True extracranial carotid artery aneurysm in a child

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Isolated true aneurysm of the extracranial carotid artery is a rare entity in the pediatric population, with nine reported cases found in the literature. Contrary to adult carotid aneurysms, which are often due to atherosclerotic disease, pediatric aneurysms are more likely to be the result of congenital malformations, connective tissue diseases, or systemic inflammatory conditions. We present the case of a 10-year-old boy with an isolated true aneurysm of the internal carotid artery and a review of the literature. (J Vasc Surg Cases 2015;1:110-2.)

Isolated true aneurysm of the internal carotid artery (ICA) is a rare entity, especially so in the pediatric population, with nine reported cases found in the literature.[1-9] We present the case of a 10-year-old boy with an aneurysm of the ICA. We have obtained consent to publish from the patient and his family.

CASE REPORT

A 10-year-old boy presented to the emergency department with a pulsatile mass in his neck. He was afebrile and had no other complaints aside from recently recovering from a mild upper respiratory infection. The physical examination was significant only for a left-sided submandibular neck mass. No skeletal abnormalities, hyperflexible joints, or abnormal facies were noted. His medical history was significant only for mild intermittent asthma, he was up to date on his vaccinations, had never had head or neck surgery, and there was no history of trauma to the area.

An ultrasound study at another facility revealed a ∼3.0-cm × 2.5-cm aneurysm of the left ICA, prompting referral to our institution. Magnetic resonance imaging confirmed the presence and location of the lesion in the proximal ICA (Fig 1, *a*) with a 5.5-mm normal distal ICA, a 7-mm common carotid artery, and a complete circle of Willis. Genetic counseling was consulted. The risks of surgery vs observation were explained to the family, including rupture, cranial nerve injury, stroke, and death. The decision was made to proceed with open surgical repair, and consent was obtained.

The patient was taken to the operating room and placed under general anesthesia. Exposure of the carotid bifurcation was obtained via an oblique skin incision made ∼2 cm below the angle of the mandible, and anatomic landmarks were as expected.

Cranial nerves X and XII were identified, carefully dissected away from the aneurysm, and protected for the duration of the repair (Fig 1, *b*). Proximal and distal control was obtained, and systemic heparinization was achieved.

The aneurysm was resected and the repair was performed with an interposition reversed great saphenous vein graft (Fig 2). Cerebral oximetry demonstrated minimal change, and no shunt was used. A technically adequate result was confirmed by completion intraoperative duplex imaging.

Postoperatively, the patient had no neurologic deficits and was discharged home, after an uncomplicated postoperative course, with aspirin therapy (81 mg daily). On routine follow-up at 2 weeks, he was healing appropriately and continued to have no neurologic deficits.

Genetic screening for Loeys-Dietz type II (*TGFBR2*) was negative. Histopathology included staining with hematoxylin and eosin, Movat, Verhoeff-Van Gieson, and Alcian blue. Findings were significant for thickened intima with destruction of the media, increased ground substance, and a lymphocytic and histiocytic infiltrate (Fig 3). Gram stain and culture of the specimen revealed no organisms. Assessment with echocardiography and follow-up carotid duplex imaging at 3 and 6 months was recommended, but the patient was lost to follow-up.

DISCUSSION

Extracranial carotid artery aneurysms are a rare entity, accounting for <1% of all carotid procedures at large referral centers.[10,11] They are even less common in the pediatric population, with <40 cases reported.[1-3,12,13] When pseudoaneurysms and mycotic aneurysms are excluded, the incidence of true extracranial carotid aneurysms in children is exceedingly low.[1] According to our review, nine cases have been reported.[1-9] The differential diagnosis for a child presenting with a neck mass should include more common etiologies, such as lymphadenopathy or cystic lesions,[14] but ruling out an aneurysm is imperative, particularly if the mass is pulsatile.

Contrary to true carotid aneurysms in adults, most of which are caused by atherosclerotic disease, pediatric patients are more likely to endure different disease processes.[1-9,15] Although data are limited to case reports and a few case series, chronic atherosclerotic disease clearly does not have the same effect on the pediatric population as it does in adults. Three aneurysms were reported in the setting
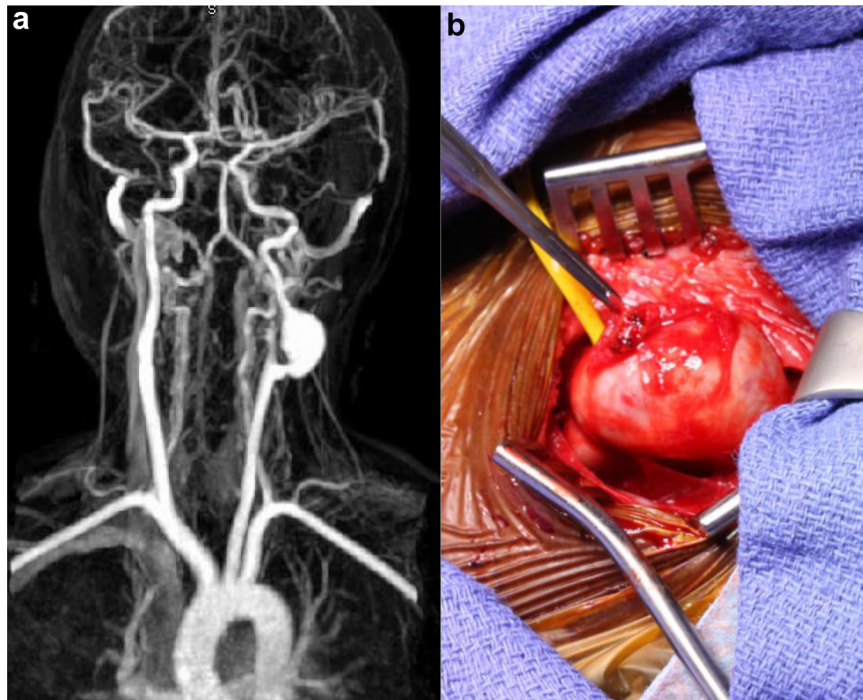
Fig 1. a, Aneurysm of the internal carotid artery (ICA) on magnetic resonance angiography. **b,** Intraoperative exposure of the aneurysm sac.
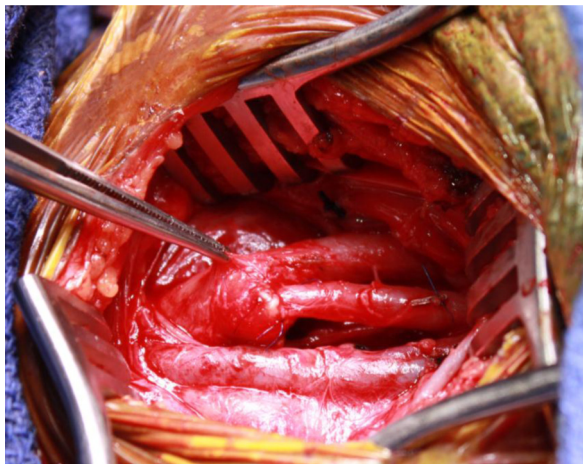

Fig 2. Repair of internal carotid artery (ICA) aneurysm with interposition reversed great saphenous vein graft.

of a pre-existing condition—Behçet disease, Loeys-Dietz syndrome type II, and type IIb hyperlipoproteinemia[3,5,7]— and the six remaining cases were attributed to a congenital anomaly or an unknown etiology.[1,2,4,6,8,9]

The initial diagnosis of a carotid aneurysm may be made with ultrasound imaging, and in the absence of life-threatening hemorrhage necessitating immediate repair, magnetic resonance imaging with angiography is helpful for operative planning. Computed tomography should be avoided to minimize exposure to radiation but may be indicated if expeditious imaging is required or magnetic resonance imaging is not tolerated. Depending on the location of the lesion, particularly if it is high in the skull base, involvement of other specialties, such as otolaryngology, may be necessary.[2]

Further etiologic workup should be continued in parallel with treatment planning, and other systemic findings, such as malaise, fever, or abdominal pain, should elicit consideration of systemic inflammatory conditions such as Takayasu arteritis, Kawasaki disease, and Behçet disease.[7] Characteristic facies and elongated or distorted extremities may suggest rare aneurysmal conditions and connective tissue disorders such as Marfan (*FBN1*), Loeys-Dietz (*TGFBR1*, *TGFBR2*), and Ehlers-Danlos (*COL5A1*, *COL5A2*, *COL3A1*) syndromes.[3] In any case, a genetic counselor should be consulted early to facilitate testing for these mutations.

Our patient was previously healthy except for a mild viral respiratory illness a few weeks prior. He no characteristic phenotypic findings to suggest a connective tissue disease, thus genetic screening was only performed for Loeys-Dietz type II (*TGFBR2*). Histopathologic findings in our patient were similar to those reported for Loeys-Dietz-related aneurysms but were ultimately non-diagnostic, especially in the setting of a negative genetic screen.[16] Our suspicion for a systemic vasculitis was low; therefore, inflammatory markers, such as C-reactive protein and the erythrocyte sedimentation rate, were not checked. In addition, an extensive workup was not performed given the lack of cutaneous findings or a family history suggestive of collagen-vascular disease.
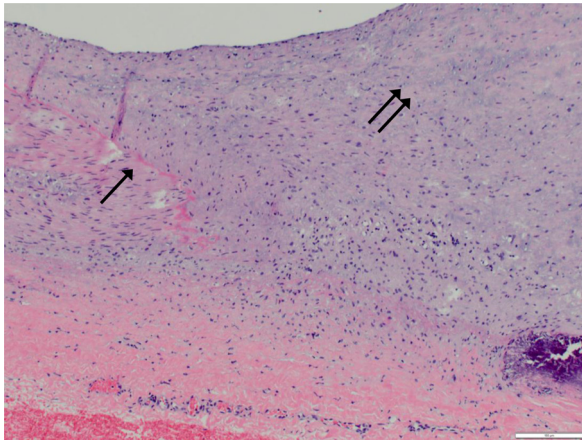
Fig 3. Minimal remnants of normal elastic media (*single arrow*) and thickened intima with increased ground substance (*double arrow*) are seen at original magnification ×20 with hematoxylin and eosin stain.

Operative repair should be performed for carotid aneurysms given the high risk of stroke and a reported rupture rate as high as 40%.[1,17,18] Although most adult patients tend to be treated with aneurysm resection and patch closure or interposition grafting, pediatric patients have been treated by more variable techniques.[1,2,10] The nine cases reviewed consisted of 3 ligations (1 after failed intervention, 1 for good collateral flow, 1 Behçet patient with multiple recurrent aneurysms), 2 vein grafts, 1 polytetrafluoroethylene graft, 1 resection with primary end-to-end anastomosis, 1 transmandibular resection (no further details available), and 1 with observation.[1-9]

Overall, long-term outcomes of pediatric carotid aneurysm repairs are unknown. Antar et al[7] reported favorable outcomes at 6 years in a patient treated with carotid artery ligation, but no other long-term data were reported. We believe that vein grafting provides a theoretical advantage over repair with prosthetic material given the potential for concomitant vessel and graft growth as the child develops. We considered using femoral vein for our repair, but vein mapping revealed an adequate saphenous vein. The radial artery was not considered given the risk of arterial vasospasm and stroke.[19] The hypogastric artery has been reported as a suitable conduit for renal artery grafting in children, but this is not as feasible for carotid repair because it would require an additional abdominal operation.[20]

Follow-up should include repeat duplex imaging and screening for additional aneurysms. Additional workup, such as echocardiography to rule out coronary aneurysms and ophthalmologic consultation to rule out ocular manifestations of connective tissue disease, should be considered if there is a high suspicion.

CONCLUSIONS

Pediatric extracranial carotid artery aneurysms are a rare entity and little is known about their long-term prognosis. Once more common etiologies of neck mass have been ruled out, the diagnosis can be confirmed and operative planning aided by ultrasound imaging and magnetic resonance imaging. High rates of rupture and stroke have been reported, and we believe that urgent repair should be performed, preferably with autologous vein grafting.

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