Management of an extracranial internal carotid artery aneurysm secondary to relapsing polychondritis

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ABSTRACT
Extracranial internal carotid artery (EICA) aneurysms make up 1% of peripheral aneurysms and less than 1% of patients who have relapsing polychondritis develop aneurysms. A 39-year-old man with relapsing polychondritis presented with right neck pain. Initial computed tomography angiography demonstrated a 16-mm right EICA aneurysm with growth to 25 mm after 2 months. A right EICA aneurysmectomy, external carotid artery to ICA transposition, and internal jugular vein patch of the common carotid artery was performed with symptom resolution. The inflammatory nature of the underlying disease, aggressive expansion, and symptomatic state warranted open repair and we recommend life-long monitoring given the rarity of this case. (J Vasc Surg Cases and Innovative Techniques 2020;6:576-9.)

Keywords: Internal carotid artery (ICA); Relapsing polychondritis; Carotid artery diseases; Extracranial aneurysm; Open surgical management

Extracranial internal carotid artery (EICA) aneurysms have approximately a 1% incidence in patients with peripheral arterial aneurysms.1 These patients typically present with pain, a pulsatile mass, or cranial nerve palsies.1 The most common underlying causes related to these true aneurysms include atherosclerosis, trauma, fibromuscular dysplasia, infection, connective tissue disorders, and large vessel vasculitis.1-3 Relapsing polychondritis is a rare rheumatologic disorder characterized by cartilage and tissue inflammation throughout the body and can present with inner ear inflammation with hearing issues, arthritis from joint inflammation, keratitis/uveitis/scleritis from visual involvement, and nasal cartilage inflammation manifesting as rhinorrhea or epistaxis.4 Fewer than 1% of these patients develop aneurysmal disease; however, reports in literature are predominantly aortic aneurysms.5 The proposed pathophysiology is thought to be antibodies to type II collagen, which are present in 50% to 60% of these aneurysms and its association with HLA-DR4.6 We present a unique case of a patient with a symptomatic EICA aneurysm in the setting of relapsing polychondritis who had no other aneurysmal disease risk factors. Consent was obtained from the patient for discussion of this case.

CASE DESCRIPTION
A 39-year-old man on multiple immunosuppressive agents (40 mg/d of prednisone, 10 mg/d of leflunomide) for relapsing polychondritis presented to an outside hospital with right neck pain. The diagnosis of relapsing polychondritis had been made for this patient several years before as he had sustained severe migraines and right eye pain with computed tomography scan demonstrating pinna thickening and evidence of scleritis consistent with this disease process.5 The computed tomography scan demonstrated a 16-mm right EICA fusiform aneurysm without intramural thrombus/inflammatory changes (Fig 1). The patient had elevation in inflammatory markers with erythrocyte sedimentation rate at 55 mm/h and C-reactive protein at 124 mg/L. Given that the aneurysmal degeneration was likely due to a vasculitis associated with his relapsing polychondritis, it was felt that an acute surgical intervention would be difficult owing to tissue friability and the risk for damage to surrounding cranial nerves.
After discussion with rheumatology, it was felt he could be stabilized and have symptom relief with modifying his immunosuppressive medications. He was given analgesics, a dose of intravenous solumedrol in the emergency department, and had an increase in his prednisone dose to 60 mg/d. Two months later, he presented with severe right neck pain, an enlarged 25-mm right EICA fusiform aneurysm, and new intramural thrombus with inflammatory changes (Fig 2). His aneurysm was presumed to have a vasculitic component refractory to immunosuppressive agents and the patient was transferred to our institution for further evaluation.

The patient was brought to the operating room and he required nasotracheal intubation and mandibular subluxation by an otolaryngologist given that his aneurysm extended to
A standard oblique incision was made on the anterior border of the sternocleidomastoid muscle extended from the sternal notch to the earlobe. He underwent a right EICA aneurysmectomy, right external carotid artery to internal carotid artery transposition, and internal jugular vein patch of the right CCA (Figs 3 and 4). Unfortunately, the patient’s specimen was misplaced so no final pathologic diagnosis was obtained; however, the patient was found to have negative blood cultures from initial presentation. Postoperatively, the patient had complete resolution of his presenting symptom, which was neck pain. He remained neurologically intact. The patient was monitored with duplex ultrasound examinations at 1 day, 1 month, 3 months, and 12 months, which all demonstrated vessel patency.

**DISCUSSION**

EICA aneurysms are treated to avoid thromboembolic events, cranial nerve compression, and aneurysm rupture. We believe this is a unique case of surgical repair of an expanding, EICA aneurysm in a patient with relapsing polychondritis. Patients with relapsing polychondritis have a propensity to develop a systemic vasculitis with involvement of large, medium, or small arteries. Aneurysms are rare and reports have been predominantly aortic aneurysmal degeneration.

Treatment options should be individualized. In this case, immunosuppressive medical management with steroids and monoclonal antibodies was the first step. To our knowledge, aneurysmal degeneration is not common on immunosuppression, so this was the initial management in the acute inflammatory phase before completing a definitive repair. Given the inflammatory nature of the underlying disease, further aneurysmal degeneration despite immunosuppressive therapy, and symptomatic state, we felt open resection was the safest and best option. At the time of repair, an autogenous venous conduit was used with internal jugular vein given the size match to the aneurysmectomy defect. In case the aneurysm was deemed to be mycotic, the vein patch allowed an immunosuppressed patient to have vein harvested from the same incision as the repair.

Previous series have reported open repair of EICA aneurysms are durable with no evidence of stroke, neurologic events, or recurrent aneurysms at 7 years of follow-up. At 12 months of follow-up in this case, the repair remains patent without any evidence of further aneurysmal dilation and the patient remains asymptomatic. Given the systemic manifestations of relapsing polychondritis and multiple symptoms this patient has exhibited, the patient will remain on immunosuppression for life.

**CONCLUSIONS**

EICA aneurysms are rare with open and endovascular treatment options available based on the underlying pathophysiology. In this case, open surgical repair without prosthetic material was possible where durability on long-term immunosuppression is unknown. Given the rarity of this case and propensity for aneurysmal degeneration, we recommend life-long monitoring with subsequent carotid duplex ultrasound examinations to be completed every 12 months. Larger series with longer follow-up will need to be established to truly understand postoperative course of patients who have had EICA aneurysms treated.
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Fig 4. Artist depiction of stepwise reconstruction of the extracranial internal carotid artery (EICA) aneurysm starting with EICA aneurysmectomy, external carotid artery to internal carotid artery transposition, and internal jugular vein patch of the common carotid artery. (Credit: Calla Heald.)