

DOLICHOECTASIA OF THE CEREBRAL ARTERIES WITH SYSTEMIC VASCULAR ABNORMALITIES: CASE REPORT

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SUMMARY

An unusual case of cerebral arterial dolichoectasia with systemic vascular abnormalities is reported. The diagnostic and clinical aspects of this entity are discussed.

KEY WORDS

Dolichoectasia, Vascular Abnormalities, Vertebrobasilar System.

INTRODUCTION

Dolichoectasic change of cerebral vessels is a rare clinical entity. Moreover, the symptoms and signs of this condition are not specific; so the correct diagnosis is not easy in all cases. Dolichoectasia of the intracranial vessels usually occurs in multiple sites. Concomitant systemic dolichoectasic changes and/or aneurysm formation of the vessels are reported with a frequency of 50 % in postmortem studies (2). However, this rate is very low in clinical studies.

In this article, multifocal dolichoectasic changes of cerebral vessels with multiple abnormalities of the aorta in a patient is reported.

CASE REPORT

A 56 year-old male patient was referred to our hospital in April 1989. His complaints were speech disturbance and difficulty in walking. Eight months prior to admission, he had an attack of paresis on the right side which resolved spontaneously. His present complaints started three months ago. Despite medical treatment, there was no improvement.

Hypertension was recorded in his past medical history. His blood pressure was 170/100 mmHg. There was a right lower abdominal mass which was hard, mobile, pain-free, pulsatile and approximately 10x15 cm. in size.

Neurological examination revealed dysarthria, bilateral dysmetria which was more prominent on the right side and ataxic gait.

The patient was evaluated in radiodiagnostic algorithm. Widening of the mediastinal shadow was seen on the chest x-ray. Ultrasonography of the abdomen revealed atherosclerotic changes and fusiform aneurysm of the aorta which arose from the level of the diaphragm and involved both iliac arteries with the largest diameter 9 cm. In cranial computed tomography (CCT), extreme widening, elongation and left-sided deviation of the basilar artery and mild hydrocephalus were shown. Digital subtraction angiography (DSA) via the intravenous route was performed to evaluate the aorta and its main branches. Aneurysmal dilatation and tortuosity of the arcus aorta, thoracic and abdominal segments of the aorta and both iliac arteries were seen (Figure 1). Selective



Fig.1 : IV Digital Subtraction Angiography shows aneurysmal dilatation and tortuosity of abdominal segments of the aorta and both iliac arteries.

cerebral angiography via left brachial artery minipuncture and catheterisation showed that there was widening and elongation in both carotid systems C1, C2 segments and in the anterior and middle cerebral arteries (Figure 2). A right vertebral arteriogram

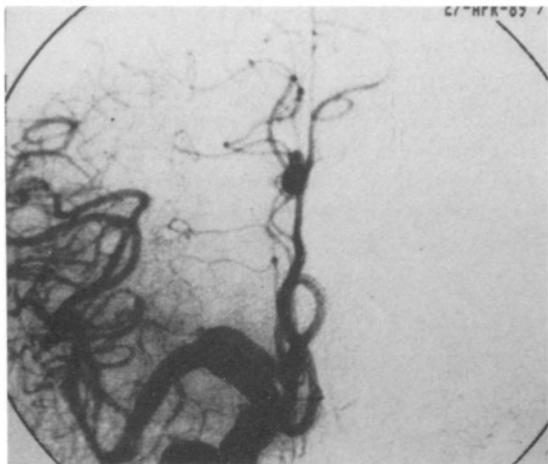


Fig 2 : Selective carotid angiography, right carotid P-A view; shows widening and elongation in carotid systems. C1, C2 segments and in anterior and middle cerebral arteries.

confirmed the CCT findings of the basilar artery (Figure 3) and revealed the widening of proximal



Fig 3 : Selective right vertebral angiography, P-A view; reveals widening extreme tortuosity and left-sided deviation of the basilar artery.

segments of both posterior cerebral arteries. The distal segment of the left vertebral artery was occluded and the basilar artery was visualised by the collateral flow from the posterior inferior cerebellar artery.

Because of the multisystemic vascular abnormalities, an operative procedure of right superficial temporal artery and vein biopsy was performed.

There were no significant changes on the vein wall, but pathological analysis of the arterial specimen revealed some characteristic findings. The arterial wall was extremely thin in some areas and these regions showed aneurysmal changes and macroscopically - visible severe tortuosity. Because of this tortuosity, some wall segments mimicking a papillary structure were seen in the vessel lumen. On the areas of normal wall thickness, microscopic examination revealed some pathological changes. Intimal thickening, weakness and destruction of the muscular layer of the vessel wall was seen. On the Haematoxyline-Eosine, Mucicarmen and Alcian blue stains of the arterial specimen, severe mucinous degeneration was found on the whole wall layers. Internal elastic membrane fragmentation and duplications; lessening of the elastic and reticular fibres was also seen (Figure 4).

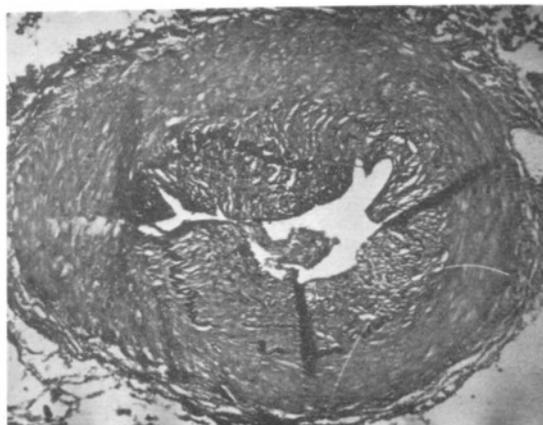


Fig 4 : Fragmentation of the internal elastic membrane; intimal thickening and irregularity are visible with elastic stain.

Ten days after admission, the patient left the hospital and did not come for follow-up.

DISCUSSION

The name "Megadolichobasilar Artery" (MDBA) has been accepted generally in the literature. However we think that this name does not cover the all specifications of this entity. At least in our case, because of the dolichoectasic changes in both carotid systems in addition to the basilar artery; "Cerebral Arterial Dolichoectasia" (CAD) seemed to be more logical.

Gautier et al. reviewed the literature and found 224 reported cases of CAD since 1922 (2). Despite the fact that vascular involvement in this disease is a systemic process, symptoms and signs of this entity are usually due to changes of the vertebrobasilar

blood flow. The most common symptoms and signs of MDBA are of: 1) cranial nerve involvement; 2) brain stem ischaemia; 3) hydrocephalus signs (1,4). All these clinical features can be explained by the elongation, widening and tortuosity of the basilar artery. In the literature we reviewed, in only one study two cases with evidence of carotid system involvement could be found (1).

However, autopsy studies revealed approximately 50 % frequency of co-existing arterial abnormalities with MDBA; especially aneurysm of the abdominal aorta (2). In studies of living cases, such multifocal involvement was very rarely reported. We could find only one case of CAD with abdominal aorta and renal artery aneurysms which resembled our case (2).

Computed tomography is helpful in the diagnosis of CAD. However angiography is a must for definite diagnosis. In the literature, the brachial route is required for cerebral angiography because of the risk factors of hypertension and atherosclerosis and probability of co-existence of other vascular abnormalities (1). Due to the presence of multifocal vascular abnormalities identified by ultrasonography and CT, left brachial artery minipuncture and catheterisation were also performed for the cerebral angiography in our case.

Because of the diffuse histopathological involvement, superficial temporal artery and vein biopsy was preferred. Typical features, especially changes in the structure of elastic elements were found as expected. Mucine - positive staining of the vessel wall and changes of the elastic elements were considered to show similar progress. Since the resistance of the vessel wall to blood pressure is basically dependent

on the elastic elements, dolichoectasia of the vessels is the unavoidable result. In our case, dolichoectastic changes were shown in a single vessel on which there was no evidence of atherosclerosis. Despite the frequent coincidence of atherosclerosis and dolichoectasia, the authors emphasize that the former is not the natural cause of the latter (3).

Our findings and the reported data support the hypothesis of systemic involvement in this entity. However the disease usually appears in the form of vertebrobasilary system involvement and other vascular abnormalities can often only be defined by autopsy. Therefore we conclude that MDBA is the most apparent form of the entity but its prognosis is relatively better than the other forms. In our opinion, superficial temporal artery biopsy with angiography is a safe and reliable method for the diagnosis. In spite of its rarity, the probability of CAD should always be born in mind in the evaluation of middle-aged, hypertensive and atherosclerotic patients.

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