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Case Report

Bubbles and Troubles: A Case of Azygous ACA with Bilateral ICA Aneurysm, Besides the Usual Distal ACA Aneurysm

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Abstract: Azygous ACA is one of the rare anatomic variants of anterior cerebral artery. Predisposition to aneurysm development is attributed to the alteration in flow dynamics. Such hemodynamic alteration is most commonly seen in the anterior circulation, causing distal ACA aneurysm to be the most commonly associated vascular pathology. Although neurosurgical literature is replete with anatomical reports of such cases, there has been no case reports where bilateral ICA aneurysms are also present with the distal ACA aneurysm. In this case, the presence of hypoplastic left A1 segment might predispose the other segments of Circle of Willis to hemodynamic alterations. Thus, whenever one comes across an azygous ACA in association with an additional anomaly he should be vigilant enough to look for other pathologies beyond the commonly encountered distal ACA aneurysm. The presence of azygous ACA also poses a technical challenge due to the risk of ischemic insult to anterior regions of bilateral hemispheres during temporary clipping of the single A2 trunk. In addition to it, the presence of a hypoplastic A1 segment also makes the approach to the giant aneurysm in ICA segment more technically demanding as cross flow is not adequate. So, the presence of multiple anatomic anomalies not just increases the number of vascular pathologies but also enhances the challenges that the surgeon has to face in planning the strategic approach to tackle all the lesions.

Keywords: Azygous ACA, Distal ACA Aneurysm, Hypoplastic A1 Segment, Bilateral ICA Aneurysm

1. Introduction

Awareness of cerebrovascular anatomic variations and the clinicopathologic entities associated with them play a key role in deciphering the pathophysiologic basis of various neurovascular disorders and ultimately devising a management plan. Azygous anterior cerebral artery is a rare anatomical variant of ACA with a reported incidence of 0.1-0.5%. [1-3] The altered flow dynamics subjects the anterior vascular territory to hemodynamic stress and thus leads to an increase in the incidence of distal ACA aneurysms in such patients, with estimated figures ranging from 13-71%. [4, 5] Although, in such cases distal end of azygous ACA is the most common site for aneurysm formation [4], other regions of anterior circulation can also be under the hemodynamic stress, especially when associated with other anatomic variations.

Medical literature has a handful of cases of azygous ACA with associated saccular aneurysm at distal ACA, but to our knowledge, this is the first reported case of an azygous ACA with bilateral ICA aneurysms, besides the usual distal ACA aneurysm.

2. Case Report

A 62 years female presented to our emergency department with single episode of loss of consciousness followed by dizziness and vomiting. She was conscious but disoriented. Noncontrast CT images showed thick bilateral sylvian SAH with IVH and ICH in the left basifrontal region (Figure 1). CT angiography depicted a giant saccular aneurysm in the

supraclinoid segment of left ICA, in association with azygous ACA and dominant right A1 segment (Figures 2, 3).

Figure 1. *Axial view of CT head showing bilateral sylvian subarachnoid hemorrhage with associated intraventricular hemorrhage and intracerebral hemorrhage.*

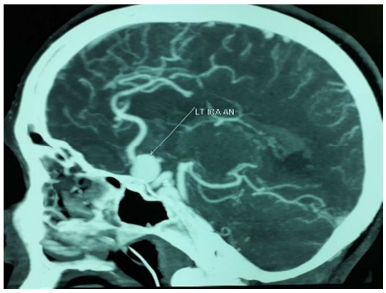

Figure 2. *Saggital view of CT angiography giant aneurysm at the supraclinoid segment of left ICA.*

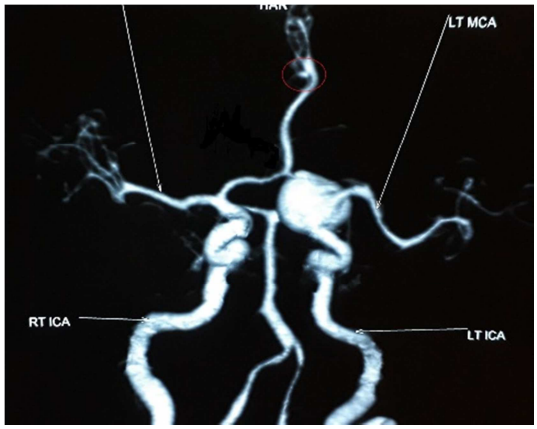

Figure 3. *3D reconstruction of CT angiography depicting the giant aneurysm in left ICA and distal ACA aneurysm (red circle).*

Diagnostic angiography through right femoral access revealed a total of three aneurysms. Besides the above mentioned giant aneurysm in the left supraclinoid ICA, there was another aneurysm in the communicating segment of right ICA and another aneurysm at the bifurcation of the azygous ACA (Figure 4). No other anatomical anomalies were detected. Carotid angiography with compression of contralateral carotid artery confirmed the presence of azygous ACA with absence of Acom (Figure 5). Patient was offered endovascular coiling for all the three aneurysms. However, the patient's bystanders did not give their consent for the procedure. The patient was discharged on request and subsequently was lost to follow up.

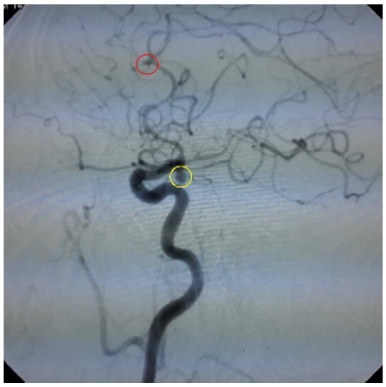

Figure 4. *Lateral view of catheter angiography showing aneurysm in the supraclinod segment of right ICA (yellow circle) and distal ACA aneurysm (red circle).*

Figure 5. *Antero-posterior view of catheter angiography through left ICA with right carotid compression, showing giant aneurysm in the supraclinoid segment of left ICA and an azygous ACA, with a small aneurysm at the bifurcation.*

3. Discussion

Since the original description of circle of Willis in 1664, various anatomic studies have been done to demonstrate the importance of paired anterior cerebral artery and anterior communicating artery in not just completing the circle geometrically but also in the embryological development of prefrontal cortex. As we descend down the ladder of phylogeny, there seems a strong association of a less developed prefrontal cortex and with either the presence of a single midline azygous anterior cerebral artery or the absence of anterior communicating artery with two anterior cerebral arteries lying in parallel. [1] In humans, its association with alteration in hemodynamics in frontal lobe leads to various clinicopathologic entities like agenesis of corpus callosum, hydraencephaly, holoprosencephaly, defects in septum pellucidum, porencephalic cysts and AVMs. [6] Baptista, through his seminal study on 381 brain specimens has classified the anatomic abnormalities in distal portions of ACA into three main types: (1) azygous anterior cerebral artery, in which a single midline anterior cerebral artery is formed by joining of bilateral A1 segments, with absence of anterior communicating segment and runs in the interhemispheric fissure, supplying the territories of both

cerebral hemispheres; (2) bihemispheric ACA, in which though there are two ACAs one is clearly dominant and supplies the contralateral cerebral hemisphere as well; and (3) triplicate ACA, in which there is a median corpus callosal artery besides the normal paired distal ACAs. [7] A "true" azygous ACA, according to Lasajaunias et al, does not divide but the "fused" variant of azygous bifurcates into separate distal ACAs along its course over the corpus callosum. [8]

In accordance with the Willis idea (1664) of equalization of blood flow to all parts of brain, the total blood flowing into the circle of Willis through the supplying arteries (carotids and basilar artery) must match with the blood flowing out through the distributing arteries (ACA, MCA, PCA). [7] The reason behind the genesis of aneurysms in anomalous vasculature is attributed either to the hemodynamic stress caused by the anomalous vessel or it develops along side the abnormal vessel. [9] This hypothesis of undue hemodynamic stress at the site of bifurcation or trifurcation of A2 is well supported by frequent clinical association of aneurysms of distal ACA with azygous, bihemispheric and triplicate ACA. [10] The bifurcation region of azygous ACA routinely serves as a zone of hemodynamic stress as a single azygous trunk is overwhelmed by an increased blood flow compared to a normally paired A2 segments of ACA. This could explain the formation of distal ACA aneurysm, and are more commonly saccular than non saccular in morphology. [11] In addition, this patient had hypoplastic left A1 segment which further predisposed the left ICA into undue hemodynamic stress, leading to the genesis of an aneurysm in the left supraclinoid ICA. The azygous trunk being fed mostly by the dominant right A1, the preferential blood flow through the right component of the circle of Willis again predisposes to the development of aneurysm in the right ICA segment, which in this case is represented by the aneurysm in the communicating segment. Although the caliber of right ICA is not significantly larger than the left one, by Poiseuille's law even a small difference can have a considerable difference in terms of blood flow. [12] Thus, presence of an associated hypoplastic A1 predisposed the other segments of Circle of Willis towards aneurysm development, in addition to the distal ACA aneurysm that is usually encountered with azygous ACA.

Presence of azygous ACA points towards the unique perfusion pattern, where a single vessel supplies the anterior portions of bilateral hemispheres. This fact that is well corroborated by clinical scenarios where thrombosis of azygous ACA has led to ischemic stroke in bilateral frontal and anterior callosal regions. [13, 14] From a neurosurgeon's perspective such an anatomy should make one more cautious while applying the temporary clips on A2 as it increases the risk of ischemic insult to bilateral basifrontal regions. [15] The presence of hypolastic A1 also removes the luxury of cross flow and one needs to be careful enough while dealing with the giant ICA aneurysm to not to occlude the main trunk. So, presence of hypoplastic A1 along with azygous ACA not only increases the number of bubbles (aneurysms) in the brain but also enhances the troubles for the treating surgeon.

4. Conclusion

The best diagnostic modality to confirm azygous ACA is catheter angiogram through one carotid artery while compressing the contralateral one. Although distal ACA bifurcation aneurysm is commonly encountered in cases of azygous ACA, it is definitely not the only vascular pathology, especially in the presence of an associated anatomic anomaly. Such a scenario should alert the treating team to diligently search for other possible vascular pathologies as well and come up with a strategic plan to tackle all the lesions.

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