

Neurologist-in-training: Painful eye-tongue syndrome

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Case vignette

A 38-year-old previously healthy man woke up with a heavy and clumsy tongue and difficulties in swallowing at breakfast. In the mirror he noted a drooping eyelid on the left side. The evening before he had perceived a dull pain on the left side of his neck. He did not remember any trauma during the past days, but he had carried heavy items the day before. He was sent to the emergency room by his family physician, suspecting a stroke. On neurological examination the patient was alert, lateral neck pain was rated minimal only, speech was slightly clumsy, he showed miosis, narrowing of the palpebral fissure and frontal hypohydrosis on the left side, left hypoesthesia of the pharyngeal wall and posterior tongue, and a loss of tone in the left half of the tongue with tongue deviation to the left on protrusion. Facial and extremity motor innervation and sensation was normal. Oculomotor exam, visual field testing and fundoscopy were normal as well.

Questions

1. Which anatomical neurological structures are affected? (Any or no answer may be correct.)
 - A Left pontomedullary brainstem.
 - B Left hypoglossal (XII) cranial nerve.
 - C Left trigeminal (V) cranial nerve.
 - D Left glossopharyngeal (IX) cranial nerve.
 - E Right corticobulbar tract.
 - F Left central descending sympathetic pathways.
 - G Left peripheral oculosympathetic fibres.

The patient wants to leave the emergency room since he has an appointment for a squash game with a friend.

2. What is your procedure and why? How do you explain it to your patient? (Only one answer is correct.)
 - A Perform emergency cranial MRI.
 - B Perform emergency cranial MRI and MRA.
 - C Perform emergency CT scan of head.

- D No emergency imaging is necessary.
- E Perform lumbar puncture.
- F Perform emergency MRI and MRA of head and neck.
- G Perform a neurovascular ultrasound exam.

The patient agrees to do an MRI, the images are shown below.

DWI brain sections disclose no ischaemia in the brain or brainstem. PWI also is unrevealing without any hypoperfusion area. T2 scans are without any abnormality.

Figure 1 shows the MRA of the supra-aortic brain supplying vessels, figure 1a shows contrast-enhanced MRA and figure 1b TOF angiography.

3. What can you describe on figure 1? And what are the consequences you take from these findings, if any?

Figure 2 shows fat suppressed MR images of the upper neck in the coronal (a and b) and axial (c and d) plane.

4. What can you describe on figure 2 and what are the consequences you take from these findings, if any?

The patient wants to know what the MRI has shown and how to proceed.

5. Do the patient's symptoms match with the MRI findings? How can you explain this to the patient?
6. Which cranial nerve is most frequently affected by ICA-dissection? (Only one answer is correct.)
 - A Accessory nerve (XI).
 - B Hypoglossal nerve (XII).
 - C Oculomotor nerve (III).
 - D Trigeminal nerve (V).
 - E Facial nerve (VII).

The diagnosis is settled, the patient is happy – but now what to do? Is any therapeutic intervention helpful? Are there any precautions to be taken or can you let him and things go their usual (spontaneous) way?

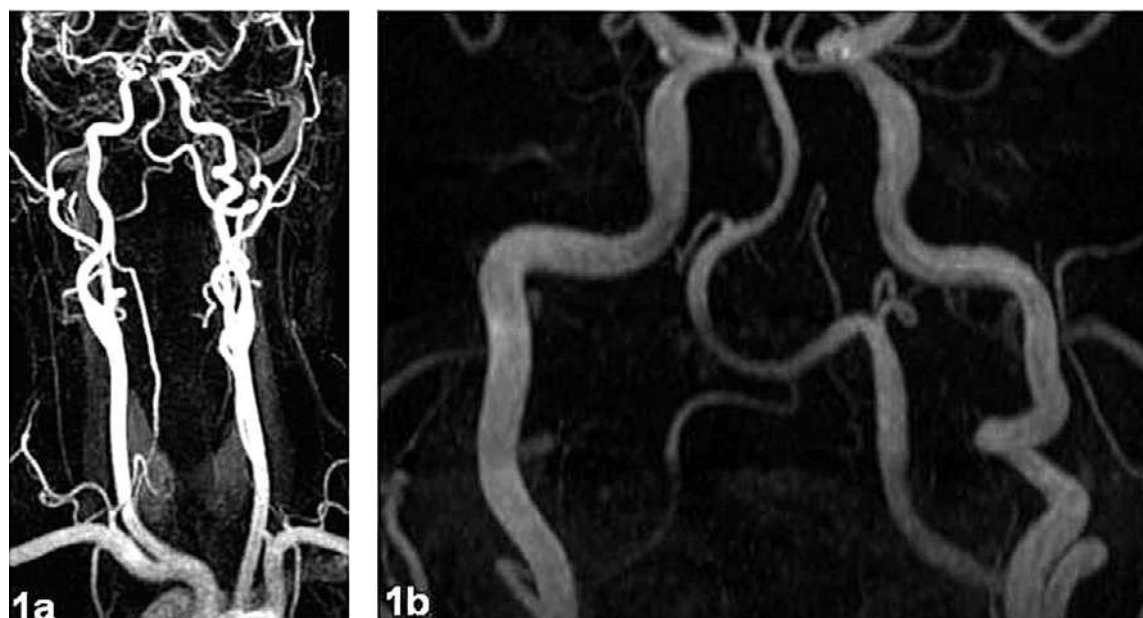
7. Which of the following statement/s is/are correct concerning the acute management of patients with cervical artery dissections? (Any or no answer may be correct.)
 - A Thrombolysis is a therapeutic option for acute ischaemic strokes.
 - B Early endovascular treatment for intracranial dissections is not recommended.

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Figure 1

MRA of the supra-aortic brain supplying vessels: contrast-enhanced MRA (a), TOF angiography (b).



- C A cervical artery dissection is a contraindication for intravenous thrombolysis.
- D Cerebral haemorrhagic complications are higher in stroke patients with cervical artery dissections than in stroke patients with other aetiologies.
- E Stenting is mandatory for pseudoaneurysms resulting from dissections.
8. Which of the following statement/s is/are correct concerning the long-term management of patients with cervical artery dissections? (Any or no answer may be correct.)
- A Long-term anticoagulation is indicated in any case.
- B Studies could not show an advantage for anticoagulation or antiplatelet therapy.
- C Residual stenosis or occlusion should be stented.
- D Residual pseudoaneurysm represents a high risk for stroke.
- E Long-term antiplatelet therapy is indicated to prevent recurrent dissections.
9. ICAD is considered a disease of the vessel wall. Several environmental and constitutional factors are associated with ICAD – which ones? (Any or no answer may be correct.)
- A Alcohol abuse.
- B Major and minor cranio-cervical trauma.
- C Fibromuscular dysplasia.
- D Marfan's syndrome.
- E Diabetes mellitus.
- F Recent infection.

Figure 2

MR images of the upper neck (fat suppressed): coronal plane (a, b), axial plane (c, d).

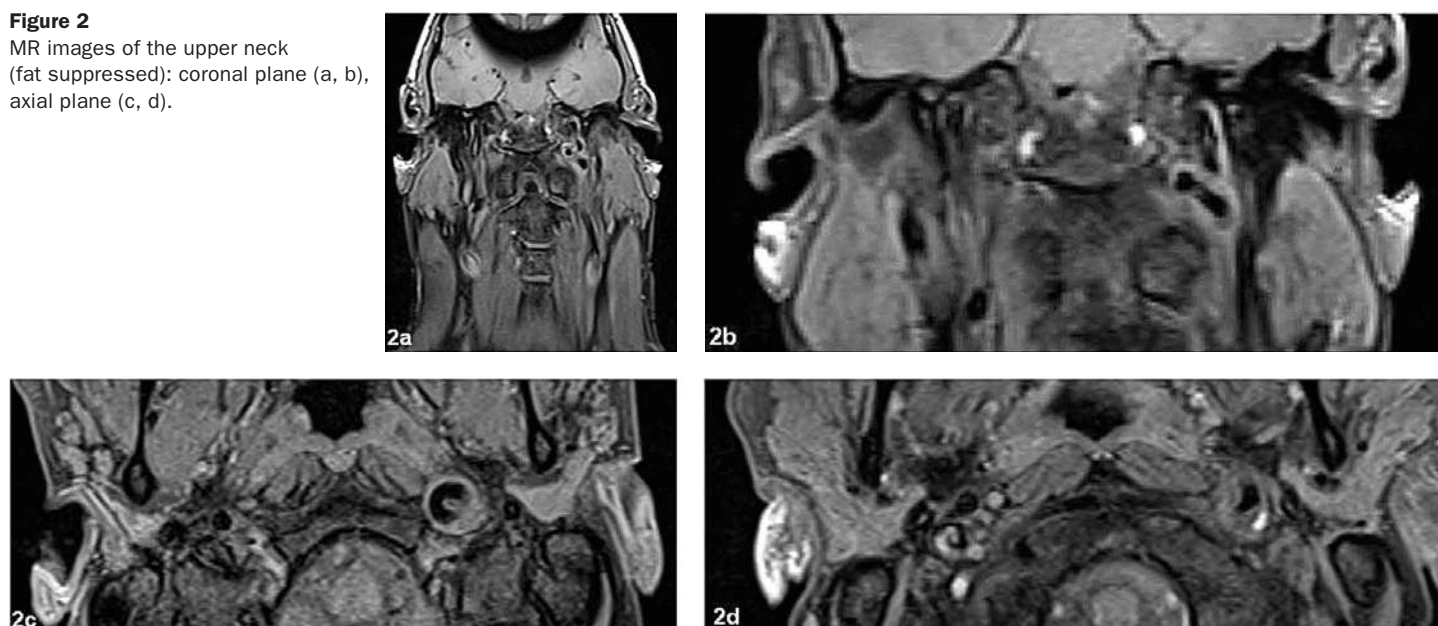
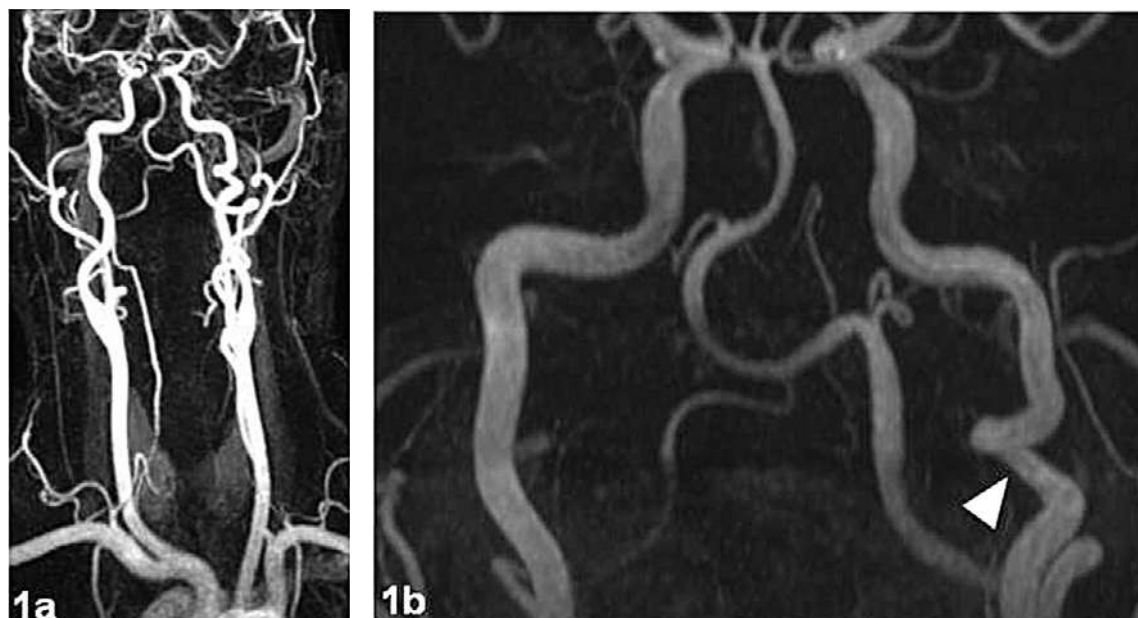


Figure 1*

MRA of the supra-aortic brain supplying vessels.



Answers

Answer 1

B, D and G are correct.

There are not “long-tract” symptoms or signs indicating a central (brain or brainstem) lesion (A, E or F).

Answer 2

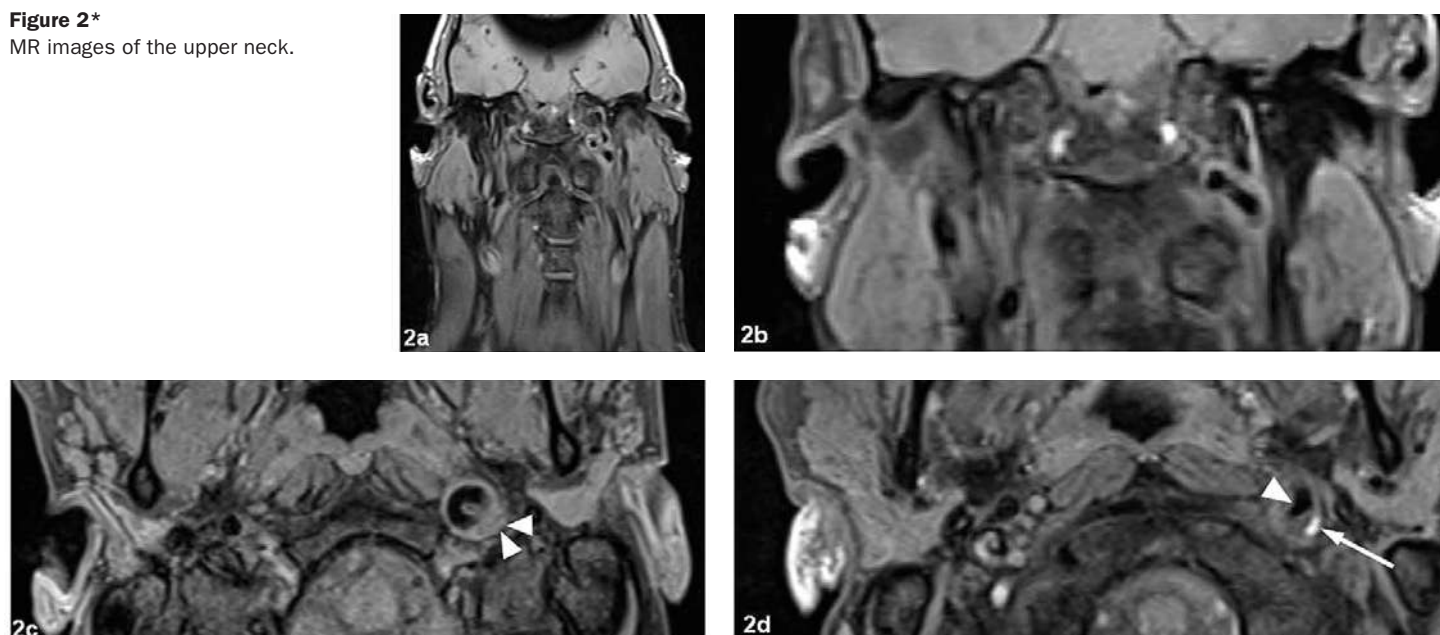
F is correct. You have left sided dysfunctions of the XII and IX cranial nerve and of the ascending sympathetic fibres from the superior cervical ganglion with ipsilateral neck pain. In this situation you must rule out left internal carotid artery dissection (ICAD). Since such a dissection usually takes place extracranially in the upper cervical region, you have to include the neck in your imaging protocol. Since one main imminent risk of ICAD is cerebral ischemia due to ar-

terio-arterial embolism you have to image the vessel lumen with MRA and the vessel wall with fat suppression sequences. In the case of dissection stroke may happen at any time; to prevent it you have to establish the diagnosis immediately – therefore this diagnosis has to be confirmed (or excluded) on an emergency basis.

This case also nicely demonstrates why ultrasound is not reliable enough (European consensus): it may demonstrate direct or indirect signs of a narrowed vessel lumen, but, as in our case, if the dissection is in the subadventitial layer with only minimal resulting stenosis (see fig. 1*), the ultrasound exam will yield a false negative result. Since ICA dissections most frequently are located in the high-cervical (retromandibular) segment (cf. fig. 1*a), the pathology is not directly accessible to B-mode imaging.

Figure 2*

MR images of the upper neck.



Answer 3

Findings: there is right vertebral artery hypoplasia, a finding without any significance; carotid artery bifurcation is normal on both sides. There is an impressive coiling of left ICA in the upper cervical region. Filling of both ICAs and intracranial vessels (MCAs) is normal. On TOF (1*b) there is a luminal narrowing in the middle branch of the coil (arrowhead).

Consequences: the unilateral ICA coiling is a predisposing factor for carotid artery dissection, especially in a young patient. The narrowed lumen on TOF, also compared to the contra-lateral ICA lumen width, is another argument that something is wrong in this part of the vessel wall. So you have to closely look for signs indicating vessel wall pathology in other MR sequences of the neck in this region. In the optimal setting you should have or order T1 fat suppressed images of that region in the axial and coronal plane to optimally disclose the mural haematoma characteristic for a dissection.

Answer 4

Coronal sections (fig. 2*a and b) disclose several vessel cuts through ICA due to the coiling. Furthermore in this region of coiling the vessel wall seems thickened.

Axial sections (fig. 2*c and d) confirm vessel wall thickening of ICA in this tortuous part and “tumor-like” appearance (two arrowheads) with only moderate lumen stenosis (arrowhead) compared to opposite side and mural hemorrhage appearing as partly hyperintense signal (arrow). Thus, ICAD is confirmed but with only minimal stenosis.

Answer 5

From (fortunately rare) histopathological examinations of dissected carotid arteries we know, that there are basically two planes in the arterial wall where mural haemorrhage may occur: the subintimal and the subadventitial layer. There is no explanation for the preference of one location. Subintimal haemorrhage more frequently causes severe vessel lumen stenosis or occlusion with subsequent embolic or (more rarely) haemodynamic cerebral ischaemic events. Subadventitial haemorrhage usually only causes minimal lumen narrowing but extension of the outer diameter of the vessel's circumference by the haematoma (tumefactive appearance) and thus compression or stretching of neighbouring structures such as the lower cranial nerves (XII, XI, X) and the pericarotid sympathetic nerve fibres taking their course from the superior cervical ganglion to the ipsilateral facial glands, eye and frontal skin. Whether the cranial nerve damage is due to compression of the nerve itself (the lower cranial nerves tightly cross the ICA in the upper cervical region) or of its nutritional vessels is debated. The sometimes rather rapid recovery (within 2–3 weeks) is in favour of a compressive (demyelinating) mechanism.

Answer 6

The hypoglossal nerve, for reasons of anatomical neighbourhood, is the most frequently affected cranial nerve in internal carotid artery dissections. The so called painful hypoglossal nerve palsy (with concomitant ipsilateral neck- and or throat pain) is a rather typical presentation of ICAD and also

other curious syndromes such as “carotidynia” probably do not exist anymore in the MRI-era.

Answer 7

Only answers A and B are correct (see comments, “management and treatment”).

Answer 8

Only answer B is correct (see comments, “management and treatment”).

Answer 9

Answers B, C, D and F are correct (see comments, “pathogenesis of dissection”).

Comments*Pathogenesis of dissection*

In most patients with ICAD, the exact pathogenesis remains undetermined. There are several constitutional or environmental risk factors associated with ICAD with a varying degree of evidence.

Environmental: major (cervical spine fracture) or minor (sports activities, such as squash or soccer; medical manoeuvres such as intubation or cervical spine manipulation) traumatic injury; drugs (sympathomimetics) or recent infection.

Constitutional: (1.) arterial disease such as fibromuscular dysplasia, cystic medial necrosis or known heritable connective tissue disorders (Marfan's syndrome, Ehlers-Danlos syndrome, osteogenesis imperfecta, pseudoxanthoma elasticum, adult polycystic kidney disease); (2.) Vasculitis (syphilis, polyarteritis nodosa), hyperhomocysteinemia, α_1 -Antitrypsin deficiency, migraine, hypertension.

Pathogenesis of symptoms

Carotid dissection may have many clinical faces. Unreflected performing a cranial MRI may miss the potentially dangerous diagnosis in the case of initial presentation without cerebral ischaemic symptoms. If dissection is suspected, the imaging must include the neck and special sequences (T1 fat suppression should be used). Within the first 2 or 3 days, MRI may not reliably show the dissection because initially the hematoma is isointense to the surrounding tissue and turns hyperintense only later, as soon as the haemoglobin converts into methaemoglobin. Therefore, vessel wall thickening or narrowing of the vessel lumen on MRA may be the only initial signs.

It is important to recognise a dissection before cerebral ischaemia has occurred because this offers the chance of stroke prevention with anticoagulant therapy. Therefore, local (neck) or distal (temporal and periocular) pain, ipsilateral oculosympathetic palsy with (Horner's triad) or without frontal hypohydrosis and ipsilateral lower cranial nerve palsies are important symptoms, whether isolated or in combination, heralding dissection. These symptoms all can be explained by compression of the cranial nerves and the sympathetic fibres by the expanding wall haematoma. Subadventitial dissection and haematoma is associated with lower cranial nerve palsies due to more outward extension

and subintimal dissection is more likely associated with stenosis.

Management and treatment

Both local intra-arterial thrombolysis and intravenous-thrombolysis are treatment options in the case of acute strokes complicating cervical artery dissection and do not differ in the risk of complications or efficacy to other stroke patients as far as studies can tell today. Usually strokes are due to arterio-arterial embolism from thrombus formation at the dissected vessel site.

Caution should be taken in patients with subarachnoid haemorrhage usually seen in the case of vertebral artery dissections extending to the intradural vessel segments.

Stenting (of the ICA) can be considered in the case of occluding ICAD with resulting embolic MCA occlusion to allow access to the MCA for local treatment and to subsequently guarantee adequate perfusion pressure. Many dissections result in a so-called pseudoaneurysm, usually located in the extracranial segments of ICA and VA. Follow-up studies have shown that these pseudoaneurysms, independent of their size, are harmless and need no (interventional or pharmacologic) treatment.

For secondary stroke prophylaxis either antiplatelet drugs or anticoagulation can be used. If the intracranial part of the vessel is affected anticoagulation should not be used because of the risk of intracranial bleeding. Currently there are no studies which could prove any benefit for one treatment over the other. We prefer anticoagulants in the case of a stenosing dissection. There is no sound indication for any long-term treatment. The primary goal is prevention of future strokes. Even in the case of persistent occlusion or stenosis the stroke risk is small. But only in this later situation long-term antiplatelet treatment can be discussed. A recurrent dissection cannot be prevented by any measure.

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