Right Basal Ganglia Germinoma: Case Report and Literature Review

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Research Article

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Abstract

Background: Among intracranial germinomas, germinoma of basal ganglia has been rarely reported. We discuss a new case of basal ganglia germinoma (BGG) and perform the literature review over the last two decades, with the aim of emphasizing the diagnosis and treatment in early-stage BGG.

Case presentation: A seven years old Chinese boy presented with 4 months history of left limb movement disorder and oblique right mouth corner. The human chorionic gonadotropin (HCG) level in cerebrospinal fluid (CSF) was slightly increased. Magnetic resonance imaging (MRI) showed ipsilateral brain and brainstem atrophy. Susceptibility weighted imaging (SWI) revealed obvious hypointensity in right globus pallidus. Pathological diagnosis on biopsy was confirmed with germinoma. The patient had a favorable relief of symptoms after chemoradiotherapy.

Conclusion: Intracranial germinoma, a potentially curable tumor, the early diagnosis is essential for the prognosis. An elevated HCG level of CSF or serum can be used as a reference indicator. MRI, especially SWI, plays an important role in early diagnosis. Patients should be treated with standardized chemoradiotherapy early rather than surgery.

Background

Intracranial germinomas (IGs), originating from residual tissue of primordial germ cells, accounting for about 0.22-3% of all intracranial tumors in adolescents and children[1]. IGs occur most frequently in the midline locations, such as the pineal and suprasellar regions. Germinomas in the basal ganglia (BG) account for only 5–10% of all primary IGs[2]. Basal ganglia germinomas (BGGs) are slow growing and the clinical manifestations are diverse, including hemiparesis, abnormal sensation, and so on. So many patients are easily misdiagnosed or missed without timely and effective treatment.

We report the clinical features, imaging and treatment of a BGG. And review and summarize the Magnetic resonance imaging (MRI) characteristics of BGGs in combination with other literatures, with the aim of emphasizing the diagnosis and treatment in early-stage BGGs.

Case Presentation

A seven-year-old Chinese boy without significant past or family medical history presented with 4 months history of left limb movement disorder and oblique right mouth corner was admitted to our institute. Nervous system examination: left central-type facial palsy, left hemiplegic gait, left limb muscle strength grade 4/5, left knee tendon reflex slightly active. The remaining physical examinations were normal. Serum human chorionic gonadotropin (HCG) level is normal, but cerebrospinal fluid (CSF) HCG level was slightly increased to 6.08mIU/ml (normal range: 0.00–5.00).

Unenhanced computerized tomography (CT) of the head (4 months after the onset of the symptom) showed hyperdensity in right BG (Fig. 1a). MRI revealed that T1 hypointensity and T2 hyperintensity in right BG and no significant space occupying effect (Fig. 1b,c) and mild atrophy of right cerebral peduncle (Fig. 1f). Contrast-enhanced MRI demonstrated irregular rim enhancement (Fig. 1d). Susceptibility weighted imaging (SWI) revealed obvious hypointensity in right BG and the size was larger than that on conventional MRI (Fig. 1e).

Pathological diagnosis on stereotactic biopsy was confirmed with germinoma. The patient received four cycles of combination chemotherapy consisting of carboplatin, ifosphamide and etoposide, followed by radiotherapy to the whole neuraxis (30.6Gy/17fractions) and a boost to the main lesion (54Gy/30fractions). After chemoradiotherapy, the clinical symptoms improved significantly and malacia of the lesion can be found on MRI (Fig. 2a,b) and the enhancing degree of the lesion (Fig. 2c) was alleviated compared with before treatment.

Discussion

Germinoma is the most common type (2/3) of the intracranial germ cells tumors[3] and 90% of cases are diagnosed before 20 years of age[4]. BG is a rare location for IGs, preceded by the pineal and suprasellar regions. BGGs are more common in Asian populations, with males predominating[5]. In Table 1, we reviewed the clinical, laboratory and imaging data of the 35 patients with biopsy-proven BGG from 2000 to 2020.(32 males and 3 females, mean age 12.98 years, range 7 to 21 years).
Table 1  
Cases of germinoma in the basal ganglia reported in literature

| References                  | Age (y) | Sex | Weakness of Limbs | Diabetes Insipidus | Increased ICP | sHCG, cHCG | CT | T1 | T2 | Contrast Enhancement | Ipsilateral brain and brainstem atrophy | SWI |
|----------------------------|---------|-----|-------------------|--------------------|---------------|------------|-----|----|----|----------------------|----------------------------------------|-----|
| Ya Takeda et al, 2004       | 11,M    | Y   | N                 | N                  | N             | Hyper      | Hyper| Hyper| N  | Y                    | N/A                                    |     |
| Noriko Oyama et al, 2005    | 10,M    | Y   | N                 | N                  | N             | N/A        | Hyper| Hypo| Hyper| Y                    | N/A                                    | N/A |
| Rodrigo V. Ozelame et al, 2006 | 14,M | Y   | N                 | N                  | N             | Hyper      | Hypo | Hyper| N/A | N/A                  | N/A                                    |     |
| Ya Takeda et al, 2004       | 13,M    | Y   | Y                 | N                  | N             | Hyper      | Hyper| Y   | N/A | Y                    | N/A                                    |     |
| Noriko Oyama et al, 2005    | 9,M     | N   | N                 | N                  | N             | Hyper      | N/A | Hetero| Y   | Y                    | N/A                                    |     |
| Joong-Seok Kim et al, 2007  | 13,M    | Y   | Y                 | N                  | N/A           | Hyper      | Hypo | Hyper| N/A | Y                    | N/A                                    |     |
| O. Klein et al, 2007        | 8,F     | Y   | Y                 | Y                  | N/A           | Hyper      | N/A | Hyper| N/A | Y                    | N/A                                    |     |
| Anita Villani et al, 2008   | 13,M    | Y   | N                 | N                  | N             | Hyper      | Hypo | Hyper| Y   | Y                    | N/A                                    |     |
| Andrea Rossi et al, 2008    | 14,M    | Y   | Y                 | N                  | N             | Hyper/Hypo| N/A | Hyper| Y   | N/A                  | N/A                                    |     |
| Shuyu Hao et al, 2009       | 13,F    | Y   | N                 | N                  | N             | Hyper      | Hypo | Hyper| N/A | Y                    | N/A                                    |     |
| Sun-To Wong et al, 2009     | 7,M     | Y   | N                 | N                  | N             | N/A        | Hyper| Y   | Y   | N/A                  | N/A                                    |     |
| En Xu et al, 2010           | 10,M    | Y   | N                 | N                  | N             | Hetero     | Hypo | Y   | Y   | Y                    | N/A                                    |     |
| D D Rasalkar et al, 2010    | 11,M    | Y   | N                 | Y                  | N             | N/A        | Hyper| Hyper| N/A | N/A                  | N/A                                    |     |
| 15,M                       | Y       | N   | Y                 | N                  | N             | Hyper      | N/A | N/A | N/A | N/A                  | N/A                                    |     |
| Soumik Goswami et al, 2013  | 13,F    | Y   | N                 | N                  | N             | Hyper      | Hypo | Y   | N/A | Y                    | N/A                                    |     |
| Takafumi Watayai et al, 2015| 9.5,M | N   | N                 | N                  | N/A           | Hyper      | N/A | N/A | N/A | N/A                  | N/A                                    |     |
| C.J. Maurer et al, 2016     | 15,M    | Y   | N                 | N                  | N/A           | Hyper      | N/A | N/A | N/A | N/A                  | N/A                                    |     |
| Yasushi Ogasawara et al, 2016| 12,M | Y   | N                 | N                  | N             | N/A        | Hyper| Hyper| Y   | Y                    | N/A                                    |     |
| Clement Vialatte de Pemille et al, 2016 | 21,M | Y   | N                 | N                  | N             | N/A        | Hyper| Y   | N/A | N/A                  | N/A                                    |     |

Note: Y, yes; N, no; ICP, intracranial pressure; sHCG, serum human chorionic gonadotropin; cHCG, cerebrospinal fluid human chorionic gonadotropin; N/A, not av
Hetero, heterogeneous; Hyper, hyperintense; Hypo, hypointense; Iso, isointense; Nor, normal; CT, computerized tomography; SWI, susceptibility-weighted imagin
T1WI, T2WI, weighted imaging; S, surgery; SB, stereotactic biopsy; WB, whole-brain irradiation; WS, whole-spine irradiation; LF, local field irradiation; CMT, chemotherapy sample source
chemotherapy, especially radiotherapy, and have the potential to be curative[11]. Radiotherapy is the mainstay of treatment for IGs. Chemoradiotherapy can
Because neurological function may be severely compromised, surgical resection should be avoided. Germinomas are highly sensitive to radiotherapy and
the SWI technique is underutilized. We cannot ignore the role of SWI and should combine SWI with conventional MRI for an early diagnosis of BGG.

studies have reported that SWI is more sensitive in detecting early-stage BGG[10]. But only 2/35 cases in the literature used the SWI technique. It is clear that
at the first MR examination, SWI revealed obvious hypointensity in right globus pallidus and the size was larger than that on conventional MRI. Previous
may occur in germinoma[10]. SWI is more sensitive in detecting hemorrhage due to it can detect blood products and biologic metal accumulation. In our case,
Approximately 50–70% of BGGs involve intratumoral hemorrhages[9]. Pathology has indicated that even at the early stage, hemorrhage and iron deposition
BGGs, which can be found in up to 33% of cases[8]. 19/35 patients in the Table
enhancement, which are less specific. If patients were misdiagnosed or untreated, the MRI findings will show large, space-occupying, multicystic, and
calcification, and/or intratumoral hemorrhage, etc, early BGGs merely show T2WI hyperintensity with slight or no space occupying effect and mild or no
The diagnosis of early-stage BGG is essential for the prognosis of patients. Whereas patients’ clinical symptoms and serum or CSF HCG were not highly
measured, 18 patients were negative and 12 patients were positive. An elevated HCG level of CSF or serum is not present in all patients with germinomas, so it
only can be used as a reference indicator. When elevated, a diagnosis of germinomas is more strongly suggested. But a negative test does not exclude the
diagnosis. Our patient had a normal serum HCG and slightly elevated CSF HCG, thus supporting the diagnosis of germinoma.

| References                        | Age (y) | Weakness of Limbs | Diabetes Insipidus | Increased ICP | sHCG, chCG | CT | T1 | T2 | Contrast Enhancement | Ipsilateral brain and brainstem atrophy | SWI |
|-----------------------------------|---------|------------------|--------------------|---------------|------------|----|----|----|---------------------|----------------------------------------|-----|
| A.N.Konovalov, et al, 2016[28]     | 15, M   | Y                | Y                  | -/-           | N/A        | Hypo | Hyper | Y  | N/A                 | N/A                                    |     |
|                                   | 13, M   | Y                | Y                  | -/-; N/A      | N/A        | Hypo | Hyper | Y  | N/A                 | N/A                                    |     |
|                                   | 14, M   | Y                | N                  | -/-; D        | N/A        | Hypo | Hyper | Y  | N/A                 | N/A                                    |     |
|                                   | 16, M   | Y                | N                  | -/-; D        | N/A        | Hypo | Hyper | Y  | N/A                 | N/A                                    |     |
| Peter Yat Ming Woo, et al, 2017[29]| 21, M   | Y                | N                  | -/-; D        | N/A        | Iso  | Hyper | Y  | N/A                 | N/A                                    |     |
| Seishiro Nodomi et al, 2017[30]   | 14, M   | Y                | N                  | -/-; D        | N/A        | N/A  | N/A  | Y  | N/A                 | N/A                                    |     |
| Jihong Yan, et al, 2019[31]       | 13, M   | Y                | N                  | -/-; D        | Hyper      | Hyper | Hyper | N  | N/A                 | N/A                                    |     |
| Eike Steidl, et al, 2019[32]      | 21, M   | Y                | N                  | -/-; D        | N/A        | Hyper | Y    | N  | Y                   | N/A                                    |     |
| Zhen-Chao Huang, et al, 2020[33]   | 12, M   | Y                | N                  | -/-; D        | N/A        | Hypo | Hyper | Y  | Y                   | Hypo                                   |     |
|                                   | 10, M   | Y                | N                  | -/-; D        | N/A        | Hypo | Iso   | Y  | Y                   | Y                                    |     |
|                                   | 16, M   | Y                | N                  | -/-; D        | N/A        | Hypo | Hyper | Y  | N/A                 | N/A                                    |     |
|                                   | 10, M   | Y                | Y                  | -/-; D        | N/A        | Iso  | Hyper | Y  | N                   | N/A                                    |     |

Note: Y, yes; N, no; ICP, intracranial pressure; sHCG, serum human chorionic gonadotropin; chCG, cerebrospinal fluid human chorionic gonadotropin; N/A, not av
Hetero, heterogeneous; Hyper, hyperintense; Hypo, hypointense; Iso, isointense; Nor, normal; CT, computerized tomography; SWI, susceptibility-weighted imagin
T2WI, T2-weighted imaging; S, surgery; SB, stereotactic biopsy; WB, whole-brain irradiation; WS, whole-spine irradiation; LF, local field irradiation; CMT, chemothera
sample source

The most common first symptom of BGGs was hemiplegia, which progressed slowly with an average age of 9.7 years[6]. In the last 20 years, 27/35 patients
reviewed came to medical attention because of symptoms of hemiplegia. The symptoms of our case were left limb movement disorder and oblique right
mouth corner too.

HCG levels are useful for the diagnosis of germinoma. In Table 1, HCG levels of CSF or serum were measured in 30 patients. Among these patients who had
measured, 18 patients were negative and 12 patients were positive. An elevated HCG level of CSF or serum is not present in all patients with germinomas, so it
only can be used as a reference indicator. When elevated, a diagnosis of germinomas is more strongly suggested. But a negative test does not exclude the
diagnosis. Our patient had a normal serum HCG and slightly elevated CSF HCG, thus supporting the diagnosis of germinoma.

The diagnosis of early-stage BGG is essential for the prognosis of patients. Whereas patients’ clinical symptoms and serum or CSF HCG were not highly
specific. Besides, the lesion is deep-seated, access to pathology by puncture is challenging, and carries a risk of bleeding. Imaging plays an important role in
early IGs diagnosis. In the early stage of the disease, abnormalities can be found on imaging. In Table 1, BGGs appear predominantly hyperdense on CT, with
the cystic component being hypodense. Although conventional MRI is able to sensitively detect signal changes in the BG, such as cystic formation,
calcification, and/or intratumoral hemorrhage, etc, early BGGs merely show T2WI hyperintensity with slight or no space occupying effect and mild or no
enhancement, which are less specific. If patients were misdiagnosed or untreated, the MRI findings will show large, space-occupying, multicystic, and
gadolinium-enhancing tumors at the site of initial lesion and/or other distant locations[7]. Ipsilateral brain and brainstem atrophy was another character of
BGGs, which can be found in up to 33% of cases[8]. 19/35 patients in the Table 1 also presented with ipsilateral brain atrophy. Just as our case, MRI revealed
slightly atrophy of right cerebral peduncle. Therefore, ipsilateral brain and brainstem atrophy can be considered as an early sign for the diagnosis.
Approximately 50–70% of BGGs involve intratumoral hemorrhages[9]. Pathology has indicated that even at the early stage, hemorrhage and iron deposition
may occur in germinoma[10]. SWI is more sensitive in detecting hemorrhage due to it can detect blood products and biologic metal accumulation. In our case,
at the first MR examination, SWI revealed obvious hypointensity in right globus pallidus and the size was larger than that on conventional MRI. Previous
studies have reported that SWI is more sensitive in detecting early-stage BGG[10]. But only 2/35 cases in the literature used the SWI technique. It is clear that
the SWI technique is underutilized. We cannot ignore the role of SWI and should combine SWI with conventional MRI for an early diagnosis of BGG.

Because neurological function may be severely compromised, surgical resection should be avoided. Germinomas are highly sensitive to radiotherapy and
chemotherapy, especially radiotherapy, and have the potential to be curative[11]. Radiotherapy is the mainstay of treatment for IGs. Chemoradiotherapy can
reduce the dose of radiotherapy, with the aim of decreasing the complications of radiotherapy, especially in children and adolescents. The therapeutic effect
of chemotherapy followed by reduced-dose radiotherapy can be similar to those of radiotherapy alone[12]. The patients with germinoma who are treated by
standardized chemotherapy and radiation therapy have long-term progression-free survival rates that exceed 90%[13]. In Table 1, 29 patients completed the treatment, 22 were treated with chemoradiotherapy, 3 with chemotherapy alone, 4 with radiotherapy alone. Clinical follow-up data were available for 17 cases with average time of 29.9 months ranging from 6 to 120 months. None of the patients presented recurrence of tumor. However, Wong et al.[6] and Kanamor et al.[14] had demonstrated that chemotherapy without radiotherapy is unable to cure intracranial germinomas. Our patient was treated with chemotherapy followed by reduced-dose radiotherapy and his clinical symptoms improved significantly.

Brain disease occurs infrequently in the BG. There are many diseases with high T2 hyperintensity in the bilateral BG, such as infarction, hypoglycemia, Leigh disease, encephalitis, MELAS, and the others[15]. The diagnosis of patients should be considered comprehensively based on age, gender, clinical symptoms, laboratory tests, imaging examinations, etc. If organic diseases cannot be completely excluded, early MRI scans and measurement of serum tumor markers and blood hormones are recommended for these patients. Although in many cases, the diagnostic sensitivity of serum tumor markers is not high, it is still considered to be valuable because of its non-invasive property and convenience. Stereotactic biopsy can determine the type of tissue and thus confirm the diagnosis. Although there is a high risk of bleeding during the procedure, stereotactic biopsy is recommended for lesions in the diagnosis of early-stage BGGs when necessary. Our case was also confirmed by stereotactic biopsy and had no complications such as bleeding. If a germinoma is suspected but a biopsy is refused or when biopsy does not reveal tumor cells, SWI is strongly recommended. Hypointense lesion in SWI, combined with patient gender, age and serum or CSF HCG level, other MR signs such as ipsilateral brain and brainstem atrophy, diagnostic treatment, such as radiotherapy or chemoradiotherapy may be performed.

Conclusion

When adolescent Oriental males develop progressive hemiplegia with slow onset, abnormal MR signal in BG with ipsilateral brain atrophy, SWI should be performed. Germinoma should be considered if the patient presents with above symptoms and abnormal imaging findings with hypointensity in SWI as well as an elevated serum or/and CSF HCG level. When necessary, a biopsy can confirm the diagnosis and patients should be treated with standardized chemoradiotherapy early rather than surgery.

Abbreviations

BG
basal ganglia; BGG: basal ganglia germinoma; CSF: cerebrospinal fluid; CT: computerized tomography; HCG: human chorionic gonadotropin; IG: Intracranial germinomas; MRI: magnetic resonance imaging; SWI: susceptibility weighted imaging.

Declarations

Ethics approval and consent to participate

The study was approved by the Research Ethics Board of the First Hospital of Jilin University. Written informed consent and any image information were obtained from the patient for publication of this case report.

Consent for publication

Written informed consent and any accompanying images were obtained from the patient for publication of this case report.

Availability of data and materials

The authors declare that all data and materials of the article are available to all readers of our article.

Competing interests

The authors declare that they have no competing interests.

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Authors’ contributions

PT and HZ performed the case analysis and drafted the manuscript. YL reviewed the literature. Radiology image reporting was performed by DL; BB and SX revised the manuscript. DL contributed to the process of review. All authors read and approved the final manuscript.

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Figures

Figure 1

Radiographic findings of 4 months after the onset of the symptom. (a) Unenhanced axial head computerized tomography (CT) demonstrates hyperdensity nodules in the right basal ganglia (BG) (arrow). MRI revealed that T1 hypointense (Fig. 1b) and T2 hyperintense signal (Fig. 1c) in right BG and no significant space occupying effect and mild atrophy of right cerebral peduncle (Fig. 1f). (d) Contrast-enhanced MR image demonstrated irregular rim enhancement of the lesion (arrow). (e) Susceptibility weighted imaging (SWI) revealed obvious hypointensity in right globus pallidus (arrow) and the size of the focus was larger than that on conventional T1WI and T2WI.
Figure 2

Radiographic findings after two cycles of chemoradiotherapy. Malacia of the lesion (arrow) can be found on T1WI (a) and T2WI (b). (c) Contrast-enhanced MR image showed the enhancing degree of the lesion (arrow) were alleviated compared with before treatment.