A 66-year-old Hispanic woman presented to the eye clinic with chronic, progressively worsening vision in the right eye over 2 years associated with right-sided temporal headaches, cutaneous allogynia of the scalp, and jaw claudication. She denied fevers, chills, joint pains, nausea, photophobia, phono-

cutaneous allodynia of the scalp, and jaw claudication. She

over 2 years associated with right-sided temporal headaches,

ical issues similar to those demonstrated by our patient: temporal

is a young woman of Asian descent with a family history of med-

collateral vessels produces the distinctive cranial angiography.

Etiologies for MMD and MMS include idiopathic, iatro-
genic (i.e., radiation), atherosclerosis, and congenital.

Twelve percent of cases are familial, and the disease has

been associated with several polymorphisms of the RNF213
gene, which encodes a gene

Poor perfusion may result in optic nerve ischemia, which can

result in the “morning glory disc” anomaly as the optic nerve
develops. This finding is characterized by chorioretinal pigment changes surrounding the optic disc, an enlarged and funnel-

shaped appearance of the optic disc, and central tuft of glia in

the center of the optic disc.3 Our patient is notable for the

angiography, leading to the diagnosis of moyamoya disease

(MMD). Neurosurgery evaluated the patient and determined

that the risks for undergoing an external carotid-internal

carotid bypass outweighed the benefits at this time.

Discussion

MMD and moyamoya syndrome (MMS) are characterized by a
diffuse narrowing of the internal carotid, middle cerebral, or
anterior cerebral artery lumen.1 The fine tangle of compensatory
collateral vessels produces the distinctive cranial angiography.

(Moyamoya is Japanese for “puff of smoke.”) The classic patient

is a young woman of Asian descent with a family history of med-
cal issues similar to those demonstrated by our patient: temporal

headache, jaw claudication, and progressive vision loss.

Etiologies for MMD and MMS include idiopathic, iatro-
genic (i.e., radiation), atherosclerosis, and congenital.

Twelve percent of cases are familial, and the disease has

been associated with several polymorphisms of the RNF213
gene, which encodes a gene finger protein.2 Although

MMD and MMS are rare, this life-threatening disease is

important to keep in the differential when a patient presents

with ipsilateral optic atrophy.

References

1. Gallagher RJ, Liverman CS, Arnold PM. Papillary thyroid
adenocarcinoma with occipital lobe metastasis 48 years after

therapies and immunotherapy for advanced thyroid cancers.
Mol Cancer 2018;17:51.

3. Stjepanovic N, Velaquez-Martin JP, Bedard PL. Ocular tox-
icities of MEK inhibitors and other targeted therapies. Ann

4. Harrison SR, Tew A, Steven N, Fisher BA. Steroid refractory
dermatomyositis following combination dabrafenib and tra-

5. van Ladingham SW, Puccetti D, Potter H, Gamm D, Diamond
EL, Lucarelli MJ. Necrotizing myositis in a rectus muscle
arising in the setting of long-standing Langerhans cell
histiocytosis and recent dabrafenib treatment. Am J Oph-

6. Karsan N, Barker R, Wren D. Myositis and metastatic mela-

Footnotes and Disclosure

The authors have no proprietary or commercial interest in
any materials discussed in this article.
Fig. 1 — Visual field demonstrates junctional scotoma; 24-2 Humphrey visual field showed decreased sensitivity more temporally in the left eye and a central defect breaking out temporally in the right eye.

Fig. 2 — Computerized tomography angiography (CTA) and angiography with stenotic internal carotid artery (ICA); axial (A), coronal (B) CTA maximum intensity projection images; 3-dimensional volume-rendered CTA reformat (C); and AP projections from right (D) and left (E) selective ICA injections from a digital subtraction angiogram demonstrate an occluded right supraclinoid ICA (blue arrow) and small caliber right middle cerebral artery (green arrow) compared to the patent contralateral left supraclinoid ICA at the carotid terminus (red arrow) and normal caliber left Middle cerebral artery (MCA) (purple arrow).
involvement of the optic chiasm, resulting in a junctional scotoma—a description that we have not seen to date in the English literature.

Medical management involves antiplatelet treatment, although the evidence supporting this practice is limited. Surgical revascularization is offered as well and is often pursued in pediatric patients who are more likely to have the progressive form of the disease.4 Revascularization can be either direct or indirect. Direct involves anastomoses created between the superficial temporal artery and the middle cerebral artery. Indirect involves the use of implanted vascular tissue to promote angiogenesis in the implanted area.5

Sean Michael Rodriguez, Kanwal Singh Matharu, Linda Epner, David Dunaway, Rod Foroozan
Baylor College of Medicine, Houston, Tex.


Correspondence to:
Rod Foroozan, MD; foroozan@bcm.edu.

Crossed-quadrant homonymous hemianopsia in a monocular patient

A 75-year-old male presented to the clinic reporting that upon waking up from a nap 4 days earlier, he noticed he could “only see in certain spots.” He has an ocular history of moderate-stage primary open-angle glaucoma in both eyes, dry eye syndrome, and pseudophakia. He had previously undergone enucleation of his blind, painful right eye due to neovascular glaucoma from a previous central retinal vein occlusion. He was a former smoker, and his medical history is significant for multiple myeloma, bladder cancer, pulmonary embolism, hypertension, and hyperlipidemia.

On initial examination, his visual acuity was 20/25-2 OS with intraocular pressure of 13 mm Hg OS and 3 out of 11 colour plates. He wore a prosthesis OD. Examination of the anterior segment and fundus of his left eye was stable from previously documented examinations, including careful examination of the optic nerve. Optical coherence tomography imaging of the retina was within normal limits. Confrontational visual field testing with a colored target revealed possible left hemi-field loss. Humphrey Central 24-2 Threshold visual field testing revealed new deficits that were not present on testing performed 3 months prior, as seen in Figure 1.

Despite the patient’s monocular status, it was believed that the automated perimetry demonstrated a combination of a left superior quadrantanopsia with a right inferior quadrantanopsia resembling a crossed-quadrant homonymous hemianopsia (CQHH). The patient was sent to the emergency department for urgent stroke evaluation and neurology consultation. Magnetic resonance imaging of the brain revealed bilateral acute occipital lobe infarcts—one infarct located along the left superior calcarine bank, and one infarct located along the right inferior calcarine bank, as demonstrated in Figure 2.

Discussion

The unique entity of CQHH or “checkerboard visual field defect” is exhibited when consecutive or simultaneous occipital lobe lesions occur superior to the calcarine fissure and inferiorly on the contralateral hemisphere.1 Automated perimetry in our monocular patient disclosed a pattern resembling a CQHH or the striking “checkerboard visual field defect”—in this case, the juxtaposition of a left superior quadrantanopsia and a right inferior quadrantanopsia. This pattern is essentially pathognomonic for bilateral occipital lesions.

A suspicion of CQHH should prompt urgent stroke evaluation, particularly in an elderly patient such as ours with vasculopathic risk factors as well as multiple myeloma and bladder cancer, further increasing his risk of thromboembolism. The most common cause of CQHH, as in our patient, is an ischemic or hemorrhagic infarct that may be associated with a thromboembolic event, dissection, or cervical vertebral trauma, but CQHH has also been reported with tumors, migraine, syphilis, necrotizing ependymomyelitis, and demyelinating disease.1–4 Most of the cerebral infarcts that cause CQHH are derived principally by embolization of the calcarine arteries from the P3 branch of the posterior cerebral artery.2 Our patient had bilateral occipital lobe infarcts

References


Footnotes and Disclosure

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