International Journal of Innovative Research in Medical Science (IJIRMS)
Volume 07, Issue 07, July 2022,
https://doi.org/10.23958/ijirms/vol07-i07/1461

Case report

Primary Intracranial Parasagittal Angioleiomyoma: A Case Report of Uncommon Location of a Rare Tumor

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Received 29 Junee 2022; Accepted 23 July 2022; Published 27 July 2022

Abstract

Background: Angioleiomyoma is a soft tissue benign tumor that occurs mainly in subcutaneous tissue, involving the lower extremities in particular. We present a rare case of angioleiomyoma invading the central nervous system. ***Case description:*** A 17-year-old Saudi male presented with two months history of headache and vomiting. MRI revealed an intra-axial mass in the right parietal area. Macroscopically, the mass measuring 4.5×4×3 cm after was resected. Both imaging and histopathological findings were consistent with angioleiomyoma. In a period of 6 months of follow-up there was no recurrence of the tumor. ***Conclusion:*** Primary intracranial angioleiomyoma is an extremely uncommon tumor, though possible to happen.

Keywords: ***Intracranial angioleiomyoma, Parietal tumor, Soft tissue tumor***

Introduction

Angioleiomyoma is a benign indolent soft tissue tumor that occurs mainly in subcutaneous tissue particularly involving the lower extremities with a predilection to occur in a middle-aged female patient [1,2]. Primary intracranial angioleiomyoma is a rare tumor, up to date only 39 cases have been reported [3,4]. It tends to occur in the cavernous sinus although a wide range of anatomical locations including parasellar region, cerebellopontine angle, internal and external auditory canals have been described [5,6]. Here we are reporting a 17-year-old boy who underwent a gross total resection of right parietal angioleiomyoma.

Case report

Patient's perspective:
A 17-year-old male patient, known case of ataxia telangiectasia who presented to our service with a history of headache and vomiting that lasted for two months ago.

Clinical findings:
Clinically patient was conscious, alert, and oriented with no neurological deficit.

Diagnostic assessment:

MRI brain scan showed a well-defined intra-axial mass in the right parietal area that is hypo-intense in T1 weighted sequence, iso- to hyper-intense in T2 weighted sequence with homogeneous enhancement in T1 post-contrast sequence (Figure 1).

Surgery and pathology:
Patient underwent right parietal craniotomy and gross total resection. The tumor was yellowish in color with a clear margin separating the tumor from the surrounding parenchyma. The tumor was vascularized with excessive bleeding, rubbery in consistency.

Macroscopically:
The mass was soft regular with smooth lobular outer surface, measuring 4.5×4×3 cm in maximum dimensions, cut section shows tan whirled surface.

Histopathological evaluation shows well circumscribed encapsulated proliferation of interlacing bundles of smooth muscle fibers with dilated vascular spaces (Figure 2 A, B). The vascular muscle wall present within the tumor (Figure 2 C), mitosis was 1/10 HPF and there was no necrosis or pleomorphism. Based on the above findings, diagnosis of angioleiomyoma (cavernous type) was made.

Follow up and outcome:
Post-operatively, patient has the same neurological status, 6 months follow-up using MRI brain scan confirmed no recurrence of the tumor.

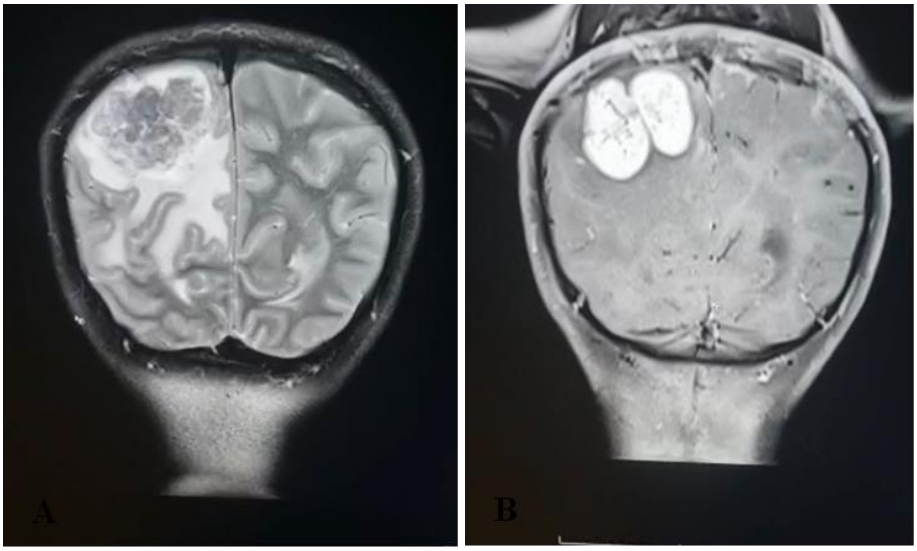

Figure 1. (A) MRI brain T2 weighted image shows right parietal intra-axial lesion with vasogenic edema. (B) MRI T1+C shows that the lesion is avidly enhancing with two nodules.

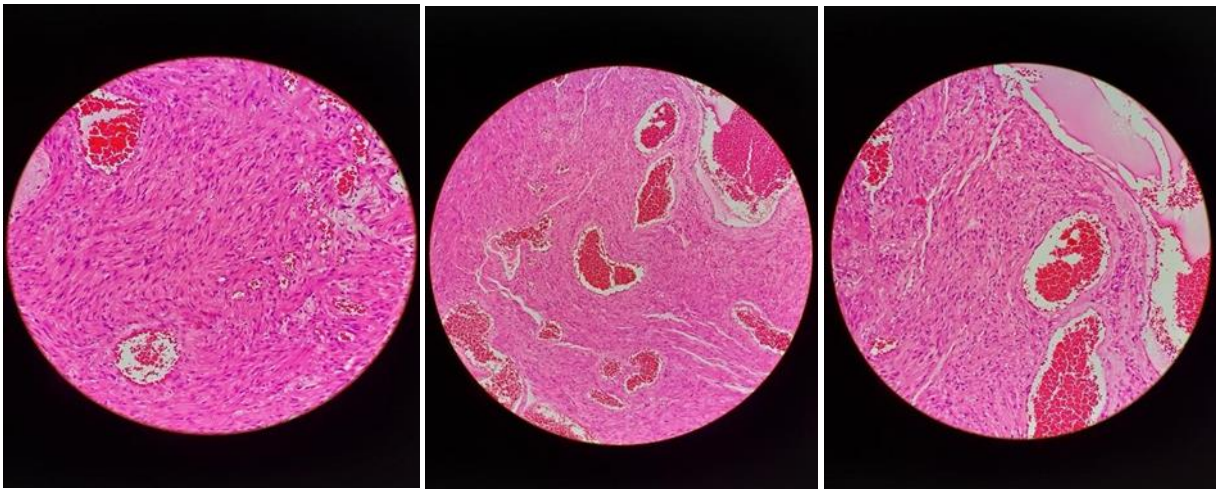

Figure 2. (A) Low power view show mixture of dilated vascular spaces and spindle cells. (B) X40 view show interlacing bundles of bland looking smooth muscle spindle cells. (C) Vascular muscular walls present within the tumor.

Discussion

Angioleiomyoma is a soft tissue tumor that occurs in lower extremities with indolent course [2]. It is thought to arise from the vascular smooth muscle cells [1,2]. The first description of intracranial involvement was by Lach et al in 1994 [7]. Further case reports and series described the occurrence of angioleiomyoma intracranially with, up to our knowledge, only 39 cases that had been reported [3-9].

The exact etiology for intracranial angioleiomyoma formation is unknown. Shinde et al suggested an immune mediated stimulation for vascular smooth muscle cells that result in its formation [10]. The tumor commonly occurred in middle-aged patients with male prediction [4]. Few cases have been reported in the pediatric age group [3].

The clinical manifestations of such cases are not specific and mainly due to the space occupying effect of the tumor and presentations are site related. However, in about 18 of the patients they were complaining of headache only. Other high ICP symptoms like nausea and vomiting are not uncommon [8]. In our reported case, the patient was complaining of headache and vomiting for a 3-month duration that increased in intensity two weeks prior to presentation.

Although the location of angioleiomyoma is commonly seen in cavernous sinus cerebellum and Sella, other anatomical locations have been described [4-6]. It includes parenchymal, intraventricular, basal ganglia, optic nerve, temporal lobe, skull base and external auditory meatus [6,11,12]. In our described case the location was intraparenchymal in the right parietal area.

The magnetic resonance imaging (MRI) features of angioleiomyoma are usually similar to meningioma with isointense signal in T1 weighted sequence and hyperintense in T2 weighted sequence with homogeneous enhancement in post contrast T1 sequence [6]. Dural tail in post contrast T1 has been described with meningioma as well as angioleiomyoma [8,13]. However, rapid maximum enhancement has been described with meningioma in contrast to angioleiomyoma which showed progressive gradual enhancement [6]. The other differentiating feature is the calcification which is seen in 25% of meningioma but has not been reported with angioleiomyoma [14].

Based on the 2021 WHO classification of CNS tumor, angioleiomyoma (vascular leiomyoma) is classified as a mesenchymal, non-meningothelial soft tissue tumor group [15]. It typically forms a varying proportion of blood vessels and smooth muscle cells, rarely associated with necrosis, atypia or pleomorphism [6]. Immunohistochemistry of angioleiomyoma is associated with positive SMA, Calponin. Vascular spaces can be highlighted with CD34 and CD31. Three histological subtypes of angioleiomyoma were described: solid, venous, and cavernous. Cavernous type is seen mainly with intracranial angioleiomyoma. In our described case, a cavernous type was found which is consistent with literature findings.

Gross total resection should be the first treatment option as suggested by Colnat-Coulbois S et al.[16] with no further adjuvant

therapy is needed. Although it is considered a WHO grade 1 [15], strict surveillance clinically and radiologically is needed to rule out progression of residual or recurrence.

Conclusion

Angioleiomyoma is a rare benign tumor involving the CNS, particularly cavernous sinus while the description of intraparenchymal location is uncommon. Diagnosis relay on the histopathological examination. Rarity of the tumor and different anatomical locations make it difficult to show its long-term outcome which is the main limitation in our study.

Patient's perspective

The patient has been followed regularly in the outpatient clinic. No recurrence of the tumor after 6 months of surgery.

Declarations

Ethical approval

Approval was obtained from the local Ethical Committee in our Hospital.
Consent to participate informed consent was obtained from legal guardians.

Funding

Not applicable.

Conflict of Interest

On behalf of all authors, the corresponding author declares that there is no conflict of interest.

Consent for publication

Patient’s guardian has been consented for submission of the case report.

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