Spontaneous Dissecting Aneurysms Of The Internal Carotid And Vertebral Arteries — Two Case Reports

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SUMMARY Two patients had acute spontaneous dissection of both internal carotid arteries and of one or both vertebral arteries. One had angiographic signs suggestive of fibro-muscular dysplasia and both were on oral contraceptives. They were treated with high dose heparin and made a good clinical recovery. A digital intravenous angiography performed two to three months later showed a complete recanalization of arteries involved. These patients are similar to those reported as “idiopathic regressing arteriopathy” and “reversible angiopathy” which probably correspond to the same entity.

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UNILATERAL SPONTANEOUS DISSECTION of the internal carotid artery (ICA) or of the vertebral artery (VA) was considered to be a rare entity a few years ago, but has recently become either less unusual or more easily recognized. However bilateral dissection of internal carotid and/or vertebral arteries remains most unusual. We report two patients with spontaneous acute dissection of at least three major cervical arteries.

Report of Cases

Patient 1

A 40 year old woman was admitted to La Salpêtrière on February 5, 1982, two weeks after the acute onset of headache, neck pain, vomiting and of a bruit in the left ear. She was seen by her local physician who found a normal neurological examination, but a high blood pressure: 190/120 mmHg. There was no history of trauma to the head or neck and her past medical and family history were not contributory. She had been on contraceptive pills for the last six years and she was advised to stop them. Over the next days, the bruit and headache increased in severity and developed over the whole of the head. Ten days after the onset of symptoms, she had a brief loss of consciousness and four days later she had a sudden episode of bilateral blurring of vision, which lasted 15 minutes. Clinical examination was normal except for the presence of a left Horner’s syndrome and a loud bruit over the left side of the head and neck. Blood pressure was 135/95 mmHg. On admission the next day, clinical findings were similar. Routine laboratory investigations and detailed coagulation studies were normal. A CT scan with and without contrast injection was normal. A percutaneous transfemoral selective bilateral carotid and subclavian angiography showed abnormalities on all 4 vessels. On the right ICA (fig. 1-A), there was a severe extensive narrowing beginning just past the origin and extending up to the siphon which was filled retrogradely by the external carotid artery via the ophthalmic artery. On the left ICA (fig. 1-B), there were slight irregularities at the origin, and distal to it, a severe narrowing extending up to the entrance into the carotid canal, with an aneurysmal dilatation at the C3 level. On the left VA (fig. 1-C), there was, distal to its origin, a moderate irregular narrowing extending up to the C6 level with two aneurysmal dilatations, one at the C6-C7 level and the other on its extracranial curve. The right VA (fig. 1-D) was hypoplastic and, in addition, showed a severe regular eccentric stenosis at the C2 level. The left VA filled both middle cerebral arteries. The intracranial circulation was otherwise normal. The abdominal aorta and renal arteries were normal. High dose Heparin calcium was started on Feb. 11. Over a period of two weeks, there was a steady improvement with a gradual...
disappearance of the bruit and headache. She was discharged on oral anticoagulants and was examined monthly afterwards. She had no further symptoms; blood pressure was always normal and the only anomaly is a left Horner's syndrome. A digital intravenous angiography (DIVA) was performed 3 months later (fig. 1-E); it showed a complete recanalization of the right ICA, right VA and left ICA, and the persistence of a sacciform aneurysm of the left ICA at C1 level. The left VA was not properly visualised but continuous wave Doppler showed normal velocity curves of both VA suggesting the absence of hemodynamically significant stenosis.

**Patient 2**

On February 20, 1982, a previously healthy 42 year old right handed woman experienced a 10 minute episode of bilateral blurring of vision. A similar episode occurred the next day and was accompanied by a severe neck pain which worsened during the following hours and lasted a week. On Feb. 28, she suddenly heard a pulsatile bruit in the right ear and developed weakness, numbness and tingling in the left leg, extending in a few hours over the whole left side of her body. The weakness worsened over the next three days, and then gradually improved. Fifteen days after the onset of symptoms, she was seen by one of us (J.M.V.) and admitted to her local hospital. Clinical examination was normal except for a slight impairment of joint position sense on the left side. A transfemoral aortography and cerebral angiography was performed on March 11: it showed a severe, irregular narrowing of the right ICA extending from the C2 level up to the entrance into the carotid canal with an aneurysm at C1 level (fig. 2-A). The left ICA was not selectively catheterized but looked extremely irregular on global aortography. The right V.A. was normal but the left V.A. showed a tight stenosis at C2 level (fig. 2-B). The intracranial circulation was otherwise normal. The extracranial portion of the right renal artery showed a narrowing with a "string of beads" appearance. On the day after angiography, she suddenly developed a speech impairment. High dose heparin sodium was immediately started and she was transferred to our hospital.

On admission (March 12), she had a moderate dysphasias involving speech, reading and writing functions. Neurological examination was otherwise normal. There was no cervical or cranial bruit. Blood pressure was 140/80 mmHg. There was no past history of head or neck trauma or of vascular disease. Contraceptive pills had been taken for the last ten years. Routine laboratory investigations and detailed coagulation studies were normal. On March 19, an ultrasound Doppler study showed a decrease in flow over the right ophthalmic artery but was otherwise normal. On the same day, a CT scan showed a small area of decreased density with contrast enhancement in the left temporal lobe, suggestive of a small infarct. The patient improved gradually; she was put on oral anticoagulants and discharged a week later. She was examined monthly afterwards. She had no further symptoms but has still some mild speech impairment. A DIVA was performed 2 months after the onset of symptoms; it showed a recanalization of carotid and vertebral arteries but the aneurysmal dilatation of the right ICA at C1 level was still visible (fig 2-C).

**Discussion**

Clinical manifestations and angiographic signs of spontaneous dissections of cervical arteries need not to be detailed since they are well documented. Clinical manifestations usually associate "local" signs such as headache, bruits and Horner's syndrome with signs of cerebral ischemia referring to the territory of the artery involved. These signs can be transient or permanent, such as in our patient 2, who had first two vertebrabasilar TIA's, then one right carotid reversible deficit and, after angiography, a left carotid completed stroke with permanent dysphasia. The diagnosis of dissection lies upon angiography. In both patients, signs of dissection were typical on at least one V.A. and on the two I.C.A. with long irregular filling defects ('the string sign'), thus eliminating other entities such as spasm, atherosclerosis and arteritis. The almost total disappearance of these anomalies in a few months is also very suggestive of dissection. Follow up angiography is therefore crucial in these patients. DIVA seems to this respect an excellent non invasive and easily repeatable investigation. It showed in the present patients a complete recanalization of carotid and vertebral arteries.

The spontaneous occurrence of a dissection is always difficult to assess with certainty. A variety of circumstances associated with abrupt change in head position, such as chiropractic manipulation, gymnastic exercises or sports activity or even "head banging", can lead to hyperextension or rotational injury to the neck and induce the formation of an arterial dissection. In the mind of the patient, these events are considered to be non traumatic and therefore are not spontaneously reported. In our cases, all these possible traumatisms, even minor ones, have been specifically looked for and denied by the patient.

Spontaneous multiple dissection of the major cervical arteries is very unusual. Among the reported cases, diagnosis rested either on pathological examination or on angiographic changes. Our two cases are clinically and angiographically

**Figure 1.** Patient 1A — Right ICA: right stenosis with pseudo-occlusion extending from 1 cm above the origin up to the siphon ($\rightarrow$). B — Left ICA: severe, irregular stenosis at C2-C1 level (•) with aneurysmal dilatation ($\rightarrow$) just below the entrance into the carotid canal. C — Left VA: Irregular stenosis from C6 to C3 with aneurysmal dilatation at C3-C2 level (•) and saccular aneurysm at C1 level ($\rightarrow$). D — Right VA: severe eccentric narrowing at C1 level (•). E — DIVA (3 months later): recanalization of the right ICA ($\rightarrow$); right VA (•) and left ICA with persistence of an aneurysmal dilatation at C1 level ($\rightarrow$).
similar to those reported as “idiopathic regressing arteriopathy,” “spontaneous bilateral recanalization in bilateral carotid artery occlusion,” or “reversible angiopathy.” It is highly probable that all these eponyms correspond to acute multiple spontaneous dissections of cervical arteries. The presence of multiple arterial involvement suggests the presence of an underlying arterial disease that could lead to weakness of the vessel wall and formation of an arterial dissection. In patient 2, the “string of beads” appearance of the right renal artery was that described in fibromuscular dysplasia (FMD). It is therefore possible that FMD was also present in cervical arteries. The association between spontaneous dissection and FMD is well documented. It has been suggested that minor injuries to the neck could be sufficient to cause dissections of arteries involved by FMD, and that elevated blood pressure due to renal FMD could be a contributing factor. Migraine has been suggested as another etiologic factor but our patients denied any history of migraine. Oral contraceptives which are known to induce intimal hyperplasia and other changes in the arterial wall have also been incriminated and it is of interest that our two patients were on oral contraceptives when the first symptoms occurred. Whatever the underlying disease, it should be stressed that the relationship between these diseases and spontaneous dissections on one hand, and the mechanism by which all four arteries are simultaneously affected on the other, remain totally unclear.

The efficacy of any treatment in a disease which is often spontaneously reversible is extremely difficult to establish. Surgical treatment has been advocated, particularly in case of progressive or repetitive neurological deficits. Anticoagulants or antiplatelet drugs have been widely used in order to prevent thrombosis of the lumen or embolization of a mural thrombus. High dose heparin was used in the two present patients because of these observations and also based on our personal experience of non surgical treatment. In a series of 24 patients with spontaneous ICA dissection followed up by angiographic (18 cases) or ultrasound studies (6 cases), recanalization was observed in 16. Of these, 10 were treated by anticoagulants, 5 by aspirin and one had no treatment. By contrast, among the 8 patients who had no recanalization,
only 2 were on anticoagulants, 2 on aspirin and 4 had no treatment. Although this was not a randomized study, it suggests a slightly better prognosis in patients treated by anticoagulants or aspirin than in patients with no treatment.

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