

PRIMARY PINEAL GLIOBLASTOMA: a case report

Onur Ozgural, MD, Resident, Ankara University Medical Faculty, Department of Neurosurgery, Sıhhiye, Ankara, Turkey

Gokmen Kahilogullari, MD, PhD, Instructor, Ankara University Medical Faculty, Department of Neurosurgery, Sıhhiye, Ankara, Turkey

Melih Bozkurt, MD, Instructor, Ankara University Medical Faculty, Department of Neurosurgery, Sıhhiye, Ankara, Turkey

Aylin Okcu Heper, MD, Associate Professor, Ankara University Medical Faculty, Department of Pathology, Sıhhiye, Ankara, Turkey

Ali Savas, MD, PhD, Professor, Ankara University Medical Faculty, Department of Neurosurgery, Sıhhiye, Ankara, Turkey

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Corresponding author: Gokmen Kahilogullari, M.D., PhD
Department of Neurosurgery
Ankara University Faculty of Medicine
Samanpazari/Ankara
TURKIYE

Phone: +90 3103333(extension: 2662)

e-mail: gokmenkahil@hotmail.com

Introduction

Tumors pineal region include germ cell tumors, parenchymal tumors and supporting tissue tumors like gliomas. Germ cell tumors are the most common tumor type of pineal region (1,16). Second frequently type is glial tumor but mostly low grade glioma and the third is parenchymal tumors as pineocytoma or pineoblastoma (1). Glioblastoma (GBM) is so rare for this region (6,12,16). We report a rare case of GBM in pineal region. This is the 18th case of primary glioblastoma in pineal region and second case that survive over two years according the literature (12).

Case report

A 60-year-old man admitted to hospital with headache and ataxia which were continuing during last 3 months. Physical examination was normal. Neurological examination revealed ataxia. There were no motor and sensory deficits. Computer tomography (CT) showed triventricular hydrocephalus and isodense rounded mass in pineal region. Magnetic resonance images revealed regular edged heterogeneous contrast enhanced tumor in pineal region (Figure 1A,1B). First of all, ventriculoperitoneal shunt applied to the patient for hydrocephalus (Figure 1C). After operation, ataxia and hydrocephalus were improved. Ten days later, serial stereotactic biopsies were applied (Figure 1C). The paraffin embedded and H&E stained sections of stereotactic biopsies were revealed a tumor composed of pleomorphic, atypical glial cells (Figure 2A,2B). Mitotic figures, necrosis and vascular endothelial proliferation were also noted. Immunohistochemical analysis demonstrated GFAP expression (Figure 2C) with high (15%) Ki67 proliferation index. More than half of the tumor cells were p53 positive. The tumor cells were negative with pan-ck. Pathology result was glioblastoma. Patient was recommended to radiotherapy (RT) and chemotherapy (CHT). The patient was treated with median 60 Gy in 2 Gy single fractions RT and for CHT regime, 75

mg/m²/per day temozolamid was used during RT. The patient is still surviving two years after biopsy and shunt operation, without deficit.

Discussion

Pineal region is framed superiorly by corpus callosum, anteriorly by third ventricle, laterally thalamus and cerebral hemisphere, posteriorly habenularum (3). Tumors of this region are rare. It comprises less than %1 in all intracranial tumors (1,6). Tumors can be derived pineal gland itself, germ cells as germinomas, teratomas and supporting tissue as gliomas. The tumors of this region may both stem from pineal gland and also posterior part of the third ventricle or quadrigeminal cistern (9). These tumors are generally irregular and compressing neighboring structures like aqueducts sylvii and frequently cause hydrocephalus (3). MRI reveals association with neighbor tissue and vascularity of tumor. Although glioblastoma is the most primary malign tumor of brain, it is seen so rare for pineal region. Only a few cases reported about GBM of pineal region (4,5,6,8,10,14,15).

The germ cell tumors especially germinoma and teratoma are the most common tumors of pineal region. It is predominant for males. It occurs generally first two decades (6,16,18). They may appear hyperdense on CT, with a strong contrast enhancement. In CT, low grade astrocytomas are seen iso- hypodense and moderate contrast enhancement. MRI may show iso-hypointense on T1-weighted images and hyperintense on T2-weighted images. MRI may show an isodense lesion. Parenchymal tumors of pineal region are pineocytoma, pineoblastoma, cysts etc. These tumors often seen in older ages and equal in male and female. Pineoblastoma is seen heterogenous contrast enhancement and generally make invasion to adjacent tissue. MRI may show an iso or hypointense lesion on T1-weighted image and hyperintense on T2-weighted images. Other tumors arising from the glial components of the pineal gland include ganglioneuroma, ganglioglioma, meningioma, and gliomas, more frequently low-grade astrocytomas (6,18).

Despite all available treatment protocols, GBM is a progressive malign tumor with aggressive course and short survival. The mean survival in patients having radiotherapy and chemotherapy after surgical resection in pineal region GBMs was shown to be 7 months (12,15). Longer survival durations were also present in the literature of pineal GBMs. One of the cases who survived 27 months operated for hydrocephalus and a shunt was inserted. The second case had radiotherapy and chemotherapy after resection and survived 18 months (6,12). These cases were both girls 5 and 12 years old respectively. The presented case underwent stereotaxic surgery and RT/CHT as treatment protocol and survived more than 2 years. This was the first adult GBM reaching a survival more than 2 years. It is known, the PG is one of unique structure that blood-brain-barrier is not present (2,7) and this knowledge may be answer of why the CHT agents could be more effectively in this region (11,13,17).

Despite of its rarity, GBM should be thought if there is a tumor in pineal region. We suggest that stereotaxic surgery may be an effective alternative way considering the difficulty of reaching this region and aggressive nature of the tumor. This technique enables pathologic diagnosis without wide resection and with minimal complication rate. We also suggest that RT/KT following stereotaxic biopsy may significantly contribute to the survival of patients having GBM.

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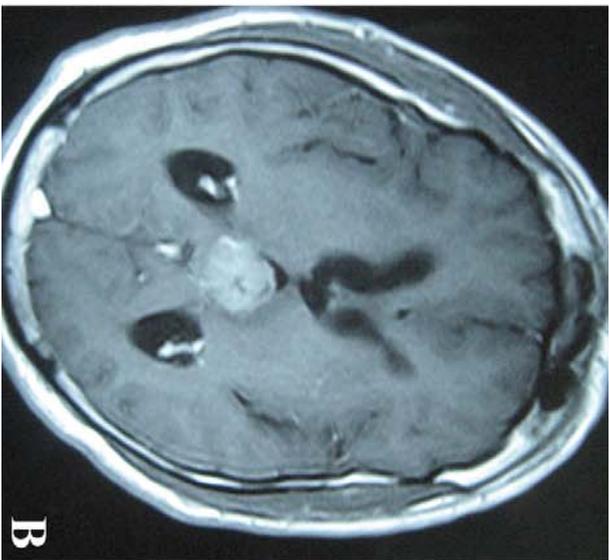
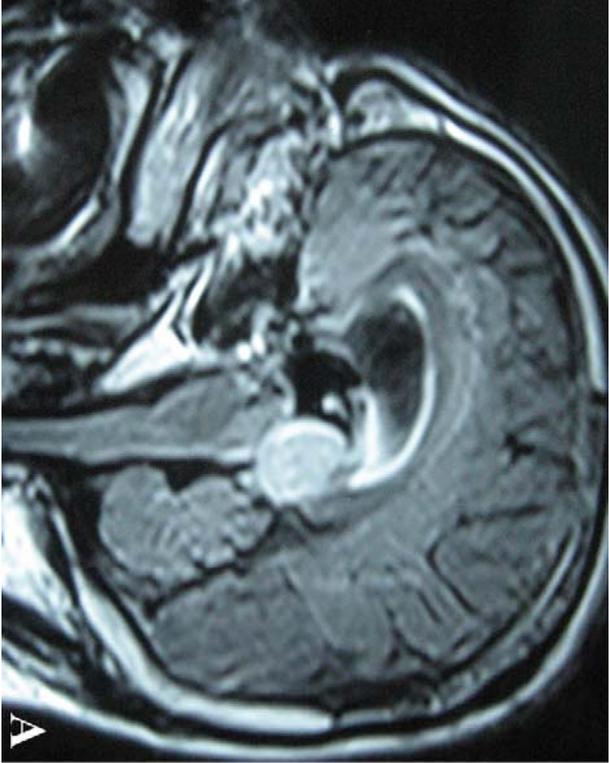
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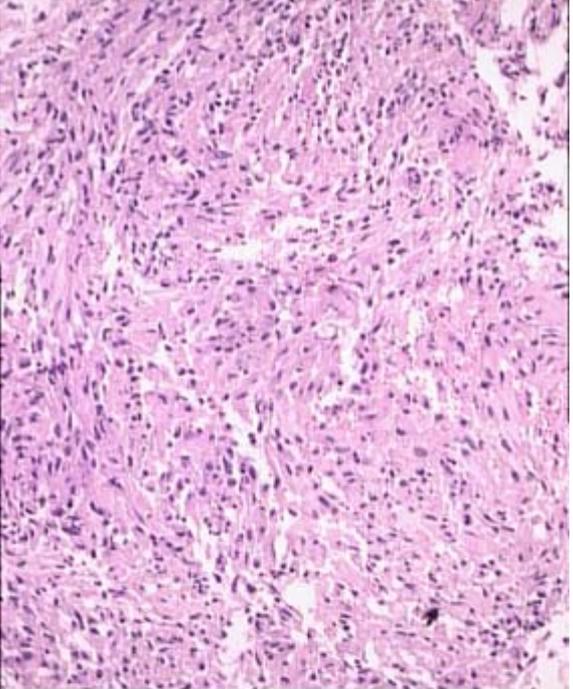
Figure 1:

A: Preoperative magnetic resonance imaging shows heterogeneous mass in the pineal region (sagittal section). **B:** Preoperative magnetic resonance imaging shows heterogeneous mass in the pineal region (axial section). **C:** Computerized tomography shows tumor of the pineal region after ventriculoperitoneal shunt and stereotactic biopsy.

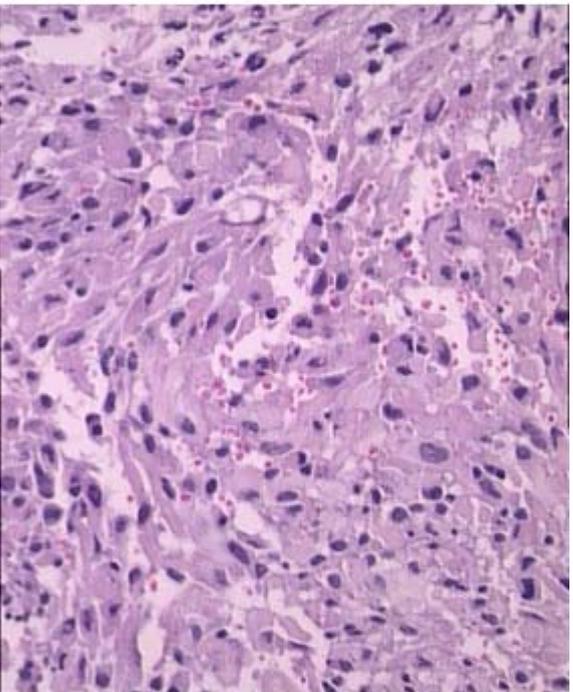
Figure 2:

A: Tumor composed of pleomorphic atypical glial cells, H&Ex200. **B:** The closer view of tumor cells, H&Ex400. **C:** Immunohistochemical assessment: The cytoplasmic GFAP expression of the tumor cells. The vascular endothelial proliferation was also present. GFAPx200.

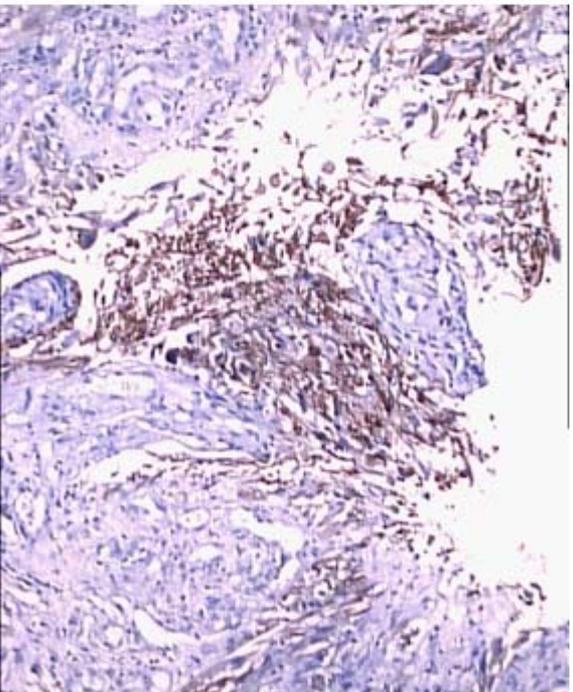




A



B



C