Nouf Al-Farsi, MD, FRCSC,[*] Adrian C Tsang,[†] Rahul A. Sharma,[†] Annick Fournier[*,†]
*The Children's Hospital of Eastern Ontario, Ottawa, Ont.; [†]The University of Ottawa Eye Institute, Ottawa, Ont.

Correspondence to: noufalfarsi@gmail.com.

References

1. Dobyns W, Pagon R, Armstrong D, et al. Diagnostic criteria for Walker-Warburg syndrome. Am J Med Genet 1989;32:195–210.
2. Beltrán-Valero de Bernabé D, Currier S, Steinbrecher A, et al. Mutations in the O-mannosyltransferase gene POMT1 give rise to the severe neuronal migration disorder Walker-Warburg syndrome. Am J Hum Genet 2002;71:1033–43.
3. Pagon RA. Ocular colobomas. Surv Ophthalmol 1981;25: 223–36.
4. Roscioli T, Kamsteeg EJ, Buysse K, et al. Mutations in ISPD cause Walker-Warburg syndrome and defective glycosylation of α-dystroglycan. Nat Genet 2012;44:581–5.
5. Edwards AO. Clinical features of the congenital vitreoretinopathies. Eye 2008;22:1233–42.
6. Vohra N, Ghidini A, Alvarez M, Lockwood C. Walker-Warburg syndrome: Prenatal ultrasound findings. Journal of Prenatal diagnosis 1993;13:575–9.
7. Brasseur-Daudru M, Vivier PH, et al. Walker-Warburg syndrome diagnosed by findings of typical ocular abnormalities on prenatal ultrasound. Journal of Padiatric radiology 2012;42:488–90.
8. Shen O, Zuckerman S, et al. Prenatal Sonographic Diagnosis of Retinal Nonattachment. Journal of Ultrasound medicine 2014;33:1095–7.
9. Drought A, Wimalasundera R, Holder S. Ultrasound diagnosis of bilateral cataracts in a fetus with possible cerebro-ocular congential muscular dystrophy during the routine second trimester anomaly scan.
10. Achiron R, Katorza E, et al. Very Early In-Utero Diagnosis of Walker-Warburg Phenotype: The Cutting Edge of Technology. Journal of Ultrasound international open 2016;02:E54–7.
11. Hakim N, Soare C, Hakim J. Very Early In-Utero Bilateral total retinal detachment at birth: a case report of Walker? Warburg syndrome. International medical case reports journal 2018;11:1–4.

Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.

Dysgeusia and amaurosis fugax: a unique presentation in spontaneous internal carotid artery dissection

Spontaneous internal carotid artery (ICA) dissection is a significant cause of ischemic stroke in young adults[1]. The presenting symptoms and signs of ICA dissection (ICAD) are variable and can be due to direct local involvement of neural structures (e.g., oculosympathetic plexus causing the ipsilateral Horner syndrome) or indirect distal ischemia to brain or eye (e.g., transient ischemic attack or stroke) from thromboembolic disease [2,3]. Transient monocular visual loss (TMVL) and the Horner syndrome are well-known presentations of ICAD and may occur in up to 50% of cases [4]. A distortion of the sense of taste (i.e., dysgeusia) in ICAD, however, is not as widely known as presenting symptoms especially among ophthalmologists. We present a case of TMVL associated with dysgeusia and describe the clinical presentation, localizing topographic anatomy, pathogenesis, and significance of dysgeusia to ophthalmologists evaluating patients with possible ICAD.

A 55-year-old white man presented with 2 episodes of TMVL of the left eye (OS) over 2 weeks associated with a "bitter taste in his mouth." He had been dining in a restaurant with his wife and complained to the restaurant manager about the taste of the food. He was seen at an outside clinic for eye pain and headache, which was diagnosed as "sinusitis" and treated with antibiotics. His medical history was significant for hypertension and hyperlipidemia. He had prior spine surgery for degenerative disc disease. He had no ocular history or allergies. His family history was noncontributory. His medications were amlodipine, vitamin C, CoQ10, losartan, and rosuvastatin.

His vision was 20/20 in both eyes. There was no temporal artery nodularity or tenderness. Pupils were briskly reactive bilaterally with neither anisocoria nor relative afferent pupillary defect. Anterior segment and fundus examinations were normal. The patient had resolution of his TMVL but persistent dysgeusia.

The patient was admitted to the stroke team at Houston Methodist Hospital. Noncontrast computed tomography (CT) of head was normal. Magnetic resonance imaging of the head was normal, including diffusion-weighted imaging. Electrocardiogram and cardiac echocardiogram were normal. Routine laboratory studies were normal, including complete blood count. Serum erythrocyte sedimentation rate was 12 mm/hr and C-reactive protein measured 0.65 mg/dL.

Contrast CT angiogram (CTA) of the head and neck showed a dissection of the distal cervical and proximal petrous portions of the left ICA (Fig. 1A–C). The patient was started on intravenous heparin. He had initially been on corticosteroids, which were discontinued. A digital subtraction catheter arteriogram showed a small intraluminal thrombus distal to the petrous left ICAD, but no other hemodynamic insufficiency was seen in the intracranial circulation. The patient was transitioned from heparin to apixaban (Eliquis) before discharge. The patient returned for follow-up 3 months later and had a

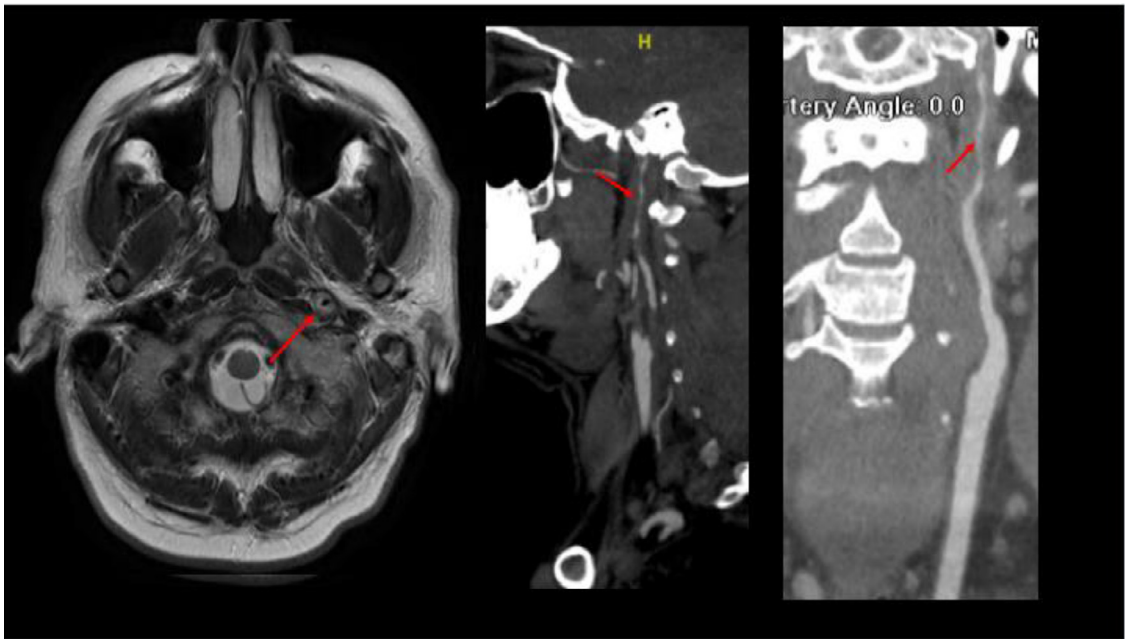

Fig. 1—(A) Coronal computed tomography angiogram (CTA) showing dissection of the cervical and petrous portions of the left internal carotid artery (ICA). (B) Oblique CTA showing filling defects in the distal portions of the left ICA. (C) Oblique digital subtraction angiogram showing long segment narrowing and intramural irregularity in the left ICA.

normal eye examination and complete resolution of his dysgeusia and TMVL episodes.

Patients with TMVL should undergo timely evaluation for embolic or nonembolic etiologies of ischemia. In older patients, evaluation and testing for giant cell arteritis is recommended (e.g., serum erythrocyte sedimentation rate and C-reactive protein, temporal artery biopsy). All patients with ischemic TMVL should undergo a full stroke work-up, including neuroimaging (e.g., acute noncontrast CT of the brain followed by magnetic resonance imaging including diffusion-weighted imaging of the brain. Vascular imaging (e.g., CT or MR angiography) of the head and neck is recommended to evaluate for vascular etiologies, including carotid dissection. In addition, echocardiogram, electrocardiogram, and a carotid Doppler are generally indicated. Older age (more than 65 years), higher blood pressure, longer duration of symptoms, and the presence of comorbidities like diabetes raise the risk of a subsequent stroke in the next 2−7 days[5].

ICADs usually occur distal to the carotid bifurcation (where atherosclerotic disease is known to occur) [2] and are typically cervical in location. Greater than 90% of ICADs arise within 2 cm cranial to the carotid bifurcation in the cervical section (C2) of the ICA, and approximately half of these extend into the petrous portion of the ICA (C3) [2,3].

The presenting symptoms of ICAD include direct local effects (e.g., neck pain, Horner syndrome) and indirect distal effects, which are ischemic thromboembolic or hypoperfusion in nature. The most common ischemic symptoms in ICAD are hemispheric transient ischemic attacks, hemispheric cerebral infarction, and TMVL (i.e., amaurosis fugax) [3,4]. In one study from the ophthalmic literature, TMVL occurred in over half of patients with ICAD [4].

Expansion and extension of the ICAD can cause pseudoaneurysm formation and intramural hematoma [6]. Common local direct symptoms of ICAD include Horner syndrome and neck pain [3,4,6]. Head or facial pain (specifically in the orbital region) or ipsilateral eye pain (as seen in our case) can occur even though the ICAD is located in the neck. This referred pain results from the general visceral afferent (GVA) pain carried on the vagus being perceived by the general somatic afferent (GSA) of the trigeminal nerve nucleus in the brainstem [4,6]. This type of referred pain is similar to the false localizing left arm pain (GSA) in myocardial infarction (GVA) or the referred visceral pain of appendicitis or kidney stones (GVA) in the back or abdomen (GSA).

ICAD can also produce lower cranial nerve palsies, specifically cranial nerves IX, X, XI, and XII [3]. Although cranial nerve IX is the closest to the petrous portions of the ICA, cranial nerve XII palsies seem to be the most common [3]. Involvement of branches of cranial nerves VII, IX, X, XI, and XII have been reported in ICAD [3]. The topographical localization of the dysgeusia in ICAD is thought to be due to glossopharyngeal nerve involvement or the chorda tympani, a branch of the facial nerve (VII) [3].

Ophthalmologists should be aware of the interesting combination of symptoms of TMVL and dysgeusia in ipsilateral ICAD. This is especially important in patients with TMVL who often have completely normal eye examinations. Patients with an ICAD may present to

ophthalmology with symptoms and signs of direct compression (Horner syndrome) or indirect distal thromboembolic/hypoperfusion events (transient ischemic attacks/TMVL). Although the combination of Horner syndrome and TMVL is a well-known constellation for ipsilateral ICAD, the combination of TMVL and dysgeusia is also a highly suggestive symptom complex for ICAD of the petrous ICA even without the Horner syndrome.

Supplementary Materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.jcjo.2020.05.004.

Preston Baker,* Shruthi Harish Bindiganavile,† Ross Taylor,† Nita Bhat,† Andrew G. Lee†
*McGovern Medical School, Houston, TX; †Houston Methodist Hospital, Houston, TX.

Originally received Apr. 5, 2020. Final revision Apr. 23, 2020. Accepted May. 17, 2020.

Correspondence to:
Andrew G. Lee, MD; aglee@houstonmethodist.org.

References

1. Debette S. Pathophysiology and risk factors of cervical artery dissection: what have we learnt from large hospital-based cohorts? Curr Opin Neurol 2014;27:20–8.
2. Downer J, Nadarajah M, Briggs E, Wrigley P, McAuliffe W. The location of origin of spontaneous extracranial internal carotid artery dissection is adjacent to the skull base. J Med Imaging Radiat Oncol 2014;58:408–14.
3. Sturzenegger M, Huber P. Cranial nerve palsies in spontaneous carotid artery dissection. J Neurol Neurosurg Psychiatry 1993;56:1191–9.
4. Biousse V, Touboul P-J, D'Anglejan-Chatillon J, Lévy C, Schaison M, Bousser M-G. Ophthalmologic manifestions of internal carotid artery dissection. Am J Ophthalmol 1998;126:565–77.
5. Givre S., Van Stavern G.P.Amaurosis fugax (transient monocular or binocular visual loss). www.uptodate.com/contents/amaurosis-fugax-transient-monocular-or-binocular-visual-loss. Accessed
6. Debette S, Grond-Ginsbach C, Bodenant M, et al. Differential features of carotid and vertebral artery dissections: the CADISP Study. Neurology 2011;77:1174–81.

Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.

Severe optic neuropathy as the presenting sign of leptomeningeal carcinomatosis from pancreatic cancer

Pancreatic cancer is one of the deadliest cancers in North America with an overall 5-year survival rate of less than 10%.[1] The majority of pancreatic solid tumours metastasize to other organs, mainly to the liver and other peritoneal organs.[1] Leptomeningeal metastasis (LM) or carcinomatous meningitis is infiltration of cancer cells to the leptomeninges and cerebrospinal fluid and is extremely rare in the setting of pancreatic adenocarcinoma with only 19 English cases reported (Supplementary Table 1, available online). There are no previous reports of an optic neuropathy from metastatic pancreatic cancer in the context of LM. Here, we report a case of pancreatic adenocarcinoma leading to LM and a severe optic neuropathy.

A 69-year-old woman with a history of pancreatic cancer presented with progressive, painless vision loss in her left eye. She was diagnosed with T2N1 well-differentiated pancreatic adenocarcinoma 9 years ago, for which she underwent a Whipple procedure followed by adjuvant gemcitabine for 5 months. Her first recurrence at 3.5 years from surgery consisted of low-volume lung nodules that were observed and, when proven indolent, resected and confirmed to be the same adenocarcinoma as the pancreas. Two years later she had radiological evidence of peritoneal metastatic disease and more lung metastasis. She commenced on standard-of-care first-line FOLFIRINOX chemotherapy. Over the next 4 years, she was treated with multiple FOLFIRINOX cycles, demonstrating radiological and biochemical partial responses followed by surveillance periods off chemotherapy and then rechallenge of FOLFIRINOX with progression. She had a repeat lung resection for dominant disease. More recently, she developed radiological signs of bone metastasis, enlarged mediastinal lymph nodes, and bilateral lung nodules consistent with progressive disease.

She developed blurry vision in her left eye 10 days before presentation, and the blurry vision gradually worsened. She had a visual acuity of 20/20 in the right eye and no light perception in the left eye. There was a left relative afferent pupillary defect, and dilated fundus examination was normal in both eyes. Humphrey 24-2 SITA-Fast visual field testing was normal in the right eye. Due to the concern for metastatic disease to the left retrobulbar optic nerve, she underwent urgent magnetic resonance imaging of the brain and orbits with contrast, which showed enhancement of both optic nerves and leptomeningeal enhancement of the corpus callosum, cingulate gyrus, and medial aspect of the temporal lobe (Fig. 1). This was consistent with metastatic disease. A