformation. In case suggesting malignant transformation, surgical exploration is necessary to confirm diagnosis. Unfortunately, the complete surgical resection of malignant transformation is usually limited to partial removal because the tumor firmly adheres to the neurovascular structures, as was the case with our case\textsuperscript{7,17}. Surgery alone is not curative and limited. Radiation therapy (external-beam radiation or GKRS) after surgical treatment may offer better control of an intracranial squamous cell carcinoma. Tamura et al.\textsuperscript{17} performed a meta-analysis for the survival of intracranial squamous cell carcinomas arising in epidermoid cysts. They are compared among different treatment modalities (surgery alone, surgery plus external-beam radiation, and surgery plus GKRS)\textsuperscript{17}. They suggested that GKRS appears to offer the best survival benefits on patients with intracranial squamous cell carcinomas\textsuperscript{7,17}. Surgery plus GKRS is recommended for residual squamous cell carcinoma\textsuperscript{4,17}. Most recently, Nagasawa et al.\textsuperscript{14} suggest that when determining the management for malignant transformation of epidermoid tumors, the combination of surgical resection and radiotherapy may be associated with improved short-term survival. Further investigation is necessary to obtain a beneficial treatment strategy against malignant transformation of epidermoid cysts.

CONCLUSION

We report a case of an epidermoid tumor in the CPA that transformed into a squamous cell carcinoma after resective surgery and GKRS. Malignant transformation is a rare cause of enhancement on MR images, as is progressive neurological deficit in a patient with an epidermoid cyst. The presence of contrast enhancement or progressive clinical symptoms within a typical epidermoid cyst along with rapid growth should alert the physician to the possibility of malignant transformation. GKRS has also been used to treat malignant transformation of epidermoids with reported shrinkage of the tumor and resolution of the patient’s symptoms, however, only short-term follow up was available. Further investigation is necessary to obtain a beneficial treatment strategy against malignant transformation of epidermoid cysts.

References

1. Abramson RC, Morawetz RB, Schlitt M : Multiple complications from an intracranial epidermoid cyst : case report and literature review. Neu-
Malignant Transformation of an Epidermoid Cyst | KH Chon, et al.

rosurgery 24 : 574-578, 1989