Radiologic Features and Surgical Strategy of Hemangioblastomas with Enhanced Cyst Wall

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■ OBJECTIVE: **Hemangioblastomas with enhanced cyst walls represent a rare radiologic presentation of hemangioblastomas with poor understandings. We aimed to summarize the clinical and radiologic features, important differential diagnosis, surgical strategy, and clinical outcome of this rare entity.**

■ METHODS: **From June 2008 to March 2017, 12 patients with cystic hemangioblastomas presenting with enhanced wall thickness on MRI were treated in our department. The clinical presentations, radiologic investigations, surgical treatment, neurologic outcome, and recurrence rate were evaluated. Important preoperative differential diagnosis and surgical strategy of this entity were discussed.**

■ RESULTS: **Twelve patients with cystic hemangioblastomas presenting with an enhanced cyst wall on magnetic resonance imaging were analyzed retrospectively. There were 5 male and 7 female subjects, with a mean age of 41.4 years (range, 13–78 years) and an average duration of symptoms before diagnosis of 2.23 months (range, 0.5–8.0 months). Radiologically, enhancement of both tumoral nodule and cyst were observed in 8 patients, while pure ring-enhanced cyst without typical tumoral nodule was found in 4 patients. Histopathologic examination confirmed the diagnosis of hemangioblastomas, and the enhanced cyst wall and mural nodule shared the same histopathologic pattern. Postoperative complications occurred in only 1 patient with postoperative cerebellar hemorrhage. During follow-up, 8 patients achieved favorable neurologic outcomes (Karnofsky score: 100) without recurrence; however, 4 patients experienced local tumor recurrence after the initial surgery.**

■ CONCLUSIONS: **Hemangioblastomas with enhanced cyst wall possess distinctive radiologic features, and they are frequently misdiagnosed preoperatively. Favorable tumor control can be achieved only when gross total resection of both the tumor nodule and cyst wall are performed. Close follow-up is necessary because of the high recurrence rate in this subset of hemangioblastomas.**

INTRODUCTION

Hemangioblastomas (HBLs) are World Health Organization (WHO) grade I central nervous system (CNS) neoplasms[1]; they can either occur sporadically or as part of the von Hippel-Lindau (VHL) disease. It is reported that 60%–70% of CNS hemangioblastomas are associated with peritumoral cysts,[2,3] and a large unenhanced cyst with enhanced mural nodule located in cerebellum is considered the typical radiologic feature of this disease.[4,5] The optimal surgical strategy of cystic hemangioblastomas is to remove the mural nodule and to leave the gliotic cyst wall undisturbed.[6,7]

Cystic hemangioblastomas with enhanced cyst walls are rarely known by people, and only 3 cases have been reported in the English-language literature.[8,9] Cystic hemangioblastomas can be misdiagnosed easily because of the atypical imaging features when presenting with enhanced cyst wall. Furthermore, little is known

Key words
- Enhanced cyst wall
- Hemangioblastomas
- Pathogenesis
- Recurrence

Abbreviations and Acronyms
CNS: Central nervous system
CTA: Computed tomography angiography
HBLs: Hemangioblastomas
MRI: Magnetic resonance imaging
WHO: World Health Organization
VHL: von Hippel-Lindau

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about the clinical and radiologic features, surgery strategy, and outcomes for this subgroup of hemangioblastomas.

In addition, there is no comprehensive large series study focusing on cystic hemangioblastomas with enhanced cyst wall. In the present study, we retrospectively analyzed the clinical features and radiologic findings of 12 patients with hemangioblastomas with enhanced cyst wall to emphasize the differential diagnosis, to optimize the treatment, and to elaborate the essence of this disease.

METHODS

From June 2008 through March 2017, 12 patients with cystic hemangioblastomas with radiologically enhanced wall thickness were treated at the West China Hospital at Sichuan University. The clinical features, radiologic findings, preoperative diagnosis, management, and outcomes of these patients were analyzed retrospectively. Data including patient age, gender, symptoms, and duration of symptoms before diagnoses were obtained. All patients underwent cranial magnetic resonance imaging (MRI), including T1-weighted, T2-weighted, and gadolinium-enhanced T1-weighted sequences. Computed tomography angiography (CTA) was performed in select patients to help with preoperative differential diagnosis. The tumor location, enhancement pattern, occurrence of hydrocephalus were recorded. All patients underwent gross total resection including both the mural nodule and enhanced cyst wall. Samples for histopathologic diagnosis were obtained from the tumor nodule and cyst wall. All patients had a pathologically diagnosed hemangioblastomas (WHO grade I), and the enhanced tumor nodule and enhanced cyst wall had the same histologic features. Follow-up was performed at 3 months, 6 months, and then yearly after surgery. During the follow-up period, MRI was used to monitor tumor recurrence, and neurologic functional outcome was recorded.

RESULTS

Patient Sample

Twelve patients of cystic hemangioblastomas with enhanced cyst wall were treated in our hospital. The general characteristics of these patients are documented in Table 1. There were 5 male and 7 female subjects, with a mean age of 41.4 years. The symptoms and signs included headache (n = 7), vertigo (n = 1), vomiting (n = 1), nausea (n = 1), ataxia (n = 1), lower limb weakness (n = 1), and hearing loss (n = 1). The duration of symptoms before diagnosis varied from 15 days to 8 months. Sporadic hemangioblastomas were diagnosed in 11 patients, whereas case 5 was in accordance with the clinical diagnostic criteria of VHL disease.

Radiologic Investigation

Tumors were located in the brainstem, cerebellopontine angle, cerebellar hemisphere, and cerebellar vermis. Cases 1, 3, 4, and 8–12 showed enhanced tumoral nodule associated with partial or entire cyst wall enhancement (Figures 1–3), whereas cases 2 and 5–7 demonstrated pure ring-enhanced cyst without typical tumoral nodule (Figures 4 and 5). CTA in case 7 (Figure 5) showed that the cyst wall was composed of abnormal vessels. Hydrocephalus were observed in all cases except case 12.

Table 1. Summary of Clinical Characteristics, Radiologic Features, Treatment Strategy, and Outcomes for 12 Patients with Hemangioblastomas with Enhanced Cyst Walls

						Radiologic Study						
Patient Number	Age (years)	Sex	Duration (months)	Signs and Symptoms	Tumor Location	Tumor Nodule	Cyst Wall Enhancement	Hydrocephalus	Operation	Recurrence	Outcome	Follow-Up (months)
1	13	Female	2	Vertigo	CV	Yes	Partial	Yes	Total	No	Improved	96
2	51	Female	2	Headache	CV	No	Entire	Yes	Total	No	Improved	96
3	78	Male	1	Lower limb weakness	CH	Yes	Entire	Yes	Total	No	Stable	24
4	48	Female	1.5	Vomiting	CH	Yes	Partial	Yes	Total	Yes	Improved	48
5	44	Male	1	Nausea	CV	No	Entire	Yes	Total	No	VHL, improved	12
6	52	Male	1.5	Headache	CH	No	Entire	Yes	Total	No	Improved	60
7	28	Female	8	Headache	CH	No	Entire	Yes	Total	No	Improved	22
8	37	Female	1.8	Ataxia	CH	Yes	Partial	Yes	Total	Yes	Improved	48
9	43	Female	2.5	Headache	CV	Yes	Partial	Yes	Total	No	Improved	36
10	30	Male	3	Headache	BS	Yes	Entire	Yes	Total	Yes	Unimproved	105
11	26	Male	0.5	Headache	CPA	Yes	Partial	Yes	Total	No	Improved	36
12	47	Female	2	Headache, hearing loss	CPA	Yes	Entire	No	Total	Yes	Improved	84

CV, cerebellar vermis; CH, cerebellar hemisphere; BS, brainstem; CPA, cerebellopontine angle; VHL, von Hippel-Lindau syndrome; Total, total resection.