Middle aortic syndrome

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A 9-year-old male patient presented for the first time with a 3-day history of headache, dizziness and vomiting. This was followed by three episodes of tonic clonic seizures that involved the right side of the body. On clinical examination his blood pressure was found to be 180/120 mmHg, pulse 86/min in the upper limbs with a very weak femoral pulse and a bruit over the abdomen. There was no history of febrile illness. No murmurs were detected on cardiac examination. Echocardiography showed a moderate aortic regurgitation and a dilated aortic root. No coarctation was detected in its classic location. No valvular stenosis was seen. There were no clinical or biochemical features of Williams or Allagile syndrome. Renal and endocrine functions were normal. The erythrocyte sedimentation rate (ESR) was 13 mm/hr, and C-reactive protein was markedly elevated (25.8 mg/l). Antinuclear antibodies were negative and there was no peripheral eosinophilia. Computed tomography (CT) scan of the brain showed infarction in the territory of the left posterior cerebral artery. An angiogram was performed, using the Seldinger technique, through the right femoral artery. Digital subtraction radiography was used. There was coarctation of the abdominal aorta beginning just below the renal arteries, with involvement of the left renal artery (Fig. 1). There was enlargement and elongation of the superior mesenteric artery, which supplied enough blood to the lower extremities to provide palpable femoral pulses (Fig. 2). There were extensive collateral vessels noted in the mesenteric circulation. The aortic arch and the branches did not show any signs of stenosis (Fig. 3). An incidental origin of the left common carotid artery from the right brachiocephalic trunk was observed. The fact that there was no evidence of stenosis of the take-off vessels in the aortic arch suggested a congenital cause for the coarctation.

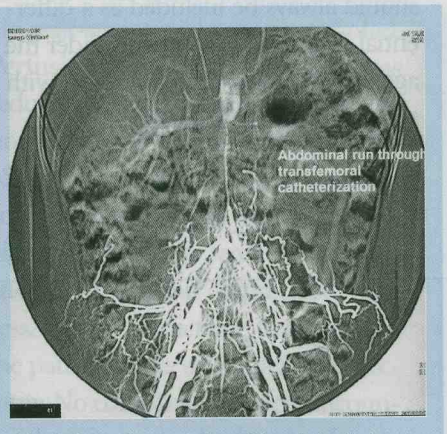

Fig. 1. Smooth coarctation of the abdominal aorta, with involvement of the left renal artery.

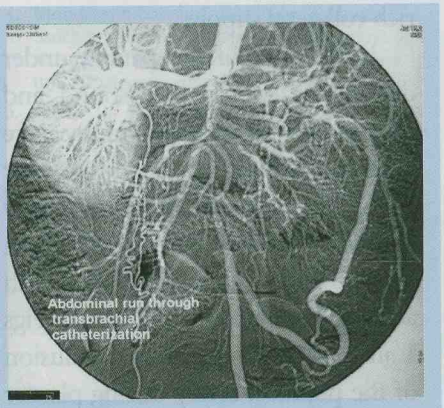

Fig. 2. Enlarged and elongated superior mesenteric artery. Note also involvement of the left renal artery.

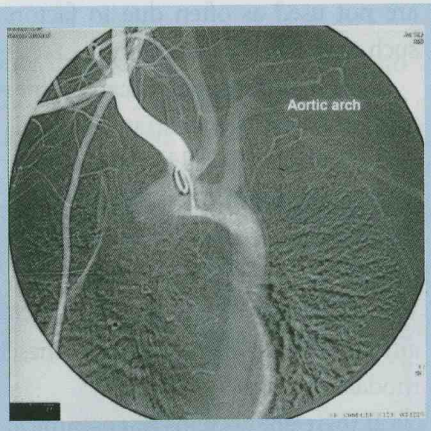

Fig. 3. The aortic arch and branches are normal with no signs of stenosis.

Discussion

Abdominal coarctation is an unusual form of coarctation characterised by segmental stenosis of the aorta and its branches. It can cause severe hypertension with its attendant risk of life-threatening complications, namely brain infarction, as in our patient. Due to disagreement regarding the aetiology of the abdominal coarctation and the fact that the name coarctation implies congenital origin, some authors prefer a more neutral name such as 'middle aortic syndrome' (MAS). The spectrum of MAS involves narrowing of the abdominal aorta and progressive involvement of the renal and visceral branches. MAS

was first described in 1963 by Sen *et al.* It accounts for only 2% of aortic coarctations and is an important cause of hypertension in children and young adults, and is associated with a high morbidity and mortality. It is thought to be the result of a primary underdeveloped aortic segment that commonly also affects the renal arteries. The histology of the affected segment shows marked sub-endothelial fibroplasias with no evidence of acute or chronic inflammation. It is thought to be the result of overfusion of the two embryonic dorsal aortas or their failure to fuse, with subsequent obliteration of one of these vessels.

The radiological differential diagnosis includes other causes of renovascular hypertension in children such as Takayasu's arteritis, fibromuscular dysplasia, neurofibromatosis, mucopolysaccharidoses, Williams syndrome, Allagile syndrome and congenital rubella syndrome. The aforementioned syndromes and genetic diseases have other specific symptoms and clinical findings, which were absent in our patient; hence the differential diagnosis was limited to Takayasu's arteritis and FMD. Takayasus's arteritis affects predominantly females and is frequently associated with systemic symptoms similar to collagen vascular disease including arthritis, myalgia, pleuritis, pericarditis, fever and rash, none of which was present in our patient. Also in its classic form it is confined to the aortic arch and its branches. The fact that there was no evidence of stenosis of the take-off vessels in the aortic arch in our patient suggested a congenital aortic origin. FMD is a congenital disorder of the connective tissue in the blood vessels and is a pathological diagnosis. There is marked intimal fibrosis and adventitial infiltrate and scarring in Takayasu's arteritis histologically, whereas in FMD there is lack of inflammation in all layers of the vessel wall and severe narrowing of the aorta and the renal arteries by intimal and medial fibroplasias. We could not obtain histological examination in our case, and because of lack of other features observed in Takayasu's arteritis, we think that abdominal coarctation in our case may result from congenital aortic dysplasia. Aortography is the only investigation diagnostic for MAS. It should be performed in all severely hypertensive patients in whom the diagnosis has not been established by initial renal imaging. Meticulous blood pressure control must be maintained during and after arteriography as this may be complicated by a hypertensive crisis.

In most patients the treatment of choice is medical control of blood pressure with a combination of antihypertensives. Renal function should be closely monitored in all patients receiving an angiotensin-converting enzyme inhibitor. The indications for surgery in MAS have not been established. Surgical mortality is high due to the extensive nature of the vascular involvement. Surgery may be limited to those patients in whom medical control of hypertension cannot be achieved and in patients with claudication or both. If surgery is to be performed then balloon angioplasty or auto transplantation for renal artery stenosis and stent implantation or bypass graft for stenosed aortic segments are the suggested therapeutic options in the treatment of MAS.

Bibliography

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