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CASE REPORT



Intraparenchymal atypical meningioma in the posterior fossa: a case report and literature review

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ABSTRACT

Intraparenchymal meningiomas without dural attachments are extremely rare. A 32-year-old female adult was admitted to our hospital, complaining of occasional dizziness. The patient had no neurological deficits. MRI demonstrated a lesion with mild edema located in the left cerebellar parenchyma. CT revealed calcification within the mass. Gross total resection was achieved. The histopathological examination indicated that the lesion was an atypical meningioma (WHO-II). We herein report an extremely rare case of an intraparenchymal meningioma located in the left cerebellar hemisphere. The significance of the differential diagnosis of lesions in the cerebellum should be emphasized.

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Intraparenchymal meningioma; posterior fossa; atypical meningioma

Introduction

Meningiomas account for 25%–30% of intracranial neoplasms.¹ They are usually attached to the dura, and are thought to arise from arachnoid cap or meningotheial cells. Occasionally, meningiomas develop without dural attachments. They are frequently located in the intraventricular, sylvian fissure, or pineal region.² The primary occurrence of meningiomas without attachments to the dura are rare,³ and meningiomas located in the intraparenchymal/subcortical region without dural attachments are extremely rare. According to the literature, intraparenchymal meningioma is a rare pathology with fewer than 40 reported cases.⁴ Here, we report a rare case of primary intraparenchymal atypical meningioma in the posterior fossa and a review of the related literature on its classification, clinical characteristics, radiological features, differential diagnosis, management and prognosis.

Case report

A 32-year-old female patient was admitted to a hospital with the complaint of occasional dizziness. Apart from this complaint, the patient had no neurological symptoms. The CT scan (Figure 1(A)) revealed an iso-to-hyperdense lesion in the left cerebellar parenchyma, 2.7 × 2.3 cm in size, with peripheral calcification. In the area of the mass lesion, MRI demonstrated (Figure 1(B–E)) low-intensity signals on T1-weighted images and high-intensity signals on T2-weighted images and images with homogeneous enhancement, and the dural tail sign was not observed. Postoperative CT (Figure 1(F)) and MRI (Figure 1(G–J)) showed that the tumor had been completely removed.

For the surgical procedure, under the guidance of ultrasound, the transcortical approach was used to explore the lesion. No structures associated with the dura matter were found. A solid avascular lesion was totally removed. During the postoperative period, the patient did not have any neurological deficits.

The histopathological examination (Figure 2(A-a,B-b)) revealed features of an atypical meningioma (WHO grade II) with the formation of psammoma bodies, the active proliferation of tumor cells, and a local tumor invading the cerebellar parenchyma. The immunohistochemistry findings showed that the tumor was positive for somatostatin receptor type 2 (SSTR2) and tumor-associated epithelial membrane antigen (EMA) (Figure 2(C,D), respectively).

Discussion

Classification

Intracranial infratentorial meningiomas without dural attachments are very rare. According to site of the tumor, meningiomas of the posterior fossa without dural attachments have been classified by Abraham and Chandy⁵ into the following three categories: (1) meningiomas arising from the choroid of the fourth ventricle and lying wholly within it; (2) meningiomas arising from the inferior tela and lying partially in the fourth ventricle and partially in the cerebellar hemisphere; and (3) meningiomas lying in the cisterna magna. Moreover, three other categories of meningiomas have been reported, which include (4) meningiomas that arise from the choroid plexus and develop in the lateral cerebellomedullary cistern,⁶ (5) meningiomas that arise from the arachnoid tissue near the foramen of Luschka,⁷ and (6) intraparenchymal meningiomas.⁸ Therefore, there are 6 types of posterior fossa meningiomas without dural attachments.

Pathogenesis

The pathogenesis of meningiomas without dural attachments remains unclear. Two main theories^{4,9–11} have been proposed to explain the possible mechanism: (1) meningiomas arise from arachnoid cells of the pia mater and enter the brain along the perforating blood vessels; (2) meningiomas that arise from the

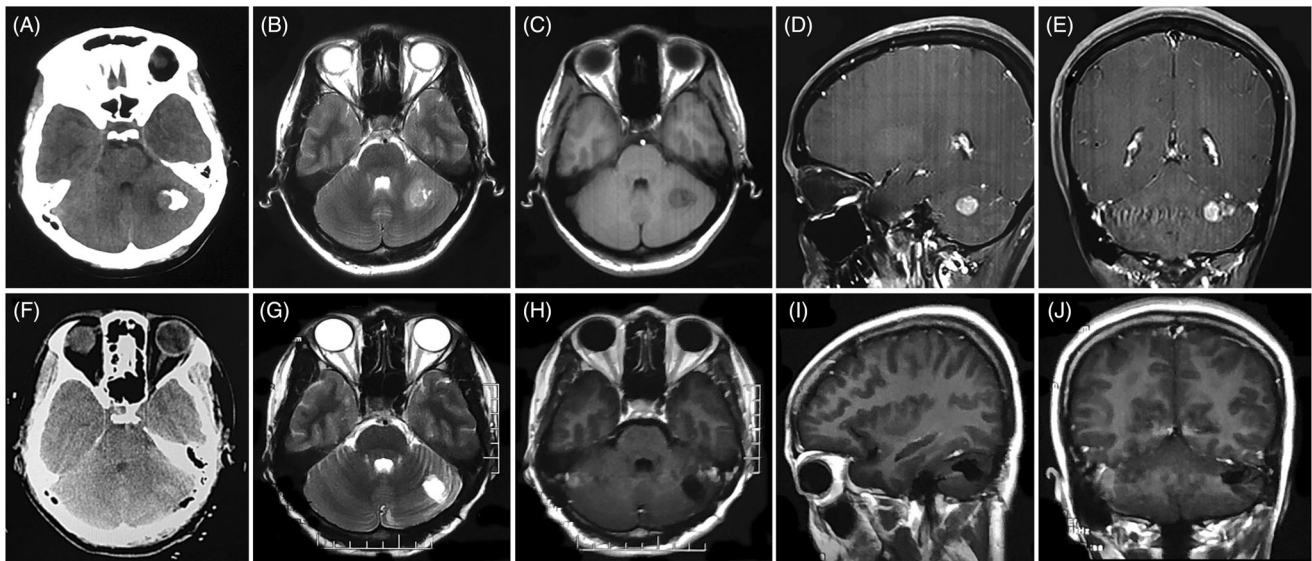


Figure 1. Intraparenchymal meningioma located in the left cerebellar hemisphere. Preoperative CT (A) showed revealed an iso-to hyperdense lesion in the left cerebellar intraparenchyma, 2.7×2.3 cm in size with peripheral calcification. MRI demonstrated (B–E) the mass lesion was low-in-tense on T1-weighted and high-in-tense on T2-weighted, with homogeneous enhancement. No dural tail sign was noted. Postoperative CT (F) and MRI (G–J) showed that the tumor was completely removed.

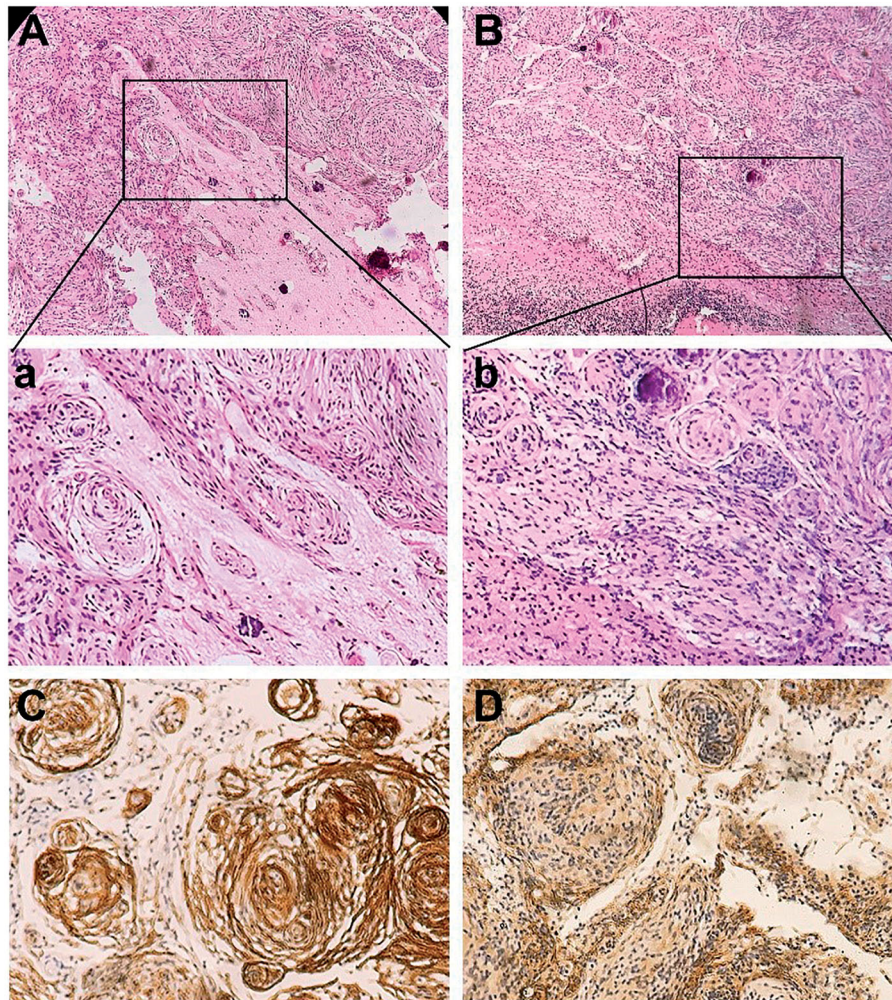


Figure 2. Histopathological features of the atypical meningioma case. HE staining (A and B, $100\times$) revealed that the tumor invades the cerebellar parenchyma (the invasion boundary is enlarged in (a), $250\times$), and the location of the tumor to the cerebellum (the junction is enlarged in (b)). Large number of psammoma bodies formation, active proliferation of tumor cells and necrotic foci were also observed. Immunohistochemistry staining showed that the tumor was positive for somatostatin receptor type 2 (SSTR2, (C), $250\times$) and tumor-associated epithelial membrane antigen (EMA, (D), $250\times$), respectively.

Table 1. Summary of 20 cases of reported meningiomas without dural attachment in posterior fossa excluding the one in the fourth ventricle.

Location	Case No.	Author/year	Age/sex	Extend of resection	Pathology
Cisterna magna ¹⁷	1	Martin <i>et al.</i> /1923	45/M		Fibrous
	2	Spurling <i>et al.</i> /1938	16/F		NA
	3	Nicoletti <i>et al.</i> /2001	53/M	GTR	Meningothelial
	4	Masaaki Kohta <i>et al.</i> /2013	36/F	GTR	Clear cell
	5	Masaaki Kohta <i>et al.</i> /2013	58/M	GTR	Meningothelial
Inferior tela corioidea	1	Olivekrona <i>et al.</i> / 1927 ⁵			
	2	Schreiber <i>et al.</i> / 1936 ⁵			
	3	Cristophe <i>et al.</i> / 1937 ⁵			
	4	Jacob Abraham. MS <i>et al.</i> /1963 ⁵	40/F	NA	Fibroblastic
	5	Giampaolo Cantore <i>et al.</i> /1983 ¹⁴	22/M	NA	Fibroblastic
	6	B. Chidambaram <i>et al.</i> /1997 ¹⁵	8/F	STR	Transitional
	7	Tae-Young Jung <i>et al.</i> /2009 ¹⁶	72/F	GTR	Fibroblastic
intraparenchymal meningiomas	1	Nakahara <i>et al.</i> /1993 ²⁰	56/M	GTR	Fibroblastic
	2	Jennifer G.C. Teo <i>et al.</i> /1998 ⁸	2/F	PR	Clear cell
	3	Jiang <i>et al.</i> /2012 ²¹	23/M	GTR	Papillary
foramen of Luschka		Our case/ 2019	32/F	GTR	Atypical
	1	Daiys Ishigaki <i>et al.</i> /2007 ⁶	14/M	GTR	Meningothelial
lateral cerebello-medullary istern	2	Pablo Miranda <i>et al.</i> /2009 ¹⁹	10/F	GTR	Clear cell
	1	Masato Shibuya <i>et al.</i> /1999 ⁷	61/F	STR	Meningioma
	2	Seong Min Kim <i>et al.</i> /2010 ¹⁸	59/F	GTR	Meningothelial fibroblastic

F: female; M: male; NA: not available; GTR: gross total resection; STR: subtotal resection; PR: partial resection.

pia mater of the brain sulcus, adhere and compress the brain parenchyma, and grow into the intraparenchymal lesion, causing the mass to be appear completely buried in the parenchyma.

Clinical characteristics

Intraparenchymal meningiomas are included in the same category of subcortical meningiomas and are located in the brain parenchyma without dural attachments, occasionally reaching the brain surface.¹² Based on the imaging and intraoperative findings, the present case should be classified as an intraparenchymal meningioma.

To our knowledge, 47 cases of meningiomas of the posterior fossa without dural attachments have been reported in the English scientific literature. Among these 47 cases, only 3 cases of intraparenchymal infratentorial meningiomas have been reported. The first case was a lesion located in the lateral region of the right cerebellar cortex, and the mass was an extramedullary tumor that had no relation with the cerebellar cortex or dura matter. The second case was located in the upper medulla and pons. Similarly, the third case was located in the pons. Herein, including present case, only 20 cases of infratentorial meningiomas without dural attachments have been reported in the literature (Table 1), excluding cases in the fourth ventricle (28 cases);¹⁰ they were excluded because their features are quite different from those of the other types of meningiomas without dural attachments.¹³ There were 7 cases originating from the inferior tela choroidea,^{5,14-16} as 3 cases were excluded from the clinical data we retrieved from the literature, as they lacked integrity; 5 cases were located in the cisterna magna,¹⁷ 2 cases were located in the cerebellomedullary cistern,^{7,18} 2 cases were located in the foramen of Luschka^{6,19} and 4 cases were intraparenchymal cases,^{8,20,21} including the present case.

The average age of the 17 patients was 35.7 years (range, 2–72 years), which was younger than the age of the patients with lesions in the fourth ventricle (44.7 years)¹⁰ but much older than that of the patients with supratentorial intraparenchymal meningiomas (less than 20 years).¹¹ The male to female ratio was similar (7:10) to that of the patients with lesions in the fourth ventricle (12:15) and differed from that of the patients with supratentorial intraparenchymal meningiomas, which mostly occur in males. Among the other types of meningiomas,

fibroblastic and meningothelial meningiomas were the most common types, but the ratio of WHO grade II and III cases (31%) seemed to be higher. Because only 16 (one case was not available) cases were included, our ability to reach a firm conclusion was limited.

Radiological features and differential diagnosis

In general, meningiomas without dural attachments in the posterior fossa do not have a characteristic clinical pattern, and intracranial hypertension and cerebellar signs are the most common clinical symptoms. The radiological features of meningiomas without dural attachments in the posterior fossa show CT scan hyperdensity, low- or iso-intensity signals on T1-weighted MRI scans, iso- or high-intensity signals on T2-weighted MRI scans, and homogeneous enhancement on both MRI and CT scans, including cystic components, calcification, large volumes and peritumoral edema. The key difference between these meningiomas and other meningiomas is the absence of a dural attachment.²² The main lesions considered in the differential diagnosis of intraparenchymal meningiomas assess by radiography includes gliomas, lymphomas and germinomas. Other lesions include choroid plexus papillomas, ependymomas, medulloblastomas, and metastatic tumors. However, it is still very difficult diagnose intraparenchymal meningiomas preoperatively.

Management and prognosis

Due to the rarity of meningiomas without dural attachments in the posterior fossa, no clear consensus has been achieved regarding the proper management. Gross total resection (GTR) is the treatment of choice.²³ Whether postoperative radiotherapy should be performed for malignant meningiomas is controversial. Some people believe that meningiomas can be induced by radiation.²⁴ Others have suggested that adjuvant radiotherapy might delay the recurrence of malignant meningiomas or the progression of residual meningiomas.²⁵ In previous studies, the follow-up period has ranged from 6 months to 7 years. The shortest time to recurrence reported is 9 months; the patient was a 23-year-old female with a meningioma in the brainstem parenchyma, reported in a study by Jiang *et al.*, and the pathology was papillary.²¹

In addition, Masaaki Kohta *et al.* reported a 58-year-old male with a meningioma with meningotheial cells in the cisterna magna without recurrence with the 7-year follow-up period. Due to the small number of cases and uncertainty of long-term outcomes, it is difficult to make a conclusion about the prognosis. In general, the prognosis is worse in pediatric populations than in adult populations, and the prognosis depends on the degree of excision, pathologic grade, tumor location and association with meningiomas.²³

Conclusion

Meningiomas in the posterior fossa without dural attachments are extremely rare, and this is the first report of an intraparenchymal atypical meningioma in the left cerebellar parenchyma; it is quite difficult to preoperatively diagnose these meningiomas. However, the tumor should be removed completely.

Patient consent

The patient provided written informed consent for the publication and the use of their images.

Disclosure statement

No potential conflict of interest was reported by the author(s).

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