Internal Carotid Artery Pseudoaneurysm as a Cause of Epistaxis: case-based update

A 43-year-old female patient reported a left hemicranial pain for six months, worsening in the last ten days. Computerized tomography (CT) revealed an expansive paramedian lesion in the left rhinopharynx, and biopsy revealed a nasopharyngeal squamous cell cancer. One month later, she was admitted with massive epistaxis causing hypovolemia. After 24 hours, the patient presented a new epistaxis, which was decided to manage surgically. The tumoral lesion was dissected and it was possible to visualize a lesion in the petrous portion of the left internal carotid artery (ICA), which the digital subtraction angiography (DSA) confirmed as a pseudoaneurysm. After a new life-threatening epistaxis in the following day, it was decided for a left ICA ligation, with DSA performed later showing left cerebral vascular permeability through the right ICA. The patient remains conscious and without new bleedings, although a focal neurological deficit. ICA pseudoaneurysms are clinically rare, accounting for less than 1% of all intracranial aneurysms, but are associated with significant morbidity and mortality. DSA is the gold standard for diagnosis of intracranial pseudoaneurysms. Treatment includes surgery, endovascular treatment or conservative therapy. Albeit uncommon, recurrent epistaxis is a red flag for ICA pseudoaneurysm in patients with rhinopharyngeal and skull base neoplasia.

Keywords: Epistaxis; Pseudoaneurysm; Internal carotid artery; Pseudoaneurysm management

RESUMO
Mulher, 43 anos, referindo dor no hemicrânio esquerdo há seis meses, com piora nos últimos dez dias. A tomografia computadorizada (TC) revelou lesão expansiva paramediana em rinofaringe esquerda e a biópsia revelou um carcinoma espinocelular de nasofaringe. Um mês depois, foi internada com epistaxe maciça causando hipovolemia. Após 24 horas, a paciente apresentou nova epistaxe, sendo decidido tratar cirurgicamente. A lesão tumoral foi dissecada e foi possível visualizar uma lesão na porção petrosa da artéria carótida interna (ACI) esquerda, que a angiografia de subtração digital (ASD) confirmou como pseudoaneurisma. Após nova epistaxe maciça no dia seguinte, optou-se pela ligadura da ACI esquerda, com ASD realizada posteriormente mostrando permeabilidade vascular cerebral esquerda pela ACI direita. A paciente permanece consciente e sem sangramentos, embora apresente déficit neurológico focal. Os pseudoaneurismas de ACI são raros, representando menos de 1% de todos os aneurismas intracranianos, mas estão associados a morbidade e mortalidade significativas. A ASD é o padrão-ouro para o diagnóstico de pseudoaneurismas intracranianos. O tratamento inclui cirurgia, tratamento endovascular ou terapia conservadora. Apesar de incomum, a epistaxe recorrente é um sinal de alerta para pseudoaneurisma de ACI em pacientes com neoplasia de rinofaringe e base de crânio.

Palavras-Chave: Epistaxe; Pseudoaneurisma; Artéria carótida interna; Manejo de pseudoaneurismas
INTRODUCTION

Pseudoaneurysms, or false aneurysms, occur when the arterial wall is formed only by the adventitial layer, extraluminal hematoma, or adjacent tissues. Massive epistaxis is rarely caused by the rupture of ICA pseudoaneurysms, but it can be life-threatening.

In this case report, the authors describe a case of a massive epistaxis caused by the rupture of an ICA pseudoaneurysm in a patient with nasopharyngeal neoplasia, as well as an update on this subject.

CASE PRESENTATION

A 43-year-old female patient reported pain in the left hemicranium for six months, worsening in the last ten days. At the physical exam, she presented with a left eye convergent strabismus. Non-contrast computerized tomography (CT) showed an expansive paramedian lesion in the left rhinopharynx extending to the skull base (Figure 1). Endonasal endoscopic biopsy was performed, showing nasopharyngeal squamous cell cancer.

One month later, she was admitted to the emergency with massive epistaxis causing hypovolemia and a Glasgow Coma Scale (GCS) score of 6, thus orotracheal intubation was performed to protect the airways. During the otorhinolaryngological exam, active bleeding was observed in the rhinopharynx, with cauteterization and apposition of absorbable hemostat and biological glue in the cauterized bed.

After 24 hours, the patient presented a new condition of epistaxis, when it was decided to perform bilateral sphenoid sinusotomy, and erosion of the inferior wall of the left sphenoid sinus was observed. The tumoral lesion was dissected and it was possible to visualize a lesion in the petrous portion of the left ICA. To control the bleeding, tamponade with absorbable hemostat and biological glue was performed at the lesion site, associated with a glove-finger nasal packing.

Digital subtraction angiography showed a pseudoaneurysm in the petrous portion of the left ICA, measuring approximately 14 mm and a neck of 5 mm with medial projection (Figure 2). The other cranial branches were normal. Functioning anterior communicating artery allowed opacification of the left middle cerebral artery in the left ICA occlusion test.

Three days later, the patient presented a new massive epistaxis with life-threatening bleeding, requiring a neurosurgical-otorhinolaryngological intervention, in which the left ACI ligation was chosen to control the bleeding. DSA performed after seven days allowed observation of left cerebral vascular permeability through the right ICA (Figure 3). The patient remains conscious and without new bleeding episodes, but evolved with dysphasia and right hemiparesis.

Figure 1. Noncontrast head CT showing an expansive lesion (arrow head) in the left rhinopharynx extending to the skull base

Figure 2. DSA showing a pseudoaneurysm (arrow head) in the petrous portion of the left ICA.
ICA pseudoaneurysms are clinically rare, accounting for less than 1% of all intracranial aneurysms, but are associated with significant morbidity and mortality. These false aneurysms are mostly caused by adjacent bone fracture due to trauma, but can also be caused by iatrogenic arterial injury during neurosurgical procedures, especially after transsphenoidal surgery (1.1%), commonly diagnosed after 90 days of the procedure by infection, especially by bacteria, tuberculosis bacilli or fungi; and in fewer cases by radiation. The present report describes the case of an ICA pseudoaneurysm, probably directly due to the infiltrative action of a tumor. According to our review, this is the first report to make this direct association in the literature.

False aneurysms occur when a blood vessel is physically or chemically damaged, and its complications, such as ruptures, usually occur immediately after or a few weeks after a vascular injury. There is a direct communication between the vessel lumen and the interior of the pseudoaneurysm, with blood circulating around the defect in the arterial wall. Thus, there is more rupture risk in a pseudoaneurysm than in a true aneurysm of similar size, due to the inadequate support of the pseudoaneurysm wall. When they are skull base located, there is an increased risk of spontaneous rupture, and symptomatic false aneurysms have a mortality rate of around 30-50%.

Epidemiologically, most ruptured ICA pseudoaneurysms that result in excessive bleeding are cavernous segment based, with rupture to the sphenoid sinus. This condition is different from the present case, which was petrous segment based but still ruptured to the sphenoid sinus.

The clinical presentation in patients with ICA pseudoaneurysms may be confused with the symptoms of the underlying condition causing the pseudoaneurysm, thus attention is needed to isolate the ICA pseudoaneurysm clinical. The most reported symptom is hemorrhage, which can lead to subarachnoid hemorrhage and carotid-cavernous fistula (CCF). There is only one report of a CCF connected via a pseudoaneurysm causing proptosis, chemosis and bruit, which, if left untreated, can lead to blindness and paralysis.

Unilateral blindness, orbital fracture and massive epistaxis compose a pathognomonic triad of symptoms related to ICA pseudoaneurysm located in the first cavernous segment, especially post-traumatic. ICA pseudoaneurysms may also present with headache, nerve paresis and facial weakness.

These false aneurysms can also cause mass effect and compression of adjacent cranial nerves, which can lead to manifestations as diplopia, ptosis, ophthalmoplegia, trigeminal neuralgia, and third nerve palsy, which can repercute in ocular movement disorders. Other broader spectrum neurological conditions such as hemiparesis can also happen. External auditory canal hemorrhage as a sign of ICA pseudoaneurysm has also been reported.

Epistaxis is a relatively rare manifestation of ICA pseudoaneurysm which, if left untreated, has high mortality, due to the usual meaning of pseudoaneurysm rupture. It is commonly caused by Kiesselbach plexus bleeding (90% of cases). The epistaxis related to ICA pseudoaneurysm, considered intermittent and persistent, is usually delayed after a traumatic incident or

Figure 3. DSA performed 7 days after ligation of the left ICA showing adequate permeability of the Circle of Willis, with satisfactory left anterior and middle cerebral arteries supply.
medical procedure. Persistent epistaxis is a strong indicator of an underlying pseudoaneurysm. All the articles used as base to this discussion were focused on epistaxis as a consequence of an ICA pseudoaneurysm.

The pseudoaneurysm diagnosis must be done as soon as possible, since there is a high mortality risk related to the rupture of the pseudoaneurysm in the ICA. In this case, the pseudoaneurysm was found in a surgical procedure, then confirmed by DSA. This method is the gold standard for the intracranial pseudoaneurysm diagnosis, since other imaging exams, such as CT, magnetic resonance imaging (MRI) or CT angiography, are not able to provide enough data to diagnose this condition.

The DSA shows a globular shape aneurysmal sac without a neck, with a delayed filling and contrast emptying. If the pseudoaneurysm is not seen initially in DSA, it is required to be repeated the exam after 1 or 2 weeks, since the development can take days or weeks after the initial cause.

DSA can also be used as treatment, with the endovascular embolization possibility. In addition, the Circle of Willis can be analyzed with the occlusion test to evaluate if there is collateral circulation able to maintain an adequate flow, which is important for some therapies.

Treatment of this condition includes surgery, endovascular and conservative therapy. In our case, it was decided to ligate the ICA due to the unavailability of endovascular treatment at our institution and to the massive new hemorrhage, which required immediate intervention. Conservative treatment includes the use of nasal packing or cauterization of active bleeding. However, care must be taken with excessive tamponade, which can cause compression injury to adjacent structures, such as cranial nerves and important vessels.

Occlusion of the injured artery is a treatment option, offering a rapid and effective treatment that can be performed through endovascular therapy with coils, stents, endovascular plugs and balloon occlusion, or through surgery, with ICA ligation. However, the carotid occlusion test should be done to assess Circle of Willis’ permeability. In case of poor patency of the Circle of Willis, extracranial to intracranial bypass surgery can be performed with ICA artery proximal occlusion. The use of an endovascular plug was reported as superior to the use of coils and stents, as it does not require associated antiplatelet treatment and allows a faster treatment. The main disadvantage of arterial occlusion is the risk of low cerebral perfusion and ischemia, even when DSA shows adequate collateral circulation. This is one of the hypotheses for the occurrence of focal deficits in our patient. In addition, this treatment method may increase the risk of aneurysm formation in the contralateral ICA or the size of an existing contralateral aneurysm. Balloon displacement and deflation are complications of using detachable balloons for arterial occlusion. Coil displacement has also been reported.

Treatment with flow preservation in the injured artery is performed with endovascular therapy, with embolization of the pseudoaneurysm using flow diverter stent or coils, or surgical treatment with microsurgical clips. The use of endovascular techniques with preservation of the parenteral vessel can present difficulties, mainly due to the weakened vascular structures in a pseudoaneurysm, different from a true aneurysm in which the arterial layers are preserved. One disadvantage of this modality is antiplatelet and anticoagulants agents necessity, increasing the risk of surgical bleeding. The use of flow diverter stents is reported as the treatment of choice for ICA pseudoaneurysm, as it has minimal complications and allows arterial patency preservation.

The prognosis is variable, depending mainly if the treatment has been performed. Due to ruptured aneurysms may have up to 50% mortality rates if left untreated. Besides, it also depends on the kind of treatment procedure used, its effectiveness, and how early that procedure was performed.

Generally, when successfully treated, the prognosis is relatively satisfactory, with well neurological recovery and still showing some sequelae. As an example, a 57-year-old patient with an ICA pseudoaneurysm treated with Pipeline-assisted coiling had her third nerve palsy completely resolved after treatment, aind still persisting the hemiparesis and aphasia.

When it is decided for a conservative procedure that does not involve the ICA sacrifice, the prognosis tends to be better, because, even if the balloon test occlusion is tolerated, there is no guarantee that complications will not be developed.
CONCLUSION

Albeit uncommon, recurrent and intractable epistaxis is a red flag for ICA pseudoaneurysm in patients with rhinopharyngeal and skull base neoplasia. Early diagnosis and treatment are essential to reduce morbidity and mortality secondary to massive epistaxis. ACI ligation is an option in the absence of adequate endovascular treatment.

REFERENCES


