Primary Intracranial Angioleiomyomas as Rare, Nonmalignant, and Distinct Neoplastic Entities: A Series of 8 Cases and a Literature Review

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Key words
- Cavernous sinus
- Central nervous system
- Neurosurgery
- Primary intracranial angioleiomyoma

Abbreviations and Acronyms
ALM: Angioleiomyoma
CK: Cytokine
CN: Cranial nerves
CT: Computed tomography
DSA: Digital subtraction angiography
DWI: Diffusion-weighted imaging
EMA: Epithelial membrane antigen
GFAP: Glial fibrillary acidic protein
GTR: Gross total resection
KPS: Karnofsky Performance Scale
MRI: Magnetic resonance image
SMA: Smooth muscle actin
STR: Subtotal resection

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OBJECTIVE: **Primary intracranial angioleiomyoma is a rare and distinct neoplasm. Only 29 cases have been reported previously, and we aimed to investigate the clinical and radiopathologic features of these lesions.**

METHODS: **Medical records and radiographs of 8 patients (7 male and 1 female; mean age: 48.7 years) at our institution were reviewed retrospectively. Patient follow-up and a literature review were performed.**

RESULTS: **The most common preoperative symptom was a visual defect (n = 2), followed by diplopia (n = 1) and abducens paralysis (n = 1). Three patients were asymptomatic. The parasellar area (particularly the cavernous sinus) was the predilection site (n = 4; 50.0%). Radiographically, all lesions were solid without cystic degeneration. All lesions appeared with T1 hypointensity and T2 hyperintensity, and they were gradually heterogeneously enhanced after the administration of gadolinium. Complete resection was achieved in 7 patients (87.5%) without recurrence after 26.8 months of follow-up. Mitosis was rarely observed, and the Ki-67 labeling index was less than 1%; pathologically, the cavernous type was the most common.**

CONCLUSIONS: **Primary intracranial angioleiomyomas were prevalent in middle-aged men, and they usually involved the cavernous sinus and were frequently pathologically identified as the cavernous type. Preoperative symptoms varied depending on lesion location. The preoperative diagnosis of primary intracranial angioleiomyomas is difficult without pathology. Digital subtraction angiography and preoperative embolization are useful for differential diagnosis and surgery. Given the indolent biology of these tumors, a favorable outcome can be achieved using total resection without recurrence. A larger sample size with long-term follow-up is needed to verify our findings.**

INTRODUCTION

Angioleiomyoma (ALM) is a relatively rare, benign, independent soft tissue tumor; it is mainly composed of well-differentiated smooth muscle cells, and it has a remarkable vascular component. ALM often appears in the skin and subcutaneous tissue of the extremities of middle-aged women.[1,2] Primary intracranial ALMs are extremely rare, and to date, only 29 cases diagnosed on the basis of pathologic examination have been reported in the literature. A favorable outcome after total resection without recurrence is reported in most studies[3]; however, prior reports are limited to single cases, and an understanding of this disease must be gleaned from small series and case reports because of the relative paucity of data. In the present study, we describe a series of 8 cases of primary intracranial ALM, evaluate the clinical radiopathologic features, and perform a literature review using the PubMed database.

PATIENTS AND METHODS

Clinical and Radiographic Data

All patients underwent surgical treatment and were pathologically confirmed to have ALM at Beijing Tiantan Hospital between February 2013 and April 2017. Two cases of ALM that were treated before February 2013 at our institute have been reported in a prior study.[4] This study was authorized by the Beijing Tiantan Hospital Research Ethics Committee.

We retrospectively reviewed the clinical data and radiographs of all the patients. The Karnofsky Performance Status (KPS) scale was used to evaluate the patients' neurologic status. Preoperative and postoperative magnetic resonance imaging (MRI) scans or computed tomography was obtained. The patients' age, sex, preoperative symptoms, symptom duration, preoperative diagnosis, tumor

Table 1. Patient Demographics and Radiopathologic Features

Patient Number	Sex	Age (Years)	Signs and Symptoms	Symptom Duration (Months)	Tumor Location	Tumor Size* (cm)	Radiology			Therapy	Immunohistochemistry	Follow-up (months)	Pathology	KPS score		
							Morphology	T1, T2, En, or CT	Progressive En					Pre-	Post-	Resent
1	Male	42	Vertigo, tinnitus, headache	0.6	Rt CPA	3.1	Solid	CT: hyper	NA	GTR	Pos: SMA, VIM, CD34, DES; Neg: EMA, PR;	37.3	Cavernous type	90	100	100
2	Male	43	Accidental	1	Rt tentorium	2.9	Solid	T1: hypo; T2: hyper; Gd: nHen	NA	GTR	Pos: SMA, VIM, CD34; Neg: DES;	29	Cavernous type	100	100	100
3	Male	58	Accidental	12	Rt parietal lobe	2.6	Solid	T1: hypo; T2: hyper; Gd: nHen	NA	GTR	Pos: SMA, VIM, CD34; Neg: DES, S-100, CK;	47.3	Cavernous type	100	100	100
4	Male	48	Diplopia	24	Rt cavernous sinus	2.9	Solid	T1: hypo; T2: hyper; Gd: nHen	Pen	GTR	Pos: SMA, VIM, CD34; Neg: DES;	46.4	Cavernous type	90	90	90
5	Male	41	Rt CN VI pals	24	Rt cavernous sinus	2.9	Solid	T1: hypo; T2: hyper; Gd: nHen	Pen	GTR	Pos: SMA, VIM, CD34; Neg: DES;	8.3	Cavernous type	90	90	90
6	Female	47	Left visual deficit	60	Lt cavernous sinus	3.1	Solid	T1: hypo; T2: hyper; Gd: nHen	NA	GTR	Pos: SMA, VIM, CD34; Neg: DES;	33.9	Cavernous type	90	90	90
7	Male	58	Left visual deficit	12	Lt sellar tuberculum	1.0	Solid	T1: hypo; T2: hyper; Gd: nHen	NA	STR	Pos: SMA, VIM, CD34; Neg: DES;	7.9	Cavernous type	90	80	90
8	Male	53	Accidental without symptom	12	Bilateral cerebral falx	3.0	Solid	T1: hypo; T2: hyper; Gd: nHen	NA	GTR	+: SMA, VIM, CD34; Neg: DES;	5.0	Cavernous type	100	100	100

En, enhancement; CT, Computed tomography; KPS, Karnofsky Performance Status; Pre-, preoperative; Post-, postoperative; Rt, right; CPA, cerebellopontine angle; NA, not available; GTR, gross total resection; Pos, positive; SMA, smooth muscle actin; VIM, vimentin; DES, desmin; Neg, negative; CK, cytokeratin; EMA, epithelial membrane antigen; PR, progesterone receptor; Gd, gadolinium; nHen, nonhomogeneous enhancement; Pen, progressive enhancement; CN, cranial nerve; Lt, left; STR, subtotal resection.

*Lesion size was evaluated as the lesion equivalent diameter $(a \cdot b \cdot c)^{1/3}$, where *a*, *b*, and *c* represent the diameters measured on axial, sagittal, and coronal magnetic resonance images, respectively.