

## Case Report

# Primary intracranial malignant ectomesenchymoma in pineal region: a rare case report and review of literature

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Received June 3, 2016; Accepted August 30, 2016; Epub December 1, 2016; Published December 15, 2016

**Abstract:** Primary intracranial malignant ectomesenchymoma (MEM) is a rare proliferative disorder that only occasionally involves the central nervous system. We presented a 16-year-old case of primary MEM located in pineal region. The patient was treated by total surgical resection followed by radiation and experienced no recurrence during the 6-month follow-up. The clinicopathological features of this case raised two points for the first record, as the first regarded the special age and pineal location as well as the second regarded the specific pathological components with malignant large round cells. The diagnosis for MEM is still challenging. The histological displays, immunophenotypical characteristics and cytogenetics arrays are still reliable evidence for diagnosis. Surgical resection has been demonstrated an effective treatment, but radiotherapy and chemotherapy have not been confirmed reliable therapeutic efficiency.

**Keywords:** Ectomesenchymoma, neuroectodermal, rhabdomyosarcoma, pathology, diagnosis

## Introduction

Malignant ectomesenchymoma (MEM) was identified as a new kind of soft tissue neoplasms in 1977, as reported by Karcioglu [1]. To date more than 50 cases have been published in the literature and only 8 cases with confirmed clinicopathological features arising from brain have been reported, as shown in **Table 1** [2-10]. MEM is a rare tumor consisting of both ectodermal elements and mesenchymal components, believed to originate from migratory neural crest and the neuroectodermal elements are the main proportion of primary intracranial MEM [7, 11]. Many MEMs occurred in the little children under 3 years old and all 8 reported intracranial cases were from 1.5 years to 10 years old and arose from cerebrum. Here, we reported a 16-year-old case of MEM located in pineal region. The malignant components of primary intracranial MEM were usually mesenchymal elements or small round cells and spindle cells [9, 11]. However, in the article we will report a rare MEM with neuroectodermal ele-

ments and special mesenchymal elements, large round cells. To best of knowledge, the present case was the first one arising from diencephalon with neuroectodermal elements and mesenchymal large-round-cell elements whose age was close to the adult in the primary intracranial cases of MEM.

## Case report

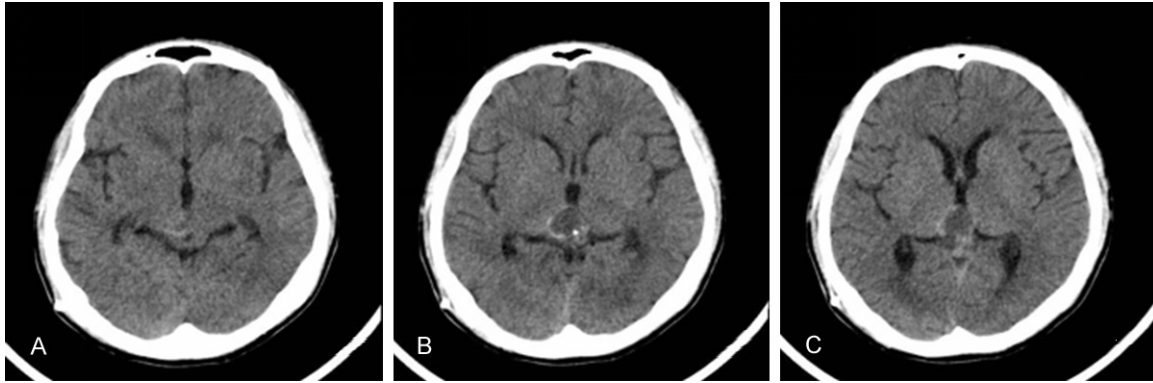
### History

A 16-year-old boy was admitted into our department with the main complaint of persistent mild to moderate headache for four months and progressive visual disturbance for half a month. Three months ago the patient presented to the local hospital and found a solid lesion in posterior third ventricle as well as expandable supratentorial ventricle, then achieved the ventriculoperitoneal shunt for the relief of hydrocephalus. After that the headache symptom had begun to relieve. However he later developed left blurred vision, and deteriorated rapidly over half a month, then was introduced to our department.

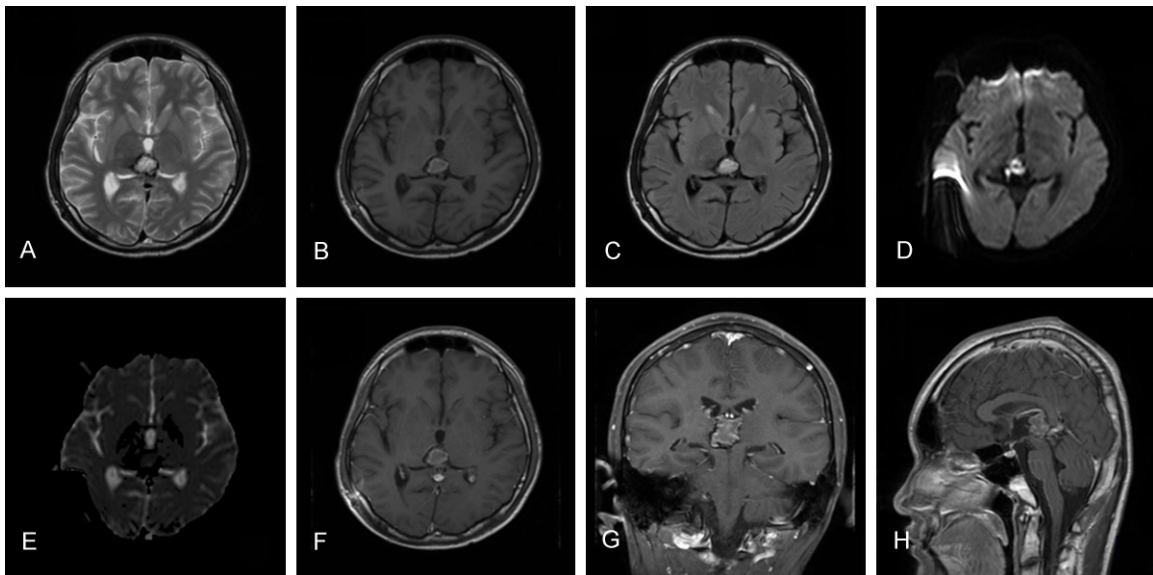
## Malignant ectomesenchymoma

**Table 1.** MEM arising from brain from 1971 to 2015

Sex	Age (y)	Location	Ectodermal component	Mesenchymal component	Surgery	Radiation	Chemotherapy	Outcomes	Year
M	15	CPA	Ganglioneuroma	Rhabdomyosarcoma	N	Y	Y	Dead/6 mo.after surgery, metastasis	1971
F	2	Frontal, corpus callosum	Ganglion cells	Rhabdomyosarcoma	Y	Y	Y	Dead/14 mo.after surgery, metastasis	1999
F	1	Temporoparietal	Neuroblastoma	Rhabdomyosarcoma	Y	N	N	Alive/10 mo.after sugery	2001
F	10	Temporo-parietal-occipital	Neuroblastoma	Rhabdomyoblastoma	Y	Y	N	Alive/6 yr.after sugery	2005
F	4	Left hemispheric	Ganglion cells	Undifferentiated sarcoma	Y	N	Y	Dead/10 wk.after surgery	2007
M	10	Falx cerebri	Ganglioneuroma	Rhabdomyosarcoma, smooth muscle cells	Y	Y	Y	Alive/1 yr.after sugery	2011
M	6	Frontal	Anaplastic neuronal cells	Less mesenchymal cells in collagenous and myxoid background	Y	Y	N	Alive/2 yr.after sugery, local relapse	2013
F	4	Frontal-parietal	Astrocytoma, anaplastic ependymoma	Rhabdomyosarcoma	Y	N	N	Alive/5 mo.after sugery	2015



**Figure 1.** The operative CT scans of the patient. CT scans demonstrated a round and smooth-edged hypodensity lesion with sporadic calcification around the circum located in right posterior third ventricle, which infringed on the right metathalamus.



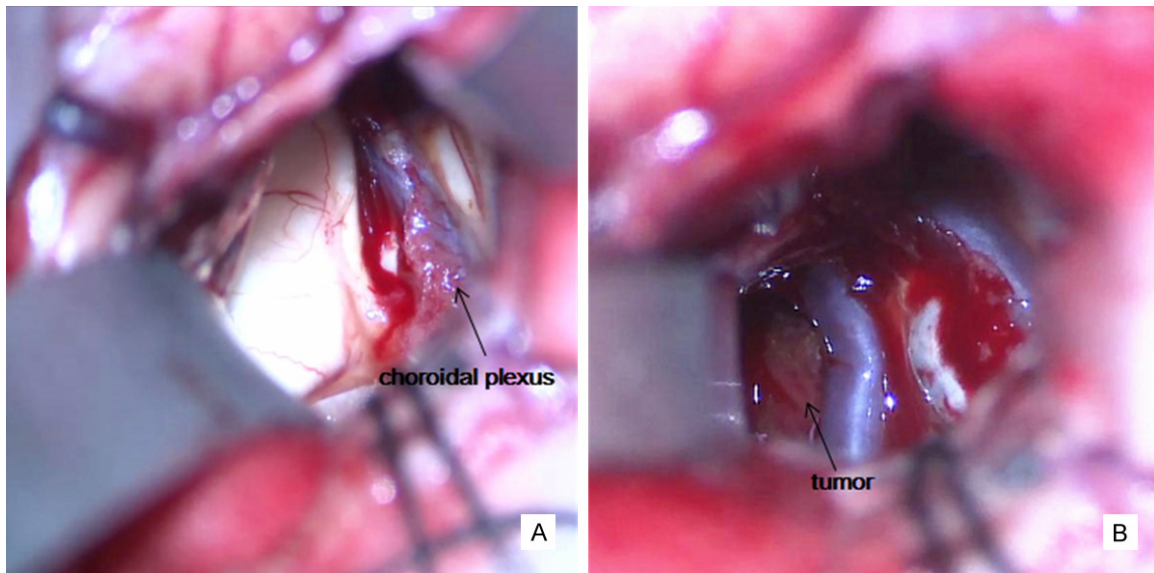
**Figure 2.** The operative MRI scans of the patient. A, B. T2-/T1-weighted axial MRI showed the lesion was hyperintense. C. Flair-MRI showed the lesion was hyperintense. D, E. The lesion without peritumor edema was hyperintense on DWI-MRI and hypointense on ADC-MRI. F-H. The lesion after administration of intravenous contrast presented heterogeneously enhanced and also obstructed the aqueduct of sylvius.

He had no significant past history or family history. There were either no systemic symptoms such as fever, leukocytosis and skin manifestations, and no lymph nodes involvement.

#### Examinations

On neurological examinations he was found to have shallow hemihypesthesia of left side and IV-grade strength of left limbs. Ophthalmological examinations identified visual acuity in the left eye of 20/100 and in the right eye of 30/50. Computed tomography (CT) scans revealed a

round and smooth-edged hypodensity lesion with sporadic calcification around the circum in right posterior third ventricle infringing on the right metathalamus (**Figure 1A-C**). Magnetic resonance imaging (MRI) scans demonstrated an irregular-shaped, calcified and solid lesion in the pineal region measuring 2.0 cm×2.0 cm×2.5 cm was hyperintense on T2-weighted MRI, T1-weighted MRI and fluid-attenuated inversion recovery (FLAIR) images (**Figure 2A-C**). The lesion without peritumor edema was hyperintense on DWI-MRI and hypointense on ADC-MRI (**Figure 2D, 2E**). The heterogeneously



**Figure 3.** The photography during operation. A. The choroidal fissure was exposed via transcortico-ventricular-choroidal fissure approach. B. The mass appeared dusty red, hypervascularized and well defined from the peripheral normal tissues.

enhanced lesion invaded the right metathalamus and upper mesencephalon, and also obstructed the aqueduct of sylvius (**Figure 2F-H**). Because of the VP shunt, supratentorial ventricle was normal. The spinal MRI study was negative for metastases. Laboratory results, including hormone levels, biochemical examinations and tumor markers such as CEA, AFP, NSE and  $\beta$ -hCG were within normal limits. Based on the history, examinations and neuroimaging results, preoperative diagnosis was made as “mixed germ cell tumor”. Operation The patient underwent a total removal of the tumor via transcortico-ventricular-choroidal fissure approach following a right frontal craniotomy (**Figure 3A**). During the surgery, the tumor was arising from and involving the dorsal midbrain and bending closely with the adjacent thalamencephalon. The mass without obvious invasion of parenchyma appeared slightly hard, dusty red, hypervascularized and well defined from the peripheral normal tissues (**Figure 3B**). Some gravel-like calcification, little cysts and multiple necroses were found.

## Pathology

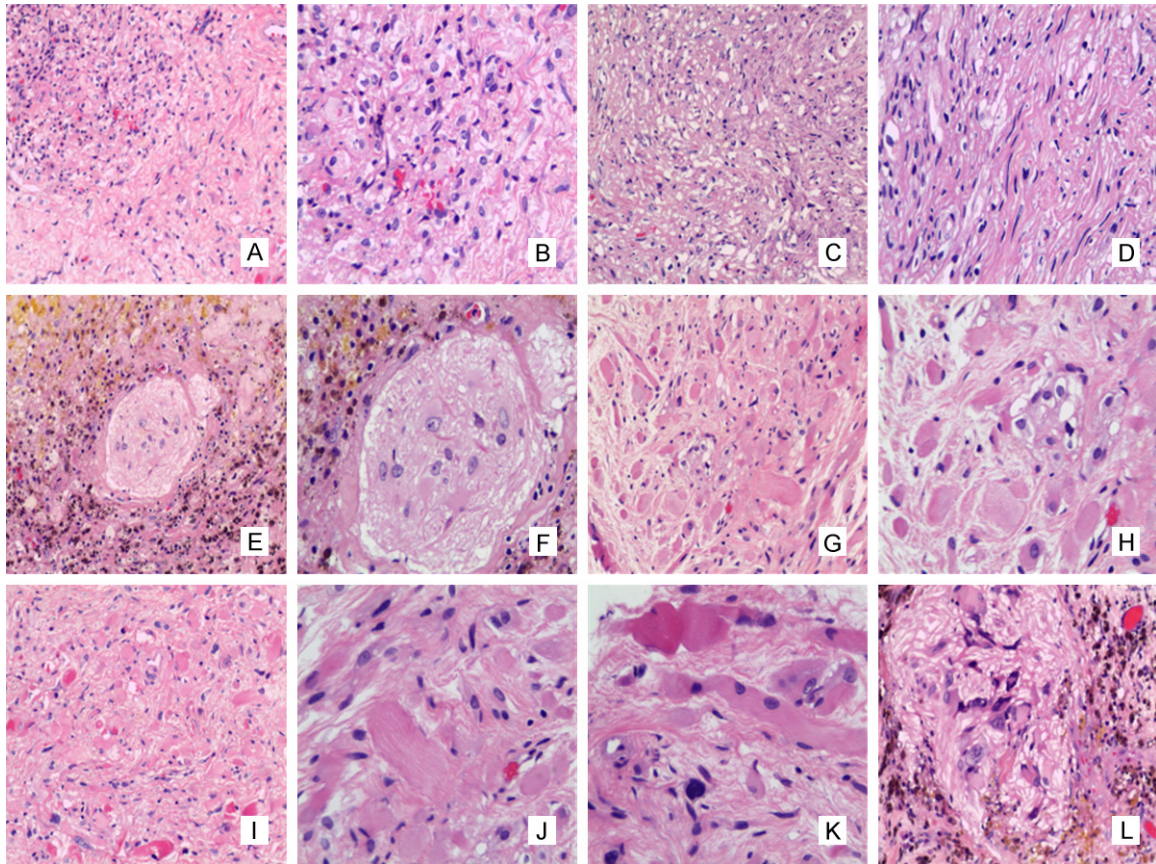
Histological morphology demonstrated the biopsy contained neuroectodermal components and mesenchymal components. The former part, neuroectodermal components consisted of immature ganglion cells and astrocytoma

cells (**Figure 4A-F**). Calcification, abundant necrosis, vascular proliferation and severe pleomorphism were found in astrocytoma. The latter parts with mesenchymal differentiation contained rhabdomyosarcoma/rhabdomyoblastoma presenting distinct striated features, hyaline degeneration of fibrous hyperplasia, extensive necrosis, new small vessels and high mitotic counts (**Figure 4G-J**) as well as large round cells with abundant plasma and obvious heteromorphism. Strips of large round tumor cells with eccentric mitosis showed severe atypia, and had an increased nuclear-cytoplasm ratio and two or more irregular-shaped, apparently dysplastic nucleus (**Figure 4K, 4L**). The mitotic rate of large round cell components was much higher than the neuroectodermal component, and the MIB-1 index was about 20%.

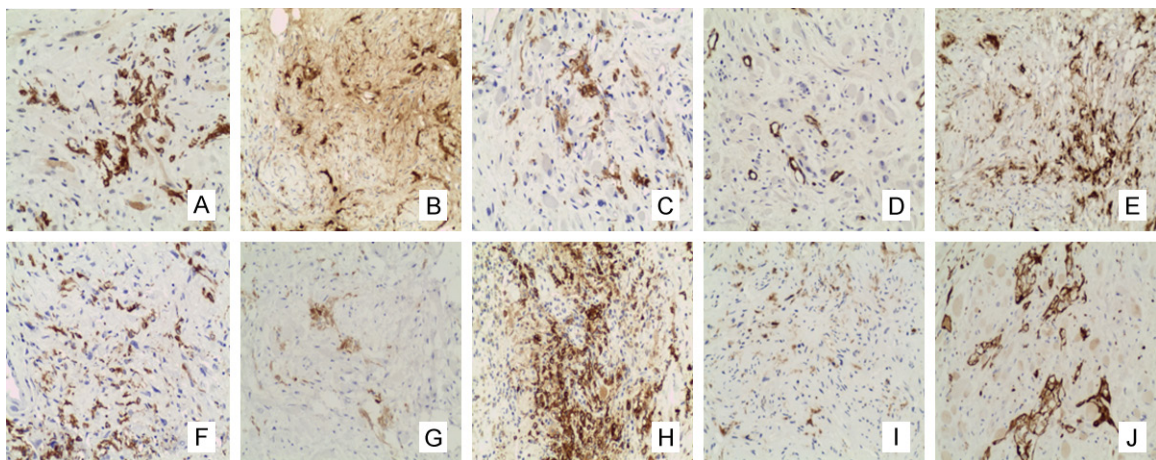
Immunohistochemical staining of astrocytoma parts displayed tumor cells were positive for GFAP, S-100 protein, Olig-2, EMA, vimentin and nestin (**Figure 5A-D**). In the ganglionic portion, Syn, NF, NSE, MAP-2, MBP and S-100 protein were partly positive, but not chromogranin (**Figure 5E-J**). The mesenchymal component cells were strongly positive for the rhabdomyosarcoma markers such as Myoglobin, MyoD1 and Myogenin (**Figure 6A-C**). In large-round-cell portions, specific mesenchymal markers including desmin, SMA and  $\alpha$ -Sarcomeric showed



## Malignant ectomesenchymoma

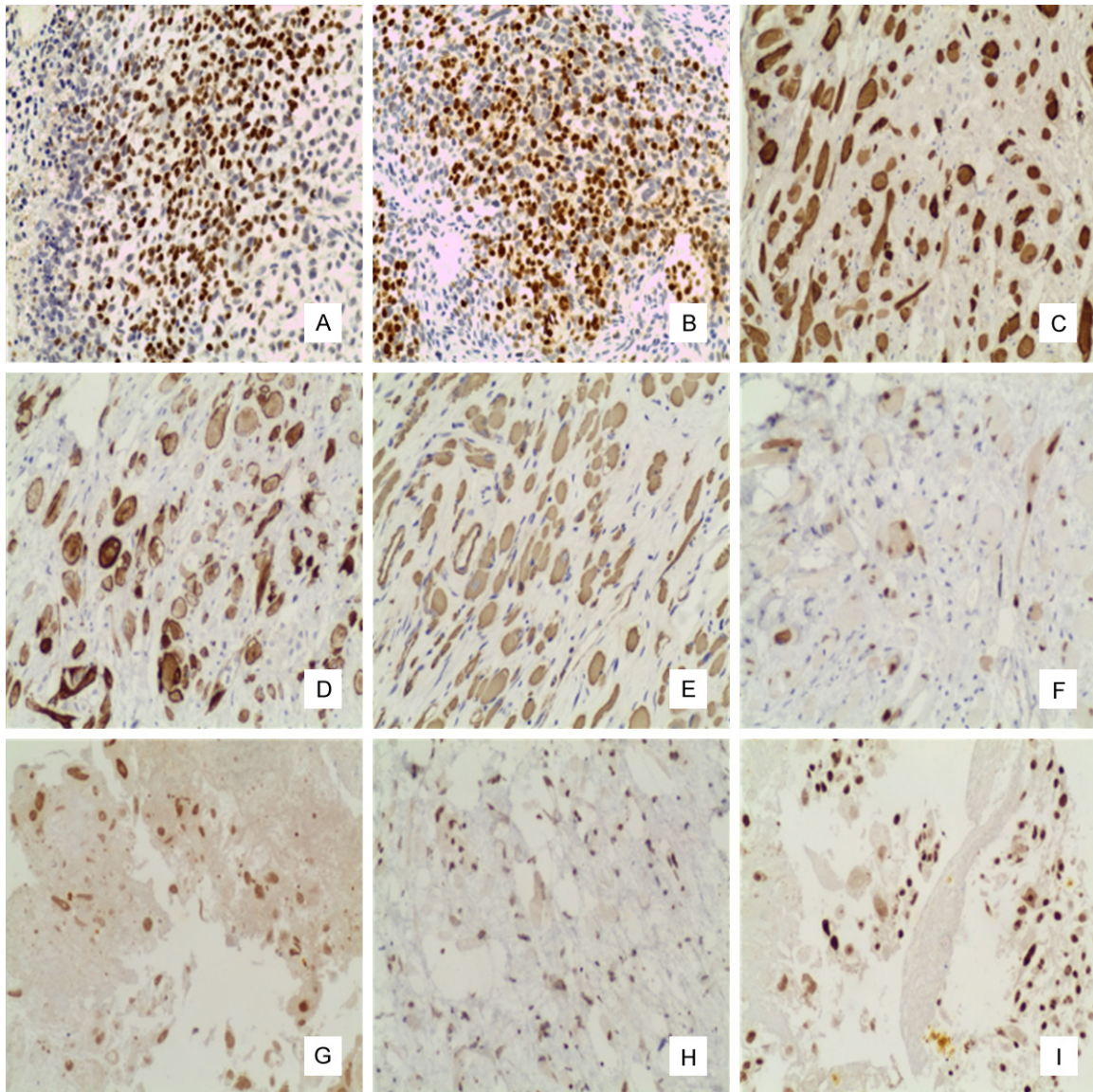


**Figure 4.** The histological results of MEM. (A, B) Neuroectodermal components consisted of immature ganglion cells and some nerve fibers. (A. 200×, B. 400×) (C, D) Neuroectodermal and astrocytoma region. (C. 200×, D. 400×). (E, F) The neuroectodermal elements showed extensive necrosis and a rich investiture of nerve fibers. (E. 200×, F. 400×). (G, H) Mesenchymal components mainly consisted of rhabdomyosarcoma presenting distinct striated features. (E. 200×, F. 400×). (I) High power photomicrograph showed the hyaline degeneration of fibrous hyperplasia in mesenchymal region. (400×). (J) Mesenchymal region of the tumor cells composed of vascular proliferation and some stale hemorrhage. (400×). (K, L) Mesenchymal region of tumor contained large round cells with eccentric mitosis and two or more irregular-shaped, apparently dysplastic nucleus. (K. 200×, L. 400×).



**Figure 5.** Immunohistochemical staining of MEM. In astrocytoma parts immunohistochemical findings were positive for GFAP (A), S-100 protein (B), EMA (C) and Olig-2 (D). In the ganglionic portion, Syn (E), NF (F), NSE (G), MAP-2 (H), MBP (I) and S-100 protein (J) were partly positive.





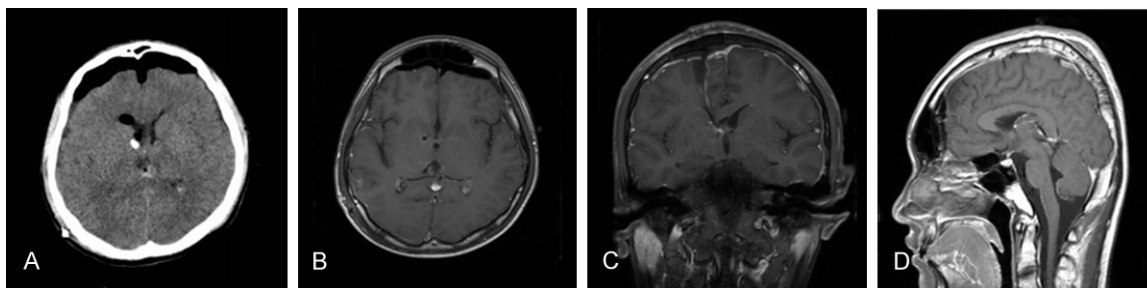
**Figure 6.** Immunohistochemical staining of MEM. The mesenchymal component cells were strongly positive for Myoglobin (A), MyoD1 (B) and Myogenin (C). In large-round-cell portions, mesenchymal markers including desmin (D), SMA (E) and  $\alpha$ -Sarcomeric (F) showed clear positivity. Positive immunostaining for INI-1 was detected in both neuroectodermal (G) and mesenchymal (H) parts. The Ki-67 immunolabeling was 10-20% in the atypical cells (I).

clear positivity (**Figure 6D-F**), but immunostaining for GFAP, Olig-2, S-100 protein, NSE and Syn was not detected. These above immunohistochemical data suggested the large round cell components showing mesenchymal differentiation. Sarcomatous areas were diffusely immunoreactive for reticular fibers. In addition, areas of probable smooth muscle differentiation were immunopositive for SMA. Positive immunostaining for INI-1 was detected in both neuroectodermal and mesenchymal parts (**Figure 6G, 6H**). The Ki-67 immunolabeling was 10-20% in

the atypical cells (**Figure 6I**). AFP, hcG, hPL, PLAP and CEA were all negative in the two populations, eliminating the possibility of germ cell tumors. Above all, these findings were consistent with a diagnosis of intracranial MEM.

#### Postoperative course

Postoperative course was relatively uneventful and there was no obvious neurological deficit. The sensation and strength were superior to the preoperative. Postoperative 3 d MRI con-



**Figure 7.** The postoperative neuroradiological results of the patient. CT scans (A) and contrast MRI scans (B-D) showed a proximal total resection of the tumor via transcortico-ventricular-choroidal fissure approach following a right frontal craniotomy.

firmed a proximal total resection of the tumor. (Figure 7A-D). Because of the aggressiveness and a little remnant bending to Galan vein, the patient received radiation treatment with a total dose of 2520 cGy/14f to the whole cranio-spinal axis and a 2540 cGy/11f boost to the tumor bed based on 3D planning promptly after operation. The radiation was completed successfully without severe acute adverse reactions. At present, the patient remained asymptomatic and returned to school over the next 6 months follow-up and had no recurrences. The long-term follow-up will be in progress.

## Discussion

The definition of ectomesenchymoma is that the tumor consists of both ectodermal elements and one or more kinds of neoplastic mesenchymal elements [12]. MEM deriving from the remnants of migratory neural crest cells belongs to soft tissue tumor and presents high malignancy with rapid progress and metastasis.

## Epidemiology

So far more than 50 cases have been reported and the tumor occurs mostly in children and adolescents and in the soft tissues of abdomen, head and neck, retroperitoneal space, perineum, pelvic cavity, limbs and CNS. In the literature, only eight cases of MEM involving the CNS have been noted, as shown in Table 1. Primary intracranial MEM was exceptionally rare, approximately 87.5% (7/8) of which were younger than 10 years of age and arose from the cerebrum. Due to the extreme rarity and complex components, the exact etiology and epidemiology are still unknown, likewise the diagnosis and treatment are still facing enor-

mous challenges. Here we described a case of primary intracranial MEM originating from the diencephalon in a 16-year-old boy, the first reported sample in such a pineal location and such an approximate adult age.

## Neuroradiology

On CT scans, MEM appears as the low-attenuated mass of isodensity or slightly hypodensity compared with gray matter. Cysts are seldom reported but calcification and necrosis are often described. The lesion tends to be heterogeneous with moderate enhancement or no enhancement after administration of intravenous contrast. On MRI scans, owing to the different and complex components it appears characteristics by heterogeneous intensity on T1-weighted and T2-weighted images. For example, the ganglion cells/ganglioglioma shows isointense to gray matter on T1-weighted images, the astrocytoma or ependymoma shows hyperintense to white matter on T2-weighted images, as well as mesenchymal elements such as rhabdomyosarcoma, fibroblast or cartilaginous elements show isointensity on T1-weighted images and hyperintensity on T2-weighted images. Calcification and hemosiderosis present hypointense on T2-weighted images and fresh hemorrhage appears hyperintense on T1-weighted images. Heterogeneous enhancement is emerged on the solid parts with the rich vascular regions and the lacking vascular areas including reticular fibers or cartilage matrix. The present tumor manifested low-density areas seen by preoperative T1-weighted images, matching the extensive necrosis on pathological examinations. These radiological features are remarkably similar to those of teratoma and not so specific that it is very difficult



to distinguish MEM from teratoma, AT/RT, Ewing's sarcoma, MPNST and others. The three most common tumors located in the posterior third ventricle or pineal region are germ cell tumors, pineal tumors and gliomas. This case told us that in differential diagnosis of tumors in this region in infancy or childhood, even senior children, MEM should be taken into consideration for the rich vascular supply during operation.

### *Pathology*

Diagnosis of MEM mainly depends on the histological characteristics. The criteria for diagnosis are still ambiguous due to the rarity of MEM. The common view supported by many experts is that the primary intracranial MEM composes of neuroectodermal represented by ganglioglioma and mesenchymal elements represented by rhabdomyoblastoma, or rhabdomyosarcoma [13]. Characterized by the two kinds of components, the common neuroectodermal components may contain ganglioglioma, neuroblastoma, schwannoma, astrocytoma, ependymoma, MPNST and neurofibroma, whereas the common mesenchymal differentiation may present rhabdomyoblastoma/rhabdomyosarcoma, chondrosarcoma, liposarcoma, meningioma, reticular fibers, cartilage tissue and smooth muscle cells. One case reported by Kun Yao [10] in our institute was the only one that anaplastic ependymoma was the main part of neuroectodermal elements. The tumor in our case was composed of immature ganglion cells and astrocytoma, as well as mesenchymal components proliferating in myxoid background and malignant large round cells with non-specific but poor differentiation. The neoplastic large-round-cell elements resulted in the up to 20% of MIB1. From the analysis of CWS [12], the most rhabdomyosarcoma belonged to the embryonal histotype, which reflected the proportion of large-round-cell components and corresponds with the promordium tendency in mesenchymal series.

Molecular pathological data on MEM showed that the EWS/FLI-1 fusion protein expressed in MEM, the same as EWS and PNET [14]. The genes which overexpressed in MEM but were absent in MPNST, EWS and rhabdomyosarcoma include MAGP2, SULT1E1, PDGERL and CXCL13 [7]. MEM with chromosomal changes often involved chromosomes 2, 8 and 11, with

numerical abnormalities (+2, +8, +9, +11 and +20) and structural abnormalities such as add (6)(p24) and t(2;13) [15]. Fluorescence in situ hybridization (FISH) was performed in some cases to determine some genes copy number, for example, PTEN, FKHR or EGFR. The cytogenesis features illustrate that gene microarrays can be employed to discriminate rare tumors like MEM.

Diagnosis of MEM should be taken into account when neoplastic neuroectodermal and mesenchymal elements are seen in pathological examinations. MEM contains both two elements, which is valuable to exclude rhabdomyosarcoma without neuroectodermal components and EWS/PNET without rhabdomyosarcoma components in differential diagnosis. It is difficult to distinguish correctly between MEM and teratoma based on radiological features, however there are no ganglion cells or neuroblastoma in teratoma. Malignant fibrous histiocytoma is negative for Myoglobin and MSA, which are important points for identifying MEM. INI-1, immunonegative for AT/RT, was positive in both components of MEM.

### *Treatment*

Due to the rarity of these cancers, the standard therapy is not available and the most favorable strategy for MEM is surgical gross-total resection, followed by comprehensive treatment including chemotherapy, irradiation and anti-angiogenesis therapy. Neurosurgeons should strive for total resection of tumor, for no one other treatment can replace the benefits of perfect surgery. In our case, the gross-total resection was achieved via transcortico-ventricular-choroidal fissure approach following a right frontal craniotomy. The advantages of this approach as follows: a. Adverse damage to normal tissues could be reduced as much as possible with the best use of natural spaces. b. The interference of Galen vein system could be excluded as a result of exposure from the front of lesion. c. Third ventriculostomy could be performed consistently to avoid obstructive hydrocephalus [16], but it was not combined in our case because of the operative ventriculo-peritoneal shunt. Chemotherapy also plays an important role in treatment of MEM because rhabdomyosarcoma is the most sensitive to drugs [17] around all kinds of soft tissues. The



right drugs should be chosen based on the components of MEM. Due to rarity of intracranial MEM, the scheme of peripheric rhabdomyosarcoma should be referred to, such as vincristine (VCR), actinomycin D (ACD), cyclophosphamide (CTX) or ifosfamide (IFO). In addition, malignant neuroectodermal components may contain astrocytoma, neuroblastoma, anaplastic ependymoma et al and the selection of drugs should also be directed against these aggressive components. The effect of radiation is still controversial according to different results in reported cases. However, the effective role of local control after incomplete resection or without operation has been supported [18] and therefore our case was recommended to receive radiation to control the fraction of remnant attaching to Galan vein.

## Prognosis

A poor prognosis has been described in some literature and factors such as size, location, resectability, dissemination, pathological elements and response to chemo-radiotherapy seem to be concerned. According to Howley S [19], the prognosis of MEM is known to be worse than rhabdomyosarcoma. Age may contribute to the prognosis, as patients aged more than 10 years old could show a worse outcome [12]. The present 16-year-old patient had no recurrences in 6-month follow-up and a longer visit should be continued.

## Conclusion

Primary intracranial MEM is an exceedingly rare disease and the diagnosis for this disease is still challenging. The histological displays, immunophenotypical characteristics as well as cytogenetic arrays are still the reliable basis for diagnosis. Surgical resection has been demonstrated an effective treatment approach and other treatments such as radiotherapy, steroid-therapy and chemotherapy still need studies to clarify the usage and efficiency.

## Acknowledgements

This work was supported by the funding from Capital health research and development of special (No. Z151100004015165), Science and technology project of Beijing municipal commission of education (No. 350011620162) and National nature science foundation of China (No. 20873999).

## Disclosure of conflict of interest

None.

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