CASE REPORT

Headache, TIA and subarachnoid haemorrhage: dissecting an unusual cause for stroke-like symptoms

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SUMMARY

We present a case of supraclinoid internal carotid artery dissection. Eleven months prior, the patient developed isolated periorbital pain and was diagnosed with giant-cell arteritis with iritis. The patient experienced recurrent spells concerning for transient ischaemic attacks and was transferred to our institution for endovascular intervention after head CT revealed an embolic infarct with a 'dense middle cerebral artery sign.' Digital subtraction angiography was negative for occlusion, instead demonstrating luminal stenosis and poststenotic dilatation. He subsequently experienced acute neurological decline secondary to massive subarachnoid haemorrhage. Non-invasive vascular imaging revealed an intimal flap and a pseudoaneurysm at the site of luminal stenosis, confirming our suspicion for intracranial carotid artery dissection. Given the moribund clinical state, the family opted to withdraw care, and he quickly expired.

BACKGROUND

Intracranial arterial dissection is a rare cause of ischaemic strokes overall.¹ It is more common in young adults. We believe our case highlights radiographic features of dissection on head CT (HCT) which may not be appreciated. The dense middle cerebral artery (MCA) sign has known false positives.^{2–3} Correct diagnosis of intracranial dissection may be elusive, as misdiagnosis has occurred despite CT angiography (CTA) and digital subtraction angiography (DSA).⁴ Intracranial dissection can manifest with recurrent ischaemic symptoms and can also produce subarachnoid haemorrhage (SAH) from arterial wall rupture.

The differential diagnosis for recurrent ischaemic symptoms includes cardioembolic phenomena, severe arterial stenosis and when visual symptoms and headache are present, giant-cell arteritis (GCA). This elderly man with multiple risk factors for cardioembolic disease presented with symptoms consistent with giant-cell arteritis for which he was treated without clinical improvement for 11 months. Initially, the diagnosis of intracranial dissection was not entertained as the patient did not meet the epidemiological characteristic. This case illustrates the necessity of keeping this rare entity on the differential diagnosis for both unexplained headache and acute stroke as the clinical ramifications of a missed intracranial dissection are catastrophic.

CASE PRESENTATION

The patient was a 77-year-old man who transferred to our institution for definitive management of an

acute ischaemic stroke. His relevant medical history included a prior left parietal lobe infarction, atrial fibrillation, complete heart block status postpermanent pacemaker implantation, peripheral artery disease, untreated obstructive sleep apnoea, hypertension, hyperlipidaemia and multiple myeloma. His CHA₂DS₂-VASc score was 6 indicating a high annual risk of ischaemic stroke. He was on Warfarin with an International Normalised Ratio goal of 2–3 as well as low-dose aspirin. He lived with his wife in an assisted care living facility. He was receiving at-home physical therapy following a recent below-the-knee amputation related to chronic peripheral artery disease.

Eleven months prior to presentation, he went to his primary medical doctor with complaints of photosensitivity and left orbital pain but in the absence of conjunctival injection, jaw claudication, scalp tenderness, fever, myalgias and hip girdle discomfort. He was started on prednisolone and homatropine drops for presumed iritis. The cephalgia remained although often masked by the opioids prescribed for his prior right shoulder arthroplasty. HCT 3 months after initial symptom onset revealed his known chronic left parietal infarct. Erythrocyte sedimentation rate (ESR) was 50 mm/hour without corroborative evidence for acute inflammation as the C-reactive protein was negative. High-dose prednisone was started 7 days prior to a temporal artery biopsy, which was negative. Nonetheless, he continued treatment for presumed giant-cell arteritis with prednisone. Despite this treatment, he continued to experience severe left orbital pain.

Four months prior to admission, he developed cellulitis in the left lower extremity. A CTA at the time revealed three-vessel disease within the extremity. He underwent angioplasty of the left anterior tibial artery. Following angioplasty, his wound progressed, requiring surgical debridement. A left proximal superficial femoral artery dissection was identified 2 months prior to admission. Following an extensive rheumatological clinical and laboratory evaluation, the clinical suspicion for vasculitis, including giant cell arteritis and infectious aetiologies, was low. One month prior to admission, he underwent left below-the-knee amputation and received antibiotics for presumed cellulitis. He was discharged 5 days after surgery and required minimal assistance with ambulation.

Eight days prior to transfer to our facility, he became somnolent, with chills, malaise and worse headache. Three days later, he went to see his

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Figure 1 CT angiogram shows clinoid and supraclinoid stenosis and irregular opacification (arrow) originally interpreted as normal.

primary doctor for these symptoms. During the visit, he developed right-hand weakness with spontaneous resolution. He was sent to a local emergency department by ambulance. No hypotension was observed. HCT and CTA were read as negative. In retrospect, the CTA reveals luminal stenosis and irregular opacification at the left supraclinoid ICA (figure 1). He was admitted for observation and warfarin up-titration as his INR was subtherapeutic. Overnight he was febrile, but no infectious source was identified. A mild hyponatraemia (128 mmol/L) was attributed to hypovolemia from poor oral intake. He was discharged home the day following admission. Three days prior to transfer to our facility, right haemiparesis and dysarthria returned for 10 min. A repeat HCT was read as negative, and a CTA was not performed. He was admitted to a local hospital to bridge anticoagulation until INR was therapeutic. On the day of admission to our facility, he developed a left gaze preference, dense right haemiplegia and aphasia. Repeat HCT and CTA were interpreted as demonstrating a left hyperdense MCA sign and clot in the left distal ICA (figure 2). In retrospect, the supraclinoid lesion has both a poorly defined border between the ICA and CSF and descends into the clinoid region. These features are atypical for the dense MCA sign. He was transferred to our institution for endovascular management.

On arrival, the National Institutes of Health Stroke Scale was 7 (one for level of consciousness questions; two for right haemianopsia; one for right facial palsy; one for mild aphasia; one for

Figure 2 Axial CT (A) shows hyperdensity in the region of the left MCA interpreted as left hyperdense MCA sign. (B) Axial CT shows extension of hyperdense lesion within clinoid space. Both poorly defined border between the dense vessel and CSF and a contiguous dense lesion extending into the clinoid portion are atypical for the MCA sign. CSF, cerebrospinal fluid; MCA, middle cerebral artery.

Figure 3 Anterior-posterior projection of left ICA showing distal ICA stenosis (arrowhead) with poststenosis dilation (arrow). CCA, common carotid artery; ICA, internal carotid artery.

mild-moderate dysarthria and one for right sensory extinction to double simultaneous stimulation). ESR was mildly elevated at 32 mm/hour. His daily dose of prednisone was 5 mg. Digital subtraction angiography revealed 60% narrowing of the supraclinoid ICA with poststenotic dilatation (figure 3). No other vessels demonstrated focal stenosis or morphology suggestive of vasculitis or inflammation within intracranial or extracranial vessels. He was admitted to the Neurosciences Intensive Care Unit without endovascular intervention as there was evidence of good flow distal to the area of stenosis. Initial blood pressure goals were liberalised because of concern for acute infarction. INR at the time of admission was 2.6. He received no antithrombotics after admission. Later that evening, he declined neurologically with worsened aphasia. Repeat HCT revealed new extensive subarachnoid blood involving the Sylvian fissures, anterior interhemispheric fissure, both anterior and lateral ventricular horns and the cerebral convexities. Anticoagulation was reversed and blood pressure parameters were reduced to a goal of <160 mm Hg systolic. Recorded blood pressure was highest 3 hours prior to HCT, noted as 176/136 mm Hg. Blood pressure at the time nearest to HCT was 158/68 mm Hg.

During preparation for repeat DSA, he became acutely unresponsive with agonal breathing and bilateral extensor upper extremity posturing. He was emergently intubated and routed instead to repeat CTA. This revealed a pseudoaneurysm in the vicinity of the previously noted 'hyperdense MCA sign' arising from the left supraclinoid ICA with a small dissection flap in the region of the dome (figure 4).

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for acute-onset haemiplegia with headache or ocular pain includes haemorrhage, ischaemia, infarction, complicated migraine, seizures or vascular anomaly.⁵

Figure 4 Sagittal CT angiogram shows a new aneurysmal dilation of the distal left ICA in the area of the previously noted hyperdense MCA sign. ICA, internal carotid artery; MCA, middle cerebral artery

Haemorrhage may be epidural, subdural, subarachnoid, intraparenchymal, or intraventricular and should be apparent on non-contrast HCT. Haemorrhage was not identified on imaging at admission. Infarction is most commonly caused by embolism from the heart, aortic arch, cervical vessels or by intrinsic disease of intracranial vessels. The clinician may appreciate evidence of acute infarction on HCT, but MRI is preferred to identify infarction early after symptom onset. Acute infarct was not seen on HCT at admission. In the proper clinical context or when not easily explained by known factors, vascular lesions such as dissection, aneurysm, dural arteriovenous fistula, arteriovenous malformation, venous thrombosis, cavernous malformation or fibromuscular dysplasia should be considered. Angiography of extracranial and intracranial vessels is often required to identify the vascular lesion. A focal region of stenosis with a poststenotic dilatation should raise concern for dissection and fusiform aneurysm. Vasculitis can produce ischaemic infarctions and intracranial haemorrhage, but its angiographic appearance consists of beading of multiple intracranial vessels.

TREATMENT

His family decided the patient would not want further aggressive medical or surgical care. No further treatment or intervention was performed. If emergent treatment had been accepted, we may have intervened with deployment of a stent. Open surgical repair with wrapping of the aneurysm, parent artery occlusion or extracranial-intracranial arterial bypass are alternative options, but their operative risk is high.¹⁶

OUTCOME AND FOLLOW-UP

He expired shortly after being transitioned to comfort care. His family declined an autopsy.

DISCUSSION

We found two prior reports of supraclinoid intracranial dissection manifesting with both cerebral infarction and haemorrhage. Severe retro-orbital pain or temporal headache followed by contralateral hemiparesis was the most common initial clinical presentation.^{7,8} The patient's advanced age and gender are atypical for ICA dissection. Further, chronic headache due to ICA dissection is unusual, but has been previously reported. In a cohort of 161 consecutive patients with ICA dissection, the median duration of headache associated with the dissection was 3 days, but ranged from 1 hour to 4 years.⁹ Despite the patient's atypical clinical course and previously negative temporal artery biopsy, we do not have pathological confirmation that the patient did not suffer an acute inflammatory arteritis at the time of presentation. Temporal artery biopsy has been documented to be negative despite final diagnosis of giant-cell arteritis in 5%–11% of cases.^{10,11} While unlikely, it is conceivable the patient also suffered from both giant-cell arteritis and subsequent dissection. We believe our patient suffered a dissection months before the stroke with a recurrence and progression of the dissection at the time of his acute presentation.

The pathophysiology of dissections involves two mechanisms that lead to either luminal stenosis or arterial rupture. Subintimal dissections allow accumulation of blood within the arterial wall thus producing luminal stenosis. Ischaemic infarctions can be caused by thromboembolism generated in the area of intimal damage or haemodynamic insufficiency when the intramural clot results in vessel occlusion or critical stenosis. Subadventitial dissections develop blood products communicating with the lumen and a layer just deep to the adventitia. Pressure against the

adventitia may lead to wall rupture and subsequent subarachnoid haemorrhage.¹² Data suggest treatment of non-aneurysmal intracranial ICA dissection with anticoagulation is safe.¹³ His therapeutic anticoagulation and prior use of aspirin may have increased his risk for haemorrhage.

Dissection is reliably diagnosed if an intimal flap, mural haematoma or double lumen is appreciated on brain imaging. Dissections may also present with a 'string sign' (narrow tapered lumen), fusiform aneurysmal dilatation, 'pearl and string' sign, occlusion, proximal and/or distal dilatation and wavy ribbon-like (ripple) appearance.¹⁴ Black blood and delay alternating with nutation for tailored excitation with three-dimensional variable flip angle turbo spin echo signal are MRI sequences recently been proposed for optimal intracranial vessel wall imaging.¹⁵

Neither a prospective study nor meta-analysis has identified differences in outcome or complications regarding long-term antiplatelet or anticoagulant use.^{16–19} Endovascular repair may involve placement of a flow-diverter stent, although the intervention has not been formally evaluated for this indication. Suggested indications for intracranial carotid artery stent placement include: (1) clinical failure of medical therapy (presence of recurrent TIAs, fluctuating neurological signs or neurological deterioration); (2) impending stroke attributable to significant stenosis/occlusion with poor collateral circulation and reduced parenchymal perfusion on the angiographic capillary phase of CT or MRI studies; (3) contraindication to anticoagulation because of intracranial or systemic haemorrhage; (4) evidence of symptomatic thromboembolic occlusion of cerebral vessels; (5) contralateral carotid stenosis/occlusion; (6) need for elective occlusion of the contralateral ICA for other pathology and (7) need to avoid flow increase through the anterior communicating artery because of an associated aneurysm.⁶ Stent placement with intracranial carotid dissection may be a predictor of poor outcome.²⁰ Observational studies of natural history have included octogenarians.²¹ There is suggestion that recurrent SAH may occur more frequently in patients with intracranial artery dissection older than 50 years.¹

Lack of angiography at the onset of headache impedes us from being certain that the patient's initial headache was caused by a dissection. The patient's initial symptoms appear better explained by dissection than by giant-cell arteritis. In retrospect, the diagnosis should have been reconsidered when the temporal artery biopsy was negative and the symptoms failed to improve with prednisone. Meanwhile, the acute presentation that eventually led to the patient's demise was certainly caused by an intracranial dissection, possibly an extension of a previous dissection months before. Careful review of the HCT and CTA, in the setting of recurrent ischaemic symptoms, might have allowed an opportunity for endovascular treatment.

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Learning points

- Intracranial dissection may present with isolated periorbital pain, while chronic symptoms are rare, they have been described.
- Radiographic diagnosis of intracranial dissection may be difficult. An atypical middle cerebral artery sign should prompt consideration of the differential diagnosis. New MRI modalities may improve sensitivity for detection.
- Supraclinoid intracranial dissection may present initially with ischaemic symptoms and then cause life-threatening subarachnoid haemorrhage.
- In cases of subarachnoid haemorrhage, endovascular or surgical repair may be appropriate.

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