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Coarctation of the Thoracic Aorta Associated with Cerebral Arterial Occlusive Disease

Harold A. Baltaxe,^{1, 2} Solomon Bloch,¹ and Paul K. Mooring³

Since coarctation of the aorta was first described by Johann Friederich Meckel in 1760 [1], a vast literature has appeared on its associated conditions and treatment. This anomaly of the descending thoracic aorta is usually located distal to the left subclavian artery and is commonly found in coexistence with intra- and extracardiac pathology. Ventricular and/or atrial septal defects [2], as well as discrete subvalvular stenosis, represent the accompanying intracardiac conditions [3, 4]. The presence of a bicuspid aortic valve was first noted by Edwards [5] and is found in 85% of patients afflicted by coarctation of the aorta [6]. Supravalvular stenosis is only rarely an associated anomaly [7], whereas the presence of patent ductus arteriosus is rather common [8]. Dilatation of the ascending aorta and aneurysms of the sinus of Valsalva combined with coarctation have also been reported [9]. Whenever the patient does not suffer from Turner syndrome [10], anomalies involving organs other than the heart and the thoracic aorta are rarely seen in coarctation. However, aneurysms of the circle of Willis were noted by Hodes et al. [11] and are the cause of cerebral hemorrhage, which is a major complication in the natural history of coarctation. Coarctation of the aorta in association with cerebral arterial occlusive disease, to our knowledge, has not been reported previously. We report two patients with such a combination. The question to be considered is whether this represents a fortuitous association or whether both entities have a common etiology.

Case Reports

Case 1

A 4-year-old boy was admitted 2 weeks after acute onset of a left hemiparesis. His father found him lying on the bathroom floor with what seemed to be choreoathetotic movements on the right, while the left side of his body was limp. He was alert and cooperative with left hemiparesis and left facial paralysis. A grade III/VI harsh systolic murmur heard at the upper left sternal border was transmitted to the back and to both carotid arteries. The pulses in the legs were

extremely poor and no blood pressure could be obtained in them. Blood pressure in his arms was 122/58 mm Hg.

Laboratory findings were noncontributory. Chest radiography revealed left ventricular prominence and a notch-shaped left contour of the descending aorta suggestive of coarctation. Electrocardiography revealed left ventricular and left atrial hypertrophy. Heart catheterization and angiocardiology revealed a 90% coarctation of the aorta distal to the left subclavian artery with poststenotic dilatation of the descending thoracic aorta. The vessels to the head appeared anatomically normal in their takeoff from the arch.

Selective cerebral arteriography was performed. Injection of the right common carotid artery demonstrated a normal extracranial course; intracranially, multiple discrete stenoses were noted mainly in the horizontal part of the right middle cerebral artery and its angular branch (figs. 1A-1C). The appearance of these stenoses was consistent with that of fibromuscular dysplasia or some other form of vasculopathy. A left carotid arteriogram with cross compression of the right common carotid artery revealed a normal right anterior cerebral artery and normal left carotid branches.

Computed tomography (CT) of the head demonstrated an area of increased uptake of contrast material in the right hemisphere that was consistent with an ischemic infarct. A brain scan supported this diagnosis.

A muscle biopsy of the gastrocnemius muscle revealed no abnormalities. The coarctation of the aorta was repaired leaving a 25 mm Hg residual gradient. Histologic examination of aortic tissue was not made.

The patient underwent reassessment of his cerebral status 5 years later. Cerebral angiography revealed an occlusion of the right middle cerebral artery at its origin. Its branches filled distally in a retrograde fashion mainly from the right posterior cerebral artery (figs. 1E and 1F). CT showed a well defined area of encephalomalacia on the right side in the middle cerebral artery distribution consistent with an old infarct (fig. 1D).

Case 2

A 2-year-old boy underwent cardiac catheterization and angiography, revealing coarctation of the aorta distal to the left subclavian artery. At age 5 years, he sustained a cerebrovascular accident resulting in left hemiparesis. He underwent surgical correction of his coarctation 1 month later. The procedure had to be repeated at

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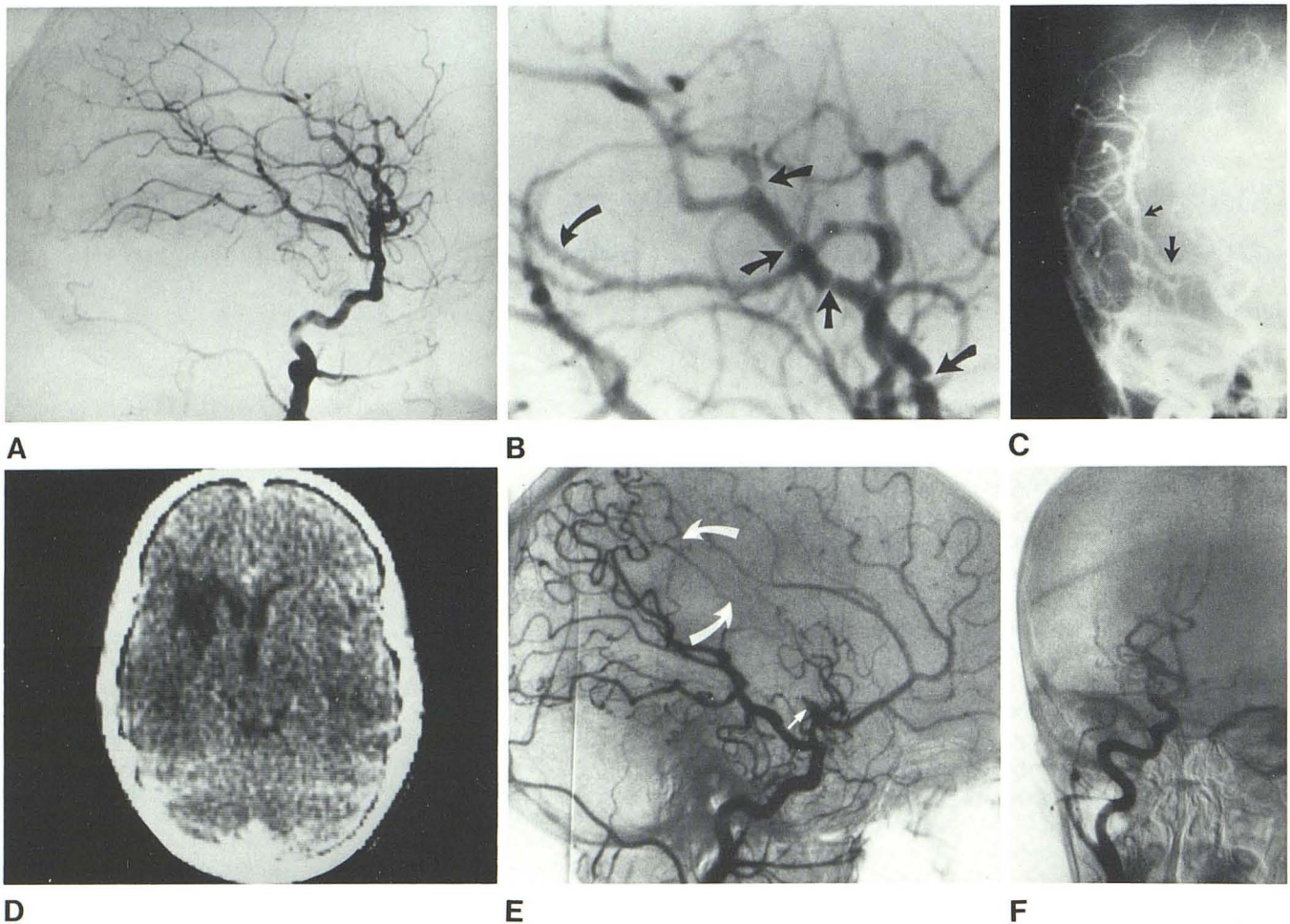


Fig. 1.—Case 1. **A**, Right lateral common carotid arteriogram. Multiple discrete linear filling defects with mild poststenotic dilations in angular branch of middle cerebral artery. Anterior cerebral artery not visible. **B**, Local magnified view. Angular branch with multiple defects (arrows). **C**, AP right carotid angiogram. Multiple discrete linear filling defects in middle cerebral artery (arrows). **D**, 5 years later. CT scan with intravenous contrast enhancement demonstrates old right frontal infarct. **E**, Right lateral common carotid

angiogram. Almost complete obstruction at origin of the middle cerebral artery (straight arrow). Distally there is early retrograde filling of distal middle cerebral artery branch via anterior and posterior cerebral arteries (curved arrows). **F**, AP right carotid angiogram. Almost complete block at right middle cerebral artery origin with opacification only of ascending frontoparietal branch.

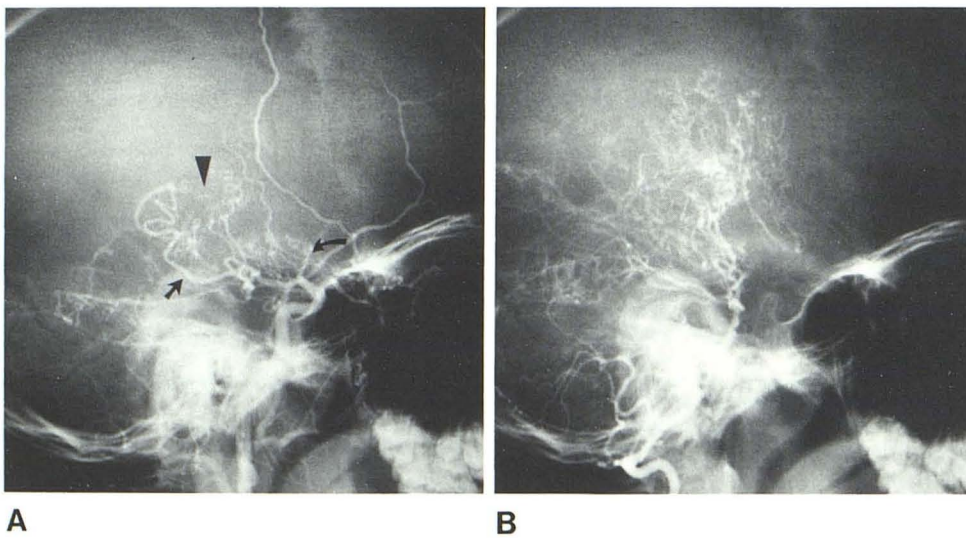


Fig. 2.—Case 2. **A**, Left lateral common carotid angiogram. Almost complete occlusion at supraclinoid part of internal carotid artery (arrowhead). Medial posterior choroidal artery (straight arrow) and perforating arteries of posterior cerebral artery show marked dilatation (curved arrow). **B**, Right vertebral angiogram. Prominent perforating vessels within basal ganglia consistent with Moya Moya syndrome.

age 9 years due to restenosis. Aortic tissue was not examined histologically. In the interim he apparently sustained one or two more cerebrovascular accidents.

At age 9 years the residual gradient across the site of his previously repaired coarctation was 30 mm Hg. Blood pressure in both arms was normal. A left common carotid angiogram revealed an almost complete occlusion at the level of the supraclinoid part of the internal carotid artery with dilatation of the perforating arterial supply to the basal ganglia (fig. 2A). A right vertebral angiogram revealed marked dilatation of the thalamoperforating arteries to the basal ganglia producing an appearance consistent with a Moya Moya syndrome (fig. 2B).

At age 14 years, he still had a left hemiparesis and appeared somewhat mentally diminished. There appeared to be no progression of the neurologic deficit.

Discussion

Whenever several pathologic conditions coexist, it is important to determine whether their association is fortuitous or whether they have a common etiology. Because coarctation of the aorta is a congenital disease, an initial assumption would be that the intracranial lesions in our patients are due to an inherent rather than an acquired defect. Nevertheless, the possibility that the cerebral arterial occlusions were secondary to the hypertension caused by coarctation must be considered also.

The exact etiology of coarctation of the aorta is unknown. In 1841, Craigie (cited in [2]) first proposed the "skodiac theory," which stipulates that the aortic narrowing is secondary to the closure of the ductus. Coarctation of the aorta, however, is often associated with a patent ductus, and, therefore, this theory seems to be invalid. In 1965, Brom [12] suggested that the tissue encircling the aorta at the site of the coarctation was similar to that of ductal tissue, and he also tried to relate coarctation of the aorta to the presence of the ductus.

Classically, coarctation is not a concentric narrowing, but primarily a posterolateral narrowing. Therefore, Edwards et al. [13] believed that this eccentric thickening was related to a developmental change of the intima. More recently Kennedy et al. [14] studied the intima in coarctation of the aorta by using light and scanning electron microscopy. They found that the intima immediately proximal to the stenosis ran in fine longitudinal folds radiating into the orifice. The intima on the distal side was grossly irregular with deep longitudinal and transverse folds producing a convoluted appearance resembling the surface of the brain. The intima immediately distal to the orifice was covered with a laminated layer distinct from the elastica, which often gave positive staining reactions for fibrin. Intimal thickness seemed to increase with age. These authors concluded that their findings were consistent with the view that the constriction had a fibroelastic component that is congenital and a fibrous part that is acquired and progressive. They believed that the progressive element is caused by deposition of fibrin on an abnormal intimal surface in a region of turbulent flow. This congenital abnormality may be similar to the mesodermal dysplasia seen in neurofibromatosis. Coarctation of the aorta and renal artery stenosis producing arterial

hypertension have been reported in neurofibromatosis [15]. The renal artery stenosis in neurofibromatosis is caused by the mesenchymal abnormality. Cerebrovascular occlusive disease has also been described in neurofibromatosis [16]. Levisohn et al. [17] reported two such cases with brain infarction diagnosed by CT, caused by occlusive intracranial disease with resulting extensive collateralization. Angiographically, the vessels presented with segmental constrictions and areas of ectasia, similar to the cerebral arteries of our patients. Neurofibromatosis, however, is a hereditary disorder with maldevelopment of the neural ectodermal tissue. Nothing suggested that our patients were afflicted by such a condition. However, it is possible that the abnormality that affects the fibroelastic tissue in coarctation may sometimes also involve the intracerebral vessels.

Coarctation of the aorta has been reported in association with aneurysms of the cerebral vessels [11]. The mechanism for the formation of these aneurysms has never been elucidated. Long-standing hypertension restricted to the upper body as seen in coarctation may produce changes in the cerebral vessels that eventually may result in the formation of aneurysms. Could hypertension have produced occlusive vascular disease in our patients? Even if long-standing hypertension would have been present, the cerebrovascular changes noted in our patients could probably not have taken place without an underlying disposition.

These two cases cannot be dismissed without suggesting the possibility of a vasculitis. Ferris and Levine [18] classified cerebral arteritides according to their etiology. Bacterial, viral, and collagen diseases must be considered when faced with intracranial vascular changes in children. Collagen diseases were excluded in our two cases. Harwood-Nash et al. [19] reviewed the cerebral arterial diseases in 40 children. None of them were associated with coarctation of the aorta and most seemed to be the sequelae of an inflammatory process resulting from a nasopharyngeal infection. No such infections were reported in our two patients.

Lastly, Slagsvold et al. [20] reported the case of 32-year-old woman in whom the carotid angiogram was consistent with fibromuscular dysplasia of the left anterior and middle cerebral arteries. Until then, it was commonly believed that fibromuscular dysplasia did not occur in cerebral vessels. Although this concept has been disproven, fibromuscular dysplasia is not known to be associated with coarctation of the aorta.

Analysis of our two cases does not permit us to suggest a single etiology for these findings. Coexistence of coarctation of the aorta and cerebral arterial occlusive disease could be fortuitous, however, our purpose was to speculate that such an association *may* not occur entirely at random.

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