Squamous Cell Carcinoma Arising in an Intracranial Dermoid Cyst

—Case Report—

Hitoshi TSUGU, Takeo FUKUSHIMA, Shuji HAYASHI,
Mitsutoshi IWAASA, and Toshihiro MATSUDA*

Department of Neurosurgery, School of Medicine, Fukuoka University, Fukuoka;

**Matsuda Neurosurgical Clinic, Fukuoka*

Abstract

A 47-year-old male presented with headache. Magnetic resonance imaging revealed an enhanced mass lesion in the cerebellar vermis and left cerebellar hemisphere and in the cisterna magna. Gross total removal of the tumor was performed. Histological examination demonstrated squamous cell carcinoma in the dermoid cyst. The patient subsequently received localized radiation therapy of total 55 Gy. He has been without tumor recurrence for 6 years since the surgery. We recommend local radiation therapy over 50 Gy following surgery.

Key words: brain tumor, dermoid cyst, malignant transformation, squamous cell carcinoma

Introduction

Intracranial squamous cell carcinoma most frequently occurs as a metastasis from tumors in distant organs or via direct infiltration from the skull base. Primary intracranial squamous cell carcinoma is extremely rare, and usually arises in an epidermoid cyst^{1,2,9,11,14)} and only rarely in a dermoid cyst.^{4,8,12)} We report a case of intracranial squamous cell carcinoma arising in a dermoid cyst.

Case Report

A 47-year-old male visited a neurosurgical clinic complaining of headache that had persisted for 1 month. Computed tomography (CT) revealed an enhanced mass lesion with calcified and cystic components in the cerebellar vermis and left cerebellar hemisphere. One month later, his headache had intensified and was accompanied by vertigo. Repeat CT demonstrated enlargement of the cystic component of the mass and associated obstructive hydrocephalus (Fig. 1).

The patient was referred to the Department of Neurosurgery, Fukuoka University, for surgical treatment. Neurological examination on admission

showed wide-based gait and truncal ataxia. Tests for serum tumor markers, squamous cell carcinoma antigen, carcinoembryonic antigen, alpha-fetoprotein, carbohydrate antigen 125, and neuron-specific eno-

Fig. 1 Computed tomography scan showing a mass lesion with both calcified and cystic components in the cerebellar vermis.

lase, were all negative. Magnetic resonance (MR) imaging showed the tumor as a slightly low signal intensity area with a cystic component on the T₁-weighted images and a heterogeneous intensity area on the T₂-weighted images (Fig. 2B). MR imaging with gadolinium administration revealed a well-enhanced solid component and marginal enhancement of the cyst (Fig. 2A, C). The mass lesion was located in the cisterna magna with continuation to the vermis and the left cerebellar hemisphere (Fig. 2A, C, D). Vertebral angiography showed elevation of the inferior vermian branch of the posterior inferior cerebellar artery (PICA) and posterolateral displacement of the tonsillohemispheric branch of the PICA without tumor staining. These neuroimaging findings suggested that the tumor was situated in the cerebellar vermis and biventral lobule of the left

cerebellar hemisphere, and extended exophytically into the cisterna magna.

Suboccipital craniectomy was performed. Opening of the dura revealed a pearly white/yellowish tumor in the cisterna magna, suggesting an epidermoid cyst. The tumor capsule was opened to expose the cheesy, cartilaginous contents, with a few gray hairs visible within the tumor, which were consistent with a dermoid cyst. The tumor extended from the cisterna magna into the biventral lobule of the left cerebellar hemisphere and the vermis. The tumor was dissected from the brain parenchyma, and grossly totally removed including the cystic wall and the surrounding brain tissue.

Histological examination of the tumor tissue found differentiated stratified squamous cells with keratinization and moderately differentiated stratified squamous mitotic cells and focal necrosis (Fig. 3A–C). Adipose tissue, and collagenous or hyalinized connective tissue were seen between the sweat gland ducts (Fig. 3D). No hair follicles were found in the specimen. Tumor cell nests were found in the brain tissue, suggesting tumor invasion (Fig. 3A). The MIB-1 labeling index was 5.2% in the area of the differentiated stratified squamous epithelium and 10.9% in the area of squamous cell carcinoma. The histological diagnosis was squamous cell carcinoma in the dermoid cyst.

The patient's postoperative course was uneventful, and he was treated with Linac radiotherapy of 55 Gy to the cerebellum. Chemotherapy was not carried out. Extensive investigation including gallium scintigraphy to detect a primary lesion outside the central nervous system was negative. The patient has been well for 6 years since the surgery without evidence of tumor recurrence.

Discussion

Primary intracranial squamous cell carcinoma is rare and usually occurs as a malignant transformation of an epidermoid cyst or a dermoid cyst, as well as *de novo* squamous cell carcinoma.^{1,2,9,11,14} Nineteen of 37 cases of primary ovarian squamous cell carcinoma were associated with dermoid cysts.¹⁵ However, malignant transformation of intracranial dermoid cysts,^{4,8,12} intraspinal dermoid cyst,⁷ or intravertebral dermoid cyst⁶ are more rare.

In our case, the squamous cell component in the dermoid cyst had transformed into squamous cell carcinoma as indicated by the coexistence of a differentiated stratified squamous epithelium and squamous cell carcinoma, the appearance of flaky keratin, sweat gland ducts, and adipose tissue as ex-

Fig. 2 A: Axial T₁-weighted magnetic resonance (MR) image with gadolinium showing intense enhancement of the solid component and cystic wall. B: Axial T₂-weighted MR image showing a heterogeneous intensity mass with cystic components. C: Sagittal T₁-weighted MR image with gadolinium showing the mass in the cisterna magna and the vermis. D: Coronal T₁-weighted MR image showing the mass in the cisterna magna with continuation into the cerebellar vermis and the left hemisphere.

Fig. 3 A: Photomicrograph showing squamous cell carcinoma with horn pearl. HE stain, original magnification $\times 48$. B: Photomicrograph revealing mild atypical epithelium (arrowheads) and moderately carcinomatous tissue (arrows). HE stain, original magnification $\times 100$. C: Photomicrograph of the cyst wall showing proliferation of dysplastic, pleomorphic, and hyperchromatic squamous mitotic cells. HE stain, original magnification $\times 60$. D: Photomicrograph showing sweat gland duct and adipose tissue. HE stain, original magnification $\times 100$.

pected in the benign dermoid cyst, and the absence of primary tumor outside the central nervous system. Cases of primary squamous cell carcinoma have resulted from malignant transformations of intracranial epidermoid cysts.^{3,13} These studies disclosed different stages of differentiation of squamous epithelium, specifically from mature squamous epithelium to carcinoma in the same specimen, which was consistent with our case.

The preferential sites of squamous cell carcinoma correspond to the main sites of both epidermoid and dermoid cysts. The cerebellopontine angle is the most common site for epidermoid cyst, and the midline for dermoid cyst.⁵ These sites also correspond to the preferential sites for intracranial squamous cell carcinoma, suggesting that intracranial squamous cell carcinoma arises from the dedifferentiation of squamous cells in epidermoid and dermoid cysts.

Epidermoid and dermoid cysts usually appear as sharply defined, low density or intensity mass lesions with no contrast enhancement on CT and MR imaging.⁵ However, intracranial squamous cell carcinoma typically appears as an irregularly-marginated mass, with intense enhancement by contrast medium.^{1,3,9,11-13} Therefore, malignant transformation should be considered in cases of epidermoid and dermoid cysts if the mass is predominantly enhanced.

The prognosis for patients with intracranial squamous cell carcinoma is poor. Mean survival time is about 4 months without postsurgical irradiation and 15.5 months with postsurgical irradiation.¹⁰ Patients who survived more than one year had all received a total radiation dose of 50 to 60 Gy.¹¹ Our patient underwent local radiation therapy of 55 Gy following surgery and has been without tumor recurrence for 6 years. Such relatively long survival is extremely

rare even after total removal of the tumor and subsequent radiation therapy. We recommend local radiation therapy over 50 Gy following surgery.

Acknowledgment

We thank Dr. Bernd W. Scheithauer in the Division of Anatomic Pathology, Mayo Clinic (Rochester, Minn., U.S.A.) for his invaluable advice concerning the histological diagnosis.

References

- 1) Acciarri N, Padovani R, Foschini MP, Giulioni M, Finizio FS: Intracranial squamous cell carcinoma arising in an epidermoid cyst. *Br J Neurosurg* 7: 565-569, 1993
- 2) Bayindir C, Balak N, Karasu A: Micro-invasive squamous cell carcinoma arising in a pre-existing intraventricular epidermoid cyst. Case report and literature review. *Acta Neurochir (Wien)* 138: 1008-1012, 1996
- 3) Dubois PJ, Sage M, Luther JS, Burger PC, Heinz ER, Drayer BP: Malignant change in an intracranial epidermoid cyst: Case report. *J Comput Assist Tomogr* 5: 433-435, 1981
- 4) Gluszczyk A: A cancer arising in a dermoid of the brain: A case report. *J Neuropathol Exp Neurol* 21: 383-387, 1962
- 5) Grossman CB: *Intracranial Neoplasms and Cysts: Magnetic Resonance Imaging and Computed Tomography of the Head and Spine*, ed 2. Baltimore, Williams & Wilkins, 1996, pp 217-218
- 6) Ino M, Nagase M, Tsuge K, Kamata M, Udagawa E: Malignant squamous cell carcinoma arising in a lumbar dermoid cyst: A case report. *Int Orthop* 19: 185-186, 1995
- 7) Keogh AJ, Timperley WR: Atypical hidradenoma arising in a dermoid cyst of the spinal canal. *J Pathol* 117: 207-209, 1975
- 8) Kobayashi T, Kuchiwaki H, Inao S, Nakashima N: A squamous cell carcinoma originated from intracranial dermoid cyst. *Neurochirurgia (Stuttg)* 36: 26-29, 1993
- 9) Lewis AJ, Cooper PW, Kassel EE, Schwartz ML: Squamous cell carcinoma arising in a suprasellar epidermoid cyst. *J Neurosurg* 59: 538-541, 1983
- 10) Mori Y, Suzuki Y, Tanasawa T, Yoshida J, Wakabayashi T, Kobayashi T: [A case report of epidermoid carcinoma in the cerebello-pontine angle]. *No Shinkei Geka* 23: 905-909, 1995 (Jpn, with Eng abstract)
- 11) Murase S, Yamakawa H, Ohkuma A, Sumi Y, Kajiwarra M, Takami T, Sakai N: Primary intracranial squamous cell carcinoma: Case report. *Neurol Med Chir (Tokyo)* 39: 49-54, 1999
- 12) Nishio S, Takeshita I, Morioka T, Fukui M: Primary intracranial squamous cell carcinomas: Report of two cases. *Neurosurgery* 37: 329-332, 1995
- 13) Nosaka Y, Nagao S, Tabuchi K, Nishimoto A: Primary intracranial epidermoid carcinoma: Case report. *J Neurosurg* 50: 830-833, 1979
- 14) Ogata N, Jochum W, Aguzzi A, Fournier JY, Yonekawa Y: Total removal of a primary intracranial squamous cell carcinoma invading the brain stem. *Surg Neurol* 46: 477-480, 1996
- 15) Pins MR, Young RH, Daly WJ, Scully RE: Primary squamous cell carcinoma of the ovary. Report of 37 cases. *Am J Surg Pathol* 20: 823-833, 1996

Address reprint requests to: H. Tsugu, M.D., Department of Neurosurgery, School of Medicine, Fukuoka University, 7-45-1 Nanakuma, Jonan-ku, Fukuoka 814-0180, Japan.