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Case Report

Carotid artery aneurysm associated with Marfan Syndrome: A case report



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1. Introduction

Incidence of extracranial carotid aneurysms is low.

Usually the lesion develops from internal carotid artery, whereas only 2% of lesions develop from the external artery.

The main causes are atherosclerosis and traumatic events.¹

Occasionally in Marfan syndrome an extracranial carotid aneurysm can occur.²

Here we describe the case of a 62-year-old woman with an aneurysm of the right internal carotid artery, and revise the techniques used to treat it nowadays.

2. Case report

A 62 year-old woman, affected by Marfan Syndrome, was admitted to our Hospital due to an aneurysm of the right extracranial internal carotid artery.

In the past medical history we found clinical features of Marfan syndrome, both cardiac (mild aneurysmatic dilation of the aortic bulb with a diameter of 46 mm, mild-severe mitral insufficiency and mild tricuspid insufficiency) and ocular (glaucoma, right cataract and previous interventions for bilateral retinal rupture and left cataract). Medical history was nonetheless not significant like family history.

At the clinical examination the patient presented a right painless, elastic and pulsing laterocervical mass with a diameter of 2.5 cm.

The lesion was completely asymptomatic.

No previous history of neurological symptoms or cervical trauma was reported. No symptoms were identified possibly related to mycotic infections.

Diagnosis was made by US scan that demonstrated the origin from the carotid bifurcation, but did not identify the vessel mainly involved.

So the patient underwent a CT scan of the sovraortic trunks, that demonstrated a very coiled kinking of the right internal carotid artery, immediately after its origin along with an aneurysm 20 mm in diameter with an intraluminal thrombus. At the same level an atherosclerotic plaque was described, without hemodynamic interference. No other vascular lesions were identified (See Figs. 1 and 2).

The patient underwent surgery under general anesthesia with intraoperative SEP (somatosensory evoked potential) monitoring. The procedure was performed through the usual laterocervical access, consisting of aneurysmectomy with end-to-end anastomosis (See Figs. 3–5).

No neurological intraoperative or postoperative events were observed.

The postoperative course was uneventful and the patient was discharged after four days with antiplatelet agents.

At five years of follow up the patient is asymptomatic and still under antiplatelet agents; the Duplex Doppler Ultrasonography demonstrates a regular flow through the carotid artery.

Up to now the patient hasn't presented new signs of Marfan syndrome.

3. Discussion

3.1. Incidence

As mentioned, incidence of extracranial carotid aneurysms is low.

DeBakey's group described only 37 carotid aneurysms out of a total of 8500 aneurysms which underwent surgery over 20 years.³

In a literature review Schechter reported 853 cases till 1977.⁴

In more recent literature only case reports of no more than 2 patients are reported.

The first one to describe a successful intervention for carotid aneurysm (a proximal ligation) was Cooper in 1808: the patient died 13 years after the procedure.

The first reconstructive intervention was performed in 1952; since then the number of cases reported has increased significantly.

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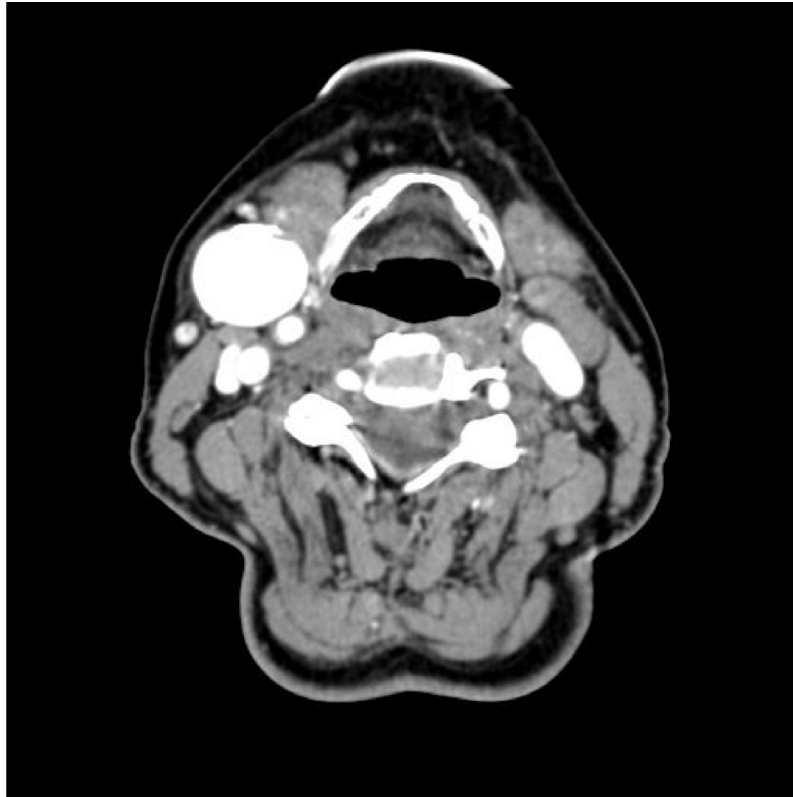


Figure 1. CT scan of the carotid aneurysm.

At present about 1% of the interventions performed on extracranial carotid artery are due to aneurysms.^{1,5}

3.2. Etiology and natural history

The most relevant causes are atherosclerosis and traumatic events.

Atherosclerotic aneurysms^{6,7} are usually found in hypertensive elderly patients. They are usually fusiform and involving the bifurcation more frequently than other sites. They can be bilateral and an intracranial carotid aneurysm with the same pathogenesis can coexist.

Traumatic events⁵ represent the second more common cause. After a blunt trauma or a distractive trauma such as in case of sudden hyperextension of the neck along with rotation there can be a damage to the middle layer of the vessel. Traumatic aneurysms can also occur after carotid surgery. Aneurysms in such cases are more often saccular and they usually involve the common carotid or the distal part of internal carotid artery beyond the base of the skull. This happens because the carotid artery is quite fixed at the cranium in its passage through it, so that the distal tract is at a higher risk of damage.⁸

Among less frequent causes there are syphilis, cystic medial necrosis, muscular hyperplasia⁹ and arteritis such as polyarteritis nodosa, Takayasu arteritis and Wegener arteritis.

We have to cite also mycotic¹⁰ aneurysms, occasionally caused by infected embolism in a patient affected by bacterial endocarditis or more frequently by cellulitis or peritonsillar¹¹ or mastoid abscess.

Carotid aneurysms can also occur after irradiation for malignant tumors of the neck; they can be rarely congenital (in this latter case they are usually bilateral).^{12,13}

Finally a carotid aneurysm can occur in Marfan Syndrome.

The loss of elastic tissue within the arterial wall with subsequent weakening is the pathogenic mechanism underlying aneurysms in Marfan syndrome.

Winslow recorded that 4 out of 5 patients treated conservatively died because of a complication: thrombosis, embolization or rupture.

3.3. Clinical presentation

As in the case here described, the more common clinical presentation of a carotid aneurysm is a pulsing, otherwise asymptomatic laterocervical mass.

Even if the aneurysm is rapidly expanding, the patient can remain asymptomatic.

In most cases, diagnosis is quite simple, but carotid aneurysms have to be differentiated from laterocervical lymphadenopathy, branchial cysts and tumors of carotid body.

In case of compression on oropharynx a carotid aneurysm can simulate a peritonsillar abscess.

Moreover every anatomical anomaly that pushes onto surface carotid bifurcation can be clinically similar to a carotid aneurysm.

3.4. Diagnosis

In the most difficult cases such as for the distal extracranial internal carotid aneurysms radiological exams are mandatory.

Diagnosis can be based on US scan and in some cases also CT scan and/or MRI.⁵

Duplex Doppler Ultrasonography is the first exam to do because it demonstrates carotid bifurcation in almost all cases. A plain X-ray often detects a soft mass possibly with calcifications on its walls.

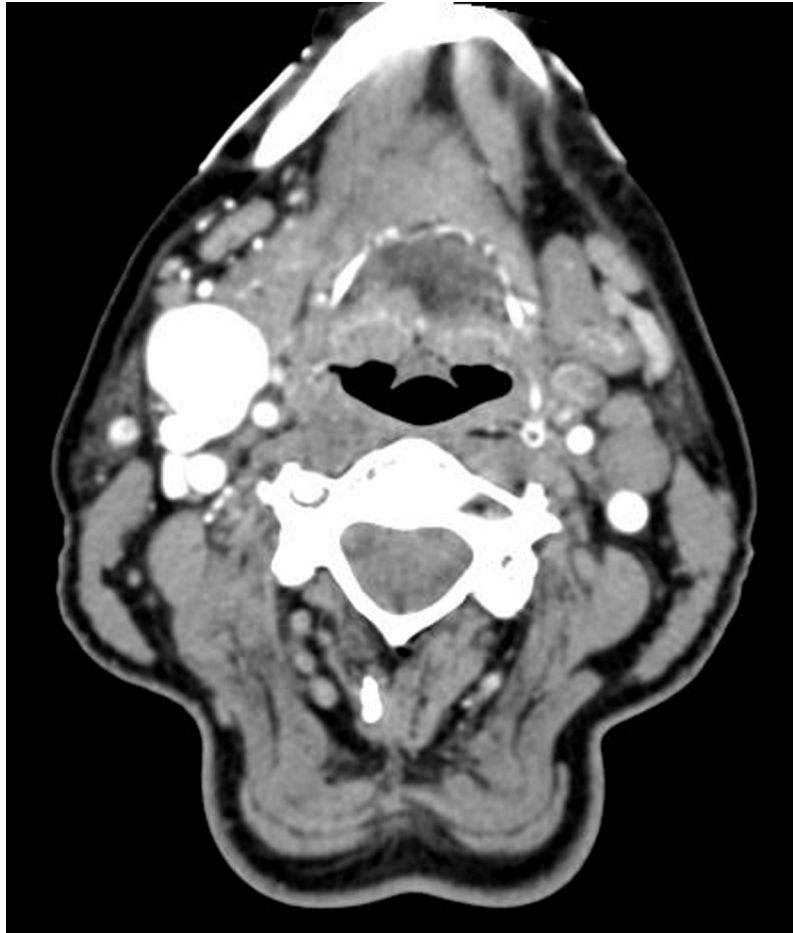


Figure 2. CT scan of the carotid aneurysm next to the bifurcation.

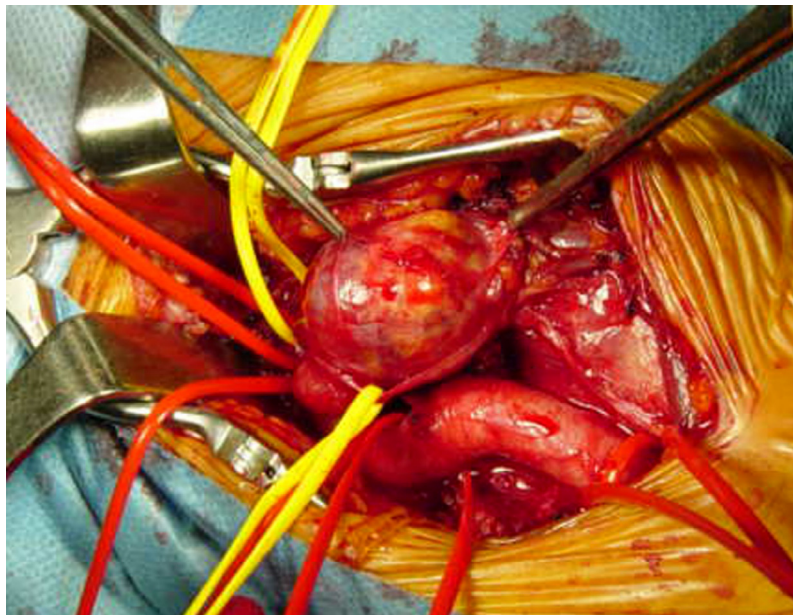


Figure 3. The carotid aneurysm after isolation of all arterial vessels in the operative field.

Even if CT scan or MRI are more important nowadays, diagnostic arteriography remains an important exam in order to establish the right kind of treatment. In fact, arteriography evaluates carotid

bifurcation, morphology of lesion, proximal and distal necks, presence of stenosis or kinking.



Figure 4. The operative field during reconstruction after resection of the aneurysm.

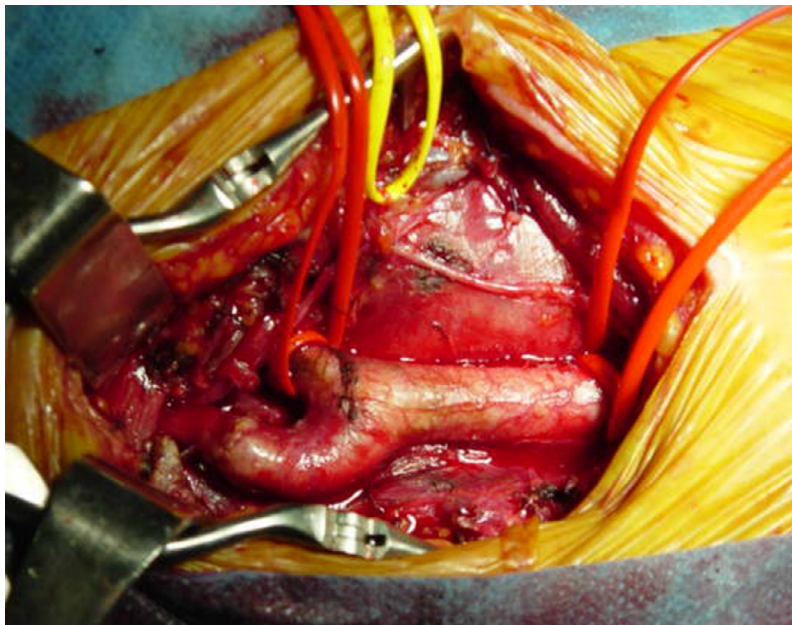


Figure 5. The final result of the reconstruction.

3.5. Treatment

Treatment of such lesion is usually surgical^{14–19}: only a few patients undergo conservative treatment (usually patients with important contraindications to surgery with asymptomatic small aneurysms). In fact conservative treatment carries important morbidity, in most cases due to neurological complications or rupture.

Surgical treatment with only ligation was successfully performed for the first time more than 2 centuries ago and it had been the only working option till the half of the last century, when reconstructive surgery was introduced.

Ligation has a high risk of postoperative stroke independently on its site. Even in more recent literature, some authors report a mortality rate greater than 50% and a relative weight of neurological morbidity which is decreasing over time.

In symptomatic aneurysms, when a distal anastomosis or suture is not technically feasible, ligation is the only option.

Two methods have been designed in order to reduce risk of neurological complications after ligation: one consists of gradual occlusion of artery with Selverstone clamp in order to gain development of collateral vessels before complete occlusion is obtained. During this period anticoagulant agents are administered and at the end ligation is performed at the same site where clamp was positioned.

The other method is trying to estimate risk of cerebral damage before ligation, performing it only in patients with low risk. McIvor reported 5 cases of stroke (2 of them fatal) out of 16 patients treated with ligation after a negative balloon test.

Thus, current opinion is that, even with cautions, the high risk of cerebral damage after ligation mandates reconstructive surgery when not strictly contraindicated.

Among different reconstructive procedures the most diffuse is aneurysmectomy with end-to-end anastomosis. It is often feasible, but it requires an adequate length of the two arterial tracts to con-

nect and a certain abundance of vascular tissue. It can permit also correction of associated lesions such as kinking (like in the case here reported). If the two arterial tracts to link after aneurysmectomy are not long enough, the carotid substitution with venous graft or prosthesis (for example PTFE) has to be performed.

Alternatively external carotid artery may be sectioned and its proximal part connected to the distal tract of internal carotid artery after aneurysmectomy.

In rare cases of mycotic aneurysm, antibiotics have to be administered and venous patch has to be preferred to prosthesis because of risk of infection.

Other procedures with limited indications have to be mentioned. Wrapping consists in enveloping the aneurysm in cellophane sleeves or plastic material in order to limit its expansion: it is an obsolete technique that doesn't reduce the risk of embolization.

Matas' endoaneurysmorrhaphy is nowadays useful when a distal anastomosis is not feasible, maintaining temporarily a shunt in the internal carotid artery.

Even endovascular procedures such as stenting or embolization with spirals have been mentioned.^{17,19,20}

3.6. Complications

Except for nerve lesions or local infections, the only serious complication after this kind of surgery is cerebral ischemic damage related to ligation, clamping or embolization.

In order to reduce this complication it's important to handle carefully the aneurysm during dissection.

Continuous intraoperative EEG and SEP monitoring, evaluation of back pressures and some anesthesiologic cautions are useful to reduce risk of cerebral damage related to clamping.

Risk of cerebral damage is significative even if its estimation is difficult: simple ligation is followed by cerebral damage in 30% of cases, but its incidence is lower after reconstructive surgery.

Neurological complications may vary from massive fatal stroke to minimal accidents with partial or complete functional recovery. Sometimes cerebral damage becomes evident after a long time since surgery. This may relate to occlusion of the reconstruction or late embolization determining distal thrombosis of internal carotid or its branches.

4. Conclusions

Extracranial internal carotid aneurysms are uncommon but they have particular relevance especially because of the risks of cerebral embolic events or (more rarely) rupture. Nowadays sur-

gery is the gold standard of treatment, with specific reference to reconstructive techniques.

Conflict of interest

The authors declare that there is no conflict of interest.

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