Aneurysm of azygous anterior cerebral artery

Case report

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This report describes a patient with a fusiform aneurysm of an azygous anterior cerebral artery (ACA) associated with a ruptured saccular aneurysm at its distal end. Gross, microscopic, and radiological documentation of this anomaly is presented. It is suggested that in cases where the question of an unpaired ACA arises, a projection paralleling the radiological baseline be employed during angiographic studies thus affording better visualization of these vessels. Clinically, in patients with pericallosal aneurysms, their frequent association with azygous ACA’s should be borne in mind, as injury to this common arterial trunk will affect both hemispheres and the corpus callosum with tragic results.

KEY WORDS □9 intracranial aneurysm □9 cerebral malformation □9 circle of Willis □9 azygous anterior cerebral artery □9 anterior cerebral artery

The occurrence of an azygous anterior cerebral artery is a relatively uncommon developmental anomaly of the circle of Willis in man. In this vascular anomaly, the distal (A2) segment of both anterior cerebral arteries (ACA’s) are represented by a single common vessel from which arise all major vessels supplying most of the medial aspect of the cerebral hemispheres and the corpus callosum. The incidence of azygous ACA’s is estimated to be less than 1%, and its association with saccular pericallosal aneurysms is high. The present report concerns a unique case of a fusiform aneurysm of an azygous ACA with an associated ruptured saccular aneurysm. In addition, the literature pertinent to aneurysms in association with this congenital defect is reviewed.

Case Report

This 48-year-old hypertensive, nondiabetic woman presented to the Emergency Room with acute onset of severe bifrontal headaches, dizziness, neck pain, and lethargy of 10 hours’ duration. Her husband observed her speech to be incoherent. These events were followed by a transient loss of consciousness for 1 minute, which was not accompanied by seizure activity or loss of continence.
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Examination. Physical examination on admission revealed an obese confused patient with normal general physical findings. Blood pressure was 160/90 mm Hg, and the patient was afebrile. There was resistance to passive neck flexion, and no evidence of head trauma. The patient was lethargic, easily arousable, and her memory function was intact. Examination of the extraocular nerves demonstrated pupils of equal size which reacted consensually to light stimuli, a right sixth nerve palsy, and right skew deviation. In addition, a left seventh nerve palsy was present. The remaining cranial nerve functions were normal. Although the patient was able to move all extremities, a right upper extremity drift and a right Babinski reflex were demonstrated. There was no impairment of brainstem reflexes. Left carotid angiography demonstrated an aneurysm arising from the ACA in the rostral portion of the corpus callosum.

Course. Conservative treatment with Amicar (aminocaproic acid), antihypertensive medication, steroids (Decadron), and antiepileptic medication was begun. However, the patient became febrile on her fifth hospital day, and a lumber puncture yielded bloody cerebrospinal fluid (CSF) under a pressure of 300 mm H2O, with a protein concentration of 106 mg%, 27,000 red blood cells (RBC’s) and 296 white blood cells (WBC’s)/ml. Cultures of the fluid were negative for the growth of organisms.

The patient's level of consciousness improved over her first hospital week and by the seventh day, she was alert and responsive but apathetic. Two days later, there was a marked deterioration in the level of consciousness, and the patient responded only to painful stimuli. A subsequent lumbar puncture performed 48 hours later revealed grossly bloody CSF under a pressure of 130 mm H2O, 18,500 RBC’s/ml, 500 WBC’s/ml, protein 265 mg%, glucose 95 mg%, and chlorides 105 mg%. Over the ensuing 5 days, she became decerebrate and required endotracheal intubation. Right carotid angiography performed just before her death disclosed a left pericallosal aneurysm with a segmental narrowing of the right middle cerebral artery (Fig. 1). Two weeks after admission, she suffered a cardiopulmonary arrest and could not be resuscitated.

Postmortem Examination. Significant findings were limited to moderate cardiomegaly (heart weight 450 gm), with no evidence of valvular anomalies, thrombosis of the left iliac vein with massive pulmonary thromboemboli, and recent small renal infarcts.
A thin film of fresh subarachnoid hemorrhage covered the base of the brain and reached into the interpeduncular fossa and extended over the convexities. The left inferior frontal gyrus and the adjoining left precentral strip were infarcted. The blood vessels showed moderate atherosclerotic changes. The circle of Willis was anomalous in its rostral portion (Fig. 2). The proximal \( A_1 \) segments of both ACA's were of equal size and joined to form a common distal \( A_2 \) trunk: the azygous artery. The major artery measured 0.5 cm in diameter and showed fusiform dilatation over its entire length of \( 2\frac{1}{2} \) cm. A ruptured lobulated saccular aneurysm, measuring \( 0.3 \times 0.4 \) cm, was situated along its distal segment 0.5 cm before its division. The wall of the left middle cerebral artery (MCA) had a dark bluish hue and its lumen was markedly narrowed over a distance of 3 cm. No other aneurysms were found.

Coronal sections of the cerebral hemispheres revealed recent infarction in the distribution of the azygous ACA involving both cingulate gyri and the corpus callosum. In addition, there was an infarct in the territorial supply of the superficial branches of the right MCA.

**Microscopic Findings.** The azygous ACA was cut in cross section at 3-mm intervals throughout its length and stained with hematoxylin and eosin, Weigert's elastic stain, and Gomori trichrome. The vessel showed marked intimal hypertrophy and contained several atheromatous plaques with cholesterol clefts. The elastica was fragmented and frayed and had disappeared completely in some areas. The media was thickened, hyalinized, and in places totally destroyed by encroaching atheromatous plaques. The saccular aneurysm revealed a thin-walled fundus with no elastica or media. The neck displayed fragments of frayed elastica and patches of hyalinized intima. Sections of the MCA revealed segmental narrowing due to atheromatous changes.

Sections of brain parenchyma obtained from the territorial supply of the azygous artery showed acute infarction with pronounced neutrophilic reaction surrounding the blood vessels, while the areas supplied by the right MCA, which were also infarcted, showed capillaries distended with numerous hyaline thrombi.

**Discussion**

The case presented here describes a unique occurrence of a diffusely dilated azygous artery associated with a saccular aneurysm at its distal segment. The rupture had occurred in the saccular aneurysm leading to extensive infarction of the medial surface of both hemispheres in addition to the subarachnoid hemorrhage. The marked degenerative changes in the walls of the dilated azygous artery, with fraying and absence of elastic in areas, were sufficient to classify it as aneurysmal. The ectasia was not merely a result of fusion of both \( A_2 \) segments since the diameter of the azygous artery exceeded the combined diameters of the \( A_1 \) segment and was greater than that of the basilar artery. Dome-shaped nodules, easily mistaken for saccular aneurysms, are known to occur along the course of a fusiform aneurysm.\(^{10}\) However, in our patient, the saccular lesion found along the aneurysmal azygous artery was definitely a berry aneurysm according to its gross and microscopic definition.

It is well known that considerable variation occurs in the configuration of the ACA cir-
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TABLE 1

Incidence of azygous anterior cerebral arteries (ACA's) in six reports

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Brains Examined</th>
<th>Cases with Aneurysms</th>
<th>Cases with Azygous ACA</th>
<th>Associated Aneurysms*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Laitinen &amp; Snellman, 1960</td>
<td>320 (angio)</td>
<td>14</td>
<td>3</td>
<td>3/3</td>
</tr>
<tr>
<td>Baptista, 1963 review</td>
<td>2153*</td>
<td>—</td>
<td>23</td>
<td>—</td>
</tr>
<tr>
<td>own series</td>
<td>381 (autopsy)</td>
<td>—</td>
<td>1</td>
<td>—</td>
</tr>
<tr>
<td>Pool &amp; Potts, 1965</td>
<td>471 (angio)</td>
<td>22</td>
<td>3</td>
<td>3/3</td>
</tr>
<tr>
<td>own series</td>
<td>471 (angio)</td>
<td>0</td>
<td>0</td>
<td>0/0</td>
</tr>
<tr>
<td>LeMay &amp; Gooding, 1966</td>
<td>107 (angio)</td>
<td>1</td>
<td>3 + 1†</td>
<td>1/3</td>
</tr>
<tr>
<td>Danziger, et al., 1972</td>
<td></td>
<td>1</td>
<td></td>
<td>1/1</td>
</tr>
<tr>
<td>Katz, et al., 1978</td>
<td></td>
<td></td>
<td></td>
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</tbody>
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*Aneurysms in anterior circulation associated with an azygous artery.
†Authors stated that the fourth case was most likely a bihemispheric artery.

Among these various anomalies of the distal anterior cerebral circulation, the unpaired ACA assumes considerable clinical importance, since this vessel supplies the medial surface of both hemispheres and a large segment of the corpus callosum. Baptista described three types of anomalies occurring in the distal ACA: 1) the azygous artery, a single artery giving off all the major branches to both hemispheres; 2) the bihemispheric lateral artery, which consists of two ACA's only one of which supplies the majority of the vessels to both hemispheres, the other being hypoplastic; and 3) triplicate artery, two ACA's plus a median artery of the corpus callosum. These anomalies can be easily identified when brains are examined grossly; however, angiographic distinction between the azygous and the bihemispheric ACA is difficult because the hypoplastic A1 segment in the bihemispheric ACA is often poorly visualized.

In our case, the fact that the aneurysm arose from an azygous ACA was not fully appreciated. Perhaps it would have been possible to determine that the ACA's in this area were fused if the anteroposterior projection had been taken with less cephalo-caudad angulation. The angulation used for the standard Towne projection in our case drastically foreshortened the segment of the azygous ACA immediately proximal to the aneurysm.

It was impossible on the Towne projection to determine whether or not the small branch to the left of the azygous ACA was an ACA or a branch of the azygous trunk (Fig. 1 left); the lateral films were of no help because of superimposition of vessels. By using a projection paralleling the radiological baseline perhaps one could have “opened up” the area, allowing this determination.

Although the unpaired ACA is a normal vascular component in lower animals, it is decidedly rare in humans. According to De Vriese, this configuration of vessels is more common in the fetus than the adult. In Lesem's series of human fetus and baby brains, evidence was found for an anatomic transition from the single ACA to the two ACA's of the normal adult circulation. Several reports indicate that this defect is more frequently encountered in association with various congenital anomalies, in particular those involving the central nervous and cardiovascular systems.

Pool and Potts have suggested that the incidence of aneurysms of the pericallosal arteries is higher in patients with an azygous ACA. The literature contains reports of 34 cases of an azygous ACA, 23 of which were presented in a series compiled from the early literature and reviewed by Baptista (Table 1), who reported one azygous ACA found in his own series of 381 autopsy cases.

report was there mention of the occurrence of aneurysms in the circle of Willis. Likewise, an extensive cooperative study on subarachnoid hemorrhage by Locksley,\(^7\) contained no report of an azygous ACA. However, in a series of 471 patients with known arterial aneurysms,\(^9\) cerebral angiography demonstrated 22 cases of distal ACA aneurysms, three of these cases were associated with an azygous artery and each had a pericallosal saccular aneurysm (Table 1). Laitinen and Snellman,\(^4\) in their study of 14 cases of aneurysms of the pericallosal artery, also encountered three azygous ACA's in their group. One case had an aneurysm on each of its pericallosal branches. Thus, in 36 patients with aneurysms of the anterior circulation, six (17\%) were associated with an azygous ACA. Since the incidence of this anomaly in the general population is less than 1\%, the high prevalence of the azygous vessel in association with the intracranial aneurysms supports the viewpoint of Pool and Potts.\(^8\)

The converse of this association is also important. In other words, what is the incidence of aneurysms in patients with azygous ACA's? LeMay and Gooding\(^5\) reviewed 107 consecutive cerebral angiograms in which both internal carotid arteries were visualized for the presence of an unpaired ACA, and found three. Of these, one had a saccular aneurysm at the proximal end close to its origin. While this study represents a limited number of cases, an associated aneurysm in one out of three is impressive. Therefore, in cases of pericallosal aneurysms, their association with an azygous ACA should always be considered, since injury to this common arterial trunk can lead to devastating complications.

**References**


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