

# Case series: Saccular aneurysm of the azygos anterior cerebral artery: report of 2 cases and review of literature

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The azygos anterior cerebral artery (ACA) is a rare variation in the circle of Willis, in which the distal ( $A_2$ ) segments of both ACAs are represented by a single common vessel, from which arise all the major vessels supplying most of the medial aspects of both anterior cerebral hemispheres and the corpus callosum.<sup>[1]</sup> Aneurysms of the azygos or unpaired ACA are extremely rare and only a few cases have been reported in literature.<sup>[1-5]</sup> This is a report of two cases of saccular aneurysm of the azygos ACA, diagnosed by angiography, the findings of which were confirmed at the time of surgical clipping. The clinical significance of the azygos ACA is highlighted.

## Case Reports

### Case 1

A 45-years-old woman presented to the emergency department with acute onset of severe bifrontal headache, dizziness, and neck pain. There was no history of seizures or loss of consciousness. There was no past or family history of hypertension or diabetes. Blood pressure was 120/80 mmHg and the patient was afebrile. Though the patient was lethargic, her memory functions were intact. The patient was able to move all her extremities, cranial nerve functions were normal, and there was no impairment of brain stem reflexes. Noncontrast CT scan of the brain showed anterior interhemispheric and intraventricular hemorrhage. Digital subtraction angiography (DSA) showed an azygos ACA, with a saccular aneurysm arising at its bifurcation [Figure 1 A-C]. In addition, the patient had another aneurysm from the basilar artery, between the left posterior cerebral artery and the left superior cerebellar artery. The anterior circulation aneurysm had ruptured and, hence, this was

operated. The azygos state was confirmed at surgery.

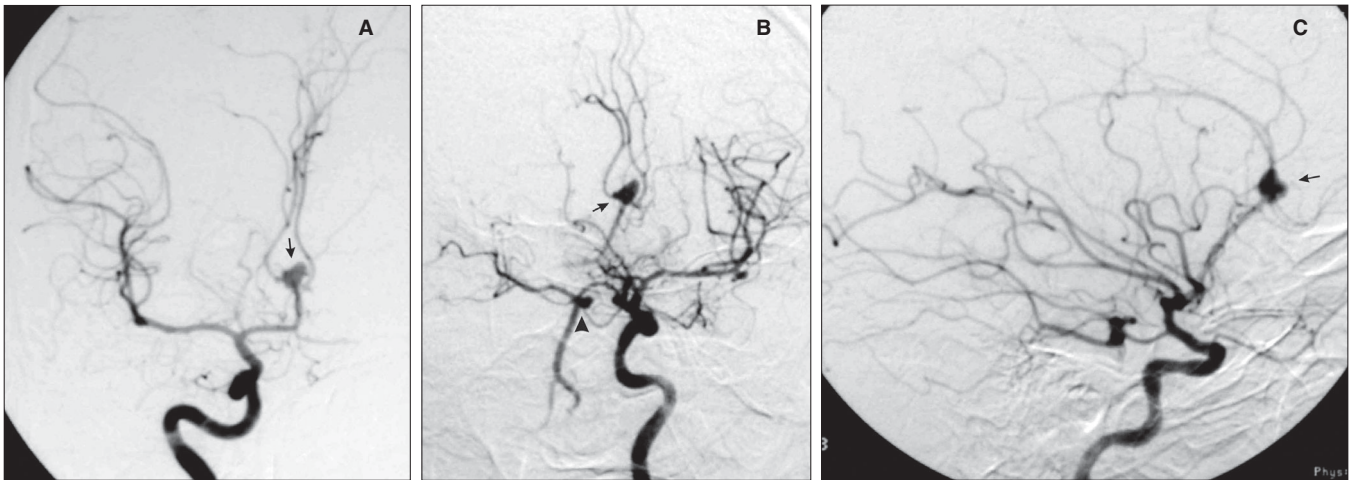
### Case 2

A 40-years-old hypertensive, nondiabetic woman developed severe headache and meningismus associated with transient loss of consciousness. A lumbar puncture at a private hospital revealed bloody CSF and the patient was referred to our institute for further evaluation. CT of the brain showed diffuse subarachnoid hemorrhage. DSA revealed a saccular aneurysm arising at the ramification of an azygos ACA; this was confirmed at surgery [Figure 2 A and B].

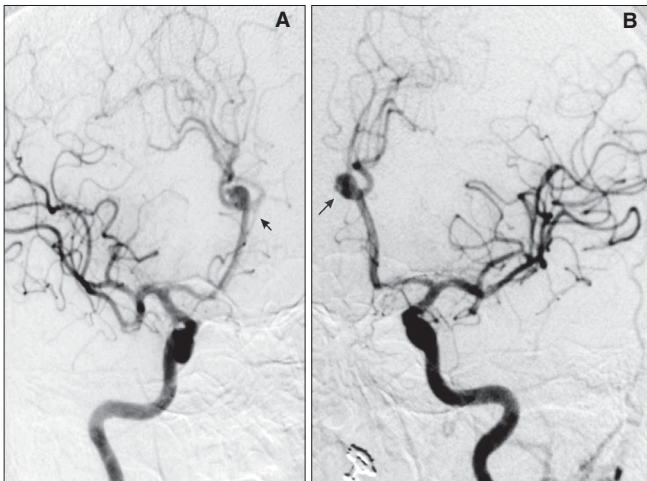
## Discussion

The ACA and the anterior communicating artery complex are an important division of the circle of Willis. Variations of the ACA deserve attention because they govern the distribution of blood to the two cerebral hemispheres, and certain clinical syndromes of cerebral vascular occlusive disease will be a function of their specific anatomic arrangement. The azygos ACA is a rare variation of the circle of Willis in which the distal ( $A_2$ ) segments of both ACAs are represented by a single common vessel, from which arise all the major vessels supplying most of both anterior cerebral hemispheres as well as the corpus callosum.<sup>[1]</sup>

A brief phylogenetic review of the ACA was presented by LeMay and Gooding in 1966, where they had stated that among the lower mammals there was no anterior communicating artery. In chimpanzees, the ACAs unite at the entrance of the longitudinal cerebral fissure and course forward to the rostrum of the corpus callosum, where the trunk then divides. This description resembles that of an



**Figure 1 (A-C):** Right internal carotid (ICA) angiogram, PA projection (A), shows the azygos ACA with the associated saccular aneurysm (arrow). PA (B) and lateral (C) projections of the left ICA angiogram, show the aneurysm of the azygos ACA (arrow). The prominent posterior communicating artery fills the basilar artery. Another small aneurysm (arrowhead in B) is seen between the left posterior cerebral artery and superior cerebellar artery.



**Figure 2 (A and B):** Oblique projections of the right (A) and left (B) ICA angiograms show the azygos ACA and the associated aneurysm (arrow).

azygos ACA.<sup>[2]</sup>

Lesem stated that in the human fetal and infantile brains which he had examined, he could make out the sequential changes marking the transition of the ACA from a single artery to two arteries. In 22 out of 33 human fetuses (68%) and in 18 out of 24 infant brains (75%), the two ACAs united to form a single artery within the longitudinal fissure.<sup>[6]</sup> DeVriese hypothesized that since there was a higher incidence of azygos ACA in fetuses than in adults, in early embryonic life either one or three ACAs may exist. If only one existed, a single stem would duplicate and in case three vessels were present, one would degenerate.<sup>[7]</sup>

According to Baptista's review of the literature, 23 of 2153

adult brains (1.0%) had unpaired ACAs.<sup>[1]</sup> Baptista described three types of anomalies occurring in the distal ACA:

a) a true azygos artery, from which all major branches are given off to both cerebral hemispheres; b) a bihemispheric ACA, where both right and left ACAs are present, but one is rudimentary and most of the major branches to both hemispheres arise from the other ACA; c) a triple ACA, with the accessory ACA arising from the anterior communicating artery.<sup>[1]</sup> Our cases were judged to belong to type (a).

The clinical significance of the azygos ACA is threefold: 1) Its association with other congenital anomalies such as holoprosencephaly; 2) The existence of an azygos ACA can explain the presence of bilateral ischemic changes in the setting of occlusion of the vessel and; 3) The high incidence of associated berry aneurysms.<sup>[2]</sup>

Aneurysms of the azygos ACA, either ruptured or unruptured, are extremely rare and only a few cases have been previously reported in the literature: three by Pool et al., three by Latinen et al., and one each by Baptista, LeMay et al., Katz et al., and Kondo et al.<sup>[1-5]</sup> Abnormal medial and intimal elements in the vessel wall of this developmental variant may account for the increased prevalence of associated saccular aneurysms.

Though these aneurysms can be noninvasively imaged either by CT angiography or MR angiography, DSA plays an important role in identifying saccular aneurysms of the azygos ACA and it is the gold standard for diagnosis. An exact preoperative diagnosis is important because extra precautions have to be taken during the surgical clipping of these aneurysms, since injury to this common arterial trunk can lead to devastating complications.<sup>[3]</sup>

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