ly reproduce either the fortification (teichopsic) or the scintillating scotoma seen in classic migraine which characteristically lasts 15–30 minutes and really cannot be differentiated from vertebro-basilar insufficiency. It was this association that led the author to name these attacks “Isolated Ophthalmic Migraine.” Others have referred to these visual episodes as Acephalgic Migraine since they do not have headache associated with them. Fischer recently reported his experience at length of “migrainous accompaniments” including transient attacks of blindness, homonymous hemianopsia and blurring in patients after the age of 40 without headache and with normal cerebral angiograms. He defended the use of the term “migraine” as used by this author. However, to save confusion, these visual attacks would probably be best referred to as Transient Binocular Blindness. Their pathogenesis may be different from the vasoconstriction as defined by Wolff or the shunting mechanism as described by Heyck. Fisher feels they are rarely caused by basilar artery disease.

The following terminology is suggested: 1) transient blurred vision as an overall designation, 2) transient monocular blindness to be divided into — a) amaurosis fugax (seconds to minutes), b) transient monocular blurring, more prolonged unilateral attacks, 3) transient binocular blindness — homonymous attacks of 5–30 minutes duration. The mechanism and the pathogenesis of these symptoms are complex and, in particular, the attacks of transient binocular blindness are poorly understood but if we all refer to them in similar terminology we may have taken a step forward.

References

FIBROMUSCULAR DYSPLASIA (FMD) is a nonatheromatous, segmental stenosing angiopathy. There are reports of involvement of nearly all systemic arteries. FMD, however, is uncommon in the intracranial circulation. Many patients with FMD harbor other vascular or developmental lesions but it has not previously been reported in association with tubular segmental aortic stenosis or coarctation. This report concerns fatal cerebral infarction in a patient with un

SUMMARY Fibromuscular dysplasia (FMD) is an uncommon finding in the cerebral circulation. We present a case of unsuspected intracranial FMD in a patient dying from a large cerebral infarct following a bypass operation for coarctation of the aorta. The need for recognizing the possible co-existence of these two lesions is emphasized.
FIGURE 1. Opened view of the aortic arch showing the cervico-cephalic arteries, coarctation of the aorta (between arrows) and the graft joining the left subclavian artery and distal aorta.

A loud precordial systolic murmur radiated to the left carotid artery. Neurological examination was normal. He had a high arched palate and small testes. No firm diagnosis of a possible congenital syndrome was recognized. Family history was unremarkable.

Admission blood pressure was normal. The electrocardiogram revealed left ventricular hypertrophy, old anterior myocardial damage, and first degree A-V block. Angiography disclosed a 5 cm. segmental narrowing of the aorta just distal to the left subclavian artery. The left common carotid and innominate arteries arose from the same trunk. The aortic "coarctation" was felt to be atypical because of the length of the stenotic segment. The carotid arteries were not included in the angiographic study.

On September 30, he underwent a left subclavian artery to aorta bypass graft. There were no intra-operative problems. Post-operatively, he was awake and following commands. In the 24 hours following surgery, there was continuous bloody drainage from his chest tubes. Hemoglobin fell to 8 grams. Blood pressure fell, briefly, to 90 systolic but there was no sustained period of hypotension. He returned to the operating room where an exploration for bleeding sites proved negative. Ten hours after the second procedure, he was able to follow commands. Six hours later, he had a dense right hemiplegia and a diminished level of consciousness. There was no period of hypotension recorded in the interim. A CT scan of the head revealed a large left hemispheric infarction with shift of the midline structures. He deteriorated rapidly. Forty hours after the second procedure, he was declared neurologically dead. He expired after withdrawal of respiratory support.

At post-mortem a 5 cm. segment of tubular aortic stenosis was seen just distal to the left subclavian artery (fig. 1). The graft was patent and the suture lines clean. There was left ventricular hypertrophy. There were no mural thrombi or valvular vegetations. Two large trunks arose from the aortic arch. The proximal one trifurcated into the right subclavian and right and left common carotid arteries. The distal trunk was the left subclavian artery. There were minimal atherosclerotic changes in both common and internal carotid systems. The supraclinoid portions of both internal carotid arteries were stenosed, the right minimally and the left significantly, to a luminal diameter of only 1.5 mm. The stenosis on the left side extended up to the origin of the anterior cerebral artery. There was a recent large hemorrhagic infarct in the territory of the left middle cerebral artery. No embolus was identified.

Serial histologic sections of the supraclinoid portion of the left internal carotid artery revealed intimal fibromuscular dysplasia (fig. 2). Similar changes were seen in other systemic arteries including the proximal internal carotid, right coronary and superior mesenteric arteries as well as the aortic coarctation (fig. 3). The renal arteries were normal.

FIGURE 2. Cross section through the midportion of the supraclinoid segment of the left internal carotid artery showing marked intimal fibromuscular dysplasia. The intact internal elastic layer (arrow) defines the thickened intima from the normal media. In serial sections, the lumen was further reduced to a diameter of 1.5 mm, as the ridges of fibromuscular tissue bridged across the lumen. There were no thromboemboli or lipid in the intima. Hematoxylin-eosin stain, x 28.
Discussion

Fibromuscular dysplasia is reported in many systemic vessels including the cervical carotid arteries. Reports of intracranial involvement are rare.245 Histological confirmation is even less common.245 FMD has never been reported in association with aortic coarctation or aortic arch anomalies. Our patient also displayed other developmental anomalies but did not represent any specific dysgenetic disorder or syndrome.

The histopathological classification of FMD is based on the locations of the major lesions within the vessel walls.1-9 Intimal, medial and adventitial forms are described. Many patients display a spectrum of lesions with a mixed histological pattern. The intimal lesions in our case represent one of the less common forms of FMD. Most cases are of the medial type. The adventitial variety is unusual but was present in the superior mesenteric artery of our case.

This patient suffered a hemorrhagic cerebral infarction. Most of these are secondary to embolization. Given the severity of the stenosis in the supraclinoid portion of the left internal carotid even a small embolus could produce complete occlusion. However, there was no identifiable source of emboli in our patient. Alternatively, a brief period of relative hypotension or altered hemodynamic state subsequent to a bypass could produce a "low flow" state with subsequent infarction in the region of the tightly stenosed vessel.

This case illustrates the unusual occurrence of intracranial FMD in association with a "coarctation" of the aorta. The latter was atypical in that it involved a longer segment of the aorta than the usual localized stenosis at the level of the ductus arteriosus. The patient also had other minor physical anomalies, including a high arched palate and small testes, and mental retardation. All these features suggest a generalized dysgenetic disorder. The aortic coarctation in this patient appears to have been part of a more widespread dysgenetic vascular disorder such as FMD. Surgical procedures are often indicated to correct symptomatic vascular lesions like aortic coarctation. In view of this possible association between aortic coarctation and FMD, particularly when the coarctation is atypical, pre-operative investigations should include cerebral angiography to rule out the co-existence of a potentially serious cerebrovascular lesion.

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