CASE REPORT

Late Presentation of Fibromuscular Dysplasia in an Elderly Female with Branch Retinal Artery Occlusion

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Case Report: Part 1

An 82-year-old female with a history of well-controlled hypertension, irritable bowel disease, mild intermittent asthma, glaucoma, and macular degeneration presented to her ophthalmologist with three days of monocular vision loss. She lives alone and is independent with all of her activities of daily living. Her ophthalmologist diagnosed her with subacute branch retinal artery occlusion (BRAO) and requested that her primary care physician see her that same day.

At her primary care physician's office, a detailed history confirmed no personal or family history of stroke, coronary artery disease, or vascular disease. She does not smoke and recent lipid profile and fasting glucose were normal. Her medications include budesonide/formoterol inhaler, albuterol inhaler, calcium-vitamin D, coenzyme Q, lisinopril, bimatoprost ophthalmic drops, a multivitamin, and red rice yeast. Physical exam revealed normal vital signs, normal heart sounds, no peripheral edema and no carotid bruits. There was no tenderness along the temporal area bilaterally, and she denied headaches or jaw pain. An electrocardiogram (ECG), echocardiogram, carotid ultrasound, MRI/MRA Brain, lipid panel, hemoglobin A1c, and ESR were ordered; she was started on clopidogrel and atorvastatin.

Discussion: Part 1

The retinal artery is a branch off the ophthalmic artery, which is the first branch of the intracranial portion of the internal carotid artery. The retinal artery is the main blood supply for the inner retina and surface of the optic nerve. A central retinal artery occlusion (CRAO) or branch retinal artery occlusion (BRAO) can cause a painless loss of monocular vision and is considered a cerebrovascular ischemic event. The incidence is 1-10 in 100,000 people.[1,2] Most patients with CRAO and BRAO have several vascular disease risk factors including hypertension, diabetes, or tobacco use. The source of the cerebrovascular event is most commonly carotid artery atherosclerosis, cardiogenic emboli, or small artery disease of the retinal artery.[3] Older patients must be ruled out for giant cell arteritis. More unusual etiologies of CRAO and BRAO include other vascular disorders such as dissection; fibromuscular dysplasia; radiation damage; or hematologic disorders, such as hypercoagulable states or leukemia; or autoimmune conditions. Treatment includes cardiovascular risk factor modification and etiologic specific therapies (i.e., anticoagulation if there is a cardioembolic source or carotid endarterectomy if there is significant carotid artery stenosis).[4] Atherosclerotic disease should be treated with a statin and anti-platelet therapy. Thrombolytic therapy has not been found to be beneficial in acute presentations of retinal artery occlusion.[5]

Case Report: Part 2

All of the patient's studies were unremarkable with the exception of the MRI/MRA Brain (Figure 1) that showed "beading of the cervical segments of both internal carotid arteries that may represent fibromuscular dysplasia. No high-grade proximal stenosis or occlusion of the intracranial vessels. The ophthalmic arteries are patent." Subsequent ultrasounds of the renal arteries revealed no evidence of fibromuscular dysplasia. Her vision returned to normal within one week of presentation.

Discussion: Part 2

Fibromuscular dysplasia (FMD) is a non-inflammatory, non-atherosclerotic arterial disease that predisposes to stenosis, occlusion, aneurysm, or dissection. The renal arteries and internal carotid arteries are most commonly involved. Clinical and physical manifestations vary depending on the arterial segment involved, but hypertension, stroke, headaches, neck pain, tinnitus, and carotid bruits are commonly reported.[6] There is a strong female predominance, and it is most commonly diagnosed in middle age.[7] The mean age at diagnosis is 51.[7] Given many patients are asymptomatic or present with vague symptoms, it is frequently not diagnosed or misdiagnosed; thus the true prevalence of the disease is yet to be determined. The differential diagnosis for FMD includes vasculitis, atherosclerotic disease, and connective tissue disorders.[8]

This patient's BRAO was felt to be secondary to FMD as opposed to atherosclerosis or small vessel disease. Her only vascular risk factor was hypertension, which had been very well controlled on Lisinopril 10mg daily. Nonetheless, given her age, atherosclerotic disease may have been a contributing factor and she was treated accordingly. It is suspected she has FMD of her retinal artery; however, this would need to be diagnosed via catheter-based digital subtraction angiography (DSA), the current gold standard for FMD.[9] This procedure was felt to be too invasive by the patient. Additionally, the results would not have changed the therapeutic management for this patient. The FMD of her internal carotid artery likely produced a downstream event in the branch retinal artery that led to the vision loss.

The treatment of FMD varies depending on the arterial segment and extent of disease. Renal artery involvement often leads to hypertension and requires stenting or balloon angioplasty. The management of carotid artery FMD is similar to the management of carotid artery atherosclerotic disease. Symptomatic lesions that are amenable to intervention are treated with percutaneous transluminal balloon angioplasty.[10] Many symptomatic patients are managed medically given the distal nature of their disease, risks of endovascular therapy, and extent of disease.[11] Aneurysms and dissection also require surgical intervention. Asymptomatic individuals should be monitored on antiplatelet therapy.

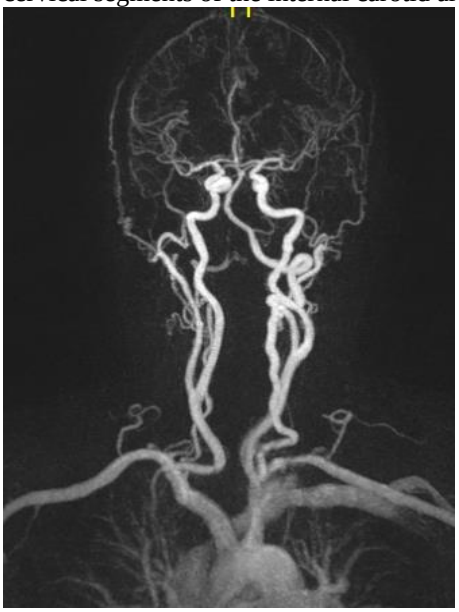
Conclusion

This patient's presentation is unusual given the rarity of seeing both BRAO and FMD in clinical practice. Additionally, FMD infrequently first presents in older age and is rarely isolated to the intra-cranial portion of the internal carotid arteries.[12,13] FMD of the retinal artery is rare; to date, only 5 case reports have been published as well as 1 case report of FMD in the celioretinal artery.[13-18]

Vision loss is not a typical presentation of a stroke and this patient warranted additional investigation given how robust, healthy, and independent she was. The diagnosis of FMD would have been missed if this patient had been judged by her age alone, assuming her BRAO was from atherosclerotic disease. However, a thorough workup to evaluate non-atherosclerotic disease was pursued and revealed an interesting etiology.

Figures and Images

Figure 1. MRI/MRA of the brain showing beading of the cervical segments of the internal carotid arteries



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