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Case Report

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DIFFUSE INTRACRANIAL ARTERIAL DOLICHOECTASIA – A RADIOLOGICAL CASE REPORT AND COMPARISON WITH VERTEBRO-BASILAR DOLICHOECTASIA

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ABSTRACT

Intracranial arterial Dolichoectasia or vertebra-basilar dolichoectasia is abnormal dilatation of the vertebra-basilar arterial system. It is phenotypically distinct from diffuse intracranial arterial Dolichoectasia which involves at least 2 segments of the cerebral arterial system. We present here a case of 70 year old male patient who underwent MRI of brain and intracranial vessels to highlight the importance of recognizing the lesser known and often missed diffuse intracranial Dolichoectasia and compare its features with vertebra-basilar Dolichoectasia. Diffuse intracranial arterial Dolichoectasia has been found to occur in older patients, with increased rates of aneurysmal growth, increased chances of concomitant aortic and visceral aneurysms with increased chances of complications such as aneurysm rupture, ischemic stroke related deaths and poor neurological recovery. Further research is needed in this direction to characterise diffuse variety of intracranial arterial Dolichoectasia so that it be diagnosed earlier and better treatment can be provided.

KEYWORDS: Diffuse Intracranial arterial Dolichoectasia, Vertebrobasilar Dolichoectasia, Aneurysm, Ischaemia.

INTRODUCTION

Intracranial Arterial Dolichoectasia (IADE) refers to abnormal dilatation, ectasia and tortuosity of the intracranial arteries most commonly the vertebra-basilar system hence also referred to as vertebro-basilar Dolichoectasia (VBDE). This entity has variable clinical sign and symptoms. It can lead to various complications such as infarction, cranial nerve compression, intracranial haemorrhage and sometimes death. Diffuse IADE is a distinct clinical entity from the more more common IADE. This involves involvement of more than two cerebral blood vessels (not restricted to a single arterial territory) and hence has higher rate of complications and fatality rates.

This article's primary purpose is to lay stress on the distinct yet seldom described clinical entity "Diffuse intracranial arterial Dolichoectasia". This article also tries to summarize the literature currently available about this less known entity and describe its risk factors, radiological characteristics and also compare it with much more commonly known entity of Vertebrobasilar Dolichoectasia.

CASE REPORT

A 70 year old male patient presented at the outpatient clinic with history of frequent falls, weakness in the left upper and lower limbs and severe headache.

Brain MRI angiography was performed using a 3D TOF pulse sequence and later viewed using a high density multi-planar MIP algorithm.

Imaging findings revealed dilatation of supra-clinoid segment of bilateral internal carotid artery (figure 1) and bilateral middle cerebral artery (figure 2). Bilateral vertebral and basilar arteries also were relatively increased in diameter. These findings are suggestive of diffuse IADE.

There were acute infarcts in left half of pons and left cerebellar hemisphere (figure 3) which were showing diffusion restriction. There was focal fusiform aneurysm in the inferior terminal branch of left middle cerebral artery (figure 2). There was also age related diffuse cerebral atrophy with chronic microvascular ischemic lesions in bilateral basal ganglia, periventricular, deep and subcortical white matter. Few focal residue of chronic bleed were also seen in pons and right parietal lobe due to long standing hypertension.

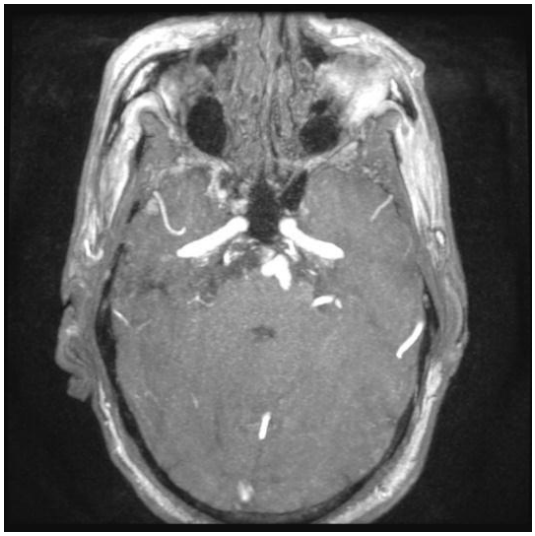

Figure 1: Dilatation of bilateral ICA (supraclinoid segment).

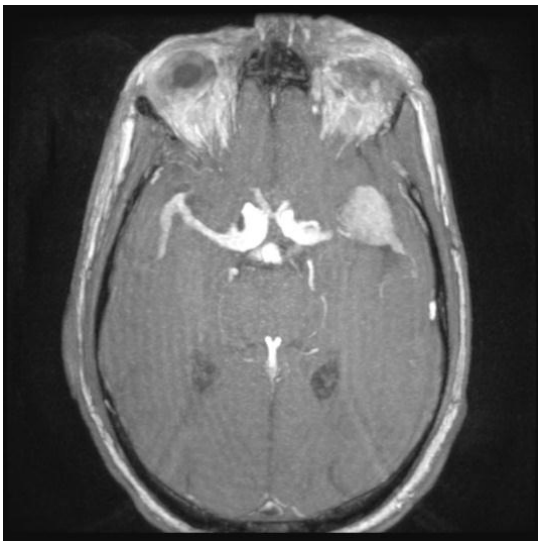

Figure 2: Dilatation of bilateral MCA and aneurysm in left MCA.

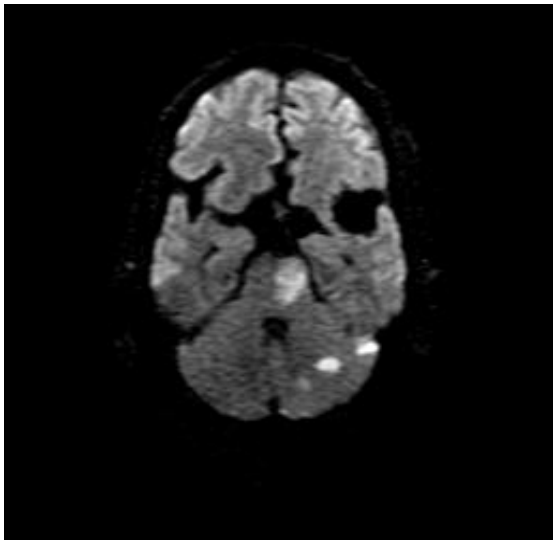

Figure 3: Acute infarcts in left half of pons and left cerebellar hemisphere

DISCUSSION

Dolichoectasia term is a derivation of two Greek words dolichos, meaning "abnormally long," and ectasis, meaning "to extend or dilate."[1] It is also known as dilative arteriopathy since ectasia of the involved artery is the key diagnostic feature.[2] Intracranial arterial dolichoectasia (IADE) refers to an unusual arteriopathy which is characterized by abnormal elongation, tortuosity and dilatation of at least one artery in the cerebral vasculature.[2,3] Diffuse IADE is a more recently recognised entity which refers to the involvement of two or more intracranial cerebral blood vessels,[4] and is more commonly associated with rupture, stroke and ischemia related deaths.

Dolichoectasia has a wide range of etiological factors such as arterial dissections, chronic high blood pressure, connective tissue disorders, infections, glycogen storage diseases and prior radiation therapy.[5,6,7,8] It is also suggested that the diffuse variant of IADE is a distinct entity as compared to the frequently encountered single vascular territory dolichoectiasia with clinically distinct presentation, association and natural history.[4]

The pathophysiology of diffuse IADE is poorly understood. It is different from the atherosclerotic endothelial injury and plaque formation. It is postulated to be the result of disruption of the internal elastic lamina associated with atrophy of the muscle in the vessel wall layer with hyalinization of the connective tissues causing abnormal dilatation of the involved arterial vessels.[3,9,10] Another theory points towards aberrant vascular remodelling and abnormal vascular connective tissue within the arterial wall due to an imbalance between the in vivo activity of tissue antiprotease and metalloprotienases.[2,11] (Figure 4).

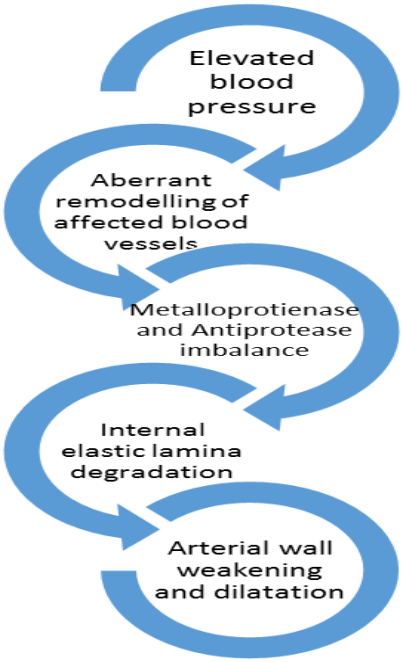

Figure 4: Pathogenesis of diffuse IADE.

IADE can affect both anterior as well as posterior circulation, however, it is more commonly seen in the vertebrobasilar (VB) system. The relative lack of sympathetic innervation of the posterior circulation giving it a less trophic support against the stresses of blood flow and blood pressure may explain the preferential involvement of the VB system.[12,13]

Brinjikji et al. conducted a study in 2014[4] comparing the patients suffering from diffuse IADE and vertebro-basilar Dolichoectasia (VBDE) and found that patient having diffuse IADE were older, having higher prevalence rates of abnormal aortic and visceral aneurysms as compared to the patients having VBDE. Also there was a higher prevalence of smoking amongst the patients of diffuse IADE. Complications were also reported to be higher in these patients with higher aneurysmal growth as well as rupture rates leading to lesser degree of neurologic function on follow up and increased incidence of aneurysm related deaths. The size criteria put forward by Brinjiki et al.[4] for diffuse IADE and its comparison with VBDE is summarised in the table 1.

Table I: ADE as compared to VBDE.

	IADE	**VBDE(14,15)**
MRI appearance	Dilatation of an entire vascular segment	Dilatation of a portion of vertebrobasilar arterial segment with any degree of tortuosity
Vessels affected	≥2 intracranial vascular beds (ie. Left anterior circulation, right anterior circulation, vertebrobasilar system)	Vertebro basilar system only
Size criteria	Cavernous ICA ≥8.5 mm[4] Supraclinoid ICA ≥8.0 mm[4] MCA ≥5.0 mm[4] Basilar artery ≥6.0 mm[4]	Basilar artery diameter of more than 5.0mm
Dilatation growth rate	10% per year[4]	7% per year[14,15]
Risk of ischemic stroke	11% per year[4]	3% per year[14,15]
Risk of aneurysm rupture	6% per year[4]	2% per year[14,15]

CONCLUSIONS

Diffuse IADE and VBDE are two distinct vascular phenotypes due to difference in difference in morbidity rates, mortality rates, coexisting features and complications. Hence, more methodological research is required to more specifically characterise genetic as well as histopathological features of the diffuse IADE so that it can be diagnosed and for exploration of newer and more advanced treatment options.

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