

Gesundheit! Sneezing, paraesthesiae and ataxia in a 35-year-old man

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A 35-year-old right-handed male chef reported three similar neurological episodes in the preceding 6 months. Each episode began with a paroxysm of sneezing. In the first, sneezing was followed by sudden onset dizziness, and then over a few minutes by double vision, left-sided paraesthesiae, left-sided weakness and then a severe left-sided headache, all resolving after a few hours. The second and third episodes, approximately 2 months apart, were less severe. During the second event, he also noticed his right pupil was bigger than the left. In the third episode, 6 weeks before his clinic visit, he developed several hours of ataxia and double vision, but no sensory or motor symptoms; this was followed by left-sided headache.

The patient underwent MRI brain and magnetic resonance angiography (MRA) of the cervical vessels (figure 1).

Question 1

What does the imaging show?

Comment

The MRA shows a dissection of the proximal right vertebral artery, causing a stenosis and a pseudoaneurysm distal to the stenosis, with thrombus in the pseudo-aneurysm. The MRI brain also shows an arteriovenous malformation (AVM) in the right temporoparietal lobe.

Question 2

What is the diagnosis? What would you do next?

Comment

The most likely diagnosis is recurrent ischaemic events due to a vertebral artery dissection, with subsequent embolisation from either the dissection or from thrombus in the pseudoaneurysm. Pseudoaneurysms develop when haemorrhage into the blood vessel wall separates the media from the adventitia during a dissection. A false lumen occurs if blood re-enters the true vessel lumen. The dissection lumen may thrombose and cause stenosis of the true lumen or there may be embolisation from the thrombus. Pseudoaneurysms develop after 10–46% of vertebral artery dissections, although they resolve spontaneously in 80%; even if they persist, they only very rarely cause a stroke.¹ Apart from antiplatelet therapy, intervention is therefore generally not recommended. Our patient had been asymptomatic for 2 months when we discussed the investigation results with him. We therefore opted for conservative management with low-dose aspirin, and further clinical and imaging follow-up.

The patient initially did not return as planned, but then returned to clinic several months later following a more prolonged episode of left-sided sensory symptoms. It emerged that there had been at least 10 further episodes over the preceding 6 months. Although not entirely stereotyped, they were all similar: the patient sneezed and would then notice diplopia, unsteadiness and left-sided paraesthesiae, although not all three symptoms on every occasion. Symptom severity and duration varied between some minutes

and several hours and appeared to be related to the force of sneezing. The patient admitted that he was becoming increasingly afraid of sneezing, apprehensive that this might set off further symptoms.

Further imaging showed a persisting pseudoaneurysm, which had slightly decreased in size. Brain imaging showed no ischaemic lesions. Nevertheless, we still felt that the most likely diagnosis was recurrent embolisation from the aneurysm, triggered by sneezing. It transpired that the patient had not taken his aspirin, due to previous peptic ulcer disease. He was now started on clopidogrel, but continued to have symptoms. He returned to hospital some weeks later after a particularly severe attack, with persisting diplopia, ataxia and left-sided sensory loss. The MRI brain now demonstrated several acute cerebellar infarcts (figure 2), strongly supporting the diagnosis of recurrent ischaemic events. As these had continued with the patient on antiplatelet treatment, we now had to consider more invasive management, as the patient was at risk of further, potentially more severe events.

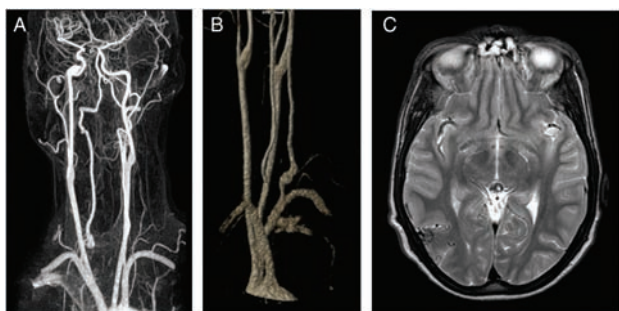


Figure 1 MRI brain and magnetic resonance angiogram at the initial presentation. The MRA (A, B) shows the dissected right vertebral artery with a proximal stenosis and a pseudoaneurysm distal to this. MRI brain (C) shows the right parietal arteriovenous malformation.

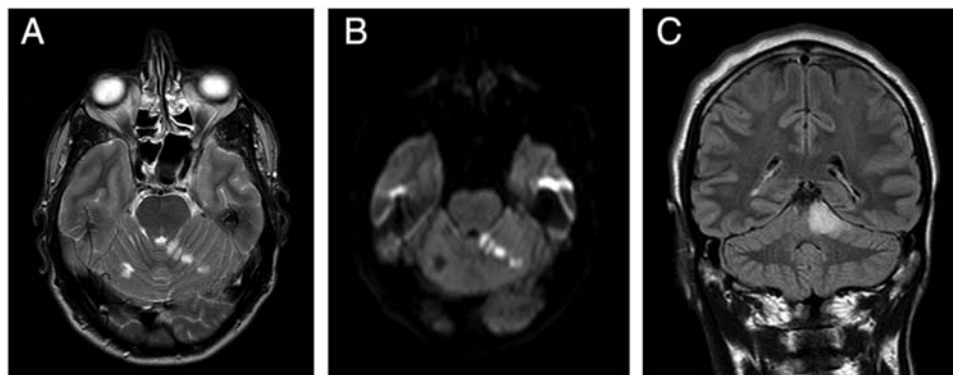


Figure 2 Acute cerebellar infarcts on T2-weighted (A) and diffusion-weighted (B) MRI brain, and coronal fluid attenuated inversion recovery images (C). Diffusion-weighted imaging shows that the right-sided cerebellar infarct (black), is older than the left-sided infarcts.

Question 3

Which other treatment options would you consider?

Comment

Further management options to reduce recurrent embolism from the aneurysm included anticoagulation and endovascular treatment. As the benefit of anticoagulation over antiplatelet therapy in this situation is uncertain, and because the patient strongly preferred intervention to long-term medication, we proceeded with endovascular treatment. We considered two options: to coil the aneurysm, preserving the vessel lumen with a stent, or to occlude the vertebral artery with coils. Inserting a stent would have been technically difficult and risky in this case because of kinking and stenosis of the vessel. Furthermore, a stent would remain a potential source of further thromboembolism and require long-term antiplatelet therapy. Coil occlusion of the parent vessel provides a definitive treatment of a pseudoaneurysm, with no further risk of embolisation and no need for long-term medication. In patients with intracranial dissecting aneurysms it is the preferred treatment if sufficient collaterals exist,² and it also appeared to be the safest and most durable treatment in our patient. He tolerated a test occlusion of his diseased vertebral artery well and underwent coil occlusion without complication (figure 3). At 6 months after the procedure, he reported no further events of

Question 4

Is the AVM related to the patient's presentation? How would you manage it?

TEST YOURSELF



Figure 3 Intra-arterial angiogram. (A) Shows the pseudoaneurysm and demonstrates the stenosis and vascular kink proximal to the aneurysm, which would have made stenting difficult. (B) The occluded vessel after coil embolisation.

neurological disturbance. More importantly, he has also sneezed on multiple occasions, without provoking symptoms.

Comment

Flow-related aneurysms close to AVMs have been described. However, in our patient the AVM and the dissection were too far apart for flow-related phenomena to have an effect. It is possible that the patient has an underlying vascular disorder that predisposed him both to the development of AVM and of arterial dissections, but this is speculative. Nevertheless, the AVM is a further management issue. AVMs are usually diagnosed when they cause symptoms, mostly seizures or haemorrhage. Our patient's AVM is asymptomatic, and the risk of future complications is uncertain. The annual risk of haemorrhage from an asymptomatic AVM is estimated to be 1–2%.³ As our patient is still young, he has a considerable lifetime risk of haemorrhage, which must be weighed against the procedural risk of any intervention, and the uncertainty that such an intervention would provide a definitive cure. The ARUBA trial (A Randomised Trial of Unruptured Brain AVMs, www.arubastudy.org) compares the outcome of conservative management with intervention in patients with AVM that have not bled. We have discussed the uncertainties of AVM management with the patient. He is keen on intervention, and after several discussions with neuroradiologists and neurosurgeons, stereotactic radiosurgery was felt to be the most straightforward and lowest risk treatment to offer him, which he is currently awaiting.

Question 5

What do you make of the association of this patient's symptoms with sneezing?

Comment

The intriguing part of this patient's presentation was the association of his symptoms with sneezing. Sneezing has been reported as a possible cause for vertebral artery dissection. The mechanism is unclear, but pressure changes or neck movements may lead to an intimal tear and dissection of the vessel wall. Similarly, they may also cause thrombus to dislodge. Given the presence of ischaemic lesions on the patient's third magnetic resonance scan, and the cessation of attacks after occluding the vertebral artery, it now appears certain that he had recurrent embolic events triggered by sneezing. However, it is surprising that sneezing was the only mechanism to set off the events: coughing or Valsalva manoeuvres never preceded the patient's neurological symptoms. We therefore considered that sneezing may have been a symptom of, rather than the trigger for, the patient's attacks. This is not inconceivable, as the 'sneezing centre' is in the lateral medulla, a frequent infarct location in vertebral artery dissection. However, although sneezing may rarely be a symptom of ischaemia, such infarcts more commonly present with an inability to sneeze. It seems unlikely that sneezing, if a symptom, would have occurred with all attacks, and only at their onset. Finally, given the high number of similar attacks, we considered that the patient might have had focal seizures originating from his AVM. Sneezing can be a seizure symptom, but it is usually associated with temporal lobe epilepsy. The AVM was in the wrong location for such seizures, and overall the attacks did not appear sufficiently stereotyped to be epileptic.

Learning points

- Pseudoaneurysms develop after 10–46% extracranial vertebral artery dissections. These are usually asymptomatic and usually resolve spontaneously.
- Embolisation from a vertebral artery pseudoaneurysm is rare and intervention beyond treatment with antiplatelet agents is generally not required.
- Further treatment options include occlusion of the parent vessel and coiling and/or stenting of the aneurysm. The best way forward will depend on the individual vascular anatomy and on local facilities and expertise.

Competing interests None.

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