

Atypical Meningioma in the Medulla Oblongata Parenchyma

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Case Report

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Abstract

Background: Glioma is the most common tumor occurring in the brainstem. A primary intraparenchymal meningioma located in the brainstem without dura attachment is rare. Meanwhile, atypical meningiomas that occur in the medulla oblongata parenchyma, and without dura coverage, are extremely rare. In this study, we report the first case of atypical meningioma in the medulla oblongata parenchyma and review the existing literature.

Case presentation: A 38-year-old female was admitted at our hospital with a 2-week history of progressive neck and occipital pain. Magnetic resonance imaging revealed a presence of a 1.5x0.9cm mass lesion, located in the left side of the medulla oblongata, which was hypointense on T1-weighted and hyperintense on T2-weighted images, and with inhomogeneous enhancement following gadolinium-diethylenetriamine pentaacetic acid (Gd-DTPA) administration. The initial preoperative diagnosis was glioma or angioblastoma. The patient underwent a total surgical resection of the left medulla oblongata tumor and the histopathological examination indicated that the lesion was an atypical meningioma. The patient returned to normal life after surgery.

Conclusions: Although glioma is the most common tumor occurring in the brainstem parenchyma, the possibility of meningioma cannot be ruled out in this area.

Introduction

Glioma is the most common tumor occurring in the brainstem [1]. However, primary meningiomas that occur subcortically in the brainstem, and without dura coverage, are rare. Meanwhile, atypical meningiomas that occur in the medulla oblongata parenchyma, and without dura coverage, are extremely rare. Our review of the literature revealed fifteen well-described cases of complete intraparenchymal meningioma. These mostly affected the cerebral hemispheres of patients aged between 14-month and 66-year-old [2–16]. Only one case occurred in the medulla oblongata parenchyma, and was identified as meningothelial meningioma in 1985. In this study, we report the first case of atypical meningioma in the medulla oblongata parenchyma and review the existing literature.

Case Presentation

Clinical history

A 38-year-old female presented with a 2-week history of progressive neck and occipital pain. In the pre-admission week, the patient experienced increased pain, vomiting, dizziness and discomfort, coughing after swallowing and drinking water, and voice hoarseness. In addition to the later symptoms, the physical examination revealed that the patient was conscious and able to reasonably answer questions. The pupil was 3.0 mm on both sides, reflecting light sensitivity. There were no central face paralysis, tongue paralysis and facial numbness. The neck stiffness suggested weak positivity, and the left limb muscle strength was grade 4 with normal muscle tension. The left side instead of right side of the Babinski sign was positive. Magnetic resonance (MR) imaging revealed the existence of a 1.5 × 0.9 cm mass lesion, located in the left side of the medulla oblongata, that was hypointense on T1-weighted and hyperintense on T2-weighted images (Fig. 1A-B). The mass also showed an inhomogeneous enhancement following gadolinium-diethylenetriamine pentaacetic acid (Gd-DTPA) administration (Fig. 1C-E). The initial preoperative diagnosis indicated glioma and angioblastoma.

Under general anesthesia, the patient underwent a total surgical resection of the left medulla oblongata tumor using the far-left lateral approach. The tumor had a rotting fish meat appearance and was completely buried in the brain parenchyma with no dura mater around the tumor. The tumor was completely removed, and the operative course was uneventful.

Pathologic findings

The histological examination revealed the presence of epithelial-like polygonal tumor cells, with eosinophilic cytoplasm. The tumor cells were actively proliferating, with moderate atypia, some necrosis and pathological mitosis. The boundaries of the tumor cells were crowded and unclear (Fig. 2A-C). The immunohistochemistry profile showed positivity for Vimentin, epithelial membrane antigen (EMA) and PR (Fig. 2D-F), but negativity for cyto-keratin (CK), the glial fibrillary acidic protein (GFAP), Oligo-2, S-100, NSE, NeuN, CD34, CD31, CD56 and the PAX-8 protein (Fig. 2H-L). The Ki-67 proliferative index was approximately 30% (Fig. 2G). These findings were consistent with atypical meningioma (World Health Organization grade III).

The most common tumor, without dura in the brainstem parenchyma, was glioma, but immunohistochemical findings showed that the tumor cells GFAP and Oligo-2 were negative in this case. On the other hand, neurogenic tumors should also be considered from the perspective of imaging, and angioblastoma should not be excluded. So we tested S-100, NSE, NeuN, CD34, CD31. The results were also negative. In addition, we considered the possibility of neuroendocrine tumor and metastatic carcinoma (transparency renal cell carcinoma, grade II), while CD56, CK and PAX-8 were all negative. The immunohistochemistry profile showed positivity for Vimentin, EMA and PR. Eventually we favored the diagnosis of meningioma based on the morphological and immunohistochemical findings.

There was no evidence of tumor recurrence on postoperative MR imaging at the patient's 3-month follow-up visit (Fig. 3A-E). The patient's physical condition significantly improved after ten months follow-up period. She can independently eat, swallow, and normally communicate with others, without hoarseness.

Discussion

After literature review [2–21], we found fifteen well-described cases of complete intraparenchymal meningioma [Table 1]. The male-to-female ratio was 11:4 and their age ranged from 14-month to 66-year-old, and the median age was 15-year-old. It is more frequently encountered in children and adolescent, with most of lesions affecting the cerebral hemispheres. We found only three primary meningioma in the parenchyma of the posterior fossa and without dural

attachment. There was only one case occurred in the medulla oblongata in all of the English-language literatures, which was a meningothelial meningioma in 1985 and our case is the first atypical meningioma in the medulla oblongata parenchyma. The patient is a 38-year-old female, who is elder than most of the patients, but younger than common sites meningioma patients.

Meningiomas that are usually attached to the dura, are thought to originate from the (meningothelial) arachnoid cells or arachnoid cap cells [2, 11]. Nevertheless, the pathogenesis of primary intraparenchymal meningiomas is unclear. It was proposed that some arachnoid cap cells were located in the arachnoid or cerebral pia mater and far from the dura mater [2]. However, the cause of meningiomas in the intraparenchymal posterior fossa is unknown. This may be due to arachnoid cells of the pia mater, which enter the brain along with perforating blood vessels, or due to an ectopy of the arachnoid cells that are localized in the white matter, or due to arachnoid cells that rest during the brain development migratory progress [5, 9]. As one gets older, arachnoid cells proliferate and form tumors.

In general, gliomas are most common tumor in the brainstem [1]. But there are no typical radiological features of gliomas in this case. During the surgery, the tumor was observed in the medulla oblongata parenchyma, without dura mater around the tumor. Thus, it was difficult to distinguish meningioma from glioma at the preoperative and intraoperative stage.

The postoperative immunohistochemical staining showed negativity for GFAP, Oligo-2, S-100, NSE, CD34, CD31 and PAX8. Gliomas, neurogenic tumors, angiogenic tumors, and high-grade clear cell carcinoma of kidney origin were excluded. Positivity for Vimentin, EMA and PR proteins confirmed the meningioma of the lesions. According to the obvious cell atypia, the high Ki-67 index, and pathological mitosis, the final diagnosis indicated an atypical meningioma in the medulla oblongata parenchyma.

Most meningiomas can be completely excised by surgery [22]. Some patients received postoperative radiotherapy due to the atypical characteristic of the meningioma and residual tumor growth [2, 5]. Whether atypical meningiomas patients need postoperative radiation therapy, remains controversial. Some studies showed that there was no significant benefit for progression-free survival or overall survival after adjuvant radiotherapy for atypical meningiomas [23–25]. Compared with benign meningioma, atypical meningioma is a tumor with a relatively poor prognosis and with a recurrence rate of approximately 29%-52% [22]. The main recurrence factors that affect are related to the extent of the surgical resection, the tumor site and invasion extent, the vital adjacent structure of the tumor and the surgeon skills [22, 25]. In this case, the tumor localized in the medulla oblongata parenchyma of the brainstem. The tumor was completely removed. Postoperatively, the patient refused radiotherapy.

Conclusions

Glioma is the most common tumor occurring in the brainstem [1]. In this case, a 38-year-old female patient was diagnosed with atypical meningiomas that localized in the medulla oblongata parenchyma of the brainstem. The lesion preoperative diagnosis was difficult due to the atypical imaging features. This study is the first to report atypical meningiomas in the medulla oblongata parenchyma, which will help clinicians and imaging doctors to better understand this disease and pay more attention to the differential diagnosis of tumors in the brainstem parenchyma. The possibility of meningioma cannot be ruled out in this area. A complete resection is important for favorable prognosis, but long-term follow-up is necessary.

Abbreviations

MR: Magnetic resonance; Gd-DTPA: Gadolinium-diethylenetriamine pentaacetic acid; EMA: Epithelial membrane antigen; PR: Progesterone protein; CK: cytokeratin; GFAP: Glial fibrillary acidic protein; NSE: Neuronal enolase

Declarations

Acknowledgements

Not applicable.

Authors' contributions

XQ designed the study, screened and analyzed radiographic images from the literature. PZ was responsible for collection of clinical data. HM provided the MRI imaging Figures. PF made the pathologic diagnosis of the primary tumor and wrote the original manuscript. All authors read and approved the final manuscript.

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Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Ethics approval and consent to participate

Not applicable.

Consent for publication

Informed consent for publication was obtained from the patient's family.

Competing interests

The authors declare that they have no competing interests.

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Table

Table 1
Literature summary of well-described cases of complete intraparenchymal meningioma.

	Author	Age (y.s.)	sexx	Symptoms	Location of Lesion	CT	MRI	Size (mm)	Surgical resection	C
1	Zhang, et al., 2007 ²	16	M	epileptic seizure	right parietooccipital region	not stated	T1.,isointense; T2.,hyperintense; heterogenous enhancement	25 × 23	Total	N
2	Reynolds, et al., 2016 ³	15	M	right knee pain, weakness and ataxic gait	left basal ganglia	a large, densely calcified mass	T1.,isointense; T2.,hyperintense; heterogenous enhancement	not stated	Subtotal	ri h a e; a
3	Jiang, et al., 2012 ⁴	23	M	nerve palsy and dysphasia	in the brainstem, pons and right cerebral peduncle	not stated	T1.,isointense; T2.,isointense; homogeneous enhancement	35 × 25 × 20	Total	N
4	Liu, et al., 2018 ⁵	8	M	headache and vomiting	right basal ganglia extending to superasellar cistern	an isodense-to hyperdense lesion	T1., isointense; T2., isointense; inhomogeneous enhancement	40 × 44	Subtotal	ir vi w cr lii
5	Karadereler, et al., 2004 ⁶	14	M	headache and seizure	right temporal region	not stated	T1.,slightly hypointense; T2., hyperintense; heterogenous enhancement	15 × 15	Total	N
6	Shimbo, et al., 2011 ⁷	10	M	vomiting, fever and seizure	left frontal lobe	an isodense mass lesion, with homogeneous enhancement by contrast medium	T1., isointense to hypointense; T2., isointense; homogeneous enhancement	20 × 22	Total	N
7	Werbrouck, et al., 2014 ⁸	13	M	loss of consciousness and generalized convulsions	right temporal lobe	a mild hyperdense lesion	T2., heterogeneous hypointense; T1,homogeneous enhancement	15 × 18 × 34	Total	N
8	Legius, et al., 1985 ⁹	14-month	M	right sided hemiconvulsions	left hemisphere	a hyperdense lesion, enhanced by contrast medium	Not stated	20 × 20	Total	N
9	Tekkök, et al., 2005 ¹⁰	54	F	headache and nausea	right temporal lobe	an enhancing intra-axial mass	T1., isointense; T2., isointense	not stated	Total	N
10	Jadik, et al., 2014 ¹¹	42	M	seizure	right parietal lobe	a hyperdense lesion with calcification	T1.,hypointense; T2.,mixed hypo- and hyperintense	15 × 15	Total	N
11	Zhao, et al., 2011 ¹²	42	M	headache and dizziness	left temporal lobe	not stated	solid component: T1., homogeneous enhancement ; T2.,low intensity	not stated	Total	N
12	Mut, et al., 2000 ¹³	20	F	temporal-type seizures	right temporal lobe	not stated	T1.,hypointense; T2.,hyperintense	not stated	Total	N
13	Matsuda,et al., 2019 ¹⁴	66	F	none	right basal ganglia	a small lesion with tiny calcifications	T1.,homogeneous enhancement ;	not stated	Biopsy	n d
14	Sakaki, et al., 1987 ¹⁵	12	M	not stated	one in the right frontal parasagittal region, another in the region of the foramen magnum	high density lesions with homogeneous enhancement	not stated	17 × 13	Total	N
15	Miranda, et al., 2009 ¹⁶	10	F	neck pain, right hemiparesis, gait instability and somnolence.	right side of the craniocervical junction	enhancement	T1.,intense contrast enhancement	40 × 35 × 45	Total	N

Author	Age (ys.)	sexx	Symptoms	Location of Lesion	CT	MRI	Size (mm)	Surgical resection	C
16 Present case	38	F	neck and occipital pain,vomiting, dizziness	left side of the medulla oblongata	no obvious lesion	T1.,hypointense;T2.,hyperintense; inhomogeneous enhancement ;	15 × 9	Total	N

Figures

Figure 1

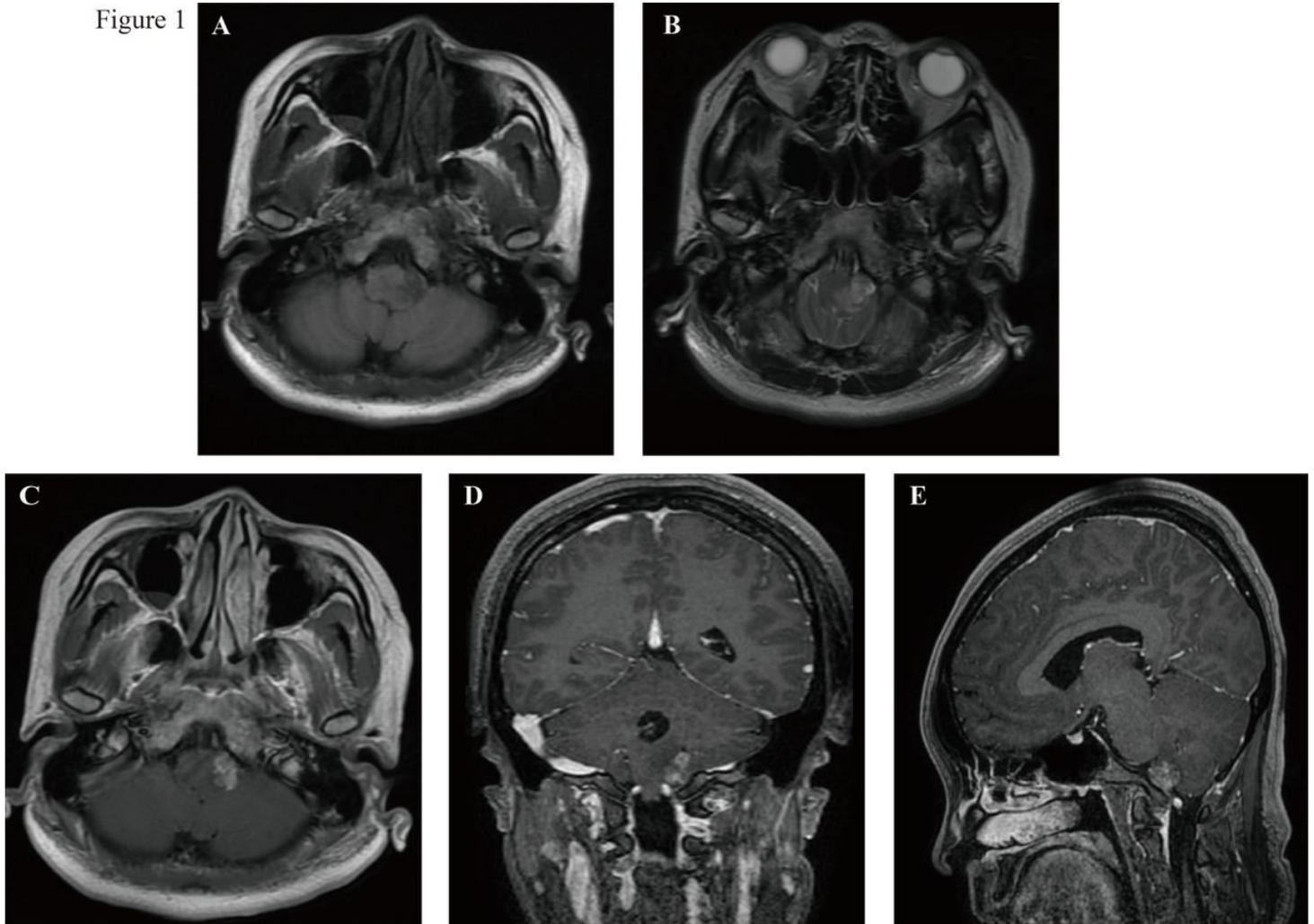


Figure 1

Magnetic resonance imaging of the left side of the medulla oblongata lesion. Axial sections from a non-contrast head MR imaging demonstrate the presence of a 1.5x0.9cm mass lesion, located in the left side of the medulla oblongata. The lesion was hypointense on T1-weighted and hyperintense on T2-weighted images (A-B), inhomogeneous enhancement following Gd-DTPA administration (C) axial, (D) coronal and (E) sagittal reconstructions.

Figure 2

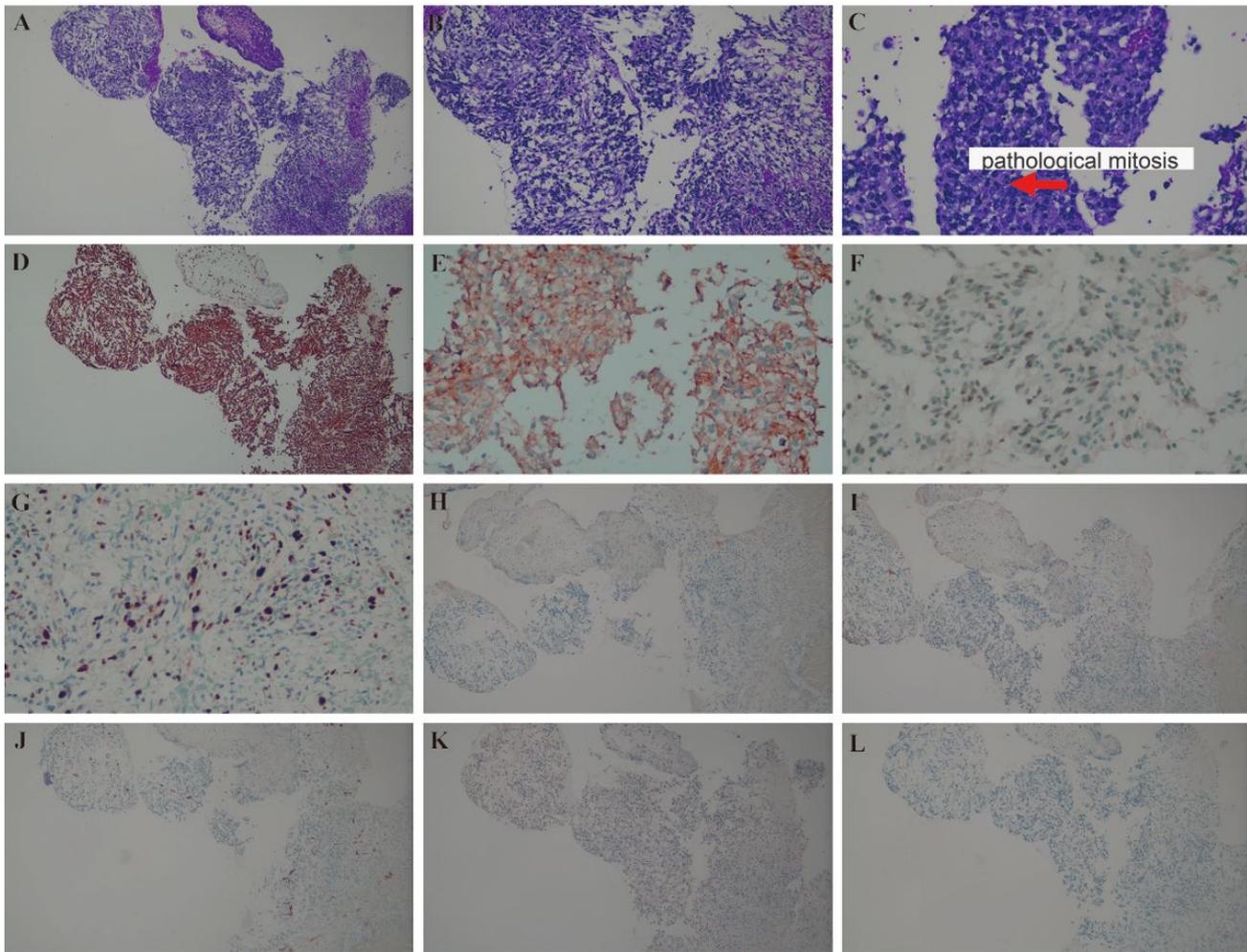


Figure 2

Hematoxylin-eosin staining exhibiting nests of tumor cell growth, with active proliferation, moderate atypia, some necrosis, and pathological mitosis (A: $\times 100$; B: $\times 200$; C: $\times 400$). Immunohistochemical staining showing positivity for Vimentin, EMA and PR (D: $\times 100$; E-F: $\times 400$), but negativity for GFAP, S100, CD34, PAX-8 and the CD56 protein (H-L: $\times 100$). The Ki-67 labeling index was approximately 30% (G: $\times 400$).

Figure 3

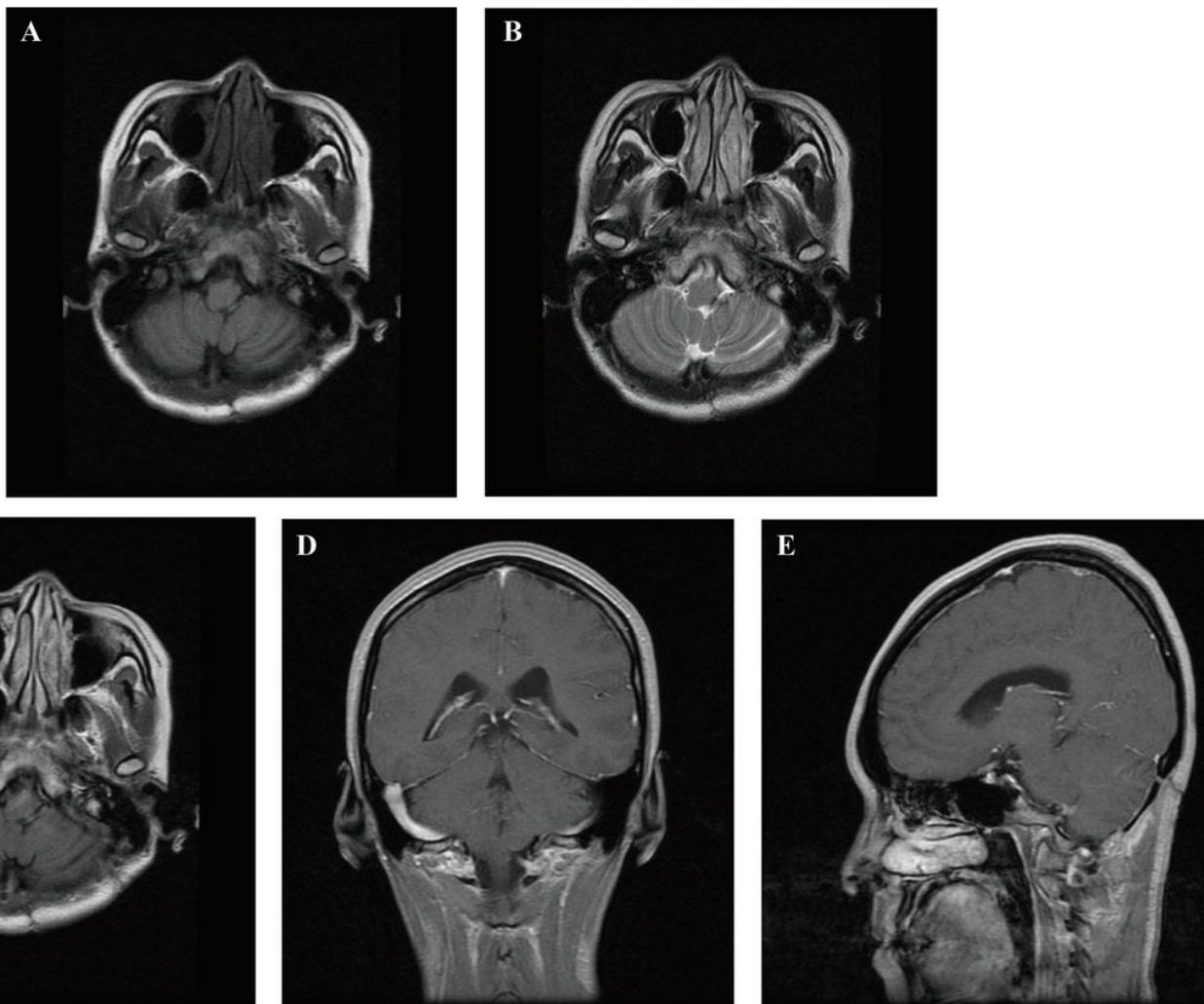


Figure 3
Postoperative MR imaging. Axial T1-weighted and T2-weighted imaging (A-B), enhancement following Gd-DTPA administration (C) axial, (D) coronal and (E) sagittal reconstructions.

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