Successful Treatment of Internal Carotid Artery Fibromuscular Dysplasia by Percutaneous Transluminal Angioplasty

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Case Report

A 35-year-old Saudi male was admitted in July 1991, following a prolonged reversible ischaemic neurological deficit (PRIND), in the form of a 2-day history of sudden severe headache and nausea associated with a decrease in vision of the right eye. The patient suffered a transient ischaemic attack 3 weeks prior to that attack.

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Figure 1. Intraarterial digital subtraction angiogram image with close-up view of the right carotid bifurcation, demonstrates severe stenosis at the proximal segment of the right internal carotid artery.
On examination the patient was not hypertensive and had no history of diabetes mellitus. There was decreased sensation over the right side of the face with minimal right-sided weakness, and normal reflexes.

Laboratory investigations revealed: cholesterol 6.7 mmol/l, triglyceride 1.7 mmol/l, PT and PTT were normal. ECG was normal. The EEG showed moderate-to-mild decrease of the slow wave, with abnormality in the right temporal area extending to the right frontal and parietal areas, indicating a significant neurophysiological disturbance. There was no evidence of an epileptic focus. He was discharged 3 weeks later.

The patient was readmitted 3 months later following a stroke attack. He exhibited a left VIth nerve palsy and a mild left hemiparesis. A lipid profile was performed: cholesterol 7.4 mmol/l; HDL cholesterol 0.64 mmol/l; LDL cholesterol 5.86 mmol/l; triglyceride 1.28 mmol/l. A CT scan demonstrated two areas of differing low density, in close proximity, within the right temporal lobe, in the area supplied by the right middle cerebral artery. The differing densities of the areas was indicative of an old infarction in the area of lowest density. A carotid Doppler ultrasound examination of the right internal carotid artery (ICA) showed an abnormal Doppler wave form suggestive of severe stenosis at the origin of the right ICA. Digital subtraction angiography of the internal carotid artery (Fig. 1) revealed a significant narrowing (80%) of the right ICA, confirming the result of the Doppler examination.

Therefore, it was decided to treat the patient by transluminal angioplasty. On 20/10/91, the patient was sedated and taken to the angiography room, where he received 5000 units of heparin intravenously and a local anaesthetic. A 6F sheath was introduced through the right femoral artery, and a 0.035 floppy guide wire was introduced, under road mapping, to traverse the stenotic segment. Once the stenotic segment had been crossed, the 5F, 3 mm balloon 2 cm long was inflated to a pressure of 6 atmospheres, for 3–5 s, a total of five times (Fig. 2). Digital subtraction angiography was performed following the procedure, and demonstrated almost complete restoration of the lumen of the ICA. The patient was returned to the ward for close neurological observation. He was fully heparinized with 1000 IU per hour in an i.v. drip for 3 days, and then commenced on dipyridamole (Persantin) 75 mg p.o. t.i.d. for a period of 3 months. The immediate postoperative course was without complication and the patient repeatedly stated that he felt better. Follow-up at 3 months showed the patient to have no neurological deterioration, and repeat Doppler examination at that time, demonstrated marked improvement, in terms of normal biphasic waveform with a peak systolic velocity of 50 cm/s. The patient is presently enjoying a normal lifestyle. A further follow-up, intravenous digital subtraction angiogram of the neck vessels, at 5 months, showed a normal right ICA (Fig. 3).

Discussion

For a long time following the description of fibromuscular dysplasia of the internal carotid
artery, in 1965, by Connett & Lansche, it was considered to be an incidental angiographic diagnosis, with a benign course. However, with recent advances in diagnostic procedures, this opinion has changed, and it is now known that it may have a progressive course and present as either a permanent, or temporary marked clinical neurological deficit. Other than the renal artery, the carotid artery is the most common arterial site for the disease. Fibromuscular dysplasia is also the second most frequent cause of extra cranial narrowing of the ICA. It is more commonly encountered in females, and usually occurs in the 3rd or 4th decades of life. Fibromuscular dysplasia has been classified according to the angiographic patterns in association with histological findings, into the following types:

I Intimal fibroplasia.
II Medial fibroplasia with aneurysms.
III Medial fibromuscular hyperplasia.
IV Subadventitial fibroplasia.
V Adventitial periarterial fibroplasia.

The most commonly occurring type (80–85%) is a lesion looking like a string of beads on angiography, which histologically corresponds to Type II or III. In 6–12% of the cases of fibromuscular dysplasia the angiographic appearance is a long segment of tubular stenosis, but in 4–6% of the cases the angiographic appearance shows ovoid outpouchings with non-circumferential web-like narrowings. These two types may be caused by any histological type of the fibromuscular dysplasia.

There are various modalities of treatment available for such cases, and each case should be individually considered, taking into account the extent of the lesion, whether it affects both carotid arteries, and whether it is associated with aneurysm, neoplasm or fibromuscular dysplasia of other arteries. The age and neurological condition of the patient should also play a role in the choice of suitable treatment. Treatment in mild cases, can be conservative, in the form of antiplatelet drugs, provided that the case is followed up closely as the disease is known to be progressive. Surgical treatment in the form of resection or carotid endarterectomy, is a possible form of treatment, where others fail. However, we feel that transluminal angioplasty should be the method of choice especially in young patients with a complete circle of Willis (as our patient), since it carries relatively low risks and has repeatedly been reported as successful. The risk of this method has been reported to be one of intimal dissection, which can be treated medically. In general, intraoperative angioplasty under direct surgical exposure of the distal common carotid artery and its branches is probably the treatment of choice. The direct vision afforded allows clamping of the distal common carotid artery and proximal external carotid arteries to allow back bleeding from the internal carotid artery after expansion of the balloon.

Follow-up monitoring of the patient after angioplasty is ideally performed by Doppler ultrasound examination which is a non-invasive accurate technique as compared to DSA.

References