

# A case report on cystic meningioma in cerebellopontine angle and recommendations for management

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## Abstract

**Rationale:** Cystic meningioma located at the cerebellopontine angle (CPA) is an extremely rare occurrence. It is frequently misdiagnosed preoperatively. Little is known about the clinical features and outcome of this rare disease.

**Patient concerns:** A 70-year-old male presenting with progressive headache and gait disturbance.

**Diagnosis:** According to the symptoms, signs, and Gd-enhanced magnetic resonance images (MRI), a preoperative diagnosis of hemangioblastoma located in left CPA was made. Finally, the histological examination revealed a meningioma.

**Interventions:** A complete resection, including the part of the solid mass together with cyst, was performed.

**Outcomes:** The postoperative course of the patient was uneventful, and no residual or recurrent tumor was found during the 24-month follow-up period.

**Lessons:** Cystic meningioma should be included in the differential diagnosis of a CPA mass with atypical radiologic features, such as a large cyst and enhanced mural nodule. By summarizing the related literature, we found that the most common pathological subtype of CPA cystic meningioma is the clear cell subtype, which belongs to WHO grade II. Gross total resection including the enhanced cyst wall is extremely important. A close follow-up is necessary because of the high recurrence rate in this subset of meningioma.

**Abbreviations:** CNS = central nervous system, CPA = cerebellopontine angle, CTA = computed tomography angiography, MRI = magnetic resonance images, VS = vestibular schwannoma, WHO = World Health Organization.

**Keywords:** cerebellopontine angle, cystic meningioma, radiological features, therapeutic strategy

## 1. Introduction

Intracranial meningioma represents the most common primary brain tumor.<sup>[1]</sup> Radiologically, they frequently behave as homogeneously enhanced dural-based masses.<sup>[2]</sup> Meningiomas with large cyst and enhanced mural nodule are a rare occurrence and are frequently misdiagnosed preoperatively.<sup>[3–5]</sup> Although

limited cases of cystic meningiomas have been reported in the literature, most of them were found in the cerebral hemisphere,<sup>[6,7]</sup> cerebellopontine angle (CPA) cystic meningioma is an extremely rare occurrence and has rarely been reported. Here, we report an extremely rare case of a cystic meningioma in the left CPA and examine previously reported cases of CPA cystic meningiomas in an attempt to provide an up-to-date summary of the condition.

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All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

The patient has consented to submission of this case report to the journal.

The authors have no conflicts of interest to disclose.

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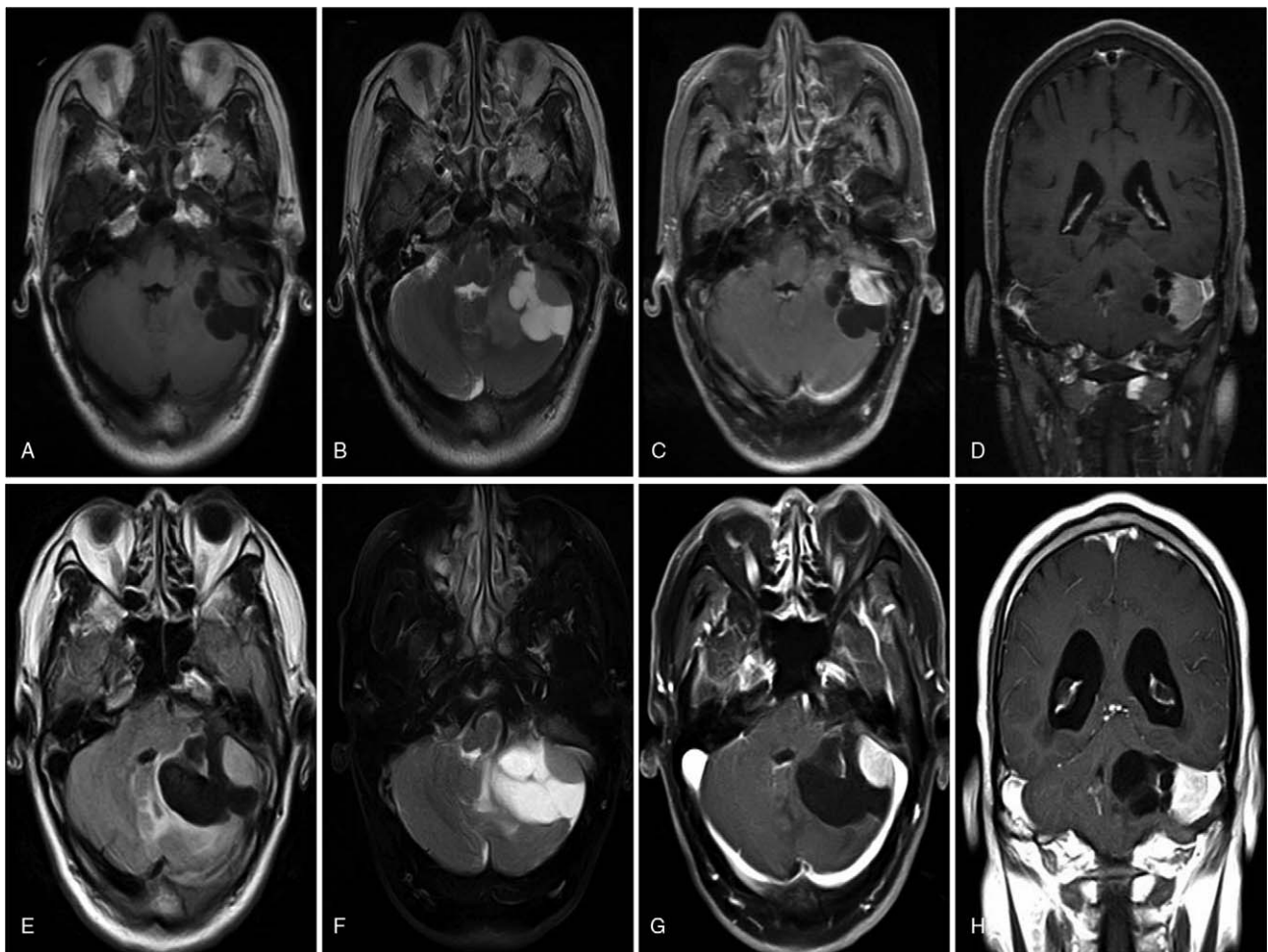
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## 2. Case report

A 70-year-old man presented to our department with a 2-year history of a headache. Postcontrast magnetic resonance images (MRI) indicated a broad based solid enhancing tumor with a multilobulated enhancing peritumoral cyst at the left CPA (Fig. 1). Because the patient refused to undergo surgery, a wait-and-see strategy and close follow-up were applied. Two years later, he suffered from a worsened headache as well as gait disturbance. While intracranial MRI showed that the tumor had spread, enhancement of the cyst wall faded (Fig. 1). Besides, preoperative computed tomography angiography (CTA) did not find any hypervascular nodule. Subsequently, a complete resection, including the part of the solid mass together with cyst, was performed. Intraoperatively, the solid part showed a rich blood supply. Postoperative histopathological examination surprisingly revealed the diagnosis of meningothelial meningioma (WHO grade I) (Fig. 2). The postoperative course of the patient was uneventful, and no residual or recurrent tumor was found during the 24-month follow-up period.



**Figure 1.** Comparison of brain MRI of a 2-year interval. (A) Axial T1-, (B) T2-, (C) enhanced T1-, and (D) coronal enhanced T1-weighted MRI showed a broad based solid enhancing tumor with a multilobulated enhancing peritumoral cyst in the left CPA. (E) Axial T1-, (F) T2-, (G) enhanced T1-, and (H) coronal enhanced T1-weighted MRI of the same patient 2 years later showed a larger tumor especially an enlarged cyst and a fading enhancing rim of the cyst. MRI = magnetic resonance images, CPA = cerebellopontine angle.

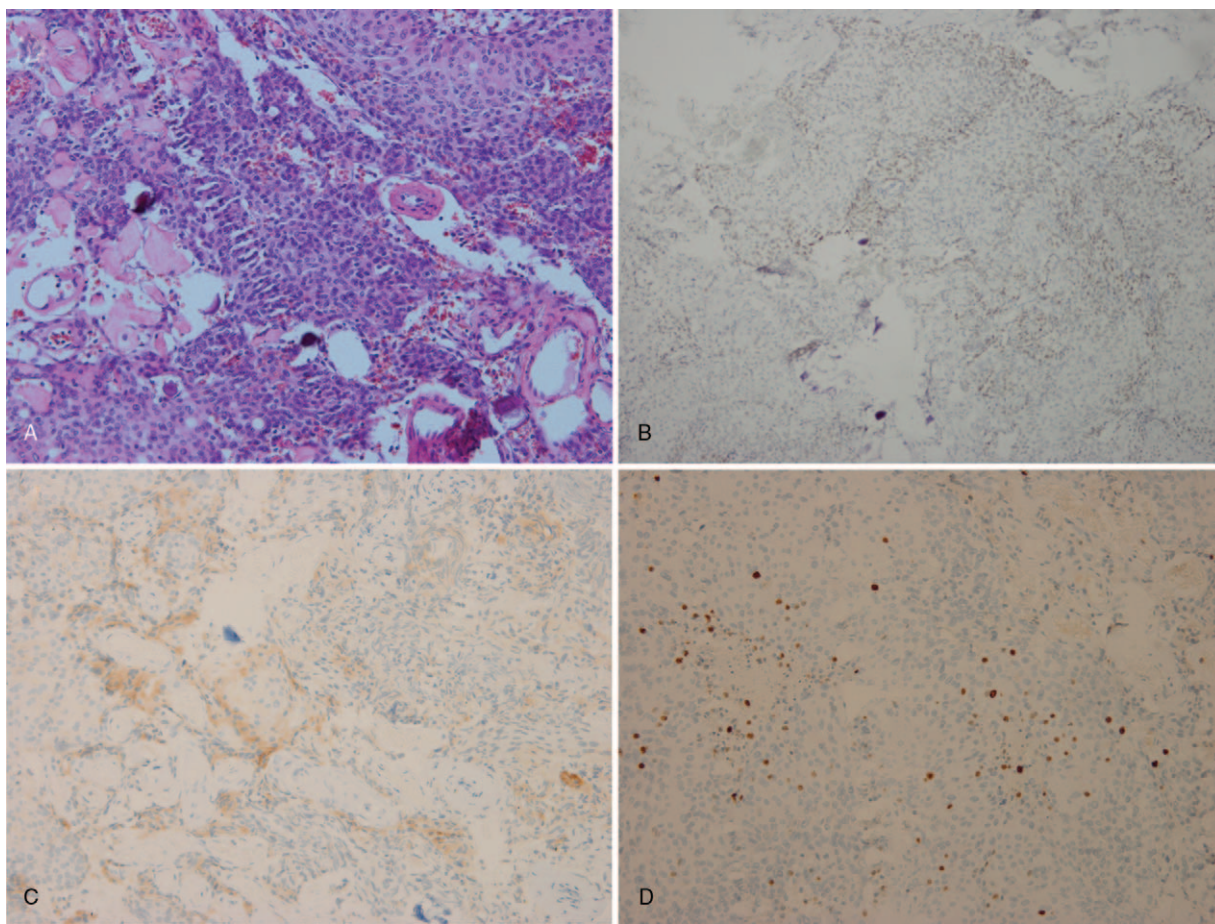
### 3. Discussion

Cystic meningiomas account for only 2% to 7% of all meningiomas and frequently occur in the cerebral hemisphere.<sup>[6,8–10]</sup> CPA cystic meningioma is a rare occurrence; only 11 cases have been reported in the English-language literature.<sup>[4,5,11,12]</sup> The pathogenesis, clinical features, and outcome of this rare disease remain undefined. In the present study, we report a case of CPA cystic meningioma with MRI findings and provide an up-to-date summary of CPA cystic meningiomas based on the literature reviews.

The pathogenesis of cystic formation in meningiomas remains controversial. Some authors believe that the etiology of cyst formation is different according to the location of the cyst.<sup>[13]</sup> Peritumoral cysts may frequently be caused by peritumoral edema into cyst, peritumoral demyelination, intratumoral hemorrhage, or entrapment of cerebrospinal fluid.<sup>[14–17]</sup> However, intratumoral cysts may result from ischemic necrosis, cystic degeneration, intratumoral hemorrhage, and active secretion of tumor cells.<sup>[2,13,14,18,19]</sup> In our opinion, as the solid part of cyst is frequently characterized by a rich blood supply,<sup>[5,12,20]</sup> we supposed that cyst formation in a meningioma might result from ultrafiltrate of tumor vessels and that increased permeability of

tumor vessels may accelerate cyst formation. As stated, there is still uncertainty as to the exact pathogenesis of this phenomenon. Further studies, like the analysis of the contents of cystic fluid, as well as exploring genetic differences, are necessary to elucidate the preferred mechanisms of cystic formation in meningiomas.

We reviewed a total of 12 cases of cystic meningiomas located in CPA (Table 1),<sup>[4,5,11,12]</sup> which exhibited several distinctions from common meningiomas. First, headache and hearing problems are the most common symptoms in CPA meningiomas, followed by gait disturbance, facial nerve involvement, and visual disorder. The duration of time is relatively short, varying from 2 to 48 months (median, 3 months). Second, different from the common meningiomas with a women predominance,<sup>[21,22]</sup> CPA cystic meningiomas prefer to occur in young patients (median age 27.5 years), and no gender predominance was found. Third, in regards to the radiological features, the size of the cystic meningioma in CPA is relatively large, with a median largest diameter of 50 mm (range 30–60 mm). Furthermore, peritumoral cysts are relatively ordinary compared with intratumoral cysts (80% vs 20%).<sup>[23]</sup> Enhancements of the cyst wall and peritumoral edema were observed in 3 of 6 (50%) and 11 of 12 (92%) patients. To our knowledge, this is the first case that



**Figure 2.** Pathological findings. A, Meningeothelial cells represented the majority of tumor stroma (HE, ×200). B, Immunoreactivity of progesterone receptor (PR, ×100). C, Epithelial membrane antigen (EMA) staining was positive (EMA, ×200). D, MIB-1 positive rate was less than 5% (MIB-1 × 200). In conclusion pathological findings indicate a meningeothelial meningioma.

**Table 1**  
**All cases of cystic meningioma located in CPA in the literature.**

Authors	Case (no)	Age (y)/sex	Duration of symptoms, mo	Clinical features	Size, mm	Histological subtype	WHO classification	Nauta et al <sup>[23]</sup> classification*	Enhancement of cyst wall	Treatment	Outcome	Follow-up, mo
Yu et al <sup>[11]</sup>	1	17/F	ND	Headache, hearing loss	55 × 50 × 40	Clear cell	Grad II	Typell	N	PR	ND	24
Ercan et al <sup>[4]</sup>	2	36/F	ND	Decreased hearing, hemiparesis	50 × 40 × 40	Meningeothelial	Grad I	Typell	Y	GTR	Improved	6
Deb et al <sup>[5]</sup>	3	58/M	ND	Headache, vomiting, gait disturbance	56 × 41 × 34	Angiomatous	Grad I	Typell	Y	GTR	ND	60
Wang et al <sup>[12]</sup>	4	23/M	2	Hearing loss	40 × 40 × 20	Clear cell	Grad II	ND	ND	GTR	Improved	63
	5	43/F	2	Gait disturbance, hearing loss	ND	Clear cell	Grad II	ND	ND	GTR	Recurrence	49
	6	13/F	3	Gait disturbance	50 × 40 × 40	Clear cell	Grad II	Typell	N	STR	Progression	47
	7	9/M	2	Right eye esotropia	30 × 20 × 20	Clear cell	Grad II	ND	ND	STR+RT	Improved	24
	8	8/M	2	Raucitas, bucking, gait disturbance	60 × 30 × 25	Clear cell	Grad II	ND	ND	STR+RT	Recurrence	18
	9	32/F	4	Headache, facial numbness, hearing loss	35 × 30 × 30	Clear cell	Grad II	ND	ND	STR+RT	Improved	15
	10	42/F	24	Headache, swallow disturbance	30 × 20 × 10	Clear cell	Grad II	ND	N	GTR+RT	Improved	13
	11	14/M	48	Headache, hearing loss	30 × 30 × 25	Clear cell	Grad II	ND	ND	GTR	Improved	11
Present case	12	70/M	24	Headache, gait disturbance	50 × 50 × 40	Meningeothelial	Grad I	Typelll	Y	GTR	Improved	12

CN = cranial nerve, CPA = cerebellopontine angle, F = female, Grad = grade, GTR = gross total resection, M = male, N = no, ND = no display, PR = partial removal, RT = radiotherapy, STR = subtotal removal, Y = yes.

\* Nauta et al classification: Type I: intratumoral cysts are centrally located within the tumor, Type II: intratumoral cysts are peripherally located within the tumor, Type III: peritumoral cysts are located within the adjacent brain, Type IV: peritumoral cysts are located between the tumor and the brain.

reports spontaneously vanishing enhancement of the cyst wall in a central nervous system (CNS) tumor after a long-term interval. We propose that the reason might be there could be normal variability in the contrast enhancement when performed at 2 different time points. Moreover, windowing may also have an influence. Fourth, the clear cell subtype is the most common pathological subtype and accounts for 9 of 12 (75%) patients. Other subtypes include meningothelial and angiomatous subtypes; as clear cell meningiomas is classified as WHO grade II,<sup>[24]</sup> with a recurrence rate of 46% to 60%,<sup>[12,25]</sup> hence gross total resection is extremely important and should be further emphasized in CPA cystic meningiomas to avoid tumor recurrence.

For cystic CPA masses with enhanced mural nodule, the most common preoperative diagnosis can be a vestibular schwannoma (VS), hemangioblastoma, and glioma.<sup>[22,26,27]</sup> However, our study suggests that cystic meningiomas should also be included in the differential diagnosis. Cystic meningiomas in CPA frequently show enhancement of the adjacent dura (7/12) and peritumoral edema (11/12).<sup>[4,5,11,12]</sup> VSs typically involve the internal auditory canal and causes widening of the porus acusticus.<sup>[26]</sup> Hemangioblastomas are usually characterized by high-flow vessels, serpentine flow void on MRI, and hypervascular nodule on CTA.<sup>[28]</sup> Gliomas frequently show a heterogeneous enhancing pattern, such as a “growing edge,” intratumoral necrosis, and vasogenic edema.<sup>[29]</sup> Thus, when dealing with tumors manifesting as enhanced mural nodule with cysts in CPA, surgeons should at least be reminded of the possibility of cystic meningiomas, and further examinations such as thinner CT scan and CTA should be considered which would be helpful for the differential diagnosis.

For cystic meningiomas, total resection is the defined standard therapy. It is established that the enhanced mural nodule and cyst wall should be resected simultaneously as much as possible; however, issues regarding the management of the unenhanced cyst wall are still under dispute. Several authors believe that it is unnecessary to remove the unenhanced cyst wall due to it containing no, or just a little amount of tumor cells.<sup>[30,31]</sup> Nevertheless, Boukobza et al<sup>[6]</sup> found that in up to 60.4% of meningiomas, the cyst wall contains tumor tissue and should be resected simultaneously. It is reported that about 20% of the nonenhanced cyst wall contains tumor tissue.<sup>[8]</sup> Furthermore, the recurrence rate is up to 9% in cases that the cyst wall was reserved.<sup>[32,33]</sup> Based on these data, we presume that for cystic meningiomas in CPA, the nonenhanced cyst wall may contain tumor tissue. We hold that multiple biopsies of the cyst wall should be implemented to assess the presence of tumor cells. With a propensity to be an atypical meningioma (WHO grade II), adjuvant radiosurgery is vital in patients who underwent incomplete resection. In addition, close and long-term postoperative follow-up should be emphasized to detect early or late tumor recurrence.

#### 4. Conclusion

For cystic tumors located in the CPA, a differential diagnosis of cystic meningioma should be considered preoperatively. The most common symptoms of cystic meningioma in CPA are headache and hearing problems. Pathologically, clear cell meningioma is the most common subtype, which has a propensity to affect young patients and a higher recurrence rate. Gross total resection is extremely important. When encountering tumors with nonenhanced cyst walls, multiple biopsies of the cyst wall intraoperatively are necessary. Furthermore, cystic meningiomas in CPA have a relatively high recurrence rate. A close follow-up for multiple years is crucial.

#### Author contributions

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**Methodology:** JiuHong Li.

**Resources:** Jinli Meng, Qiguang Wang, Wenke Liu, Xuhui Hui.

**Supervision:** Jinli Meng, Qiguang Wang, Yanchao Wang, Wenke Liu.

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**Visualization:** Jinli Meng, Xuhui Hui.

**Writing – original draft:** JiuHong Li.

**Writing – review & editing:** JiuHong Li, Xuhui Hui.

#### References

- [1] Campbell BA, Jhamb A, Maguire JA, et al. Meningiomas in 2009: controversies and future challenges. *Am J Clin Oncol* 2009;32:73–85.
- [2] Buetow MP, Buetow PC, Smirniotopoulos JG. Typical, atypical, and misleading features in meningioma. *Radiographics* 1991;11:1087–106.
- [3] Guan TK, Pancharatnam D, Chandran H, et al. Infratentorial benign cystic meningioma mimicking a hemangioblastoma radiologically and a pilocytic astrocytoma intraoperatively: a case report. *J Med Case Rep* 2013;7:87.
- [4] Ercan Ö, Yücesoy K, Çitak G, et al. Cystic meningioma imitating vestibular schwannoma at the cerebellopontine angle: case report. *J Neurol Sci (Turkish)* 2007;24:84–7.
- [5] Deb P, Sahni H, Bhatoo HS. Cystic angiomatous meningioma in the cerebellopontine angle mimicking hemangioblastoma. *J Cancer Res Ther* 2010;6:560–3.
- [6] Boukobza M, Cebula H, Pop R, et al. Cystic meningioma: radiological, histological, and surgical particularities in 43 patients. *Acta Neurochir (Wien)* 2016;158:1955–64.
- [7] Zhang D, Hu LB, Zhen JW, et al. MRI findings of intracranial cystic meningiomas. *Clin Radiol* 2009;64:792–800.
- [8] Jung TY, Jung S, Shin SR, et al. Clinical and histopathological analysis of cystic meningiomas. *J Clin Neurosci* 2005;12:651–5.
- [9] Parisi G, Tropea R, Giuffrida S, et al. Cystic meningiomas. Report of seven cases. *J Neurosurg* 1986;64:35–8.
- [10] Chen TY, Lai PH, Ho JT, et al. Magnetic resonance imaging and diffusion-weighted images of cystic meningioma: correlating with histopathology. *Clin Imaging* 2004;28:10–9.
- [11] Yu KB, Lim MK, Kim HJ, et al. Clear-cell meningioma: CT and MR imaging findings in two cases involving the spinal canal and cerebellopontine angle. *Korean J Radiol* 2002;3:125–9.
- [12] Wang XQ, Huang MZ, Zhang H, et al. Clear cell meningioma: clinical features, CT, and MR imaging findings in 23 patients. *J Comput Assist Tomogr* 2014;38:200–8.
- [13] Carvalho GA, Vorkapic P, Biewener G, et al. Cystic meningiomas resembling glial tumors. *Surg Neurol* 1997;47:284–9.
- [14] Awada A, Scherman B, Palkar V. Cystic meningiomas, a diagnostic and pathogenic challenge. *Eur J Radiol* 1997;25:26–9.
- [15] Worthington C, Caron JL, Melanson D, et al. Meningioma cysts. *Neurology* 1985;35:1720–4.
- [16] Russell D. Meningeal Tumours: a Review. *J Clin Pathol* 1950;3:191–211.
- [17] Rengachary S, Batnitzky S, Kepes JJ, et al. Cystic lesions associated with intracranial meningiomas. *Neurosurgery* 1979;4:107–14.
- [18] Rotondo M, D’Avanzo R, Natale M, et al. Atypical cystic meningiomas arising from the trigeminal nerve: surgical and neuroradiological consideration. *Clin Neurol Neurosurg* 2012;114:179–81.
- [19] Yamada SM, Fujimoto Y, Kawanishi Y, et al. A cystic meningioma misdiagnosed as malignant glioma by radiologic and intraoperative histological examinations. *Brain Tumor Pathol* 2010;27:111–5.
- [20] Chen TC, Zee CS, Miller CA, et al. Magnetic resonance imaging and pathological correlates of meningiomas. *Neurosurgery* 1992;31:1015–21.
- [21] Li P, Yang Z, Wang Z, et al. Clinical features of clear cell meningioma: a retrospective study of 36 cases among 10,529 patients in a single institution. *Acta Neurochir (Wien)* 2016;158:67–76.

- [22] Friedmann DR, Grobelny B, Golfinos JG, et al. Nonschwannoma tumors of the cerebellopontine angle. *Otolaryngol Clin North Am* 2015;48:461–75.
- [23] Nauta HJ, Tucker WS, Horsey WJ, et al. Xanthochromic cysts associated with meningioma. *J Neurol Neurosurg Psychiatry* 1979;42:529–35.
- [24] Louis DN, Ohgaki H, Wiestler OD, et al. The 2007 WHO classification of tumours of the central nervous system. *Acta Neuropathol* 2007;114:97–109.
- [25] Tao X, Dong J, Hou Z, et al. Clinical features, treatment, and prognostic factors of 56 intracranial and intraspinal clear cell meningiomas. *World Neurosurg* 2018;111:e880–7.
- [26] Bonneville F, Savatovsky J, Chiras J. Imaging of cerebellopontine angle lesions: an update. Part 1: enhancing extra-axial lesions. *Eur Radiol* 2007;17:2472–82.
- [27] Bonneville F, Savatovsky J, Chiras J. Imaging of cerebellopontine angle lesions: an update. Part 2: intra-axial lesions, skull base lesions that may invade the CPA region, and non-enhancing extra-axial lesions. *Eur Radiol* 2007;17:2908–20.
- [28] Cheng J, Liu W, Zhang S, et al. Clinical features and surgical outcomes in patients with cerebellopontine angle hemangioblastomas: retrospective series of 23 cases. *World Neurosurg* 2017;103:248–56.
- [29] Baehring JM, Bi WL, Bannykh S, et al. Diffusion MRI in the early diagnosis of malignant glioma. *J Neurooncol* 2007;82:221–5.
- [30] Weil RJ, Lonser RR, DeVroom HL, et al. Surgical management of brainstem hemangioblastomas in patients with von Hippel-Lindau disease. *J Neurosurg* 2003;98:95–105.
- [31] Lonser RR, Vortmeyer AO, Butman JA, et al. Edema is a precursor to central nervous system peritumoral cyst formation. *Ann Neurol* 2005;58:392–9.
- [32] Fortuna A, Ferrante L, Acqui M, et al. Cystic meningiomas. *Acta Neurochir (Wien)* 1988;90:23–30.
- [33] Inoue T, Kuromatsu C, Sawada K, et al. Recurrent cystic meningioma. *Surg Neurol* 1986;26:399–404.