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Isolated giant cerebral varix – A diagnostic and therapeutic challenge: A case report

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Abstract

Background: Isolated giant cerebral varix (IGV) is an uncommon vascular lesion that represents a diagnostic challenge and requires dynamic vascular studies for its characterization. The IGV is considered a benign, low-flow venous lesion with very low risk of bleeding that might cause secondary symptoms mainly due to compression of the adjacent parenchyma.

Case Description: A 12-year-old female patient with non-contributory medical history presented with headache for the last 2 months. Upon admission, her neurological examination was unremarkable. Magnetic resonance imaging (MRI) and computed tomography angiography (CTA) images demonstrated a large varicose dilation of the superficial Sylvian vein, located anterior to the left temporal pole, with no evidence of abnormal arteriovenous connections or tumoral lesions. This finding was considered incidental and unrelated to her symptoms. In this case, we considered that the combination of CTA and MRIs was enough to establish an accurate diagnosis, excluding the need to perform invasive imaging studies. Taking into account these considerations, the patient was managed with conservative treatment and has been followed up for 1 year, remaining asymptomatic.

Conclusion: Cerebral IGVs are rare vascular lesions that are treated conservatively when asymptomatic and surgically in the case of rupture or compression of adjacent structures. Given our observation of a high unlikelihood of vascular connections to arteries, and the information obtained with non-invasive imaging techniques such as CTA and MRI was enough to make a clinical decision and avoid the evaluation with invasive procedures.

Key Words: Arteriovenous malformation, cerebral varix, developmental venous anomaly, venous aneurysm

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INTRODUCTION

Cerebral varices are frequently associated with other vascular abnormalities such as arteriovenous malformations (AVMs) and developmental venous anomalies (DVA). Their etiology is usually explained by an increase in venous drainage pressure.[6,15] Isolated cerebral varix is an uncommon, rarely documented

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phenomenon with an unclear etiology. Within few reported cases, the isolated giant cerebral varix (IGV) is usually described as an incidental finding in patients without associated neurological deficit.[6,15] In addition, due to its minor clinical manifestations, it is often misdiagnosed as a cystic or tumoral lesion, making its diagnosis undoubtedly challenging.

CASE REPORT

A 12-year-old female patient without relevant medical history complained of 2 months of right, unilateral, intermittent headache, associated with photophobia and hyperacusis that improved with the administration of analgesics. Upon admission, her neurological examination was unremarkable. Magnetic resonance images (MRIs) were performed, revealing an elongated well-defined extra-axial abnormality, located in the anterior aspect of the middle cranial fossa, showing high signal on T2-WI, intense homogeneous enhancement after intravenous contrast administration, and absence of flow voids or pulsation artifacts [Figure 1]. Computed tomography angiography (CTA) images of the head and neck with arterial and venous phases demonstrated dilation of a venous structure corresponding to the superficial Sylvian vein (SSV), with affluence from the superficial temporal veins, as well as the basal frontal vein, draining into the sphenoparietal sinus. The M1 and M2 segments of the middle cerebral artery were adequately visualized without an evidence of arteriovenous communication to the lesion or dilatation that suggested hyper flow [Figure 2]. Furthermore, the absence of dilation of the external carotid artery branches, specifically the middle meningeal artery ruled out the presence of an arteriovenous fistula [Figure 3]. As a result, the diagnosis of a cerebral IGV was made. The characteristics of the headache were compatible with migraine. The absence of neurologic deficit in the physical examination made unlikely the compression of the adjacent cerebral parenchyma by the venous dilation. Consequently, conservative management with medical treatment was prescribed for the migraine achieving complete resolution of the headache. The patient was discharged and followed up as an outpatient.

DISCUSSION

Cerebral varices are often associated with other vascular malformations, more frequently with AVMs and DVA.[3,6,7,15] The association with AVMs is believed to be secondary to vascular changes induced by a high-flow arteriovenous shunt pressure, which in turn, lead to a high risk of bleeding.[2,14] DVAs, on the other hand, have been most commonly described as early childhood congenital malformations, which can also lead to varix formation.[3,13]

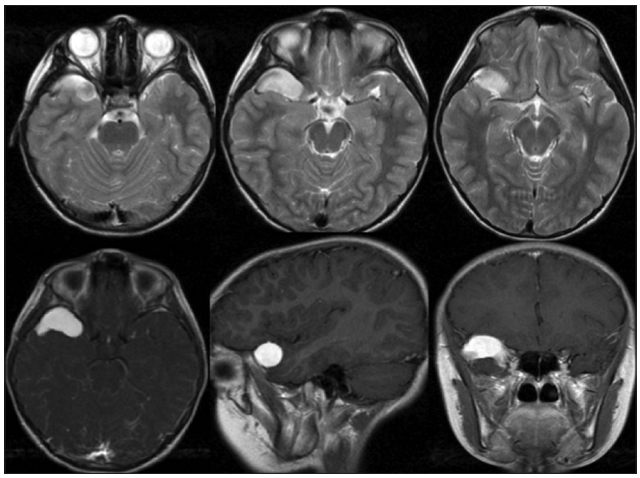

Figure 1: Cerebral magnetic resonance image. Top row: Axial T2-weighted images. Bottom row: Axial, sagittal, and coronal T1-weighted images with intravenous contrast. A well-defined elongated extra-axial lesion located anterior within the right middle cranial fossa, with slightly heterogeneous high signal intensity on T2-weighted images, and high homogeneous contrast enhancement

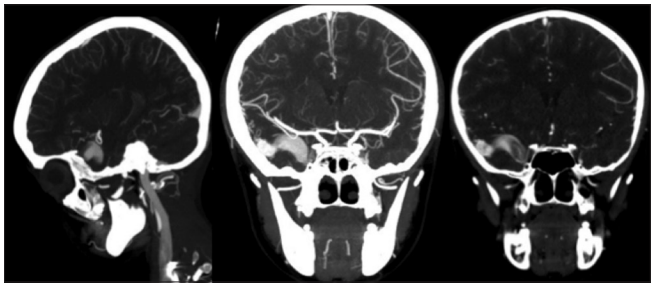

Figure 2: Cerebral computed tomography-angiography images with maximum intensity projection reconstructions in venous phase demonstrating the absence of connections between branches of the middle cerebral artery and the superficial Sylvian vein lesion

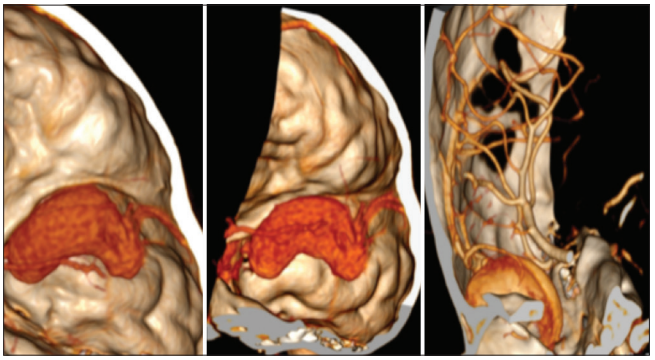

Figure 3: A three-dimensional reconstruction of cerebral computed tomography angiography venous phase images showing a large dilation of the superficial Sylvian vein. No abnormal arterial connection is noticed

Isolated presentation of a giant cerebral varix is a highly unusual finding that was first reported in 1987.[1] The low incidences of reports in the literature of this phenomenon have led to fierce debate regarding its etiology.[10] Lasjaunias *et al.* believe that persistence of embryologic

venous drainage leads to structural weakness of venous walls.[6] The histologic description of a varix wall includes a single layer of fibromuscular tissue adhered to the vascular endothelium, which predispose to dilation and supports this theory.[2]

In most of the reported cases, isolated cerebral varices are subclinical entities discovered incidentally.[1,3,5,7,10,11] On those rare occasions in which they produce symptoms, they are usually associated with hemorrhage due to rupture, thrombosis, or mass effect on neighboring structures.[9,12] There is currently not enough information in the literature to determine the risk of rupture and bleeding in isolated cerebral varices.

The diagnosis of an IGV is based on radiologic findings. On CT images of the brain without contrast, cerebral varices appear as extra-axial lesions with similar attenuation to the surrounding parenchyma. When the varix has contact with the skull, it causes bone remodeling, which allows ruling out the differential diagnosis of meningioma, which would cause hyperostosis in the adjacent bone.[10,11,15] On MRIs, cerebral varices are described as extra-axial, cystic, and well-circumscribed lesions. Occasionally, they have been reported as intraaxial lesions, which can lead them to be mistaken with bleeding cystic tumors or thrombosed vascular malformations.[5] They have intermediate signal intensity on T1-WI, high signal intensity on T2-WI, and enhance intensely after contrast administration, like in our patient, the homogeneous enhancement, and absence of flow voids are features that suggest a low flow vascular abnormality, consistent with the diagnosis of a venous varix. Before the advent of MRI, the imaging study of choice for cerebral varices was the conventional cerebral angiography because of its accurate depiction of vascular abnormalities. Recently, MRI has surpassed angiography because it provides information regarding the etiology and allows an accurate differential diagnosis in most of the cases.[4,8,10] In addition, MRI-angiography allows the visualization of arterial and venous phases without the administration of contrast media, ruling out abnormal arteriovenous communications.

In our case, CTA images of the brain with three-dimensional reconstruction allowed the complete evaluation of the variceal lesion, and the determination of possible arterial pedicles, dilations, or arteriovenous communications that would suggest an AVM or cranial and spinal dural fistula. We were able to determine that the dilated vein was a right SSV, and we ruled out arterial communication or high-flow signs, concluding with the diagnosis of a giant cerebral varix. Invasive procedures such as conventional cerebral angiography were not considered due to patient age and the adequate evaluation of the lesion without invasive imaging techniques. CT imaging is seldom used in the diagnosis of this type of malformation, only found in a single case report by Asano *et al.* in which a subepicranial varix was diagnosed with CT imaging of the brain, showing a better diagnostic value than conventional cerebral angiography.

To the best of our knowledge, there is limited information in the literature regarding the definitive treatment of cerebral varices. Because these lesions are usually diagnosed incidentally without associated symptomatology, conservative treatment is generally considered the most appropriate course of action.[1,3,6,15] Surgical or endovascular treatment is only considered in patients with symptoms arising from hemorrhage secondary to varix rupture or compression of adjacent structures,[9,12] otherwise it is not suggested because the varix may drain to a normal venous territory, as occurred with the DVAs.

CONCLUSION

Cerebral IGVs are extremely rare and, to the best of our knowledge, there is insufficient information available in the scientific literature. The incidence and the risk of rupture remain unclear, making this entity a diagnostic and therapeutic challenge. Currently, conservative management is the most appropriate treatment for asymptomatic lesions; however, in case of rupture or compression of adjacent neurovascular structures, endovascular or surgical treatment is warranted.

Given the MRI and CTA findings, the observation of a highly unlikelihood of abnormal connections to arterial feeders was sufficient to make a clinical decision and avoid interventional angiography procedures. We emphasize the use of noninvasive imaging techniques such as CTA and MRI for the diagnosis of this type of lesions, when AVMs and dural fistulas can be reliably excluded, avoiding the risk of invasive procedures particularly in children.

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Conflicts of interest
There are no conflicts of interest.

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