



Mechanical Thrombectomy for Azygos Anterior Cerebral Artery Occlusion: A Case Report and Literature Review

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Objective: Azygos anterior cerebral artery (ACA) is a well-known anomaly of the second segment of the ACA. Although cases of intracerebral aneurysms related to this anomaly have been reported, acute ischemic stroke (AIS) related to the azygos ACA is extremely rare.

Case Presentation: An 84-year-old man developed disturbance of consciousness (Glasgow Coma Scale [GCS] E3V1M5), quadriplegia and aphasia, with a National Institutes of Health Stroke Scale (NIHSS) score of 32. Magnetic resonance imaging (MRI) showed no early ischemic changes, although a head magnetic resonance angiogram (MRA) demonstrated a single A2 trunk without any A3 branches that were suspected bilateral ACA occlusions. Mechanical thrombectomy for the occluded A2 trunk with contact aspiration using a Penumbra 4MAX aspiration catheter was performed, and the clot was retrieved and complete recanalization was achieved after two attempts (Thrombolysis in Cerebral Infarction scale 3) without any complications (onset to recanalization time: 187 min). The final angiogram demonstrated the recanalization of the single A2 and bilateral A3 branches, so we diagnosed as azygos ACA occlusion. MRI performed the next day revealed several small infarctions in bilateral frontal lobes, but ischemic symptoms gradually improved. NIHSS score decreased to two in 2 weeks and modified Rankin Scale (mRS) score at 90 days was one.

Conclusion: In this case, occlusion of the azygos ACA led to a large ischemic penumbra that spread widely and bilaterally in the ACA area, resulting in sudden onset of severe ischemic symptoms, including quadriplegia and aphasia. However, due to complete and rapid recanalization with contact aspiration, a large part of the ACA territory bilaterally was salvaged and the patient recovered extremely well.

Keywords ► mechanical thrombectomy, endovascular treatment, azygos anterior cerebral artery, acute ischemic stroke, contact aspiration

Introduction

The azygos anterior cerebral artery (ACA), which consists of a single A2 trunk supplying bilateral ACA territories,¹⁾ is

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a rare, but known, anatomical anomaly with an incidence of 0.3%–2%.²⁾ Although several cases of intracranial aneurysm associated with azygos ACA have been reported,³⁾ acute ischemic stroke (AIS) due to azygos ACA occlusion is extremely rare, and only five such cases have been previously reported,⁴⁻⁸⁾ only one of which was treated by mechanical thrombectomy (MT).⁸⁾ This report presents a case of AIS due to azygos ACA occlusion, who presented with quadriplegia and total aphasia that was treated by contact aspiration thrombectomy and reviews the previous reports.

Case Presentation

An 84-year-old man with known hypertension and independence in activities of daily living suddenly developed disturbance of consciousness, quadriplegia and a speech

disturbance at night. He was transferred to the emergency department of our hospital within 39 min of symptom onset. His National Institutes of Health Stroke Scale (NIHSS) score was 32, and Glasgow Coma Scale (GCS) score was 10 (E3V1M5). He showed right complete paralysis, left paresis, total aphasia, and left conjugate deviation of the eyes. Head plane computed tomography (CT) revealed no early ischemic changes or intracranial hemorrhage. Diffusion-weighted head magnetic resonance imaging (DW-MRI) also showed no early ischemic change in bilateral cerebral hemispheres or the brain stem (**Fig. 1A** and **1B**). However, a head magnetic resonance angiogram (MRA) demonstrated a single A2 trunk without any A3 branches that was suspected bilateral ACA occlusion (right A2 proximal and right A2 distal occlusions) (**Fig. 1C**).

Since DW-MRI showed no ischemic core and a large viable penumbra occupied ACA territories bilaterally, intravenous tissue-plasminogen activator (t-PA) injection

(IV t-PA) followed by MT was performed; the time from symptom onset to groin puncture was 98 min. First, using a right femoral artery approach, we tried to navigate a 9-Fr balloon guiding catheter, OPTIMO (Tokai Medical Products, Aichi, Japan) into the left internal carotid artery (ICA) because of the dominant left A1. However, due to the presence of a bovine aortic arch, it was impossible to navigate the guiding system via the femoral artery (**Fig. 1D**). Hence, we switched to a right brachial artery approach. We directly introduced a 9-Fr OPTIMO into the right brachial artery with a sheath-less method⁹⁾ using a 6-Fr long dilator, and easily navigated the OPTIMO into the left ICA (**Fig. 1E**). Since left ICA angiogram showed occlusion of the single A2 segment (**Fig. 1F** and **1G**), a Penumbra 4MAX (Penumbra Inc., Alameda, CA, USA) aspiration catheter was navigated to the occlusion site with a coaxial system using a Velocity microcatheter (Penumbra Inc.) and a 0.014 inch micro guidewire (ASAHI CHIKAI, ASAHI INTECC Co., Ltd.,

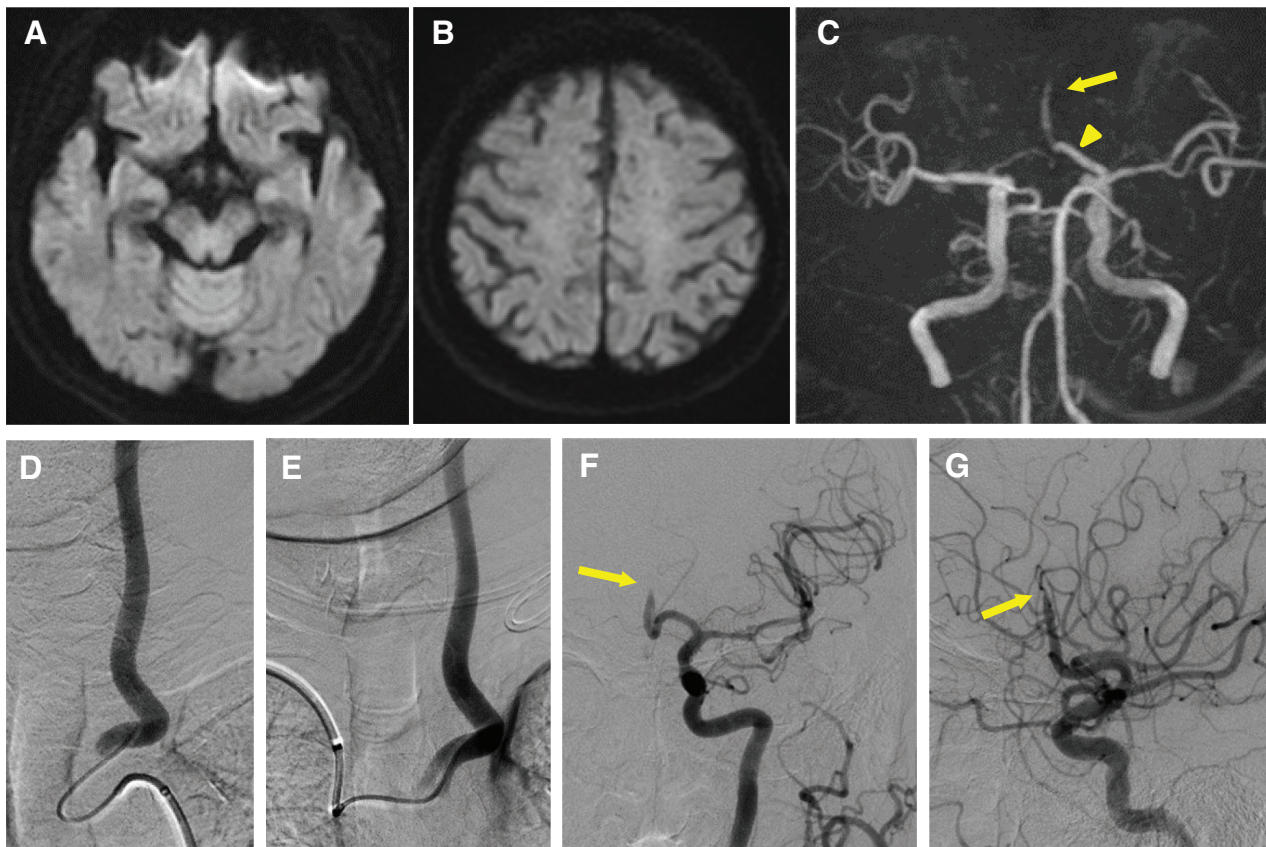


Fig. 1 (A, B) DW-MRI showed no early ischemic changes in the cerebral hemispheres bilaterally or in the brain stem. (C) MRA showed occlusion of the azygos ACA A2 segment (arrow), with a dominant left A1 segment (arrowhead). (D) DSA of the left CCA, AP view, showed a bovine type aortic arch and it was impossible to navigate the balloon guiding catheter into the left CCA from the femoral artery. (E) DSA of the left CCA, AP view: the catheter was

easily navigated via the right brachial artery. (F, G) DSA of the left ICA (F: AP view, G: Lateral view) showed complete occlusion of the azygos ACA (arrow). ACA: anterior cerebral artery; AP: antero-posterior; CCA: common carotid artery; DSA: digital subtraction angiogram; DW-MRI: diffusion-weighted magnetic resonance imaging; ICA: internal carotid artery; MRA: magnetic resonance angiogram

Aichi, Japan) (**Fig. 2A** and **2B**). After the 4MAX catheter contacted the clot, aspiration was commenced with a MAX pump (Penumbra Inc.) for 120 s, and the 4MAX was withdrawn into the OPTIMO. A second attempt resulted in complete recanalization of the single A2 trunk and bilateral A3 branches originated from the single A2 trunk (Thrombolysis in Cerebral Infarction scale 3) (**Fig. 2C** and **2D**), so we diagnosed as Azygos ACA occlusion just after final angiogram. The three pieces of thrombus was retrieved (**Fig. 2E**) and the time from symptom onset to recanalization was 187 min. Head CT immediately after the procedure showed no sign of intracranial hemorrhage. DW-MRI performed the next day showed several small cerebral infarctions in bilateral frontal lobes (**Fig. 2F**), although MRA showed good recanalization of the azygos ACA (**Fig. 2G**). The patient's aphasia and quadriparesis improved gradually after the procedure, and GCS score also improved to 15 (E4V5M6) and NIHSS score decreased

to two in 2 weeks. On the 19th day after the procedure, he was transferred to a rehabilitation hospital and recovered sufficiently to be able to walk and live his daily life independently. His modified Rankin Scale (mRS) score was one at 90 days.

Discussion

Azygos ACA is a rare anatomical anomaly that consists of a single midline A2 trunk¹⁾ and provides blood supply to bilateral ACA territories.¹⁰⁾ Gunnal et al.¹¹⁾ divided this anomaly into five subtypes. Classic or true azygos ACA (Type I) is described as a single artery that does not divide into two distal ACA branches, and instead gives bilateral cortical branches. Azygos ACA Type II usually divides into two pericallosal arteries close to the genu of the corpus callosum, as was seen in our case. The other three types give rise to other small A2 branches in addition to the main A2

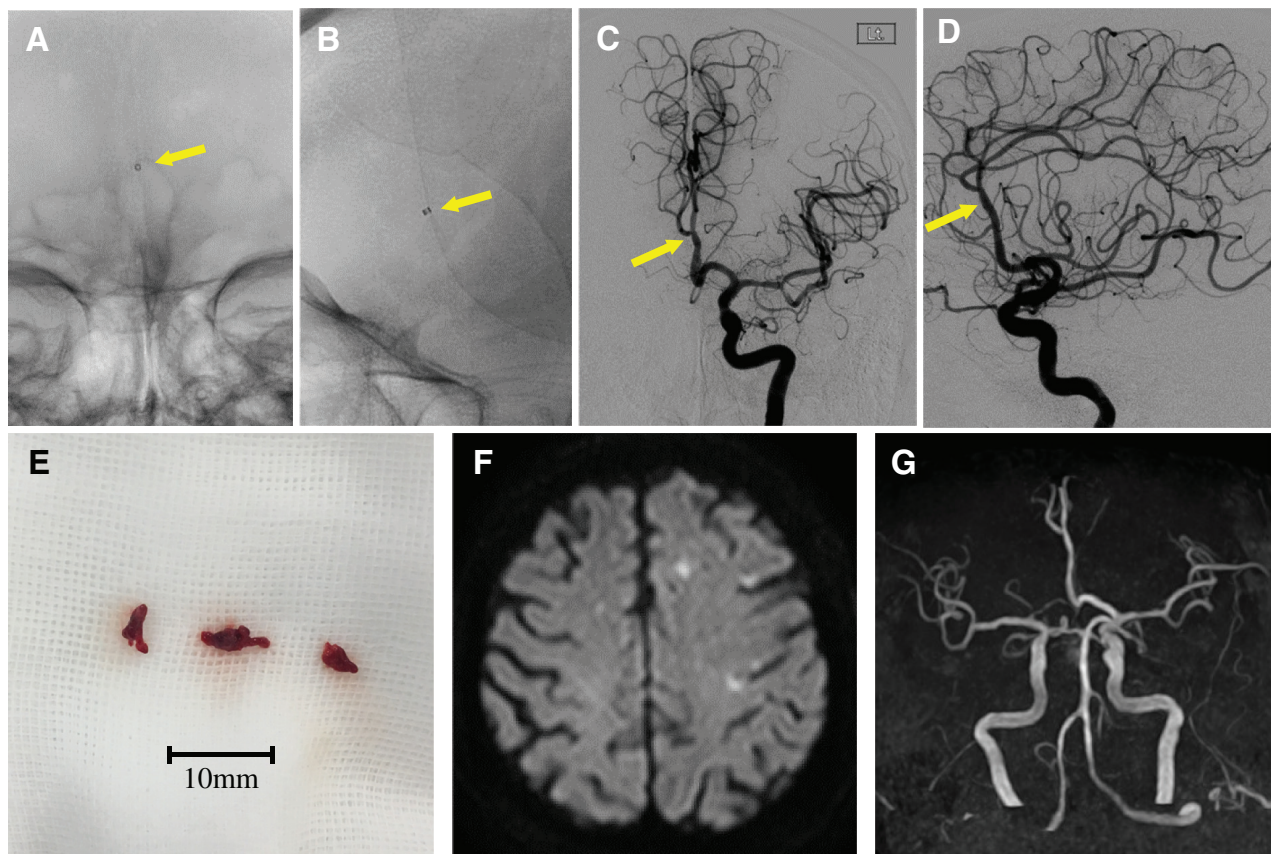


Fig. 2 (A, B) Digital angiogram (A: AP view, B: lateral view): the Penumbra 4MAX catheter was navigated to the occluded azygos ACA A2 segment (arrow). (C, D) DSA of the left ICA (C: AP view, D: lateral view): complete recanalization of the azygos ACA was achieved successfully. (E) The clots retrieved with contact aspiration thrombectomy using the 4MAX catheter. (F) DW-MRI showed

several small ischemic infarctions in bilateral frontal lobes on the next day. (G) MRA showed recanalization of the azygos ACA on the next day. ACA: anterior cerebral artery; AP: antero-posterior; DSA: digital subtraction angiogram; DW-MRI: diffusion-weighted magnetic resonance imaging; ICA: internal carotid artery; MRA: magnetic resonance angiogram

trunk. Although several cases of intracranial aneurysm associated with the azygos ACA have been reported,³⁾ AIS due to occlusion of an azygos ACA is extremely rare.

This case presented unusual symptoms of anterior circulation stroke including quadriplegia and disturbance of consciousness. AIS in anterior circulation could cause quadriplegia like stroke of posterior circulation due to bilateral frontal infarctions with unpaired ACA occlusion. Stroke mimics also could present quadriplegia. Seizure, syncope, sepsis, migraine, space-occupying lesions, functional disorders, and metabolic conditions are common differential diagnoses of acute stroke.¹²⁾ Spontaneous cervical epidural hematoma also present quadriplegia that should be rapidly diagnosed and properly treated.¹³⁾ Stroke should be suspected with sudden onset neurological symptoms and neuroimaging such as MRI/MRA or CT angiogram is useful to distinct between AIS and stroke mimics.¹⁴⁾

Five case reports of azygos ACA occlusion were previously published⁴⁻⁸⁾ (**Table 1**). Since the azygos ACA supplies both hemispheres, complete occlusion might result in large bilateral frontal infarctions and severe symptoms, such as quadriplegia, disturbance of consciousness and aphasia. In our case, complete right-sided paralysis, weakness of the left

lower limb, total aphasia, and left conjugate deviation of the eyes were present, suggesting that the ischemic state was more dominant in the left ACA territory than the right hemisphere because of difference in collateral blood flow from the middle cerebral artery (MCA) or posterior cerebral artery. In four of the previous cases,⁴⁻⁷⁾ the patients were diagnosed as ischemic stroke in the late phase and head CT revealed low intensity lesions suggestive of a large ischemic core, and hence, MTs were not indicated. The clinical outcomes of these four cases were poor, with no improvement of weakness in bilateral lower limbs. On the other hand, Rangel-Castilla⁸⁾ reported a case of MT for azygos ACA occlusion using a stent retriever (SR) and aspiration catheter that resulted in complete recanalization after second attempt. The patient's symptoms showed good improvement and NIHSS score was two at 3 days after the procedure. In our case, complete recanalization was achieved with contact aspiration thrombectomy using a Penumbra 4MAX reperfusion catheter, along with good clinical recovery, suggesting that MT is effective for azygos ACA occlusion, as for ICA and MCA occlusion, with the widespread ischemic penumbra. The Aster trial¹⁵⁾ and Compass trial¹⁶⁾ showed that there was no difference in

Table 1 Summary of previous cases of acute ischemic stroke due to azygos anterior cerebral artery occlusion

Author	Year	Age (years)	Gender	Symptoms	NIHSS	Occluded portion	IV t-PA	MT	Technique	Post-TICI	Number of pass	Outcome
Rajasekharan C et al ⁴⁾	2012	50	Male	Akinetic mutism quadriplegia	ND	ND	-	-	-	-	-	mRS 5 at 3 months
de Sousa CS et al ⁵⁾	2017	63	Male	Coma	ND	A2 distal	-	-	-	-	-	Dead
Khaliq A et al ⁶⁾	2018	50	Male	Disturbance of consciousness aphasia paraparesis	ND	A2 distal	-	-	-	-	-	mRS 4 at discharge
Saleh C et al ⁷⁾	2018	62	Female	Disturbance of consciousness quadriplegia eye deviation to the right	ND	ND	-	-	-	-	-	mRS 4 at discharge
Rangel-Castilla L et al ⁸⁾	2019	59	Male	Right hemiparesis aphasia	20	A2 distal	-	+	SR + Aspiration	3	2	NIHSS 2 at 3 days
Present case	2020	84	Male	Disturbance of consciousness quadriplegia aphasia eye deviation to the left	32	A2 distal	+	+	Aspiration	3	2	mRS 1 at 90 days

IV t-PA: intravenous tissue-plasminogen activator injection; mRS: modified Rankin Scale; MT: mechanical thrombectomy; ND: no data; NIHSS: National Institutes of Health Stroke Scale; SR: stent retriever; TICI: Thrombolysis in Cerebral Infarction

results between SR and contact aspiration for large vessel occlusion in the anterior circulation. However, since the target vessels of these trials were the ICA or proximal MCA, it is not clear whether the results can be extrapolated to azygos ACA occlusion. In our case, pre procedural MRA (Fig. 1C) showed that this occluded A2 segment was similar to M2 superior trunk just only in terms of the distance from the guiding catheter to the lesion and the diameter of the occluded artery. Hence, we chose contact aspiration thrombectomy with the Penumbra 4MAX, as we usually perform for M2 occlusion.¹⁷⁾

However, Chung et al¹⁸⁾ reported that because ACA had a more tortuous vascular course and acute angles compared with MCA, it was difficult to navigate the aspiration catheter and dissection, vessel perforation or vasospasm might easily occur during the procedure.

Since the 4MAX catheter is longer than other large bore aspiration catheters, and accessibility of the distal lesion is also better using the Velocity microcatheter, we were able to carefully navigate the 4MAX to the distal azygos A2 lesion and aspirate the clot. There are two more advantages of contact aspiration thrombectomy for azygos A2 occlusion like as other distal lesions. One is that the procedure does not require blind passage of the microcatheter through the occluded lesion without information about distal vascular anatomy. The azygos ACA usually divides into two pericallosal arteries, although there are also several other anatomical types,¹⁰⁾ and cerebral aneurysms might be present at the bifurcation of bilateral A3 branches. Second, there are two sharp curves between the terminal ICA and azygos A2 segment. The SR might stretch the ACA, leading to a higher risk of subarachnoid hemorrhage as compared to contact aspiration.

Conclusion

Occlusion of the azygos ACA results in large bifrontal ischemic infarctions and severe neurological symptoms, such as disturbance of consciousness, quadriplegia, or aphasia. In our case, contact aspiration thrombectomy using a Penumbra 4MAX catheter was effective for complete recanalization, allowing the patient to recover sufficiently to be able to live his daily life independently.

Informed Consent

The authors received written informed consent regarding of this case report from the patient.

Disclosure Statement

None of the authors have conflicts of interest in relation to this article.

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