Germinoma Involving Bilateral Basal Ganglia: a Rare Case Report

Jang-Chun Lin, Da-Tong Ju, Chang-Min Chen, Hsing-Lung Chao, Chun-Shun Lin, and Yee-Min Jen

1Department of Radiation Oncology; 2Department of Neurological Surgery, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan, Republic of China

Germinoma that occurs to the bilateral basal ganglia is exceedingly rare. The diagnosis can be difficult because of non-specific clinical presentation and ill-defined abnormalities without a definite tumor mass on neuroimaging. Here, we presented a 24-year-old male who suffered from bizarre behavior and affective disturbance. At emergency room, the psychiatrist was consulted to evaluate his psychiatric abnormalities. Computed topography (CT) scan of brain revealed several cystic lesions with rim-like enhancement involving bilateral basal ganglia. Magnetic resonance (MR) imaging of brain showed multiple cystic lesions with irregular rim-like enhancement involving bilateral basal ganglia. Accordingly, stereotactic aspiration biopsy was performed by neurological surgeon, and pathological result revealed bilateral germinoma of basal ganglia. Then, the patient received chemotherapy followed by craniospinal irradiation. After craniospinal irradiation, MR imaging of brain was performed again, which showed complete regression. In conclusion, when the patient presented with bizarre behavior and affective disturbance with lesion located in basal ganglia, the differential diagnosis of germinoma should be considered.

Key words: bilateral germinoma, basal ganglia

INTRODUCTION

Germ cell tumors include germinomas, teratomas, embryonal carcinomas, endodermal sinus tumors, choriocarcinomas, and mixed germ cell tumors. Kim et al., pointed out that the incidence of intracranial germ cell tumor is approximately 2% in primary intracranial neoplasms in patients below 20 years of age. However, the incidence has been reported to be as high as 6.5% in Japan. Germinomas account for approximately two-thirds of germ cell tumors and usually present in the midline of the suprasellar and pineal regions. Germinomas involving the bilateral basal ganglia are exceedingly rare. We reported a 24-year-old male with bilateral basal ganglia germinomas presenting non-specific symptoms. Computed topography (CT) scan and magnetic resonance imaging findings were not specific. Early diagnosis is usually difficult to make and the mainstay of management is chemotherapy followed by radiotherapy with craniospinal irradiation.
Germinoma of bilateral basal ganglia

CASE REPORT

A previously healthy 24-year-old male suffered from bizarre behavior and affective disturbance for two weeks. The patient was sent to our emergency room, and initially the psychiatrist was asked to evaluate this patient. His neurological examinations revealed no abnormal finding except decreasing deep tendon reflex. CT scan of brain revealed cystic lesions with rim-like enhancement involving bilateral basal ganglia. (Fig. 1). MR imaging studies of brain also showed cystic lesions with irregular rim-like enhancement involving bilateral basal ganglia. (Fig. 2A, 2B and 2C). Germinoma, pilocytic astrocytoma, cryptococcal meningitis, tuberculosis meningitis and brain abscess should be included for differential diagnosis. Laboratory examinations revealed no remarkable contribution including β-human chorionic gonadotropin (β-HCG) and α-fetoprotein (AFP) within normal range. He underwent stereotactic aspiration biopsy of basal ganglia. Pathological examination revealed sheet-like pattern of large neoplastic tumor cells, characterized by well-defined cell border, and clear cytoplasm. (Fig. 3A) Immunohistochemical staining showed positive for placental alkaline phosphatase (PLAP) (Fig. 3B) and C-kit. (Fig. 3C) consistent with germinoma. Then, the patient received chemotherapy with cisplatin followed by craniospinal irradiation. The patient received regular follow-up for three months, and MR imaging of brain showed complete regression.

DISCUSSION

Germinomas of the basal ganglia are believed to arise from ectopic totipotential cells that derived from the mid-
line during a very early stage of rostral neural tube development.\textsuperscript{2} Intracranial germinoma accounted for 0.5-2.1% of all intracranial tumors. The peak incidence presented in the latter half of the second decade of life.\textsuperscript{3} Moreover, intracranial germ cell tumors usually arise from the pineal and suprasellar regions. Germinomas situated in the basal ganglia are rare. Their involvement in bilateral basal ganglia is exceedingly rare (five cases reported to date).\textsuperscript{4}

Pineal germinoma produces discrete masses that are often detected early because of characteristic symptoms of paralysis of the upward gaze and eyelid retraction, but basal ganglia germinoma produces ill-defined infiltrations that may long remain undetected because of non-specific clinical presentation, such as hemiparesis, mental deterioration, ocular motor palsy, involuntary movement, seizures, and diabetes insipidus.\textsuperscript{1} The clinical course is usually slow progression, with the duration of clinical symptoms ranging from 1 month to 4.5 years.\textsuperscript{5}

It is very difficult to confirm the diagnosis of basal ganglionic germ cell tumors at an earlier stage, because of its non-specific clinical features, and lack of mass lesions on CT scans and MR images. To make a prompt correct diagnosis, early imaging findings of basal ganglia germ cell tumors have to be identified. The early CT features of basal ganglia germinoma have been described as an ill-defined, homogeneous or inhomogeneous, isodense to slightly hyperattenuated area without mass effect.\textsuperscript{7} Early MR imaging findings of basal ganglia germinomas have been described as low signal intensity on T1-weighted images and high signal intensity on T2-weighted images of the basal ganglia.\textsuperscript{2} T1- and T2-weighted images in our cases were similar to theirs. By contrast, Okamoto \textit{et al}.\textsuperscript{7} stated that high signal intensity on T1-weighted images and a small hyperintense lesion on T2-weighted images were seen in the basal ganglia. Various signal intensity patterns may result from different cell densities and amounts of tissue necrosis among individual lesions.

AFP can be detected in endodermal sinus tumor, mixed germ cell tumor, embryonal carcinoma, and immature teratoma. $\beta$-HCG is identified in choriocarcinoma and various types of germ cell tumors. It is well known that $\beta$-HCG and AFP increase in CSF and serum of patients with germ cell tumors. Wong \textit{et al}.\textsuperscript{8} reported the clinical significance of elevated $\beta$-HCG in pure basal ganglia germinoma is unknown. Immunohistochimically, the neoplastic cells show cytoplasmic membranous pattern, sometimes diffuse cytoplasmic pattern, of staining for placental alkaline phosphatase.\textsuperscript{9} Immunopositivity for the proto-oncogen C-kit appears to be specific for germinomas.\textsuperscript{10} Activating \textit{KIT} mutations may contribute to tumorigenesis in germinoma.\textsuperscript{11} Mutation of c-kit gene at exon 11 (W557C) or 17 (D816V, D820V, and N822Y) was found in some germinomas but no statistically significant difference in any clinicopathological factor was found between patients with or without mutations.\textsuperscript{12}

Though germinomas respond well to radiation therapy, conventional radiation therapy carries risks of neurologic sequelae in young patients. Reduction of posttreatment sequelae and preservation of neurocognitive functions are important because the prognosis of cerebral germinoma is excellent. Early diagnosis can prevent progressive of neurologic deficit. Decrease in radiation field and dose can contribute to reduce posttreatment sequelae and preserve neurocognitive functions.

In conclusion, although germinoma arising in the basal ganglia are usually unilateral, they can involve bilateral basal ganglia. Clinical findings are non-specific, such as bizarre behavior and affective disturbance. It is important to recognize the early MR imaging findings of germ cell tumors in bilateral basal ganglia to improve the outcome. When a young patient presented psychological symptoms accompanied with bilateral basal ganglia lesions, the differential diagnosis of germinoma should be considered.

**DISCLOSURE**

The authors declare that they have no conflict of interest.

**REFERENCES**


Germinoma of bilateral basal ganglia


