Isolated Extra-cranial Internal Carotid Artery Aneurysm in a Young Adult with Eale’s Disease

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Abstract
A young male patient, a known case of Eale’s disease presented with a large isolated aneurysm of the extra-cranial part of internal carotid artery. Detailed investigation failed to establish the etiology of the aneurysm, and it is possible that the underlying pathology may be the same as that of Eale’s disease. Eale’s disease is an idiopathic vascular disease of the peripheral retina and we hypothesize that aneurysm in this patient may be associated with Eale’s disease.

INTRODUCTION
Isolated aneurysm of the extra-cranial part of internal carotid artery (ICA) is very rare in comparison with the cranial part. It is commonly caused by atherosclerosis, in an elderly individual, who is typically a smoker and hypertensive. Common presentations include pain, embolism and rupture. Management includes surgery or covered stents. We report here a case of large isolated extra-cranial internal carotid aneurysm in a young individual who earlier had loss of vision in the right eye due to Eale’s disease. Cause could not be identified, possibly aneurysm may be associated with Eale’s disease.

CASE REPORT
A 19 years male presented to the surgeon with a slowly growing pulsatile mass in the right side of the neck of one year duration. The patient earlier had two episodes of gradual onset loss of vision. First episode, occurred 24 months back, and was diagnosed to have Eale’s disease (Fig. 1) by an ophthalmologist. He was treated with steroids and empirical antitubercular drugs, and vision improved to near-normal in two months of time. The last episode, was two months prior to noticing swelling in the neck. Ultrasonogram, revealed a 7.5 x 2.6 cm aneurysm of extra-cranial part of ICA along with multiple mural thrombi and intimal calcification (Fig. 2). Patient was referred to us for angiography. Physical examination
revealed a healthy appearing normotensive young male. A 5x2 cm non-tender, pulsatile mass was palpable in the right superior cervical region immediately below the angle of mandible. There was no similar mass elsewhere in the body. Ophthalmologic examination, this time revealed a scar in the retina and vision was normal. Oral and nasopharyngeal examinations were entirely within normal limits, as were those of neurological and systemic examinations. Routine biochemical and hematological investigations were normal. Antinuclear antibodies, rheumatoid factor, and ANCA were negative.

Carotid angiography was performed. A large aneurysmal dilatation of the right ICA was documented 5-6 mm distal to the carotid bifurcation extending up to the base of skull (Fig. 3). Intracranial segment was normal. Left internal carotid artery was normal. Diffuse nature of the disease precluded endovascular stent graft implantation. Patient refused to undergo surgery. He was prescribed, aspirin and warfarin. On follow up, one year later, the size of the aneurysm remained same and was painless.

**Discussion**

Aneurysm of the extracranial internal carotid artery is quite uncommon in comparison to the intracranial carotid artery and of aneurysm arising elsewhere in the arterial system. Several pathological conditions may result in the development of the aneurysm, such as atherosclerosis, trauma (including prior surgery) dissection, infection, inherited tissue disorders such as Marfan’s syndrome, Ehler-Danlos syndrome, and cystic medial necrosis, large vessel vasculitis such as Takayasu’s arteritis, Behcet’s disease, and fibromuscular dysplasia. Fibromuscular dysplasia, in particular, demonstrates a predilection for the carotid vessels.

Rarely, extra-cranial carotid artery aneurysms have been reported, without any of the above pathological process. Unilateral, single vessel involvement is the rule in these cases in contrast to systemic bilateral vessel disease. Such patients are also characteristically young to middle aged adults, where as atheromatous and fibromuscular dysplasia typically present in the 40 to 70 year age group.

Most patients are asymptomatic or present with pulsating mass in the neck. However, focal neurological symptoms causing, TIAs amurosis fugax or retinal infarction have been reported in 15 to 67% of patients. Symptoms are usually attributed to embolization.

Although the literature about the natural history of this disease remains limited, treatment of aneurysm is recommended because of the potential for morbidity and mortality from a neurological event, local pressure complication and rupture.

Surgery is the mainstay of therapy, which includes, carotid ligation, resection with reconstruction using saphenous vein or synthetic grafts and cervical to intracranial carotid artery bypass. However one can attempt endovascular techniques in suitable cases. Our patient had diffuse disease involving the whole extent of extra-cranial segment rendering it unsuitable for endovascular stent grafting. The patient refused to undergo surgery.

Eale’s disease is an idiopathic obliterative vasculopathy that usually involves the peripheral retina of young adults. It is believed to be primary non-inflammatory disorder of the peripheral retinal vessels namely shunt vessels and causes recurrent hemorrhages in the retina, vitreous humor and ischaemic changes in the eye. Patient commonly presents with blurring or loss of vision in one eye and is more common in India. Associations with tuberculosis and multiple sclerosis have been suggested. There is no specific treatment. Steroids and antitubercular therapy have been tried with varying success rate. Extra-ocular manifestations of Eale’s disease are rare and limited to the central nervous system, association with myelopathy and stroke has been reported.

Thorough Medline search did not reveal association with an arterial aneurysm. It is possible that the underlying pathophysiology of aneurysm may be similar to Eale’s disease, that vasculopathy of vasa vasorum in the tunica media of internal carotid artery may have resulted in ischaemia, weakness of the arterial wall and subsequently aneurysm formation.

**References**

3. Schievink WI, Piepgras DG, Mc Caffrey TV, et al. Surgical treatment of extra cranial internal carotid artery dissecting...


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**Announcement**

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