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Case Report

A rare differential diagnosis of intra-sella tumor: angioleiomyoma

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Abstract: Angioleiomyoma (ALM) is a rare, benign soft tissue tumor commonly occurring in middle-aged individuals with female predominance, whose main occurrences are in the subcutaneous tissue of the extremities. Intracranial ALM is particularly rare. Here we present the third intra-sella ALM proved by pathology, with a bleeding complication in a 65-year-old male. Imaging examination showed an oval-shaped tumor occupying the left side of the sella, with an intact pituitary. The tumor was isointense on T1-weighted magnetic resonance imaging (MRI) and extremely hyperintense on T2-weighted imaging scan, which was similar to the cerebrospinal fluid, and progressively flame-like enhancement after gadolinium injection. The computed tomography (CT) scan revealed a slight hyperdensity intra-sella tumor invading into the left cavernous sinus. Subtotal resection was performed using an endonasal transsphenoidal approach with a microscope and neuronavigation. Postoperatively, the patient immediately recovered from his symptoms, and there was no evidence of recurrence during the past 2-year follow-up. In this case, we observed that surgery on a rare case of intra-sella ALM with typical radiological manifestation was difficult and risky, early diagnosis before operation and proper treatment with precaution of intra-sella ALM was particularly important, and it was worthy considering to ALM in the differential diagnosis list of an abundant blood-supply mass in the intra-sella region and the anatomical relationship of the tumor and peripheral structure should be examined carefully by the surgeons to avoid blood loss and post-operative endocrinological complications in future.

Keywords: Angioleiomyoma, intra-sella, cavernous sinus, computed tomography (CT), magnetic resonance imaging (MRI)

Introduction

Angioleiomyoma (ALM) also known as vascular leiomyoma or angiomyoma, is a relatively rare, benign soft tissue tumor which is composed of well-differentiated smooth muscle cells with a prominent vascular component [1]. ALM is recognized as an independent tumor entity in accordance with the 2002 World Health Organization classification of soft tissue tumors [2]. And normally excision can be curative owing to the benign nature of ALM [3]. After reviewing the relevant literature, it has been found that there are only 25 reported cases of intracranial ALM [4-21] (**Table 1**). Among these cases, only two intra-sella cases are recorded [10, 15]. Here we present the third intra-sella ALM, and to the best of our knowledge, this is the first case report to specify both computed tomography (CT) and magnetic resonance imaging (MRI) findings of ALM located in the sellar region. And it is the second one occurring with a bleeding complication, but the only one surviving from the complication and having no evidence of recurrence during the past 2-years follow-up. We have reasons to doubt that surgery on ALM in sella is difficult and risky despite the relatively favorable prognosis. Therefore, early diagnosis before operation and proper treatment with precaution of intra-sella ALM is particularly important.

Case report

A 65-year-old man was admitted to The Second Affiliated Hospital of Zhejiang University School of Medicine (Hangzhou, China) on November 20, 2014 following a seven-month history of

Table 1. Literature review for the intracranial angioleiomyoma

Author	Age	Sex	Symptoms	Location	Surgical treatment	Follow-up
Calle et al. [4]	43	M	Light-headed, syncope	Anterior falx cerebri	GTR	NA
Delgado-Fernandez et al. [5]	43	M	Hearing loss	Infratentorial	GTR	2 years, NR
Lin et al. [6]	36	M	Headache, diplopia, nausea, vomiting	Right cavernous sinus	STR	20 months, NR
He et al. [7]	46	F	Headache, blepharoptosis in the right eye	Cavernous sinus	GTR	72 months, NR
He et al. [7]	57	M	Headache, right vision loss accompanied by diplopia	Cavernous sinus	GTR	57 months, NR
He et al. [7]	48	F	Headache, right ptosis	Cavernous sinus	GTR	47 months, NR
He et al. [7]	35	F	Headache, decreased left vision accompanied with diplopia	Cavernous sinus	GTR	14 months, NR
Lescher et al. [8]	40	M	Focal epilepsy	Falx cerebri	GTR	NA
Li et al. [9]	23	F	Fmenorrhea, impaired right visual acuity	Left cavernous sinus	STR	NA
Li et al. [9]	62	M	Hypophrasia, hypomnesis, mild disturbance of consciousness	Right temporal lobe	GTR	NA
Sun et al. [10]	51	F	Visual deterioration	Intra-sella	GTR	Died of delayed hemorrhage
Sun et al. [10]	49	M	Weakness of lower limbs	Infratentorial	STR	1 year, NR
Sun et al. [10]	77	M	Headache	Left temporal diploic space	GTR	1 year, NR
Teranishi et al. [11]	52	F	Eye discomfort	Cavernous sinus	GTR	NA
Zhou et al. [12]	62	M	A sudden seizure	Middle cranial fossa	NA	7 months, NR
Conner et al. [13]	42	M	Headache	Infratentorial	GTR	23 months, NR
Conner et al. [13]	36	M	Headache	Falcine	STR	26 months, NR
Shinde et al. [14]	60	M	Headache, seizures, irritability	Right putamen	No surgery	Died of recurrent seizures and septicemia
Xu et al. [15]	53	M	Headache, visual deterioration	Intra-sella	GTR	NA
Colnat-Coulbois et al. [16]	50	M	Vertical diplopia	Left cavernous sinus	GTR	6 years, NR
Gasco et al. [17]	43	M	Headache, blurred vision, dizziness, gait abnormalities	Left cerebellum	GTR	NA
Figueiredo et al. [18]	52	M	Horizontal diplopia, headache, facial numbness, impaired visual acuity	Right cavernous sinus	GTR	NA
Karagama et al. [19]	47	F	Hearing loss	Left auditory meatus	GTR	1 year, NR
Ravikumar et al. [20]	12	F	Headache, seizures, left hemi-dystonia, apraxia of eyelid closure	Right head of caudate	GTR	20 months, NR
Lach et al. [21]	47	M	Gait abnormality with a right-sided limp	Leptomeninges of the right parietal lobe	GTR	4 years, NR
Current case	65	M	Dizziness, headache	Intra-sella	STR	2 years, NR

Abbreviations: GTR, gross total resection; STR, subtotal resection; NA, not available; NR, no recurrence.

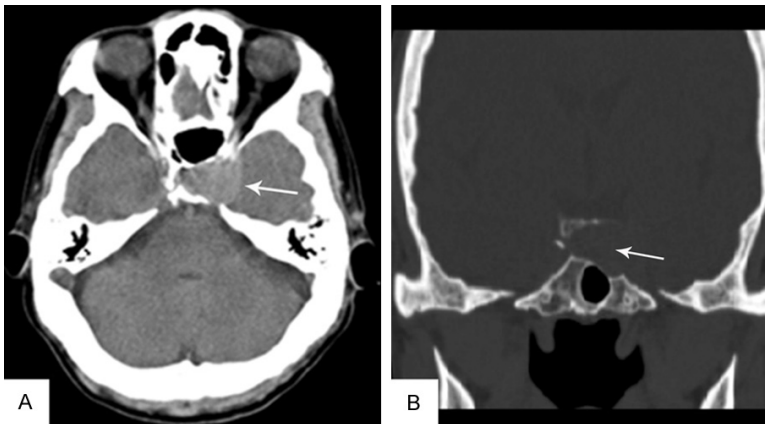

Figure 1. CT (A. Axial scan, soft tissue window. B. Coronal reconstruction, bone window) showed a 2.1 cm × 1.7 cm, well-defined, slightly hyperdense (mean CT attenuation value of 54 HU) mass (arrow) located in the left sellar region without calcification, hemorrhage or edema, and the local sphenoidal saddle bone was absorbed and oppressed.

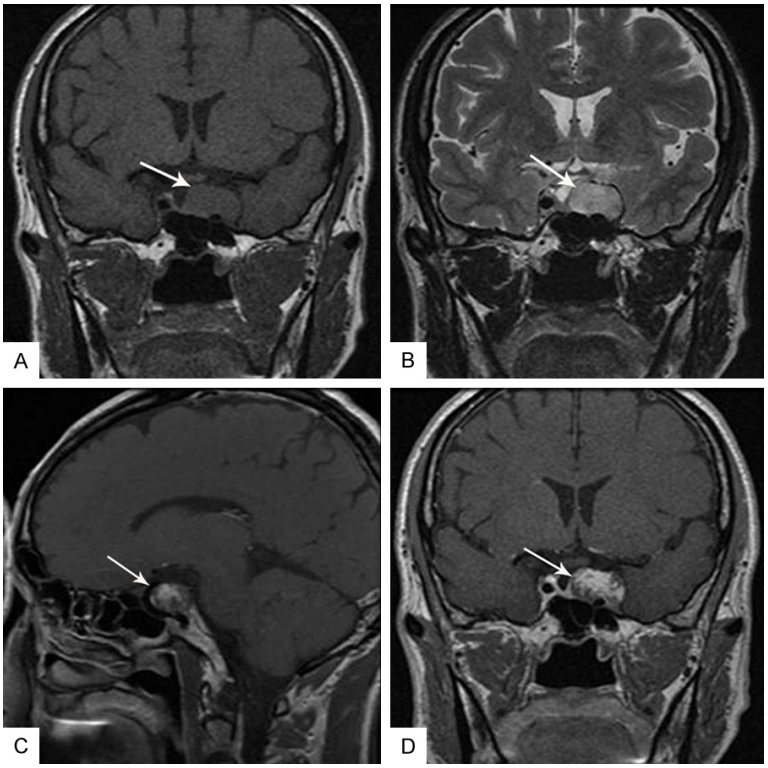

Figure 2. The mass (arrow) was homogeneous iso-signal on T1-weighted MRI (A) and heterogeneous hyper-signal on T2-weighted MRI (B). With gadolinium contrast (C, D), the mass was obviously unevenly and partially enhanced, progressively from the central region to the peripheral region, with linear and curvilinear areas interspersing throughout the tumors, and the peripheral region of the lesion was hardly enhanced.

recurrent dizziness with mild headache, progressively worsening for 3 months. There were no serious illnesses in the patient's past medical history. And his neurological physical examination was unremarkable.

CT (Somatom Sensation 16; Siemens, Munich, Germany) findings of the tumor were as follows (**Figure 1A**, **1B**): i) Location, the tumor was located in the left sellar region; ii) Shape, the tumor appeared as a small (2.1 cm × 1.7 cm), smooth, oval mass; iii) Composition, the tumor appeared heterogeneously slight with hyperdensity on a plain CT examination, with a mean CT attenuation value of 54 Hounsfield units (HU); iv) Neighborhood, the adjacent cavernous sinus was compressed, and sphenoidal saddle bone absorption and oppression was found. Based on these findings, a pituitary tumor was the initial presumption.

MRI studies of the sellar region were performed with 3 mm scans in sagittal and coronal planes and included T1-weighted imaging (T1WI), T2-weighted imaging (T2WI), and T1WI after administration of gadolinium (Gd). T1WI showed an iso-signal homogeneous tumor (**Figure 2A**), and T2WI revealed a heterogeneous hyper-signal (**Figure 2B**). With gadolinium contrast, the lesion was clearly heterogeneously and partially enhanced, and the enhancement modality was progressive from the central region to the peripheral region, with linear and curvilinear areas interspersing throughout the tumors. Furthermore, the peripheral region of the lesion was hardly enhanced (**Figure 2C**, **2D**). The intrasella tumor sized 2.1 cm × 1.8 cm × 1.2 cm, the sella fossa was obviously enlarged, and the left internal carotid artery (ICA) cavernous segment

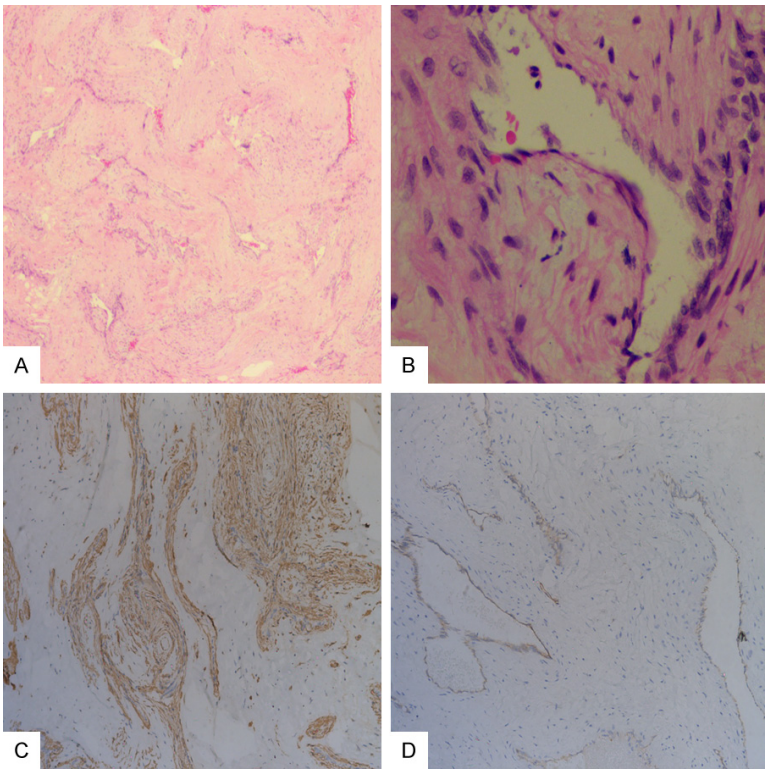

Figure 3. A. (hematoxylin-eosin stain; magnification, × 40) showed the overall view, tumor tissue composed of blood vessles and spindle cells. B. (hematoxylin-eosin stain; magnification, × 400) showed bundles of spindle shaped smooth muscle cells arranged irregularly surrounding small thick-walled vessels. C. (immunohistochemistry of smooth muscle actin (SMA), magnification, × 100) showed the spindle cells were strongly positive for SMA as shown by the brown color. D. (immunohistochemistry of CD34, magnification, × 100) showed the vascular endothelial cells were positive for CD34 as shown by the brown color.

was involved. Pituitary tumor, meningioma, fibrosarcoma, and cavernous hemangioma were initially considered.

An endonasal transsphenoidal approach with a microscope and neuronavigation was used for surgical resection. After the sinus was opened, the capsule of the tumor was exposed. The tumor tissue around the sellar region and the intact pituitary gland was presented. The tumor revealed a white, tough and rich in blood supply mass. With partial resection achieved, the tumor adhered tightly to the left cavernous sinus. After subtotal resection, severe bleeding occurred, packed hemostasis with gelatin sponge and surgical was performed. Approximately 300 ml of blood was lost altogether.

Microscopically, the tumor was composed of bundles of spindle shaped smooth muscle cells and dilated vascular channels, which were generally uniform in size and shape. Smooth muscle cells were arranged irregularly surrounding small thick-walled vessels. They were closely compact and intermingled with each other (**Figure 3A**). An immunohistochemical study of the tumor revealed: smooth muscle actin (SMA) (+++), CD34 (+), CD31 (+), neuron specific enolase (NSE) (+), desmin(-), S100 (-), P53 (-), neurofilament protein (NF) (-), synaptophysin (Syn) (-), Ki-67 < 1% (**Figure 3B**, **3C**). Pathological diagnosis of ALM in sella was ultimately ascertained.

Postoperatively, the patient immediately recovered from his symptoms, and there was no evidence of recurrence during the past 2-year follow-up since being discharged from the hospital on November 30, 2014.

Discussion

ALM is an independent tumor entity which was recognized by the World Health Organization [2], and first described by Lach et al. in 1994 [21]. ALMs are benign soft tissue tumors commonly involving the subcutaneous tissue of the lower extremities with predominant occurrence in middle-aged women [1]. Primary intracranial ALM is extremely rare and, to the best of our knowledge, only 25 cases have been reported worldwide [4-21]. Among these, there are only 3 intra-sella cases recorded in the literature to date including the present case.

After reviewing the English literature, it was revealed that the occurrence of intracranial ALMs exhibits a male predominance, which is in contrast to the subcutaneous counterpart [4]. The present article describes a case of a 65-year-old male with intracranial ALM, which is comparable with the literature on intracranial

ALMs. The most common presentation of patients with ALM of the sellar region is a non-specific headache and visual deterioration [10, 15].

Histologically, the characteristics of ALM are still controversial, it is composed of an abundant number of vascular channels separated by loosely organized smooth muscle bundles with a variable amount of collagen [1, 12-16, 18, 19, 22-24]. Most researchers assumed that ALM arose from blood vessels, while other researchers suggested that the tumor was probably in a process of continuous proliferation of smooth muscle cells [22]. Immunohistochemically, SMA as a bio-marker of muscle cells, and CD34 for the endothelium of vessels, are essential for diagnosis. The histogenesis of these lesions is not clear [1]. The immunohistochemical results of the present study were mainly consistent with previous observations.

Only 2 cases of radiological findings of intra-sella ALM have previously been described [10, 15]. According to the literature on intra-sella ALM, radiological manifestations are variable [10, 15]. Xu et al. [15] reported a cystic mass with a nodular solid component. There was a homogeneously hypointense tumor in the sellar region with a slightly bright nodule inside the posterior borderline of the tumor on T1WI, that was heterogeneously hyperintense with a hypointense nodule on T2WI. Sun et al. [10] identified a solid mass of intra-sella ALM. MRI scans revealed an iso-intense tumor on T1WI, homogeneous hyper-intense on T2-fluid-attenuated inversion recovery-weighted sequences (T2-FLAIR), with distinctive partial enhancement after gadolinium injection, which arose from the right cavernous sinus. It was flame-like in shape, with a bushlike edge. The tumor was close to the medial part of the right cavernous sinus, the sella fossa was obviously enlarged and the tumor had invaded the sphenoid sinus.

However, as far as we know, CT and contrast-enhanced multiple-phase MRI findings of intra-sella ALM have not been well addressed. In the present case, CT scans showed a slightly hyperdense mass located in the left sellar region without calcification, hemorrhage, which was similar to the previous reports of intracranial ALMs [6, 7, 9, 13, 17, 18]. The sella fossa was clearly enlarged and the left ICA cavernous segment was involved. MRI studies were dramatically similar to the case described by Sun L et al. [10], which showed a homogeneously iso-signal tumor on T1WI, and heterogeneously hyperintensity on T2WI. With gadolinium contrast, the lesion was clearly heterogeneously progressive enhancement from the central region to peripheral region, with linear and curvilinear areas interspersing throughout the tumors, and the peripheral region of the lesion was hardly enhanced, namely a “flame-like” appearance. This enhancement pattern can also be seen in other cases [7, 16]. Some authors explained it as “delayed progressive enhancement” and they thought the whole tumor would be enhanced if enough time was given [16]. The exact reason of the enhancement pattern is unclear. We hold the opinion that it may be due to the histopathological characteristics of the tumor that the blood vessels are surrounded by smooth muscle cells irregularly. In brief, the “flame-like” or “delayed progressive” enhancement pattern may be valuable in distinguishing ALMs from other tumors in the sellar region. No doubt, further studies are required to prove this opinion.

As is with the case described by Sun et al. [10], the intra-sella ALM tumor tissue revealed a rich blood supply, and the tumor had invaded the cavernous sinus and involved the ICA. But the case described by Sun L et al. died of pseudoaneurysm of the left ICA cavernous segment postoperatively, and the present case presented severe bleeding due to the difficulty of stripping the tumor from the left caverous sinus. According to the surgery results, it is vital that radiological manifestations have been previously described before surgery.

Surgery on ALM in sella is difficult and risky, early diagnosis before operation and proper treatment with precaution of intra-sella ALM is particularly important. ALM has unique manifestation on enhanced MRI scan with “flame-like” or “delayed progressive” enhancement pattern, which are considered to be helpful for diagnosis before surgery. There are some differential diagnoses that must be considered, including pituitary adenoma, meningioma, fibrosarcoma, and cavernous hemangioma. MRI is necessary for demonstrating the anatomical relationship of the tumor and pituitary grand, stalk and cavernous sinus for the neurosurgeon to prevent uncontrollable bleeding in the surgery.

In this case report, a rare case of intra-sella mass of ALM with typical radiological manifestation was presented. The authors emphasized on that ALM should be considered in the differential diagnosis list of an abundant blood-supply mass in the intra-sella region; the anatomical relationship of the tumor and peripheral structure should be examined carefully by the surgeons to avoid blood loss and post-operative endocrinological complications.

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Disclosure of conflict of interest

None.

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