

SHORT COMMUNICATION

Middle cerebral artery dolichoectasia in a young woman with a previous stroke

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We observed a 32-year-old female who had suffered from a left hemisphere ischemic stroke with right hemiparesis at the age of seven. At that time, a CT scan demonstrated a left ischemic lesion in nucleo-capsular region and a cerebral angiogram documented a complete occlusion of the supraclinoid segment of the internal carotid artery. When we observed the patient neurological examination demonstrated a moderate right brachio-crural hemiparesis. A brain MRI showed an old ischemic lesion involving the left nucleo-capsular and 'flow voids' suggestive for a vascular malformation in the left sylvian region. A cerebral rotational angiogram with 3-D reconstructions demonstrated a dolichoectatic left middle cerebral artery with an unusual 'corkscrew' aspect. Middle cerebral artery dolichoectasia is a rare pathological condition that may manifest with a stroke. The patients with intracranial arterial dolichoectasia (IADE) are most often hypertensive elderly men, and, to the best of our knowledge, an ischemic stroke associated with IADE has never been reported in children.

Case report

We observed a 32-year-old female who had suffered from a left hemisphere ischemic stroke with right hemiparesis at the age of seven. At that time, a CT scan demonstrated a left ischemic lesion in nucleo-capsular region and a cerebral angiogram documented a complete occlusion of the supraclinoid segment of the internal carotid artery.

At an office visit, the general practitioner prescribed a brain MRI that showed the old ischemic lesion in the left nucleo-capsular region and 'flow voids' suggestive for a vascular malformation in the left sylvian region (Fig. 1). For this reason, she was referred to our out patient-clinic: neurological examination demonstrated a moderate right brachio-crural hemiparesis. A cerebral rotational angiogram with 3-D reconstructions was obtained with a standard Integris V5000 system (Philips medical Systems, Best, The Netherlands); iodinated contrast agent (320 mg/ml) was injected at a flow rate of 4 ml/s. 3-D subtraction images were produced by the workstation within 8 min. Reconstructed images such as maximum intensity projection, shaded surface display and virtual endoscopy were obtained from the data. This study showed the presence of a dolichoectatic left middle cerebral artery with an unusual 'corkscrew' aspect (Fig. 1); an arterio-venous malformation was definitely excluded. Laboratory tests excluded throm-

bophilic conditions. Extensive cardiac investigation was normal. Normal levels of the alpha-galactosidase enzyme ruled out a Fabry disease, a condition that may be associated with arterial dolichoectasia (IADE) even in heterozygote females [1].

Discussion

The nosological definition of IADE is complex and not well understood; furthermore, the nomenclature regarding dolichoectasia, fusiform or dissecting aneurysms is very confusing.

Intracranial arterial dolichoectasia affects more frequently the vertebro-basilar system and the carotid arteries [2,3]; a more peripheral location involving the anterior and the middle cerebral arteries is less frequent [2,4–6]. The pathophysiology of IADE is unknown; it has been suggested that IADE is an arteriopathy distinct from atherosclerosis [3].

In our patient, that we observed 25 years after the stroke, the absence of any other risk factor suggests that the stroke might be related to the artery abnormality. However, because there was no strict correspondence between the left middle cerebral artery dolichoectasia and the site of arterial occlusion documented by the angiographic study performed in the acute phase of stroke, being the site of the occlusion much more proximal than the site of IADE, it can be speculated that an initial infectious, inflammatory or immunological cause or an artery dissection determined the stroke and that IADE developed subsequently as a consequence of the same artery abnormality. A variety of cerebral artery

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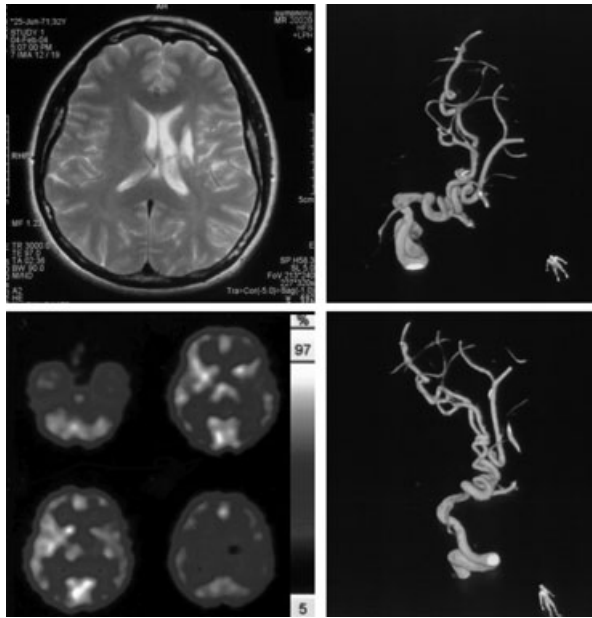


Figure 1 Axial T2 weighted brain MRI (left upper and lower images), and cerebral rotational angiogram with 3-D reconstructions (right). The MRI shows an old ischemic lesion involving the left nucleocapsular region (right upper image) and 'flow voids' (right lower image) suggestive of a vascular malformation. The 3-D angiogram shows the presence of a dolichoectatic left middle cerebral artery with an unusual 'corkscrew' aspect.

abnormalities such as moyamoya syndrome, stenooclusive arteriopathy, arterial dissection or arterial maldevelopment have been reported as a cause of stroke in children [7]. Arterial wall injury may account for up to 85% of arterial strokes in childhood, after the exclusion of congenital heart disease [8]. A monophasic transient cerebral arteriopathy probably because of a vasculitis induced by infections seems to be the most common type of arteriopathy in children with ischemic stroke [8]. A classification system based mainly on clinico-radiological features has been proposed in two recent papers in order to standardize the diagnosis of cerebrovascular ischemic diseases in childhood [7,8]. The frequency of cerebral arterial abnormalities is probably to have been underestimated in the past because many patients have not had cerebrovascular imaging. The availability of brain MRI and magnetic resonance angiography allows for an accurate parenchymal and arterial investigation after an ischemic stroke; conventional angiography is indicated in selected cases.

The role of intracranial arterial dissections as a cause of ischemic stroke in younger patients has been recently discussed [9,10]. Intracranial arterial dissections are less common than extracranial forms, affect younger patients, are associated with larger strokes and can cause dilatations of the surrounding arteries [10,11].

The aetiopathogenesis of spontaneous cervicocerebral dissections is unclear, although minor traumatic events, infections or an underlying arteriopathy has been considered [10]. The process of dissection may be multifactorial and may include congenital and acquired factors, a traumatic injury is the most frequent cause of dissection [8]. Congenital vascular processes such as fibromuscular dysplasia and α 1-antitrypsin deficiency have been associated with intracranial dissections [12]. The theory of an underlying vasculopathy, however, cannot explain the specific site of predilection of the lesions, their low recurrency rate and the rarity of familiar cases [10]. Neurological symptoms can be related to haemodynamic failure, caused by luminal stenosis and thrombus formation or by an artery to artery thromboembolism [10]. A significant association of spontaneous carotid dissections with internal carotid redundancies (coils, kinks and loops) has been described, but a clear causal relation has not been established, although this can support the theory of an underlying arteriopathy [13]. Intracranial arterial dissections may produce spontaneous fusiform intracranial aneurysms [14] and involvement of the middle cerebral artery has been frequently observed in children and adolescents [9,14]. Intracranial arterial dissections are often associated with intramural haemorrhages between elastica and media. Some authors consider a rupture of the vasa vasorum of the media as the initial event in dissection; others postulated that an intimal tear allows the penetration of blood under pressure into the vessel layers [9,10]. Intramural or intraluminal thrombosis or an intimal flap may produce symptomatic stenosis or occlusion with subsequent ischemic episodes. The pulsatile and expansile forces in a growing direction may be dissipated by elongation and dilatation, with gradual enlargement of the dissecting segment; it may expand laterally and longitudinally to create larger focal dilatations [14]. The affected arterial segment can undergo to progressive focal dilatation and eventually evolve into a fusiform aneurysm [14].

In our case, an arterial dissection could have been the cause of the internal carotid occlusion and of the stroke at the age of seven; no further ischemic episodes were reported thereafter. The involvement of the ipsilateral middle cerebral artery with the unusual corkscrew appearance could be explained by the mechanisms proposed by Day and co-workers [14].

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