JPSIM Journal of Pakistan Society of Internal Medicine

Case Report

Azygous Anterior Cerebral Artery Stenosis: An Unusual Cause of Bilateral Frontal Lobe Cerebral Infarctions

Muhammad M Javaid,[1] Shruti G Iyer,[1] Rama D Penumarty,[2] Asif Hussain[2]

[1]Department of Medicine, South West Healthcare, Warrnambool, & [2]Monash University Victoria, Australia

Abstract

An azygous anterior cerebral artery is a rare anatomical variant of the Circle of Willis. It is characterised by the absence of the anterior communicating artery and the union of proximal segments to form a single vessel supplying both cerebral hemispheres. Occlusion or stenosis of the single vessel and lack of collateral circulation can lead to bilateral frontal cerebral infarctions, an otherwise relatively uncommon presentation. Knowledge of such an anatomical variant is vital for adequately investigating and managing patients presenting with bilateral anterior cerebral artery territory strokes.

Keywords: Anterior cerebral artery, azygous anterior cerebral artery, anatomical variants, Circle of Willis, infarction

How to cite this:

Javaid MM, Iyer SG, Penumarty RD, Hussain A. Azygous Anterior Cerebral Artery Stenosis: An unusual cause of bilateral frontal lobe cerebral infarctions. J Pak Soc Intern Med. 2022;3(1):66-68

Corresponding Author: Dr. Muhammad M Javaid **Email:** mmjavaid@doctors.org.uk

DOI: https://doi.org/10.70302/jpsim.v3i1.2214

Introduction

Azygous anterior cerebral artery (AACA) is a rare anatomical variant of the Circle of Willis, affecting 0.3- 2% of the population. It is characterised by the absence of the anterior communicating artery and union of the two proximal segments to form a single vessel supplying both cerebral hemispheres (figure 1).[1,2] The AACA is a significant predictor of bilateral frontal strokes, especially if occluded or stenosed. The knowledge of this variation is crucial for appropriate diagnosis and management. Moreover, the condition has also been associated with multiple anomalies (Table I) including lobar holoprosencephaly, agenesis of the corpus callosum, porencephalic cysts, septo-optic dysplasia, arteriovenous malformations, and saccular aneurysms which can affect as many as 13 – 70 % of such patients.[1,2]

Table 1: *Table I: Associated Anomalies with Azygous Anterior Cerebral Artery*

Aneurysms of the distal part of anterior cerebral artery
Bilateral frontal lobe infarctions
Agenesis of corpus callosum
Porencephalic cysts
Hydranencephaly
Lobar holoprosencephaly
Septo-optic dysplasia (blindness, optic nerve atrophy, absence of septum pellucidum)

Case Presentation

A 47-year-old Caucasian male presented to the accident and emergency department with sudden onset of right lower limb weakness. He had previously been well. His only significant past medical history included hypertension which had been reasonably controlled in the recent past. His regular medications included Aspirin 100 mg daily, Telmisartan 80mg daily, and Amlodipine 10mg daily. He was a smoker with a 30-pack-year smoking history. He did not drink alcohol. He lived at home with his wife and worked as a crane operator.

On examination, he was orientated in time, place, and person. He had a pulse rate of 84 beats per minute with a regular rhythm. Blood pressure measured 150/83 mm Hg. The temperature was normal. He had a moderate (grade 3/5) weakness of the right lower limb (hip and knee flexion and extension). The sensation of light touch was reduced in the right upper and lower limbs. Cranial nerves and facial sensations were intact.

Initial investigations showed haemoglobin of 162 g/L. White cell and platelet counts were normal, measuring 9.4 X 109/L and 206 X 109/L, respectively. His kidney function tests, liver function tests, and coagulation profile were also within normal limits.

A non-contrast computed tomography (CT) scan of the brain did not show any significant abnormality. CT angiogram showed no defects in the vertebral and

common carotid arteries. The basilar and internal carotid arteries appeared normal. At The Circle of Willis, anterior, middle, and posterior cerebral arteries were preserved, and no abnormality was noted in the anterior and posterior communicating arteries. No enhancing lesion was identified concerning the brain parenchyma.

A Magnetic Resonance Imaging (MRI) scan of the brain showed small focal areas of restriction abnormalities in the right frontal lobe superiorly towards the vertex. The left frontal and posterior parietal regions had similar appearances. The appearance was suspicious of an embolic phenomenon.

Electrocardiogram (ECG) was normal. A transthoracic echocardiogram (TTE) did not reveal an embolic source. Further investigations including anti-nuclear antibodies (ANA), anti-nuclear cytoplasmic antibodies (ANCA), extractable nuclear antigens (ENA), antiphospholipid antibodies, anticardiolipin antibodies, and cryoglobulins were all normal.

Within twenty-four hours, his symptoms spontaneously resolved, and he was discharged from the hospital with a plan to perform an outpatient Transesophageal Echocardiography (TOE). Aspirin was changed to Clopidogrel 75 mg a day, and Atorvastatin 80 mg a day was initiated. Atenolol 50 mg a day was also added on top of his usual antihypertensive medications.

Thirty-six hours after his discharge from the hospital, he presented with a recurrence of right lower limb weakness with new symptoms of right upper and lower limb anaesthesia. A repeat CT scan of the brain showed small lesions in the right frontal and left frontoparietal regions. The appearance was suggestive of infarctions, vasogenic oedema or metastasis. An MRI scan was recommended to help clarify if changes were due to infarction.

An MRI scan showed worsening of the previously noted embolic phenomenon primarily affecting both frontal lobes. MR angiogram (MRA) was performed to evaluate further the findings, which showed a single anterior cerebral artery with a short segment of stenosis and multifocal infarcts in the anterior cerebral artery territory bilaterally (figure 1).

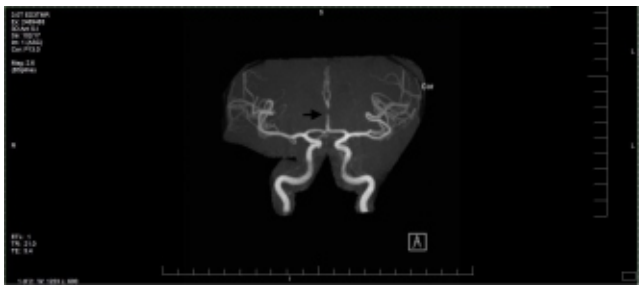

Figure 1: *Magnetic Resonance Angiogram Showing Stenosed Azygous Anterior Cerebral artery*

A final diagnosis of bilateral frontal lobe infarctions secondary to the stenosed azygous anterior cerebral artery was made. With the aetiology of these infarcts confirmed, a trans-oesophageal echocardiogram was not performed.

The patient was managed medically. He was commenced on dual antiplatelet therapy (Clopidogrel 75mg daily and Aspirin 100mg daily) and was advised to continue his Atorvastatin, Atenolol, Telmisartan, and Amlodipine. He was also encouraged to quit smoking and improve his lifestyle.

Over the next few days, his symptoms gradually resolved, and he made a good neurological recovery. At a follow-up three months post-discharge, he had remained stable and had not had any further symptoms. He had managed to stop smoking, and his blood pressure was well controlled at 127/77 mmHg.

Discussion

The Circle of Willis, situated at the base of the brain, is a complex anastomotic network between the internal carotid arteries and vertebrobasilar systems. It provides a crucial connection between the blood supply to the forebrain and the hindbrain. It is vital in maintaining cerebral circulation in case of compromised blood flow in one of its component vessels.[3]

The internal carotid arteries bifurcate into the anterior cerebral arteries (ACA) and the middle cerebral arteries (MCA) bilaterally to form the anterior part of the circle of Willis. The two ACAs extend in medial and anterior directions to supply the most midline portions of the frontal lobes and superior and medial parietal lobes. A single anterior communicating artery connects them to complete the anterior part of the circle. The MCAs, not considered a part of the Circle of Willis per se, constitute morphological extensions of the internal carotid arteries, and extend laterally to supply most of the lateral surface of the hemisphere and the inferior portion of the temporal lobe and occipital lobe.[1,4]

The two vertebral arteries join intracranially to form the basilar artery, which bifurcated into two posterior cerebral arteries (PCA) to constitute the posterior part of the circle of Willis. The PCAs primarily supply the occipital lobes and inferior portions of the temporal lobes and are connected to the anterior part of the circle via two posterior communicating arteries which branch from internal carotid arteries just before its terminal bifurcation into ACA and MCA.[1,4]

However, a complete circle of Willis is rarely seen radiographically in its entirety and is only present in some individuals. Anatomical variations are common. A well-developed communication between each of its parts is recognised in less than half of the population.[3] AACA

is a rare variant of the normal cerebral circulation and can have implications of bilateral cerebral ischaemia if blocked.

ACA territory infarctions are relatively uncommon, accounting for a considerably small share of the total number of ischemic infarcts.[5,6] Bilateral anterior circulation infractions are rare and are often secondary to vasospasm complicating subarachnoid haemorrhage due to the rupture of one or more aneurysms of the anterior communicating arteries or distal ACA.[7] Congenital variations of the anterior cerebral circulation may also cause bilateral frontal lobe ischemia with a lesion affecting a single vessel. Thrombosis or embolus affecting a dominant ACA supplying both cerebral hemispheres due to aplasia or hypoplasia of the contralateral ACA, occlusion of an azygous ACA or an accessory vessel arising from the anterior communicating artery can result in bilateral ischemic strokes in both ACA territories.[8]

Our patient was managed medically with antiplatelet agents and modification of cardiovascular risks. Thrombolytic therapy was not considered due to delayed presentation outside the stroke thrombolysis window period. Endovascular intervention was not considered a first-line option due to the higher risk of complications, including vessel rupture or occlusion, which could have had severe implications for the patient in the context of a single ACA supplying both cerebral hemispheres. However, this option was reserved for the future if his symptoms recurred despite maximum medical therapy.

Conflict of Interest: None

Funding Sources: None

References

1. Makowicz G, Poniatowska R, Lusawa M. Variants of cerebral arteries-anterior circulation. Pol J Radiol. 2013;78(3): 42-7.
2. de Sousa CS, de Miranda CL, Avelino MC, Bastos BB, Moreira de Sousa RS, de Miranda CV. Thrombosis of the Azygos Anterior Cerebral Artery. Case Rep Radiol. 2017;doi: 10.1155/2017/5409430
3. Krabbe-Hartkamp MJ, van der Grond J, de Leeuw FE, de Groot JC, Algra A, Hillen B, Breteler MM, Mali WP. Circle of Willis: morphologic variation on three-dimensional time-of-flight MR angiograms. Radiol. 1998; 207(1):103-11.
4. Harrigan MR, Deveikis JP. Handbook of Cerebrovascular Disease and Neurointerventional Technique. Humana Press; 2013.
5. Toyoda K. Anterior cerebral artery and Heubner's artery territory infarction. Front Neurol Neurosci. 2012; 30(2): 120-2.
6. Kubis N, Guichard JP, Woimant F. Isolated anterior cerebral artery infarcts: A series of 16 patients. Cerebrovasc Dis. 1999;9(3):185-7.
7. Menezes BF, Cheserem B, Kandasamy J, O'BrienD. Acute bilateral anterior circulation stroke due to anomalous cerebral vasculature: a case report. J Med Case Repor. 2008;2(1):188
8. Kunal Kumar, Daniel Strbian, Sophia Sundararajan. Acute cerebral infarction presenting with weakness in both legs and one arm. Stroke. 2015;46(6): e134-6.