Pineal Dermoid Cyst Developing 18 Years after Gross Total Removal of a Pineal Mature Teratoma

—Case Report—

Tatsuo MORIMURA, Hiroaki KUBO, Juji TAKEUCHI, and Ba JII

Department of Neurosurgery, National Utano Hospital, Kyoto

Abstract

A 21-year-old male presented with a pineal dermoid cyst manifesting as headache and diplopia. He had undergone gross total removal of a pineal mature teratoma 18 years before and had done well until recently. Diffusion-weighted magnetic resonance imaging showed a region of high signal intensity due to a round mass in the pineal region and extending into the trigone of the right lateral ventricle. Subtotal excision of the tumor was achieved. Histological examination showed an epidermoid cyst consisting of keratinized stratified squamous epithelium and connective tissue. Intraoperative observation had detected black hairs, so the diagnosis was dermoid cyst. The dermoid cyst may have arisen from a microscopic remnant of the cyst wall of the original pineal mature teratoma. The regrowth of a dermoid cyst 18 years after gross total removal of a mature cystic teratoma in the pineal region is exceptional. However, careful follow-up of patients who undergo gross total removal of a pineal teratoma is recommended for a period more than that of the patient’s age at surgery plus 9 months.

Key words: pineal dermoid cyst, diffusion-weighted magnetic resonance imaging, pineal mature teratoma

Introduction

The pineal region is the most common site for the occurrence of intracranial germ cell tumors. The most frequent types are germinomas (55.6%) followed by teratomas (16.3%), but others include gliomas, pineoblastomas, pineocytomas, choriocarcinomas, epidermoids, and dermoids. Intracranial teratomas are rare and comprise only 0.5% of all intracranial tumors. The frequency is higher in Japan, like those of germ cell tumors in general. Intracranial teratomas account for 2.6% of intracranial tumors in Japan in children younger than 14 years of age. There is a male preponderance during the first two decades of life. The prognosis for patients with mature teratomas is good after surgical excision, but malignant transformations of included teratomatous tissue do occur. However, long-term follow-up of these patients after total removal of mature pineal teratomas is very rare.

We describe a case of pineal dermoid cyst developing 18 years after gross total removal of a pineal mature cystic teratoma.16)

Case Report

A 21-year-old right-handed male student presented complaining of headache and diplopia persisting for 3 months. He had suffered from acute intussusception at the age of 7 months. He underwent surgery at age 3 years for a pineal cystic teratoma at a university hospital, after transfer by ambulance when semicomatose with left hemiparesis. Gross total excision was carried out after emergent implantation of a ventriculoperitoneal shunt. Postoperative computed tomography confirmed gross total removal of the tumor. The tumor was fairly large (5 cm in diameter) and the cyst contained a central solid portion, which was pearly white. Histological examination revealed a mature teratoma consisting of epidermis, hair follicles, sebaceous and sweat glands, columnar gland, bone, cartilage, muscle, adipose tissue, nervous tissue, and connective tissue. No immature components or malignant features were noted. The histology was typical of mature cystic teratoma. Postoperatively, the patient showed left abducens nerve palsy, positive Parinaud’s sign, and

Received November 5, 1997; Accepted February 24, 1998
left spastic hemiparesis but these symptoms improved within 2 weeks after the operation. No routine follow-up radiological examination was performed after a few years postoperatively because his parents reported yearly that he was developing normally. Eighteen years after gross total removal, the patient consulted a local physician complaining of headache and diplopia. Magnetic resonance (MR) imaging revealed a round mass in the pineal region associated with hydrocephalus. The tumor was low intensity on the T₁-weighted image, high intensity on the T₂-weighted image, but with no apparent contrast enhancement (Fig. 1). The patient was admitted to our hospital for further treatment.

Neurological examination at admission revealed no deficit except for symptoms of increased intracranial pressure. His extracranial movement was full and there was no Parinaud's sign. The valve of a ventriculoperitoneal shunt, which was implanted 18 years ago, was dented after repeated flushing. Diffusion-weighted MR imaging revealed a high signal intensity mass (5 × 5 × 3 cm) compatible with epidermoid cyst (Fig. 2 left).

A right occipital craniotomy using the previous occipital, transtentorial approach with the patient in the three-quarter prone position was used to perform subtotal excision of the tumor. The tumor, which was well demarcated and had a shiny "mother of pearl" appearance, was removed piecemeal (Fig. 3). Tumor content was granular, grumose, and flaky but there were also black hairs. The right occipital horn was opened and the capsule, which was tightly adherent to the pial surface, was left partially intact. The right posterior choroidal artery, which was embedded in the tumor, was spared. After subtotal removal, thick arachnoid membrane of the quadrigeminal plate was recognized. It was thought that the tumor had grown up from the pineal gland compressing right occipital lobe and trigone. Histological examination revealed an epidermoid tumor consisting of keratinized stratified squamous epithelium and connective tissue (Fig. 4). There was no other teratomatous or germinomatous component. Based on the operative finding of black hairs in the tumor, the diagnosis was dermoid cyst.

The patient had transient left hemiparesis after the operation but recovered. Occasional fever due to chemical meningitis occurred during the postoperative period. He now visits the outpatient clinic without help and receives steroid and anticonvulsant medication. Postoperative diffusion-weighted

![Fig. 1](image1.png)  
**Fig. 1** T₁-weighted magnetic resonance images, showing a low intensity mass in the pineal region extending into the right trigone (left: axial view), and no apparent contrast enhancement (right: coronal view).

![Fig. 2](image2.png)  
**Fig. 2** Preoperative (left) and postoperative (right) diffusion-weighted magnetic resonance images, showing a high signal intensity mass compatible with epidermoid cyst.

![Fig. 3](image3.png)  
**Fig. 3** Operative photograph through the right occipital transtentorial approach showing a pearly tumor.
MR imaging showed no apparent mass (Fig. 2 right).

**Discussion**

The dermoid cyst in our patient must have arisen from a microscopic remnant of the cyst wall of the mature teratoma 18 years after the solid portion of the tumor was removed en bloc, although some of the cyst wall was possibly left intact. The period of regrowth is unknown but may have been equivalent to the patient’s age plus 9 months. The patient presented with symptoms of increased intracranial pressure due to the slowly expanding tumor and shunt malfunction. Epidermoid and dermoid cysts of the pineal region are usually part of a well-differentiated teratoma. Mature teratomas are typically benign, firm in texture, and clearly demarcated, so complete resection is easily achieved. However, half of the patients in one series died within the 1st postoperative year, and three patients survived for 8, 12, and 13 years, with the latter patient developing a symptomatic recurrence at 8 years, which was again resected.

The World Health Organization classification of intracranial teratoma includes three histological variants: mature, immature, and malignant. The correct diagnosis of mature teratoma requires extensive sectioning of the surgical specimen to exclude mixed and/or malignant elements. Some germinomatous components may be present in mature teratomas. In fact, malignant transformation or selection of malignant tumors has occurred in teratocarcinoma, and leiomyosarcoma in a teratoma of the pineal area or ectopic (septum pellucidum) teratocarcinoma after the total removal of pineal mature teratoma. Suprasellar germinomas have developed several years after total removal of pineal teratomas.

The potential for phenotypic differentiation and cellular maturation within immature teratomas of the ovary and testis is well known. Differentiation of experimental tumors was studied by injecting single cells of mouse malignant teratoma into the peritoneal cavity of other mice. The successfully implanted tumors had both embryonal cancer cells and many well-differentiated cells of various types. Therefore, maturation of malignant teratomas is possible and the embryonal cell is a multipotent cell capable of producing various well-differentiated daughter cells. A case of primary intracranial germ cell tumor occurred in a 5-year-old Chinese boy. The initial diagnosis was pineal immature teratoma because of elevated serum and cerebrospinal fluid levels of α-fetoprotein and β human chorionic gonadotropin in addition to the histological examination of a biopsy specimen. After chemotherapy consisting of cisplatin, etoposide, and bleomycin, total excision revealed only a mature teratoma containing components of all three germ line elements. Therefore, multifocality in intracranial germinomas and teratomas cannot be dismissed. Recently, diffusion-weighted MR imaging, equivalent to a physiological MR cisternogram, has been helpful to characterize and distinguish between arachnoid and epidermoid cysts and is the only method that can distinguish between epidermoid cyst recurrence and postoperative encephalomalacia. The preoperative diagnosis of this patient was an epidermoid cyst in the pineal region based on the diffusion-weighted MR image. Careful follow-up observation of patients who undergo gross total removal of pineal mature teratoma should continue for a period exceeding the age at the time of surgery plus 9 months.

**References**


Address reprint requests to: T. Morimura, M.D., Department of Neurosurgery, National Utano Hospital, 8 Ondoyama-cho, Narutaki, Ukyo-ku, Kyoto 616-8255, Japan.