Cystic Pineocytoma
—Case Report—

Nobuaki MOMOZAKI, Kiyonobu IKEZAKI, Masamitsu ABE, Masashi FUKUI**, Kiyotaka FUJII and Takashi KISHIKAWA*

Departments of Neurosurgery and *Radiology, Saga Medical School, Saga; **Department of Neurosurgery, Neurological Institute, Kyushu University Faculty of Medicine, Fukuoka

Abstract

Pineocytoma and pineoblastoma, originating from pineal parenchyma, are rare and usually solid. An unusual case of totally cystic pineocytoma in a 37-year-old female is reported. The tumor showed neuronal differentiation and had a good outcome. Prominent calcification associated with pineocytoma and pineoblastoma is a useful finding to differentiate these from benign pineal cysts.

Key words: pineal tumor, pineocytoma, cyst, computed tomography

Introduction

Pineocytoma and pineoblastoma both have incidences of 0.2% or less of all intracranial neoplasms, and originate from pineal parenchymal cells. More common pineal tumors are of germ cell origin. Histopathological and radiological studies show that a cystic tumor in the pineal region is commonly an epidermoid or dermoid tumor. We report a rare case of pineocytoma with a large cyst.

Case Report

This 37-year-old female complained of headache, occasionally accompanied by nausea and vomiting 3 months before admission. After 1 month, she also developed diplopia. Neurological examination found bilateral papilledema and lateral gaze paresis. All laboratory tests including alpha fetoprotein, beta human chorionic gonadotropin, and carcinoembryonic antigen were within normal limits. Precontrast computed tomographic (CT) scans disclosed a pineal cystic mass with marginal calcification and obstructive hydrocephalus. Postcontrast CT scans showed a ring-like enhancement (Fig. 1). Cerebral angiograms showed no tumor stain or compression of the vein of Galen.

The well-defined, grayish cystic tumor was found under the splenium of the corpus callosum via a right occipital transtentorial approach. The tumor was drained of about 5 ml of yellowish, slightly viscous fluid. The collapsed tumor was almost completely removed except for a small part adhered to the tegmentum of the midbrain. The tumor resembled a dermoid cyst intraoperatively.

Histological study of the cyst wall specimens, however, revealed no epithelial component. The tumor consisted of a highly cellular area of medullary proliferation and a less densely cellular area. The tumor cells had small round nuclei and scanty cytoplasm. Large rosettes, with nuclei grouped around a central eosinophilic area, were often observed in the less densely cellular area (Fig. 2). The cyst wall showed spongy degeneration among loosely arranged tumor cells. There were some foci of rounded calcium deposits which often aggregated to form large calcified masses. Immunohistochemical studies revealed tumor cells positive to neuron-
specific enolase (NSE) in the less densely cellular area. Slightly larger, strongly positive cells suggesting neuronal differentiation were also found near the pineocytomatous rosettes (Fig. 3). Tumor cells were negative for S-100 protein and glial fibrillary acidic protein (GFAP). The histological diagnosis was a transitional form between pineoblastoma and pineocytoma with neuronal differentiation.

The postoperative course was uneventful. She received 50-Gy whole brain irradiation. There was no evidence of tumor recurrence or dissemination 5 years later.

Discussion

Russell and Rubinstein\(^{16}\) classified pineal cysts into neoplastic and non-neoplastic cysts. Non-neoplastic small cysts without mass effect are common incidental findings at autopsy. Recently, magnetic resonance (MR) imaging has suggested a relatively high incidence of benign pineal cysts (or glial cysts) which are important to distinguish from other neoplasms.\(^{9}\) On CT scans, pineal cysts are characteristic of dermoid and epidermoid tumors.\(^{4}\) Pineocytoma and pineoblastoma are usually solid and homogeneously enhanced by contrast media.\(^{6}\) Jooma and Kendall,\(^{8}\) however, reported a pineocytoma appearing as a cystic mass with marginal enhancement. Other cases of cystic pineocytoma have also been reported.\(^{12,18}\) Histological studies have demonstrated frequent cystic degeneration in this type of tumor.\(^{10}\) Prominent calcification is associated with pineocytoma and pineoblastoma,\(^{5,18,19}\) although frequently present in pineal teratomas.\(^{2,6}\) Therefore, a cystic appearance with prominent (ring-like) calcification may be, retrospectively, characteristic of pineocytoma and pineoblastoma. Benign pineal cysts usually have no abnormal calcification or ring-like enhancement on CT scans or MR images.\(^{10,11}\)

Neurol Med Chir (Tokyo) 32, March, 1992
The histological study revealed a transitional form between pineoblastoma and pineocytoma, with features of neuronal differentiation. Pineocytoma with neuronal or neuronal and astrocytic differentiation may indicate a relatively benign clinical course. Malignant features such as papillary pattern and fleurettes were not observed. Cystic degeneration and focal bleeding often occur in pineocytoma, so the mechanism of cyst formation was probably such secondary changes.

It is important to identify pineocytoma with specific markers and electron microscope studies. Okeda et al. concluded that NSE was present in both normal pineocytes and tumor cells. S-100- and GFAP-positive cells within the tumor may indicate astrocytic differentiation or pre-existing normal astrocytes. Pineocytoma with neuronal differentiation have crowded clear and/or dense-cored vesicles, microtubules, and microfilaments in the processes.

The effectiveness and indication for radiation therapy of pineocytomas are still controversial. Vaquero et al. did not recommend radiation therapy for pineocytoma with neuronal differentiation because the prognosis is good. Nakatsukasa et al. reported a case of disseminated pineocytoma with a pineoblastoma component in which chemotherapy and radiation therapy were effective. We suggest that radiation therapy is indicated if the tumor contains a pineoblastoma component, and residual tumor or dissemination is suspected. Symptomatic cystic lesions in the pineal region should be managed surgically and precise histological diagnosis is necessary to decide further therapy.

Acknowledgments

We thank Dr. Keith L. Black (UCLA) for his critical reading of this manuscript. We also thank Mr. T. Tanamachi and Mr. Y. Tateishi for their photographic assistance.

References

6) Herrick MK: Pathology of pineal tumors, in Neuwelt EA (ed): Diagnosis and Treatment of Pineal Region Tumors. Baltimore, Williams & Wilkins, 1984, pp 31-60

Address reprint requests to: N. Momozaki, M.D., Department of Neurosurgery, Saga Medical School, 5-1-1 Nabeshima, Saga 849, Japan.

Neurol Med Chir (Tokyo) 32, March, 1992