

Case Report

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Saccular Aneurysm of the Azygos Anterior Cerebral Artery : Three Case Reports

The azygos anterior cerebral artery, a rare anomaly in the circle of Willis in which only a single vessel supplies the medial aspects of both anterior cerebral hemispheres, is closely associated with saccular aneurysms. We present three cases of azygos anterior cerebral artery aneurysms among the 781 cerebral aneurysms surgically treated at our institution in an 11-year period. These three cases all involved elderly women who presented with subarachnoid hemorrhage. Conventional cerebral angiography and CT angiography revealed small saccular aneurysms at the distal ends of the azygos anterior cerebral arteries. These aneurysms were clipped successfully using a bifrontal interhemispheric approach. Hence, the pathogenesis of these particular aneurysms relating to hemodynamic change, associated anomalies, and surgical pitfalls is discussed with review of literature.

KEY WORDS : Azygos anterior cerebral artery · Saccular aneurysm · Anomaly.

INTRODUCTION

The azygos anterior cerebral artery (ACA) is a very rare anatomic anomaly with an incidence of about 1.1%^{8,16}. This variation is closely associated with saccular aneurysms and has significant impact on the arterial hemodynamics of the frontal lobe. Azygos ACAs also often accompany other malformations, such as agenesis of the corpus callosum, hydranencephaly, saccular aneurysm and arteriovenous malformation (AVM)⁹.

Several authors have reported that the rate of occurrence of an azygos ACA aneurysm between 13% and 71%^{11,13}. An azygos ACA has twice the blood flow and hemodynamic pressure of a normally paired A₂ region of the ACA, which increases its susceptibility to aneurysm formation. Development of an azygos ACA saccular aneurysm is associated with blood flow restriction and pressure on the arterial wall in the distal portion of the parent artery.

We reviewed 781 consecutive patients with aneurysmal subarachnoid hemorrhage (SAH) who were treated at our hospital between 1995 and December 2006. We found three cases (0.38%) of azygos anterior cerebral artery aneurysms. These cases all involved elderly women (mean 70 years old; range 60-78 years) who presented with SAH. Angiographic findings revealed saccular aneurysms located at the distal ends of the azygos anterior cerebral arteries. All patients underwent a bifrontal craniotomy and interhemispheric approach. We will report on these cases and compare them to the relevant literature.

CASE REPORT

Case 1

A 60-year-old woman presented with headache, dysarthria and drowsiness. The patient's history revealed that she had been diagnosed with hypertension and had been taking an antihypertensive drug for ten years. Upon neurological examination, she showed a slightly drowsy mental status and did not display any other neurological focal signs besides dysarthria. The brain computerized tomography (CT) showed a SAH in the basal cistern and sylvian fissure, a hematoma in the interhemispheric fissure and intraventricular hemorrhage; there was no evidence of hydrocephalus. The CT and conventional angiography showed the azygos artery and the saccular aneurysm at the distal end of the A₂ segment (Fig. 1). After preparation of free bone flap from the right frontal area, the unpaired A₂ and saccular aneurysm in the transition area between A₂ and A₃ were detected with an interhemispheric approach.

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After dissection in the proximal portion of the aneurysm, a temporary clipping was performed, which was followed by a permanent clipping. On the second postoperative day, the patient exhibited severe drowsiness and motor weakness in the right lower extremities. The CT did not show brain swelling, cerebral infarction, or hydrocephalus. The patient's status continued to improve postoperatively, with a full recovery of consciousness and mild motor weakness three months after the operation.

Case 2

A 74-year-old woman presented with sudden onset of headache and nausea. She had been taking an antihypertensive drug for three years. A neurological examination showed mental alertness and no focal neurological deficiencies. The brain CT revealed a SAH in the basal cistern and a hematoma in the interhemispheric fissure. However, there was no evidence of intraventricular hemorrhage, hydrocephalus or cerebral infarction. The CT with angiography did not show the left A1, but detected the unpaired status

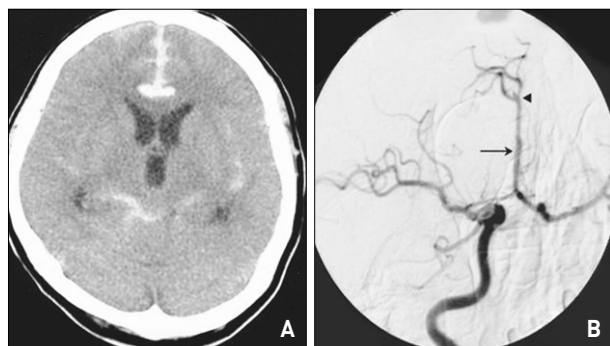


Fig. 1. A : The initial Brain computed tomography (CT) shows subarachnoid hemorrhage within the basal cistern and sylvian fissure with hematoma within interhemispheric fissure. B : The unilateral carotid angiography with contralateral carotid compression shows the unpaired distal anterior cerebral artery (arrow head) and saccular aneurysmal dilatation on the A2 & A3 junction (arrow). Other vascular anomalies are not seen on the angiogram.

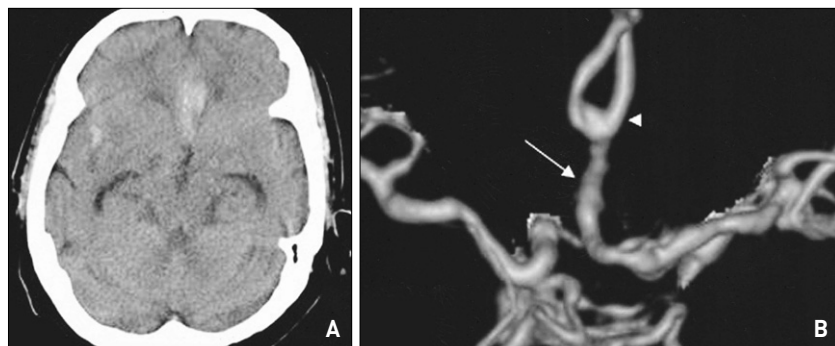


Fig. 2. A : The initial brain computed tomography (CT) shows subarachnoid hemorrhage within the basal cistern and interhemispheric fissure. There is no evidence of hydrocephalus. B : The CT angiogram shows an unpaired distal segment of the anterior cerebral artery (arrow head) and aneurysmal dilatation (arrow) on its origin site.

of the A₂ region of the ACA, and revealed the saccular aneurysm in the transition area between A₂ and A₃. It also did not show any evidence for vasospasm (Fig. 2). After dissection in the proximal portion of the aneurysm, a temporary clipping was performed, which was followed by a permanent clipping. The patient recovered completely without any neurological focal damage and was transferred to the inpatient ward. However, on the tenth postoperative day, the patient developed abdominal pain, diarrhea and a drowsy mental status. She presented with respiratory difficulties on the 16th postoperative day and continued having diarrhea and severe drowsiness on the 22nd postoperative day. The patient was diagnosed with pseudomembranous colitis, and her general condition deteriorated. She developed aspiration pneumonia and septic shock on the 75th postoperative day, and died on the 77th postoperative day.

Case 3

A 78-year-old woman presented with sudden onset of severe headache, nausea, vomiting and disturbance in consciousness. She had been taking an antihypertensive drug for 20 years. Examination revealed a drowsy and confused mental status but no focal neurological deficits (Hunt and Hess Grade III). A non-contrasted CT scan of the brain revealed thick and diffuse SAH involving the basal cistern, both sylvian and interhemispheric fissures and intraventricular hemorrhage without hydrocephalus (Fisher Grade 4). The initial cerebral angiogram and contrast enhanced computed angiography revealed a small outpouching in the ACA bifurcation as well as an azygos ACA (Fig. 3). An operative intervention was planned immediately, but the operation was performed after external ventricular drainage via Kocher's point due to hydrocephalus and mental deterioration. The proximal portion of the aneurysm was dissected using an inter-

hemispheric approach, and a temporary clipping was performed, followed by a permanent clipping. The interhemispheric approach allowed us to recognize the unpaired A₂ and saccular aneurysm in the transition area between A₂ and A₃. The patient exhibited a stuporous mental status on the second postoperative day. The CT did not show cerebral infarction but indicated hydrocephalus. We performed a ventriculoperitoneal shunt on her 28th postoperative day, and her status improved postoperatively.

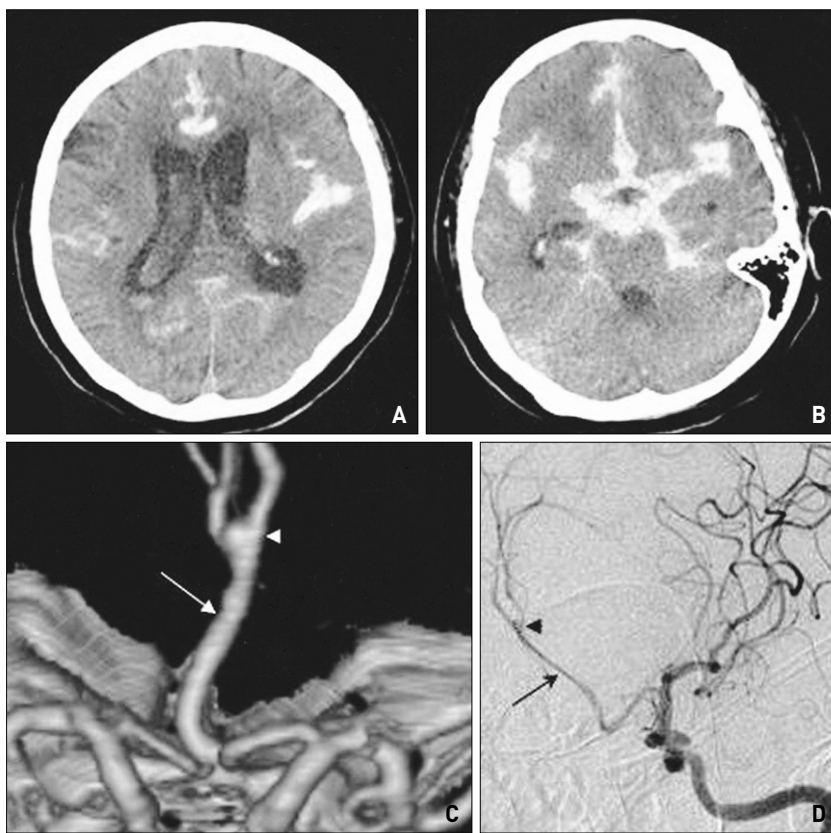


Fig. 3. A, B : The initial brain computed tomography (CT) shows subarachnoid hemorrhage within the basal cistern and interhemispheric fissure with intraventricular hemorrhage. C, D : The CT angiogram and carotid angiogram shows an unpaired distal segment of the anterior cerebral artery (arrow head) and aneurysmal dilation (arrow) on its origin site.

DISCUSSION

In 1885, Wilders described *arteria termatica*, the formation of one artery from fusion of the A_2 in the anterior cerebral artery. He also called it the azygos artery of the pericallosal artery^{2,16}. The azygos anterior cerebral artery (ACA) is a very rare anatomic anomaly with an extremely low incidence, anywhere from about 0% to 5%^{1,6,8,11}. However, Osborn has reported that the azygos ACA has an incidence of about 10%¹⁵.

The unusual fusion of the paired A_2 in the ACA originates either from the medial branch of the olfactory artery at the initial 16 mm stage of embryogenesis or the continuation of the median artery in the corpus callosum at the 20-24 mm stage. It can also be generated by a lack of development or regression of the ACA^{7,10,17}. Baptista divided abnormalities of the distal portion of the ACA into three groups : 1) single unpaired ACAs, in which a single ACA feeds into the medial surface of both cerebral hemispheres; 2) bihemispheric ACAs, in which there are two ACAs, but one is clearly dominant with branches extending into the contralateral hemispheres; 3) accessory

ACAs, in which a third, or median artery, is distributed to either one or both hemispheres¹¹.

Among the three types, the azygos, or single unpaired ACA, is the most important. The azygos artery is frequently associated with other malformations of the central nervous system, such as porencephalic cysts, agenesis of the corpus callosum, hydranencephaly, saccular aneurysms, and AVM. Among them, the incidence of aneurysms is about 13-71%. There are two generally accepted mechanisms for the formation of such aneurysms : the aneurysms develop alongside a congenital anomalous artery, or the aneurysms result from hemodynamic stress caused by the azygos artery^{6,8,9,11}.

In this study, other than the saccular aneurysm in distal portion of the fused A_2 , no other central nervous system malformations were detected. The clinical significance of the azygos artery is great; it is intimately associated with the formation of aneurysms and possible neurological deficiencies. These deficiencies can be caused by ischemia

in both hemispheres brought on by arterial damage or occlusion during an operation for an aneurysm^{4,5}. Although there are no guidelines on the optimal tolerance time for the temporary clipping of the azygos artery during an operation, Normes et al. reported that it would be helpful to use the temporary clipping method when performing an MCA aneurysm operation^{12,14}.

Even though a diagnosis of azygos ACA is possible with cerebral angiography, we used CT with angiography in one case and cerebral angiography in other two cases. In the latter two cases, we were able to diagnose the fused A_2 with a contrast media infusion to the unilateral carotid artery by compressing the contralateral carotid artery while performing the 4-vessel cerebral angiography. All cases were confirmed upon operation. Without compressing the contralateral carotid artery, it can be difficult to ascertain the exact number of arteries or arterioles with a single carotid artery angiography. Therefore, blood flow of the opposite A_1 should be restricted through compression of the contralateral carotid artery for an exact diagnosis. An azygos ACA diagnosis can also be confirmed directly during operation^{3,10,13}.

CONCLUSION

Although the incidence of azygos ACA aneurysm is rare, cerebral angiography and CT angiography should be carefully examined in order to avoid missing the diagnosis, to evaluate the form and projection of the aneurysm, and to detect associated vascular anomalies. During the operation, temporary clipping in the proximal A₂ is an essential adjunct intervention and should be performed vigilantly in order to avoid ischemic damage in both hemispheres by vascular deficit.

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