Infratentorial angioleiomyoma: case report and review of the literature

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Introduction. Intracranial angioleiomyomas are extremely rare lesions. Only 22 intracranial angioleiomyomas have been described in the literature and only three were infratentorial.

Case report. We report a case of an infratentorial angioleiomyoma in a 43 year-old-man, who underwent a brain computer tomography because of hipoacusia. The MRI showed a 1.4 cm tumor, initially described as a meningioma, with progressive enhancement after gadolinium injection, an augmented apparent diffusion coefficient and a generalized metabolite decreased in the spectroscopy. The lesion was surgically removed through a suboccipital approach with a good evolution and without postoperative complications. In the inmunohistological study, the lesion was mainly composed of multiple vessels and the immunohistochemistry was positive for actina and caldesmon. Two years after surgery, no recurrence has been found in the MRI.

Conclusion. Angioleiomyomas diagnostic may be complex, but some radiological features could help in the differential diagnostic. Angioleiomyomas are benign tumors associated with favorable outcomes after total resection, that in our case, did not show a significant bleeding risk.

Key words. Angioleiomyoma. Apparent diffusion coefficient. Infratentorial. Progressive enhancement. Intracranial tumor. Spectroscopy.

Introduction

Angioleiomyomas (ALMs) are benign solitary smooth muscle tumors with prominent vascular component. They usually present as soft tissue tumors in middle-aged women in the subcutaneous tissue of the lower extremity, between the third and the fifth decades of life [1-3]. ALMs may also present in parenchymatous organs in association with large vessels [2,4]. Primary smooth muscle tumors involving the central nervous system are very rare; only twenty-two cases have been reported, most of them involving the dura. Until now, only three cases of ALM have been described at the infratentorial dura [5-7], three in the cavernous sinus dura [4,8,9], two in the sella [7,10], three in the auditory meatus [11-13], two in the bone diploic space [7,14], one in the parietal lobe leptomeninges [15], one in the temporal lobe leptomeninges [16], three in the falx cerebri [5,17,18], one in the middle fossa [19], one occupying both sella and cavernous sinus [16] and two multicentric cases [20,21].

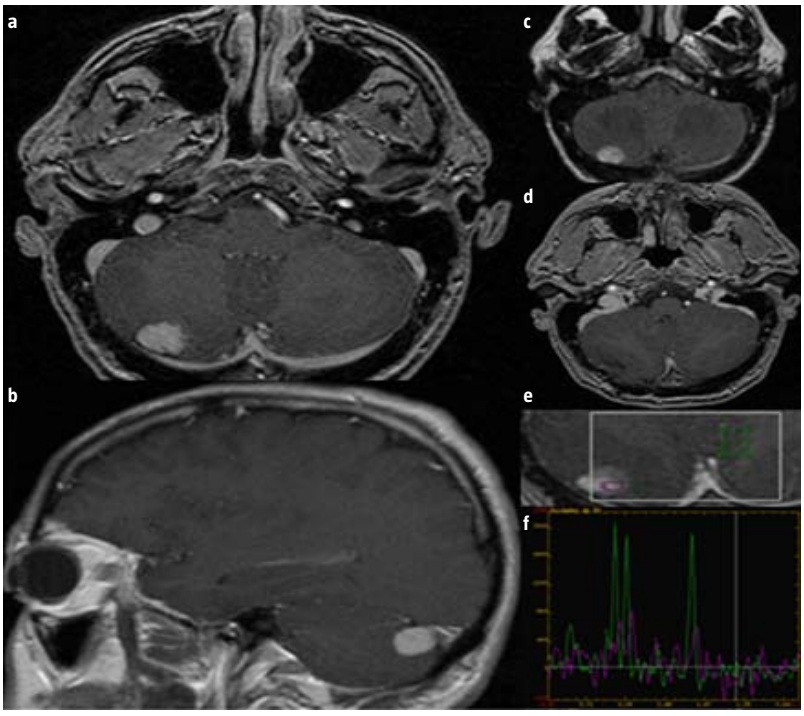
Case report

We present a case of a 43 year-old-man with a 6-month history of right ear hearing loss. CT showed an incidental hyperdense lesion with contrast enhancement without surrounding edema, and brain MRI revealed infratentorial isointense tumor 14.7 × 13.5 × 14.4 mm with broad dural contact, hyperintense on T_2 weighted sequences, near the right transverse sinus without invasion or thrombosis. After contrast administration it has a progressive homogeneous enhancement in the latest sequences. No dural 'tail sign' appears with gadolinium contrast. There was no surrounding edema or mass effect. An increased apparent diffusion coefficient (ADC) was shown in relation with the normal tissue, and a generalized metabolite decrease appeared in the echo time (TE) long spectroscopy MRI (Fig. 1). The tumor was interpreted as a meningioma in the first place, and the patient was followed with yearly MRIs for three years, where a slow tumor growth (1.1 mm/year) was demonstrated and surgical treatment was proposed.

A paramedian suboccipital craniotomy was performed. The lesion was a 1.5 cm vascularized mass that suggested a meningioma implanted on the inferior surface of the tentorium. A piecemeal resection was made and 4 fragments were obtained, the biggest measuring 1 × 1 cm. The lesion was smooth, soft and brown-colored. It was a well vascularized lesion without significant bleeding and a good plane of dissection. The postoperative course was uneventful and postoperative CT demonstrated gross total resection (GTR) without any complications. At 2-years follow-up there are no signs of recurrent or residual tumor on MRI (Fig. 1).

Microscopically, the tumor was essentially composed of blood vessels of different sizes, generally dilated and congestive surrounded by a thickwall of spindle cells without nuclear atypia or necrosis. No mitoses were found. Spindle cells were arranged in an irregular pattern with abundant eosinophilic cytoplasm (Fig. 2). In the immunohistochemical study spindle cells were positive for muscular markers such as actin and caldesmon (Fig. 3). A diagnosis of solid type ALM (WHO grade I) was made.

Figure 1. a, b) Magnetic resonance imaging, axial and sagital preoperative T_1-w MRI with contrast showing a well-defined lesion adhered to the transverse sinus; c) Preoperative T_2-w FLAIR MRI; d) Postoperative T_1-w MRI with contrast; e, f) Spectroscopy MRI showing a generalized decreased in metabolites.



Discussion

ALM is a benign tumor composed of smooth muscle and endothelium in which the predominant feature is an abundant number of vascular channels. It occurs most commonly in the lower extremities as a painful, solitary tubercule in the subcutaneous tissue [1,2]. Intracranial ALM are extremely rare; the first case was reported by Lasch et al in 1994 [15]. Since then 22 cases have been described. In contrast with subcutaneous ALMs, intracranial ALMs are more frequent in males [1,7,9,16,17]. From all the cases described 7 were in females with a ratio male/female 2/1 and the average age was 44.3 ± 16.9 years. Symptoms depend on the localization and size (Table I).

ALM seems to be benign, growing slowly without recurrence or malignant transformation. Colnat-Coulbois et al reported a case that was followed for six years after surgery without recurrence, which is the longest described in literature [8], although none of them have reappeared after surgery. Our case is the fourth one described in the posterior fossa. As described by Sun et al all reported cases seem to develop from dura matter near large venous vessels, like venous sinus or skull diploic structure [7]. Six cases are described as para-cavernous sinus lesions, three involve the falx cerebri [5,17,18] and three were described in posterior fossa [5-7].

Figure 2. Histologically we observed a tumor composed of vascular channels of variable sizes, dilated and congestive. Vascular channels are bordered by a wall showing numerous fusiform cells without cytological atypia.

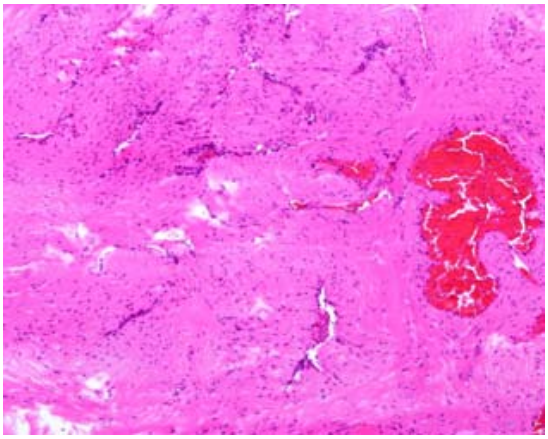
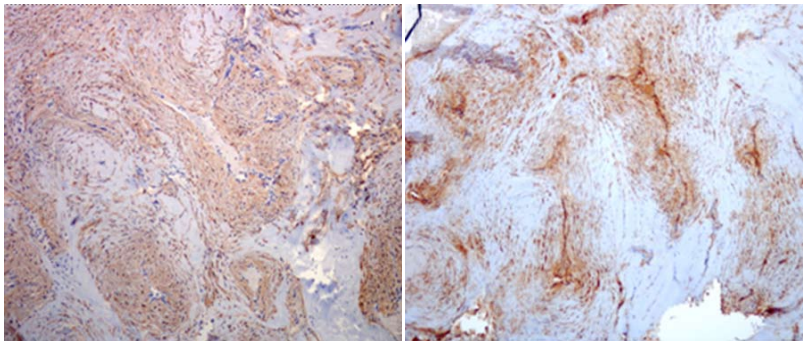


Figure 3. The immunohistochemical study showed that the spindle cells express muscle markers (caldesmon and actin).



Other locations described are less frequent, like cranial bone [7,14] or auditory meatus [11-13]. Two cases were reported as multiple lesions. Ravikumar et al reported two lesions one large right cystic lesion in the head of caudate and another solid one in the left globus pallidus [20]. Shinde et al describe another multicentric ALM in the right putamen, left hippocampus, bilateral optic nerves with thickened meninges [21].

ALM is difficult to diagnose and differential diagnosis include all extra-axis enhancing tumors such as meningiomas, hemangiomas, schwannomas and angiopericytomas [7]. In CT, ALMs are more frequently described as hyperdense lesions that enhance homogeneously after contrast administration. In three cases it was described as hypodense [14,15,21] taking into account that two of them were intraxial or in the diploic bone. Edema was not significant except in the case described by Li et al [16].

In our review, typically ALMs are isointense on a T_1 weighted MRI, but it is also described as hypointense in some cases. On T_2-weighted MRI it is usually hyperintense in a non-homogeneous pattern and is homogeneously enhanced after gadolinium administration. Sun et al described a flame-like enhancement after injection of gadolinium called 'enhancement flame' that seemed to rise from the tumor base [7]. Colnat-Coulbois et al has also described this phenomenon previously as a 'delayed progressive enhancement' [8]. In our case we could also see this progressive enhancement that could help in the diagnosis of ALM, which was also mentioned by Teranishi et al [9], but it has not been emphasized in previous literature.

The dural tail sign has also been described in some cases that may be difficult to diagnose. In four cases, dural tail was described and the lesion was misinterpreted as a menigioma [13,16,17,19]. In these cases, we consider that the progressive enhancement should help in differential diagnosis (Table II).

In our case, ALM presented an increase on the ADC, which was also described by Zhou et al as a hypointensity on diffusion weighted imaging (DWI) [19]. We also have described a generalized metabolite decrease in spectroscopy and, to our knowledge, this is the first time that these parameters were analyzed in an ALM. Lescher et al described that signal-time curves showed a prolonged signal decrease after contrast administration, and was then followed by a marked slow signal increase. This effect could explain the 'delayed progressive enhancement' that could differentiate ALMs from meningiomas [18].

According to the 2002 World Health Organization classification, ALM is recognized as an independent tumor entity. The typical morphology consists in abundant ectatic thick-walled vascular channels intermingled with spindle-shaped cells and bands of collagen without elastin. In our case, dilated and congestive vessels were surrounded by elongated spindle-shaped cells positive for smooth muscle actin. There were no signs of mitosis, cytological atypia or necrosis. Some cases [1,8,16,19] were described as having some described adipose tissue as a component of ALM. Also Colnat-Coulbois et al described the presence of hemorrhage, hyalinization and calcification [8]. We did not find any of these signs.

ALMs seem to be benign and potentially cured by surgery, and once the ALM is removed no cases of recurrence have been described during the follow-up (range: 7 months-6 years) for all but two cases. Only two of the described cases had a bad evolution [7,21]. Sun et al [7] published a large seized lesion that had an extremely rare complication that resulted in a fatal outcome because of the rupture of a pseudo-aneurysm of the internal carotid artery. Shinde et al [21] reported a case of a patient who died because of septicemia and recurrent seizures before any surgery was taken into account. According to some authors, given the potential hemorrhagic risk due to its high vascular component, GTR should be recommended, even though in some cases [4,5,16] subtotal or piecemeal resection was performed without major bleeding complications, as we have seen in our own experience. Also Sun et al [7] or Li et al [16] propose that digital substraction angiography or preoperative emboliza-

Table I. Clinical findings of patients presenting angioleiomyoma in the literature.

	Age (years) / Sex	Symptoms (duration)	Location (size)	Surgery	Follow up
Lasch et al [15]	47 / M	Abnormal gait with a right sided limp (14 months)	Leptomeninges right parietal lobe (2 × 2 × 2.7 cm)	Approach not specified, GTR	4 years, NR
Ravikumar et al [20]	12 / F	Headache, diplopia, seizures, left hemidystonia (2 months)	Right head of caudate, left globus pallidus (NA)	Right: frontal craniotomy, GTR	20 months, NR; left lesion quiescent
Kohan et al [12]	NA	Hearing loss, tinnitus (NA)	Internal acoustic meatus (NA)	Retrosigmoid, GTR en bloc	NA
Figueredo et al [4]	52 / M	Headache, horizontal diplopia (2 years); visual deficit, facial numbness (6 months)	Right cavernous sinus (6 × 6 × 5 cm)	Frontotemporal, GTR piecemeal resection	NA
Karagama et al [11]	47 / F	Hearing loss (12 months)	Internal acoustic meatus (1 cm)	Translabyrinthine, GTR	1 year, NR
Colnat-Coulbois et al [8]	50 / M	Headache (8 years) and diplopia	Left cavernous sinus (NA)	Frontotemporal intradural, GTR	6 years, NR
Vijayasaradhi et al [14]	10 / F	Headache (4 months)	Frontal intradiploic space (4 × 3 cm)	Supraorbital, GTR	NA
Gasco et al [6]	43 / M	Headache, blurred vision, dizziness, gait abnormalities	Left cerebellar hemisphere (4.4 × 3.9 × 3.9 cm)	Suboccipital, GTR in three pieces	NR
Chongxiao et al [17]	50 / M	Headache, seizures (6 months)	Falx cerebri (4 × 3 × 3 cm)	Bilateral parafalx approach, GTR	18 months, NR
Pepper et al [13]	13 / F	Hearing loss, headache (1 year)	Internal acoustic meatus (0.7 cm)	Transtemporal approach, GTR	6 months, NR
Xu et al [10]	53 / M	Headache (3 months); visual deficit (1 month)	Sella (< 1 cm)	Transphenoidal approach, GTR	NA
Shinde et al [21]	60 / M	Headache, seizures, irritability (2 months)	Right putamen (2 cm), left hippocampus, bilateral optic nerves	Biopsy	Died
Conner et al [5]	42 / M	Headache (8 years)	Right cerebellar hemisphere (0.8-1 cm)	Suboccipital, GTR	1 year and 11 months, NR
	36 M	Headache (8 years)	Falx cerebri (2.5 cm)	Interhemispheric, STR	2 years and 2 months, NR
Zhou et al [19]	62 / M	Seizure	Middle fossa (3.5 × 3.5 × 3.7 cm)	NA	7 months, NR
Li et al [16]	23 / F	Primary amenorrhea (9 years); visual deficit (8 years)	Cavernous sinus and sella (5.5 × 5.5 × 7 cm)	Fronto-temporo-zygomatic, STR	3 months, NR
	62 / M	Hypophrasia, hypomnesis, alteration of consciousness (2 months)	Temporal lobe (2.5 × 3.5 × 2.5 cm)	Temporo-parietal, GTR	NA
Teranishi et al [9]	52 / F	Right eye discomfort (6 months)	Cavernous sinus (2.3 cm)	Extradural temporopolar, GTR	NA
Sun et al [7]	51 / F	Visual deficit (2 months)	Sella (3 × 2.5 × 2.5 cm)	Transsphenoidal, GTR	Died
	49 / M	Weakness of lower limbs (1 year)	Sub-tentorium (4.2 × 4.6 × 5.7 cm)	Suboccipital, GTR	1 year, NR
	77 / M	Headache (5 months)	Left temporal diploic space (1.6 × 3.1 × 3.9 cm)	Parieto-temporal, GTR	1 year, NR
Lescher et al [18]	40 / M	Seizure (2 years), headache	Falx cerebri (NA)	NA, GTR	NA
Present study	43 / M	Hearing loss (6 months)	Sub-tentorium (1,4 × 1,3 × 1,4 cm)	Suboccipital, GTR piecemeal resection	2 years, NR

F: female; GTR: gross total resection; M: male; NA: not available; NR: no recurrence; STR: subtotal resection.

Table II. Radiological findings of angioleiomyoma described in the literature.

	Computerized tomography	Magnetic resonance imaging			Relation with dura	Delayed progressive enhancement
		T_1	T_2	Gadolinium		
Lasch et al [15]	Hypod, little edema, Hen	NA	NA	NA	No attachment	NA
Ravikumar et al [20]	Right: hypod, Hen mural nodule, cystic, no edema. Left: hyperd	NA	NA	NA	No attachment	NA
Kohan et al [12]	NA	NA	NA	NA	NA	NA
Figueredo et al [4]	Hyperd, no edema, Hen	Iso	Hyper	Hen	NA	NA
Karagama et al [11]	NA	Iso	Hyper	Hen	NA	NA
Colnat-Coulbois et al [8]	NA	Hypo	Hyper	Hen	NA	Pen
Vijayasaradhi et al [14]	Hypod, no enhancement with contrast	NA	NA	NA	NA	NA
Gasco et al [6]	Hyperd, no edema, Hen	Iso	Hyper	Hen	Broad dural contact	NA
Chongxiao et al [17]	Hyperd, no edema, Hen	Hypo	Hyper	Hen	Dural tail sign	NA
Pepper et al [13]	Expansion of right IAC	Iso	Iso	Hen	Dural tail sign	NA
Xu et al [10]	NA	Hypo	nH Hyper	NA	NA	NA
Shinde et al [21]	NA	Iso-hypo	Hyper	Hen	Bilateral dural enhancement	NA
Conner et al [5]	NA	NA	Hyper	nHen	NA	NA
	Hyperd, no edema, Hen	NA	NA	NA	NA	NA
Zhou et al [19]	NA	Hypo	nH Hyper	nHen	Dural tail sign	NA
Li et al [16]	Hyperd	Hypo	Hyper	nHen	NA	NA
	Hyperd, edema	Iso	Iso	nHen	Dural tail sign	NA
Teranishi et al [9]	Nodular tumor	Iso-hypo	Hyper	nHen	NA	Pen
Sun et al [7]	NA	Iso	Hyper	nHen	NA	Pen
	NA	Hypo	Hyper	nHen	NA	Pen
	NA	Iso	Hyper	Hen	No dural tail sign	NA
Lescher et al [18]	NA	Iso	Hyper	Hen	Small dural contact	Pen
Present study	Hyperd, no edema, Hen	Iso	Hyper	nHen	Broad dural contact, no dural tail sign	Pen

Hen: homogeneous enhancement; Hyper: hyperintense; Hyperd: hyperdense; Hypo: hypointense; Hypod: hypodense; IAC: internal carotid artery; Iso: isointense; nHen: nonhomogeneous enhancement; NA: not available; Pen: progressive enhancement.

tion would reduce intraoperative bleeding and decrease surgery risk. Shinde et al [21] also have proposed immune-target therapy in multicentric ALMs.

In conclusion, intracranial ALMs are extremely rare tumors that seem to have a good outcome after surgery without recurrence after total resection. Frequently ALMs seem to appear in relation with the dura but could appear as intra-axial lesions. The differential diagnosis with other entities could be difficult but some MRI special features like progressive enhancement may be helpful, and an alternative possibility this lesion should be considered. Histology is the only way to have a definitive diagnosis. Surgery resulted in good outcomes in most of the procedures, but risk of bleeding and tumor size should be taken into account when planning their treatment.

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Angioleiomioma infratentorial: caso clínico y revisión de la bibliografía

Introducción. Los angioleiomiomas intracraneales son lesiones muy poco frecuentes. Sólo se han descrito 22 casos en la bibliografía hasta la actualidad, únicamente tres de ellos de localización infratentorial.

Caso clínico. Varón de 43 años con un angioleiomioma infratentorial descubierto de forma incidental tras la realización de una tomografía computarizada por hipoacusia. La resonancia magnética mostró una tumoración de 1,4 cm, descrita inicialmente como un meningioma, con un realce progresivo tras la administración de gadolinio, un aumento del coeficiente de difusión aparente y un descenso generalizado de metabolitos en la espectroscopia. La lesión se resecó quirúrgicamente mediante un abordaje suboccipital con buena evolución y sin complicaciones postoperatorias. En el estudio histológico, la lesión presentaba un abundante componente vascular, y en la tinción inmunohistoquímica era positiva para actina y caldesmona. Dos años después de la cirugía, el paciente no presentaba recurrencia en la resonancia magnética de control.

Conclusión. El diagnóstico de los angioleiomiomas puede ser complejo, pero algunas de sus características radiológicas pueden facilitarlo. Los angioleiomiomas son tumores benignos asociados con un resultado funcional favorable tras su resección completa, que en nuestro caso no presentó un alto riesgo de sangrado.

Palabras clave. Angioleiomioma. Coeficiente de difusión aparente. Espectroscopia. Infratentorial. Realce progresivo. Tumor intracraneal.