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ABSTRACT

Background. Intracranial dolichoectasia (IADE) is a rare vascular disease characterized by distension, elongation and tortuosity of an artery. IADE rarely involves paediatric aged groups. It is either asymptomatic or manifests as ischemic or haemorrhagic attacks.

Case description. A healthy, 30-year-old, female teacher presented with recurrent attacks of bi-frontal headaches associated with dizziness and dropping attacks of two-week duration. She was referred by her general physician to our institution of Neurosurgery Teaching Hospital in Baghdad, Iraq with a suspicion of medial temporal lesion on a cranial computed tomography (CT) scan. Magnetic resonance imaging study excluded the diagnosis suggesting a dolichoectatic middle cerebral artery that was confirmed by CT-angiography.

Conclusion. Dolichoectasia of the middle cerebral artery is a rare and benign lesion. However, it can masquerade as cerebral cavernous malformation or intracranial arterial aneurysm. Thus, careful radiological evaluation with the suggested diagnostic criteria are of paramount importance to prevent its misdiagnosis.

BACKGROUND

Intracranial arterial dolichoectasia (IADE) is sporadic angiopathy characterized by dilatation, elongation and tortuosity of an intracranial artery. IADE is approximately 0.1-6.5% prevalent in the general popula-

Keywords

dolichoectasia,
middle cerebral artery,
anterior circulation,
arteriopathy,
tortuous artery



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tion and 12 % in stroke patients [1]. IADE preferentially involves the basilar artery and posterior circulation; anterior circulation IADE constitutes only one third of all cases [2]. IADE is usually an incidental finding but symptoms may arise due to ischemia, hemorrhage or cranial nerve compression [3]. Although uncommon, hydrocephalus was also reported as a manifestation of IADE due to the obstruction of cerebrospinal fluid flow through the foramen of Monro or the cerebral aqueduct [4]. IADE may co-exist with several vascular pathologies such as abdominal aortic aneurysms, intracranial aneurysms and coronary artery disease [5]. A diagnostic criterion for dolichoectatic basilar artery was suggested by smoker et al, but no criteria for dolichoectasia in other intracranial arteries have been validated thus far [6].

Herein, we present a case of middle cerebral artery dolichoectasia, initially diagnosed as cerebral cavernous malformation along with proposed diagnostic criteria based on reviewing the available literature.

CASE SCENARIO

A healthy, 30-year-old, female teacher presented with recurrent attacks of pulsatile bifrontal headache of two-week duration. Each attack lasts for more than one-hour and significantly impacted her daily activities. The headache is associated with profound dizziness and “dropping attacks”; assumed to be seizures by her general physician. The patient’s EEG and video EEG were both normal. She was referred to the neurosurgery outpatient clinic at our institution with a suspicious medial temporal lesion on a cranial CT scan. On examination, the patient was pale but fully oriented with no remarkable neurologic deficits. Her brain CT scan showed a small rounded heterogeneous lesion with calcification at the medial temporal area; there was no evidence of peri-lesional edema. Initially, the diagnosis of uncal cavernous malformation was pondered. (Fig.1). In order to confirm this diagnosis, brain MRI with T2-gradient echo and MRA studies were ordered. The T2-weighted MRI study revealed lesional flow-void, the T2-gradient echo was negative and the MRA showed an abnormal vascular loop of the MCA within the lesion. These findings excluded the diagnosis of cavernous malformation and strongly suggested MCA aneurysm or dolichoectasia (Fig.2). Next, a computed tomography angiography (CTA) of

the brain was obtained. The CTA revealed that the proximal part of the MCA was dilated, elongated, tortuous and formed a superior blind loop; findings that were consistent with the diagnosis of MCA dolichoectasia (Fig.3). Moreover, catheter cerebral angiography can be used to confirm the diagnosis in such situation but it is not feasible in our facility nor in many neurosurgical institutions around the world. Thereafter, patient was reassured that she had a “benign variation in the brain circulation rather than a critical pathology and that no intervention was indicated at the time”. Laboratory investigations normal except for iron deficiency anemia. The patient was discharged and a follow-up was scheduled with her physician. At 6-month follow-up, patient was generally well, had already resumed her normal daily activities and received treatment for anemia. The headache was both minimal and occasional at this stage. Both CTA and MRI revealed no new significant findings.

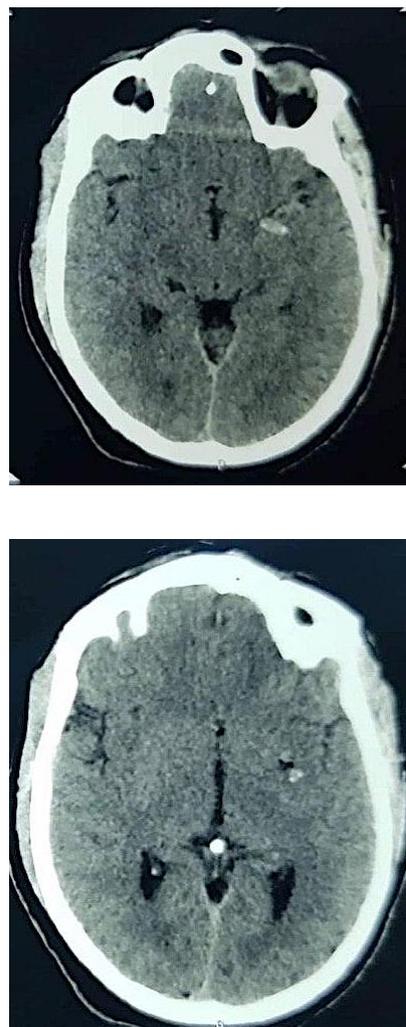
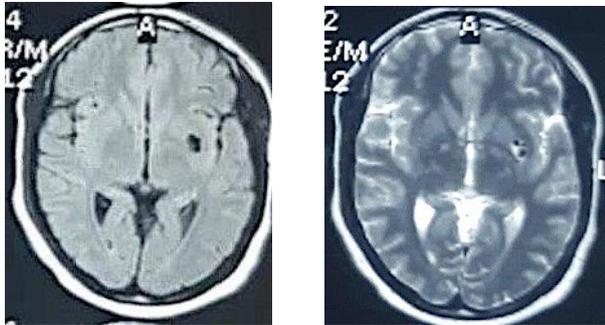
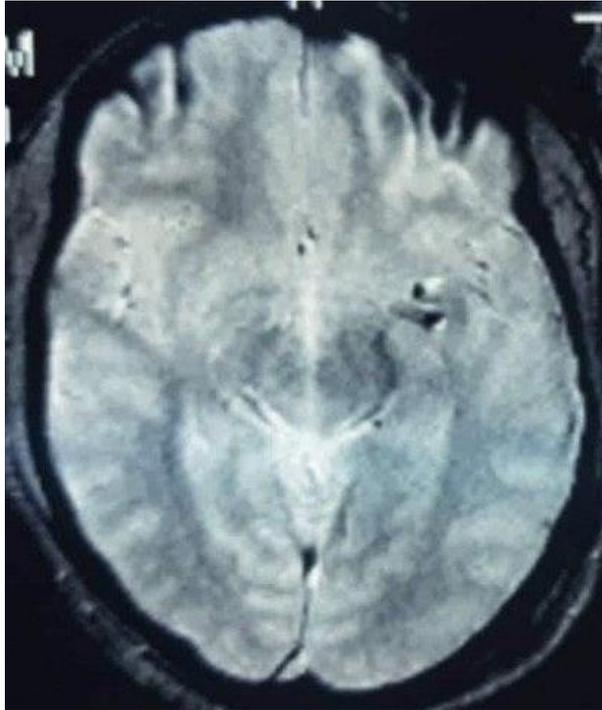


Figure 1. Cranial CT scan showing a heterogeneous, deep left-sided, temporal lesion of mixed density with no perilesional edema. Here, the initial diagnosis was cerebral cavernous malformation.



A. **B.**



C.
D.

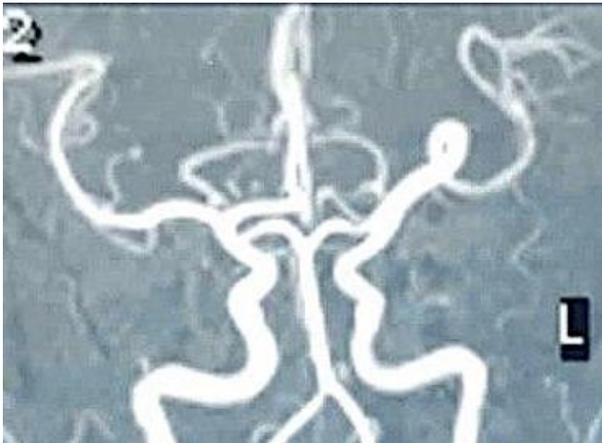


Figure 2. Brain MRI showed deep left temporal lesion. **A:** FLAIR axial section: The lesion is hypointense with no perilesional edema. **B:** T2 axial section: The lesion contains signal voids that denoted the presence of vessels within the lesion. **C:** T2-

Gradient Echo axial view showing the absence of blood clots. **D:** MRA showing an enlarged and tortuous left MCA (sphenoidal segment) as compared with the right MCA with a superiorly projecting loop or a possible aneurysm.



Figure 3. Brain CT angiography 3D reconstructed image showing an enlarged and tortuous left MCA (sphenoidal segment) as compared with the right MCA with a superiorly projecting loop, confirming the diagnosis of left MCA Dolichoectasia and excluding the presence of an aneurysm.

DISCUSSION

the vertebrobasilar system is the most common site of IADE followed by internal carotid artery. IADE is a disease of all ages although its presentation in the pediatric age group is rare. It affects females more than males [2]. IAED may be asymptomatic or masked by an array of inconclusive manifestations such as headaches, strokes, seizures and focal deficits [4]. In this report, a 30-year-old female presented with recurrent bouts of headache and dizziness. A brain CT-scan revealed a calcified lesion, leading to a provisional diagnosis of an uncus cavernous malformation. Further imaging studies including MRA and T2-gradient ECHO- provided a better visualization of the vasculature and the brain parenchyma narrowing the differential diagnosis to an aneurysm or a dolichoectasia. Finally, CTA images showed dilated, distended and tortuous vessels and excluded the differential of an aneurysm.

IADE is commonly mistaken with other vascular pathologies, such as dural fistulas or Arteriovenous malformations [7]. Therefore, multiple imaging modalities are often required to reach the definitive diagnosis of dolichoectasia especially if it is located distally in the anterior circulation. Nakahara et al

reported a case of MCA dolichoectasia, initially diagnosed as a terminal-ICA, saccular aneurysm using antero-posterior and lateral CTA views. However, the reverse waters view suggested the correct diagnosis of a dolichoectatic MCA rather than an ICA aneurysm; this study highlights the importance of multiple CTA views in the diagnosis of IADE [8]. Several efforts attempted to establish a solid platform for diagnosing anterior circulation dolichoectasia depending on a multi-modal diagnostic approach. Some authors have recommended the co-utilization of additional radiological techniques such as digital subtraction angiography to capture the real-time flow properties of the aberrant vessel [9,10]. Dolichoectatic MCA is a benign lesion in general. However, treatment is usually indicated when there is a co-existing pathology. Surgical manipulation of the enlarged vessel may lead to complications such as hemorrhage or ischemia [10]. In this study, we

treated the patient conservatively using simple analgesics which enabled her to resume her normal activities. Our approach is comparable to the one described in the literature of MCA dolichoectasia (Table 1). However, the initial false interpretation of medial temporal cavernous malformation, then MCA aneurysm rendered this case report to be of a critical value regarding differential diagnosis of MCA dolichoectasia [8,10]. Based on the aforementioned literature analysis, we suggest 3 features that can be considered as alarming criteria for the diagnosis of MCA dolichoectasia, these include: atypical clinical presentation, enlarged parent vessel and unusual location of the lesion.

We recommend a multi-modal diagnostic approach to determine the most appropriate management along with long-term clinical and radiological follow-up for monitoring of a less likely but possible lesion enlargement.

Table 1: Literature review of dolichoectatic middle cerebral artery

Author	Sex	Age, year	Presentation	Radiologic findings	Treatments
Brinjkji et al [11]	F	19	Asymptomatic, found incidentally	MCA dolichoectasia with superimposed multilobulated aneurysm, mild preceding stenosis	Conservative
Feliciano et al [12]	M	42	Headache associated with left-sided weakness and intermittent nausea and vomiting	MCA dolichoectasia and a cluster of aneurysms with right basal ganglia hemorrhage	Conservative
Abe et al [13]	M	32	Asymptomatic, found incidentally after a motor-vehicle accident	MCA dolichoectasia	—
Nakahara et al [8]	F	59	Left hyposmia and mild intermittent occipitalgia	MCA dolichoectasia	No surgery, no medication
Kanemoto et al [14]	F	41	Seizures and anxiety	MCA dolichoectasia with cavernous hemangioma	Cavernous hemangioma resection

Tokunaga et al [15]	F	58	Right hemiplegia, homonymous hemianopia, hypertension	Left putamen hematoma and bilateral MCA dolichoectasia	—
Puca et al [16]	F	32	Ischemic stroke at the age of 7 y	Old ischemic lesion and MCA dolichoectasia	No surgery, no medication
Maruya et al [17]	F	40	Subarachnoid hemorrhage	A saccular aneurysm on a dolichoectatic MCA	Surgical clipping of the aneurysm
Guo et al [10]	M	43	Ischemic stroke 3 y ago	MCA dolichoectasia	No surgery, antiaggregating therapy
Current study	F	30	Headache and dizziness	MCA dolichoectasia	Conservative
M = Male; F = Female; MCA = Middle cerebral artery					

CONCLUSION

MCA dolichoectasia is a rare and benign lesion. However, it can masquerade as cerebral cavernous malformation or intracranial aneurysm. Thus, careful radiological evaluation along with the suggested diagnostic criteria are of a paramount importance to prevent its misdiagnosis.

ABBREVIATIONS

IADe; Intracranial arterial dolichoectasia, EEG; electroencephalogram, CT; computed tomography, MRI; Magnetic resonance imaging, CTA; computed tomography angiography, MCA; Middle cerebral artery.

DECLARATIONS

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