Nonsaccular aneurysms of the azygos anterior cerebral artery

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Object. The azygos or undivided anterior cerebral artery (ACA) is a rare variant, and aneurysms associated with this variant are particularly rare. Most reported azygos ACA aneurysms are saccular, but the authors encountered four patients with this variant who had nonsaccular aneurysms. A review of the management of these lesions and this morphological distinction is presented.

Methods. A retrospective review of patients with aneurysms treated over a 6-year period identified five Type I (according to the Baptista classification) azygos ACA lesions, of which four were nonsaccular. Aneurysms associated with other ACA variants (Baptista Types II and III) were excluded.

Azygos ACA aneurysms accounted for 0.5% of all treated lesions and 1.7% of all ACA and anterior communicating artery aneurysms. One lesion in this series was located proximally at the azygos ACA origin, and three were located distally. All four aneurysms were large (> 10 mm in diameter), and two were thrombotic. All aneurysms were treated with microsurgical clip occlusion.

Conclusions. Azygos ACA aneurysms are rare, and may have unusual nonsaccular anatomy (for example, fusiform shape, broad base, complex branching, and/or thrombus in the lumen). The nonsaccular morphology of these aneurysms may render them unsuitable for endovascular coil placement, and may complicate their microsurgical management.

KEY WORDS • intracranial aneurysm • azygos anterior cerebral artery • nonsaccular aneurysm • fusiform aneurysm • microsurgical clip occlusion

The azygos or undivided ACA is a single, midline vessel arising from the confluence of the A_1 ACAs, representing a rare variant of the usually paired A_2 vessels.^{2,3,11,18,19,23,25,27,28,30,31}

It supplies the medial aspects of the frontal and parietal lobes. Based on angiographic and autopsy studies, this variant has an incidence ranging between 0.3 and 2%. The azygos ACA divides into bilateral PerAs and CMAs at variable distances from its origin, typically distal to the genu of the corpus callosum.^{17,23} The anatomy at this branch point is also variable, with bifurcations, trifurcations, and even quadrifurcations having been reported.^{3,22,26,29}

After reviewing 381 brain specimens, Baptista³ distinguished the azygos ACA from other variants by defining three types of distal ACA anomalies. He referred to the azygos ACA as the “unpaired ACA” (Type I anomaly), and observed it in only one brain in his collection of specimens. He defined a “bihemispheric ACA” as an A_2 segment of the ACA that sends branches across the midline to both hemispheres, usually in the presence of a contralateral A_2 segment that is either hypoplastic or that termi-

nates early in its course toward the genu of the corpus callosum (Type II anomaly). Type II anomalies were observed in 45 specimens (12%). The most common anomaly (50 specimens, 13%) was the “accessory ACA,” defined as a third artery originating from the ACoA, in addition to the paired A_2 ACAs, usually in the midline, and with branches to one or both hemispheres (Type III anomaly). This accessory ACA varied in caliber from a small remnant of the median artery of the corpus callosum to a hyperplastic vessel that can resemble an azygos one when the two A_2 ACAs are small in caliber and terminate early.

Aneurysms of the azygos ACA have been reported previously, but with minimal attention given to their morphological features. In most of these reports the authors describe saccular aneurysms. We encountered four cases of azygos ACA aneurysms with nonsaccular anatomy, and present our management of the disease in these patients along with a review of this morphological distinction.

CLINICAL MATERIAL AND METHODS

A retrospective review of the aneurysm database maintained at the University of California, San Francisco was conducted. Between August 1997 and July 2003, a total of 876 aneurysms were treated microsurgically by the senior

Abbreviations used in this paper: ACA = anterior cerebral artery; ACoA = anterior communicating artery; AP = anteroposterior; CMA = callosomarginal artery; MR = magnetic resonance; OphA = ophthalmic artery; PerA = pericallosal artery.

author (M.T.L.), of which 231 lesions involved the ACA or ACoA. Of these, five aneurysms were associated with an azygos ACA (Type I). Four of the azygos ACA aneurysms displayed nonsaccular morphology. Nonazygos ACA aneurysms (Types II and III) were excluded from this study. Medical records, angiograms, other neuroimaging studies, and intraoperative photographs were reviewed.

RESULTS

Azygos ACA aneurysms accounted for 0.5% of all treated lesions in this group, and constituted 1.7% of all ACA and ACoA aneurysms. One of the azygos ACA aneurysms was located proximally at the origin of the artery, and three of them were located distally at its branch points at the level of the genu of the corpus callosum. One distal aneurysm was located at a bifurcation, one at a trifurcation, and one at a quadrifurcation. All four aneurysms were large (> 10 mm in diameter) and had nonsaccular morphology. Two lesions were thrombotic.

The proximal azygos ACA aneurysm was exposed via an orbitozygomatic approach, and distal aneurysms were exposed through a bifrontal craniotomy via an interhemispheric approach. For the interhemispheric approach, patients were positioned with the head turned laterally 90° and angled upward 45° , placing the right hemisphere in the dependent position to allow gravity to retract the frontal lobe and open the interhemispheric fissure.²¹ All aneurysms were eliminated with surgical clip occlusion.

ILLUSTRATIVE CASES

Case 1

History. This 46-year-old woman presented with sudden, severe headache. Admission computerized tomography scans of the brain revealed no subarachnoid hemorrhage, but an interhemispheric abnormality was noted and subsequent brain MR imaging (Fig. 1A) and MR angiography (Fig. 1B) revealed an aneurysm. Cerebral angiography demonstrated an azygos ACA in which a fusiform aneurysm measuring approximately 1 cm in diameter was located at the distal quadrifurcation at the level of the genu of the corpus callosum (Fig. 1C). The patient was referred for endovascular therapy, but the anatomy of the lesion precluded this modality.

Operation and Postoperative Course. The patient underwent a bifrontal craniotomy and interhemispheric approach to the aneurysm, which had an afferent azygos ACA, two efferent PerAs, two efferent CMAs, and no discrete neck. A temporary clip was applied to the azygos ACA for 8 minutes to soften the aneurysm, and a series of clips was applied to reconstruct a neck. After removing the temporary clip, Doppler ultrasonography flow measurements confirmed patency of the branches. Postoperative angiography demonstrated complete obliteration of the aneurysm and patency of the branch arteries (Fig. 1D). The patient remained neurologically intact.

Case 2

History. This 67-year-old woman presented with chron-

Fig. 1. Case 1. Neuroimages obtained in a 46-year-old woman presenting with sudden headache. A: Axial T_2 -weighted MR image of the brain revealing a hypointense lesion in the interhemispheric fissure anterior to the genu of the corpus callosum. B: Admission MR angiography study demonstrating an azygos ACA with a fusiform aneurysm at the distal quadrifurcation. C: Oblique view, left carotid angiogram demonstrating the approximately 1-cm aneurysm of the azygos ACA. D: Postoperative AP angiogram of the left carotid artery demonstrating no residual filling of the azygos ACA aneurysm.

ic headaches. Her workup included an MR image of the brain that demonstrated two incidental aneurysms. Catheter angiography revealed a 7-mm-diameter left OphA aneurysm, a Type I azygos ACA, and a 1.5-cm-diameter azygos ACA aneurysm located distally at a trifurcation (Fig. 2A and B). The OphA aneurysm was treated successfully with endovascular coil occlusion, but the complex anatomy of the branches at the base of the azygos ACA aneurysm made this lesion unsuitable for coil placement.

Operation and Postoperative Course. The patient underwent a bifrontal craniotomy and interhemispheric approach to the aneurysm, which had an additional thrombosed lobe on its left side that did not opacify on the angiogram. There was one afferent azygos ACA, an efferent trunk on the left that bifurcated 1 cm distal to the aneurysm into the PerA and CMA, and a PerA and CMA that originated from the aneurysm on the right. A temporary clip was placed on the azygos ACA to soften the aneurysm, the final dissection of the branches was completed, and the lesion was reconstructed with a series of 45° -angle fenestrated clips in tandem. The aneurysm walls were atherosclerotic and thickened, requiring that a tentative clip (a permanent clip that is applied imperfectly across the neck of the aneurysm and is later removed or readjusted) be placed initially to close the aneurysm and serve as a template above which additional clips were placed. The tentative clip was then removed after the oth-

Fig. 2. Case 2. Neuroimages obtained in a 67-year-old woman with chronic headaches. A and B: Left carotid artery angiograms demonstrating a 1.5-cm azygos ACA aneurysm and an approximately 7-mm OphA lesion in oblique (A) and AP (B) views. C and D: Postoperative lateral (C) and AP (D) projection angiograms demonstrating no residual aneurysms.

ers had been used successfully to close the aneurysm. A total of 6 minutes of temporary occlusion was required. Blood flow in the branch arteries was confirmed both grossly and with Doppler ultrasonography flow measurements. Postoperative angiographic studies demonstrated complete occlusion of the aneurysm and patency of the branch arteries (Fig. 2C and D). The patient remained neurologically intact.

Case 3

History. This 57-year-old man in whom a right-sided vestibular schwannoma had been resected 5 years previously underwent a follow-up MR image of the brain that demonstrated a large thrombosed ACoA aneurysm that had not been present on previous imaging studies (Fig. 3A and B). The aneurysm was 2 cm in diameter, with a luminal diameter measuring only 1 cm. Catheter angiography demonstrated an azygos ACA with an aneurysm arising at its origin, where the two A₁ segments converged (Fig. 3C and D). The left A₁ ACA was dominant.

Operation and Postoperative Course. The patient underwent a left orbitozygomatic-pterional craniotomy. The aneurysm did not have a discrete neck, and the base was tentatively clipped. The dome was then opened, thrombus was removed down to the base of the aneurysm, additional clips were placed above the tentative one to reconstruct a neck, and the tentative clip was then removed. An intraoperative angiogram demonstrated complete elimination of the aneurysm and preservation of flow in the azygos ACA. The patient remained neurologically intact.

Fig. 3. Case 3. Neuroimages obtained in a 57-year-old man with a distant history of a right-sided vestibular schwannoma and no new neurological deficits. A: Axial contrast-enhanced T₁-weighted MR image of the brain revealing incomplete contrast enhancement of an interhemispheric, suprasellar lesion consistent with a partially thrombosed aneurysm. B: Sagittal T₁-weighted MR image revealing a hypointense lesion situated between the optic chiasm and the genu of the corpus callosum. Diagnostic AP (C) and oblique view (D) angiograms reveal a 1-cm aneurysm located at the origin of an azygos ACA.

Case 4

History. This 39-year-old man presented with an incidental arteriovenous malformation located in the left occipital lobe that was diagnosed on a brain MR image. Catheter angiography demonstrated a Spetzler–Martin Grade V lesion, and conservative management was recommended. The angiogram also demonstrated a fusiform azygos ACA aneurysm measuring 1.3 cm in diameter that was located distally at a bifurcation into two efferent trunks that each branched into the PerA and CMA. The distal left PerA contributed to the arteriovenous malformation.

Operation and Postoperative Course. The patient underwent a bifrontal craniotomy and interhemispheric approach to the aneurysm. The azygos ACA was temporarily occluded to soften the aneurysm, and two clips were used to close the aneurysm base and to reconstruct a bifurcation between the two efferent trunks. Blood flow in the branch arteries was confirmed both grossly and with Doppler flow measurements. Postoperative angiography demonstrated complete occlusion of the aneurysm and patency of the branch arteries. The patient remained neurologically intact.

DISCUSSION

These four cases demonstrate that azygos ACA aneurysms can have nonsaccular morphology that can complicate their microsurgical treatment. The nonsaccular mor-

phology of these aneurysms has not been emphasized in previously published reports. In a review of the literature identifying 50 cases of surgically treated azygos ACA aneurysms, 35 were categorized as saccular but nine had an unspecified morphology that might have been nonsaccular (Table 1). Furthermore, six of these aneurysms were giant and one was large, probably with the nonsaccular anatomy encountered in our experience. The most common treatment reported for these aneurysms was direct clip placement (26 of the 50 patients in the published literature), but treatment was unspecified in 16 patients and alternative therapies were used in eight (proximal ligation in four, medical management in three, and wrapping in one patient), possibly implying nonsaccular morphology. Our experience indicates that these aneurysms can be fusiform or broad based, with multiple efferent arteries that require complex clip applications to reconstruct a neck.

The high incidence of nonsaccular morphology that we observed may reflect the bias involved in referral to a tertiary neurosurgical center. Nevertheless, in the literature others have proposed several reasons why these azygos ACA aneurysms might have a propensity for nonsaccular morphology. An azygos ACA has twice the blood flow and hemodynamic stress of the usual paired A₂ ACA, increasing its susceptibility to aneurysm formation.²⁵ Complex bi-, tri-, and quadrifurcations arising from the distal azygos ACA may also contribute to unusual aneurysm morphology.^{3,22,26,29} All four lesions in this series were

large, and progressive enlargement of an aneurysm can convert simple saccular to complex nonsaccular anatomy.⁷

Considering this propensity for and underreporting of nonsaccular morphology, neurosurgeons treating azygos ACA aneurysms should be prepared for complex clip applications. Multiple clips are often required to close the aneurysm and contour the reconstruction between branches on opposite sides of the base. Temporary clip placement, performed after administration of barbiturate drugs and induction of mild hypothermia and hypertension for cerebroprotection, is an essential adjunct that softens the aneurysm for the final dissection and clip application. Placement of temporary clips requires proximal control of the azygos ACA, which necessitates a low trajectory into the interhemispheric fissure below the genu of the corpus callosum. Establishing this low trajectory is particularly important with ruptured aneurysms, because the interhemispheric approach can bring the neurosurgeon to the dome of the aneurysm early in the dissection.

Tentative clip placement is another useful adjunct, placing an imperfect clip to close the aneurysm while other permanent clips are applied before removing the tentative one. Case 2 demonstrates the use of fenestrated clips with overlapping blades, which were applied from the deep to the superficial portion of the neck, as an effective way of occluding a large aneurysm with atherosclerotic walls. Improper clip placement in azygos ACA aneurysms may place both hemispheres at risk for ischemia, making intraoperative Doppler ultrasonography measurements and intraoperative angiography useful checks.

In this era of evolving endovascular technology and techniques, it is important to emphasize that these four azygos ACA aneurysms were all deemed unsuitable for endovascular therapy because of their nonsaccular morphology and the difficulty visualizing in two dimensions the complex branching of the efferent arteries. Endovascular therapy remains an appealing option for saccular azygos ACA lesions. It is particularly appealing for patients with ruptured aneurysms and swollen brains, in whom an interhemispheric dissection might be difficult. In elderly patients, endovascular management would spare the patient a bifrontal craniotomy that might pose an increased risk of superior sagittal sinus injury. Nevertheless, endovascular coil occlusion has limitations that should be recognized and respected.

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TABLE 1

*Literature review of azygos aneurysms treated in 50 patients**

Authors & Year	No. of Lesions	Morphology	Treatment
Adachi, 1928	1	NA	NA
Huber, 1960	1	NA	NA
Laitinen & Snellman, 1960	4	saccular	clip occl (2), ligation (2)
Pool & Potts, 1965	3	NA	NA
LeMay & Gooding, 1966	1	saccular	NA
Handa, et al., 1971	1	saccular	med mgmt
Katz, et al., 1978	1	saccular	med mgmt
Kondo, et al., 1979	1	saccular	clip occl
Huber, et al., 1980	7	saccular (all 7)	NA
Fujimoto, et al., 1981	1	saccular	wrapping
Niizuma, et al., 1981	2	saccular	ligation
Benedetti & Curri, 1983	1	saccular	clip occl
Yaşargil, 1984	4	NA	clip occl
Hayashi, et al., 1985	2	giant	clip occl (1), med mgmt (1)
Kobayashi, et al., 1986	1	saccular	clip occl
Yamagami, et al., 1986	1	giant	clip occl
Lightfoote, et al., 1989	1	saccular	NA
Schick & Rumbaugh, 1989	1	saccular	clip occl
Hashizume, et al., 1992	1	giant	NA
Hernesniemi, et al., 1992	2	saccular	clip occl
Baykal, et al., 1996	2	saccular	clip occl
Dietrich, et al., 2000	1	saccular	clip occl
Kanemoto, et al., 2000	1	giant	NA
Miyazawa, et al., 2000	9	1 giant, 1 saccular (large), 7 saccular	clip occl

* Med mgmt = medical management; NA = not available; occl = occlusion.

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