

Case Report

Seung-Jae Hyun, M.D.
 Seung-Won Park, M.D.
 Taek-Kyun Nam, M.D.
 Sung-Nam Hwang, M.D.

Department of Neurosurgery
 College of Medicine, Chung-Ang
 University, Seoul, Korea

A Case of Pineal Ependymoma

A 64-year-old man was admitted to our hospital in semicomatous consciousness. Brain computed tomography scans demonstrated 2.6×2.5 cm sized hyperdense mass in the pineal region with multiple punctate calcifications and hydrocephalus. Brain magnetic resonance imaging demonstrated a pineal mass which was heterogeneously enhanced with gadolinium. After external ventricular drainage, the patient regained consciousness. The mass was totally removed via occipital transtentorial approach. No consequent ventricular shunt was needed and the patient recovered without any neurological deficit. Final pathologic report of the tumor was ependymoma.

KEY WORDS : Pineal tumors · Ependymoma · Surgery · Occipital · Transtentorial approach.

INTRODUCTION

Pineal region tumors account for less than 1% of primary brain tumors¹⁰. These tumors are of various histological types reflecting the origin from the various adjacent anatomical sites, the different cell populations within the pineal gland (pineocytoma and pineoblastoma) and the midline inclusions of multipotent germinal cells and more mature derivatives of one or more of the germ cell layers (ectoderm, mesoderm and endoderm) in the pineal region^{2,6,15}. Earlier confusion regarding the nomenclature of pineal region tumors has now been eradicated by the widespread acceptance of the histological classification of pineal tumors by Russell and Rubinstein¹² (Table 1). We present here a patient with pineal ependymoma which has very rarely been reported in the literatures.

CASE REPORT

History and examination

A 64-year-old man was admitted via emergency room in semicomatous consciousness after drinking in the funeral. Headaches, nausea and vomiting preceded before he lapsed into unconsciousness. At admission he was semicomatous and both pupils were fixed at 3 mm in diameter with papilloedema.

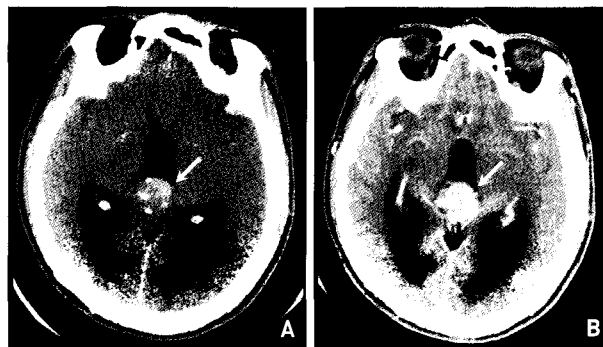


Fig. 1. Computed tomography scans of the brain at the level of the third ventricle (A) before and (B) after intravenous contrast enhancement. There is an enhancing mass in the posterior third ventricle (arrow) and secondary dilatation of the ventricular system. Multiple calcified nodules are visible in (A).

Non-enhanced brain computed tomography (CT) scans showed 2.6×2.5 cm sized hyperdense mass with multiple punctate peripheral calcifications in the pineal region and obstructive hydrocephalus of the third and lateral ventricles (Fig. 1). On the following post-

Table 1. Histological classification of pineal region tumors (after Russell & Rubinstein, 1977)

Tumors of germ cell origin	Tumors of pineal cell origin	Other cellular origin
Teratoma		Glioma
Geminoma	Pineocytoma	Ganglioglioma
Embryonal cell carcinoma	Pineoblastoma	Ganglioneuroma
Choriocarcinoma		

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 • Address for reprints :
 Sung-Nam Hwang, M.D.
 Department of Neurosurgery
 College of Medicine, Chung-Ang
 University, 65-207 Hangangno 3-ga
 Yongsan-gu, Seoul 140-575, Korea
 Tel : +82-2-748-9565, 9965
 Fax : +82-2-748-2818
 E-mail : tarheelk@hanmail.net

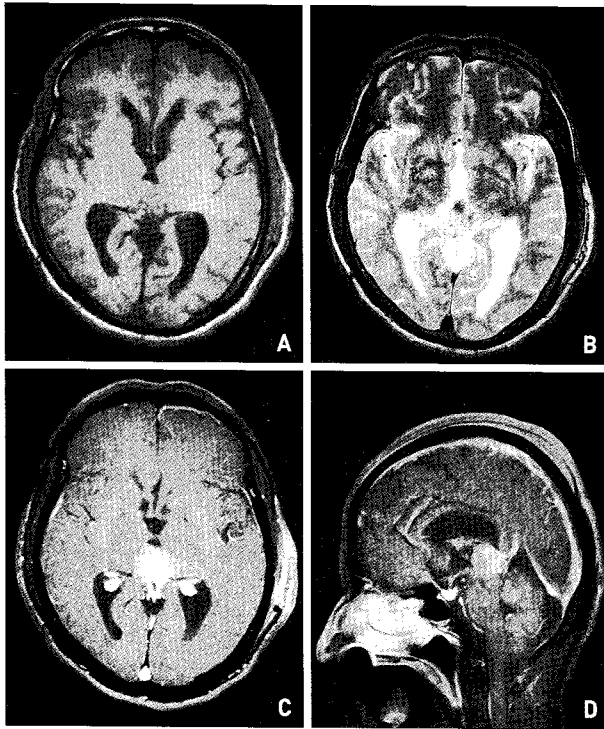


Fig. 2. A : Spin echo T1-weighted axial magnetic resonance (MR) image shows a heterogeneous mass lesion with multiple punctate calcifications in the pineal region. B : Turbo spin echo T2-weighted MR axial image shows no marked cyst formation. Contrast-enhanced T1-weighted axial (C) and sagittal (D) MR images show strong heterogeneous enhancement.

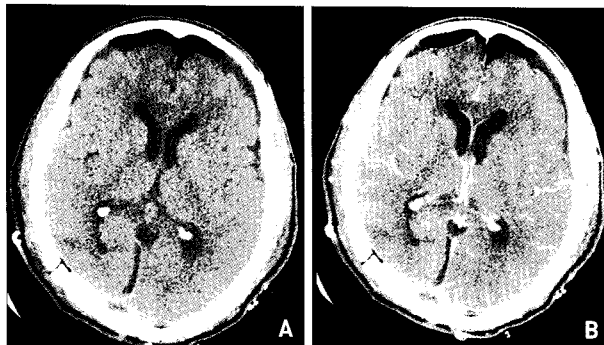


Fig. 3. Computed tomography (CT) scans obtained 3 days after the operation (A) before and (B) after intravenous contrast enhancement. Follow-up CT scan reveals no hydrocephalus and no visible tumor.

contrast CT scan, this mass lesion showed strong enhancement. Brain magnetic resonance imaging (MRI) demonstrated a pineal mass, which was heterogeneously enhanced after gadolinium administration (Fig. 2). Cerebro-spinal fluid cytology and serologic examination of human chorionic gonadotrophins (HCG) and alpha-fetoprotein (AFP) were all negative.

Operative procedure and Pathologic finding

Following external ventricular drainage (EVD) the patient regained consciousness very shortly. Soon after regaining

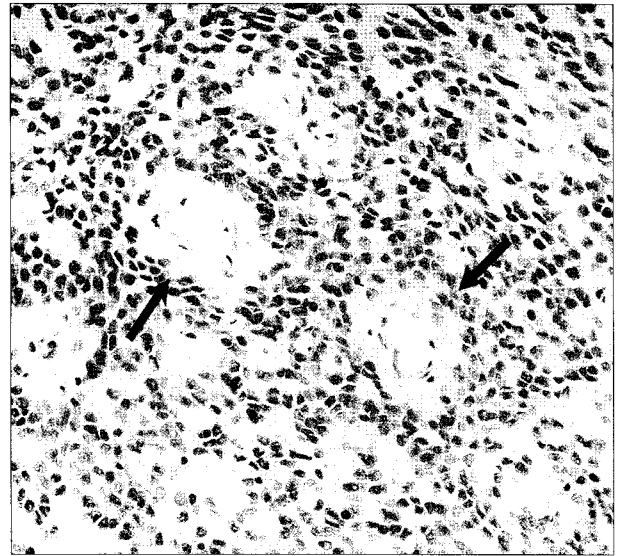


Fig. 4. Photomicrograph demonstrating perivascular pseudorosettes (arrow) of varying sizes. Between the pseudorosettes the cells are somewhat spindled with abundant cytoplasm. (H&E, original magnification $\times 400$).



Fig. 5. Staining for pancytokeratin (AE1/AE3) shows perivascular accentuation of labeling (original magnification $\times 630$).

consciousness, occipital transtentorial approach was performed and a well demarcated tumor which was densely attached at the left part of the quadrigeminal plate of the midbrain was totally excised. Histopathologically, the tumor displayed a pseudopapillary growth pattern of dyshesive cells forming perivascular pseudorosettes (Fig. 4). Cells contained round, uniform nuclei with exceedingly rare mitotic figures. Immunoreactivity for glial fibrillary acidic

protein was not detected, but pancytokeratin stain (AE1/AE3) showed perivascular accentuation of labeling, characteristic of an ependymoma (Fig. 5).

Post-operative course

Following surgery he became fully orientated. After removal of the EVD catheter three days after the operation, the patient tolerated well and follow-up CT scan revealed no hydrocephalus and no visible tumor (Fig. 3). The patient was discharged without any neurological deficit.

DISCUSSION

The occurrence of supratentorial ependymoma in the pineal region was well recognized as early as in 1954¹¹. In 1998, Cho et al.⁴ reported pineal region tumors treated at Seoul National University Hospital. Among the pineal region tumors, there was one pineal ependymoma. However, there is no published case report on pineal ependymoma in Korea. The ependymoma, a layer of inactive cells that retains its epithelial character, provides lining of the various parts of the nervous system such the ventricles, choroid plexus, central canal of the spinal cord, and filum terminale³. It is generally agreed that it is very difficult to predict an accurate histological diagnosis of pineal tumors on the basis of the CT scan or MRI alone⁷. Germinomas account for more than 50% of pineal region tumors, in common with other germ cell tumors at this site, they occur almost exclusively in males in the 2nd or 3rd decade^{8,9,15}.

Tumors arising from the pineal parenchymal cells are less common than germ cell tumors and account for only 20% of pineal region tumors^{6,15}. They occur with equal incidence in both sexes and at any age and could, from the appearances on CT, be included in the differential diagnosis of pineal tumors, as it is not clear whether the tumor is engulfing or displacing the pineal gland^{8,9}.

Glial tumors may arise from the fibrillary astrocytes of the pineal gland or the quadrigeminal plate or wall of the third ventricle. They displace the pineal gland anteriorly and superiorly suggesting an extrapineal origin^{8,15}. Astrocytoma is usually hypodense on plain CT scans and calcification is unusual that it is unlikely to be an astrocytoma as in this case⁸.

Supratentorial ependymoma is an uncommon tumor and is usually not included in the differential diagnosis of pineal region tumors. However, its occurrence at this site has been rarely reported in the literatures^{11,13}. The majority of intracranial ependymomas occur infratentorially in children¹⁴. Most of the supratentorial ependymomas are situated intraparenchymally in the fronto-parietal region¹

and it is rare to find in the midline parenchyme or in the ventricles, most of the infratentorial ependymomas occur in the fourth ventricle¹⁴. Supratentorial ependymomas are usually large in diameter¹. On the contrary, infratentorial or pineal ependymoma are smaller due to earlier development of hydrocephalus before growing large^{1,5}.

Dense, punctate calcification is seen in about 50% of supratentorial ependymomas^{1,3}. Tumors of pineal cell origin characteristically engulf a calcified pineal gland while those of extrapineal origin displace it. There is general agreement that the supratentorial ependymoma is often cystic although there was no marked cyst formation in this case, possibly a reflection of its early presentation. Contrast enhancement is generally marked with no dominant pattern^{1,14}.

CONCLUSION

It is suggested that ependymoma, although rare at this site, should be considered in the differential diagnosis of a pineal region tumors containing dense, punctate calcification and displaying marked contrast enhancement without any positive tumor markers.

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