

Carotid Mycotic Aneurysm associated with Persistent Primitive Hypoglossal Artery. Case Report and Literature Review

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ABSTRACT

Carotid mycotic aneurysm is extremely rare and even more unusual when it is associated with a persistent primitive hypoglossal artery. This artery is the second most common of the embryonic carotid-vertebrobasilar anastomoses. It originates from the cervical internal carotid artery and enters the cranium through a widened hypoglossal canal before anastomosing with the basilar artery. We report a case of an elderly man with a rare Salmonella-induced mycotic aneurysm associated with a persistent primitive hypoglossal artery. Surgical resection of the mycotic aneurysm was complicated by a posterior circulation stroke. To the best of our knowledge, there was no previous report of a carotid mycotic aneurysm associated with a persistent primitive hypoglossal artery thus far in the literature. Owing to the high mortality rate of the carotid mycotic aneurysm, it is imperative to be familiar with the vascular and imaging anatomy prior to surgery particularly in the presence of an embryonic carotid-vertebrobasilar anastomosis. In this report, we highlight the imaging characteristics and treatment options for this rare mycotic aneurysm together with a literature review.

CASE REPORT

CASE REPORT

A 74-year-old man with a history of diabetes mellitus and hypertension presented to the emergency department with progressive worsening dysphagia, hoarseness of voice and intermittent fever of 2 months duration. Physical examination revealed a pulsatile mass and erythema on the right side of the neck. A bruit was detected on auscultation. His laboratory results showed abnormal liver function tests [Alkaline phosphatase 142 U/L (reference range, 40-129 U/L), and

alanine amino-transferase 99 U/L (reference level, <41 U/L)], elevated total white cells of 16,440/mm³ (neutrophils 95%), erythrocyte sedimentation rate of 125 mm/h (reference range, 2-10 mm/h) and C-reactive protein (CRP) level of 74 mg/L (reference level, <3.0 mg/L). Blood cultures grew group D Salmonella sensitive to cefotaxime, ceftriaxone and levofloxacin. A systemic search for a potential primary source of infection was performed but no definite source was identified. The patient was treated with cefotaxime intravenously.

Computed tomography (CT) of the neck and subsequent CT carotid angiogram revealed a 2.8 x 2.5 cm large saccular aneurysm arising from the proximal right extra-cranial internal carotid artery (ICA) just distal to the carotid bifurcation (Fig. 1A). A persistent primitive hypoglossal artery (PPHA) was seen arising distal to the origin of the aneurysm at C2 level, entering the cranium through the hypoglossal canal and anastomosing with the basilar artery (Fig. 1B, C & D). The PPHA was the main supply to the posterior circulation as both vertebral arteries were hypoplastic (Fig. 1B & C). The proximal extra-cranial ICA and the aneurysm sac were surrounded by inflammatory soft tissue and collection (Fig. 1E). The patient was counselled regarding the best treatment option which comprised surgical resection of the aneurysm and in-situ graft replacement under cardio-pulmonary bypass with hypothermic circulatory arrest. Endovascular treatment with coil embolization or stent graft deployment was not considered a viable treatment option in this case due to the high risks of infection and thromboembolism caused by foreign material in the presence of an infected surgical bed.

Operative procedures – A sub-sigmoid osteotomy of the right ramus of the mandible was performed to allow access to the right common carotid artery (CCA) and for securing proximal control. Hypothermic circulatory arrest was performed during surgery. The carotid mycotic aneurysm was isolated from the surrounding inflammatory surgical bed and excised. This was followed by an end-to-end anastomosis and carotid artery repair using a cadaveric homograft.

Microscopic examination of the excised aneurysm sac revealed the presence of inflammatory cells infiltration and atherosclerotic plaque which was compatible with a mycotic aneurysm. Subsequent bacterial culture however did not yield any growth.

Postoperative condition – The patient was comatose and suffered a left hemiplegia. Follow-up CT carotid angiogram revealed severe narrowing of the right carotid stent graft with severe luminal narrowing and reduced flow along the extra- and intra-cranial right carotid arteries as well as the PPHA (Fig. 2B). MRI brain revealed multiple foci of acute infarctions notably in the pons, posterior circulation and arterial territory of the right middle cerebral artery (Fig. 2 C-H). After 4 weeks of acute hospital care, the patient was transferred to a nursing home. He was readmitted six months later and died in the hospital from pneumonia and sepsis.

DISCUSSION

Etiology & Demographics:

PPHA is one of the four persistent embryonic anastomoses between the carotid and vertebrobasilar circulations [1]. The other primitive anastomoses include the persistent primitive trigeminal artery (PPTA), persistent otic (acoustic) (POA), and persistent proatlantal intersegmental artery (PIA) [1]. Among the four embryonic anastomoses, PPHA is the second most common carotid-vertebrobasilar arterial anastomosis with a prevalence of 0.02%–0.10% [2]. The PPTA is the most common of the four carotid-

vertebrobasilar anastomoses with a prevalence of 0.5-0.7%. The POA is extremely rare with only a handful of previous reports in the literature. The PIA is extremely rare and approximately 40 cases have been reported [2, 3].

There were over 100 cases of PPHA being reported in the literature of which 20 were associated with aneurysms while a smaller number of them were related to ICA stenosis [4]. Our patient with PPHA had non-typhoid Salmonella bacteremia which predisposed him to a carotid mycotic aneurysm. To the best of our knowledge, there was no previous case report of a carotid mycotic aneurysm associated with PPHA, thus far in the literature.

The carotid mycotic aneurysm is extremely rare with an incidence of 0.03% of all arterial aneurysms and 5% of all mycotic aneurysms [4, 5]. It occurs more frequently in males than in females with a ratio of 6:4. The disease has a predilection for the elderly between the 6th and 7th decades of life. Mycotic aneurysms usually occur in the setting of systemic sepsis with spread from a distant infectious source (e.g. endocarditis). It is prevalent amongst the immunocompromised with predisposing conditions such as intravenous drug abuse, dental extractions and angiographic procedures [5]. Direct spread of endovascular infections may occur from the head and neck region following carotid surgery or trauma [5]. The most common pathogens responsible for these infections are Staphylococcus aureus followed by Salmonella, Streptococcus, Klebsiella and Escherichia coli in the order of decreasing frequency [5, 6]. These organisms were found to inoculate atheromatous plaque at the carotid bifurcation, leading to inflammation and mycotic aneurysm with high risks for rupture [6].

In our patient, advanced age and diabetes mellitus were likely to contribute to the atherosclerotic disease which were the risk factors predisposing to endovascular infection following the Salmonella bacteremia [7]. The reported incidence of endovascular infections with Salmonella bacteremia ranges from 9 to 10% in western countries to 16.2% in a Taiwan publication [7]. According to the literature, the common Salmonella species responsible for endovascular infections were group C (47%), group D (32%), and group B Salmonella (21%) [4,7]. These species have a predilection for invading damaged endothelium in the heart and arterial walls leading to a spectrum of cardiovascular infections including mediastinitis, pericarditis, endocarditis, aortitis, mycotic aneurysms, and infection of cardiac devices [4]. Failure to consider such unusual complications of Salmonella bacteremia can lead to a catastrophic outcome.

Clinical & Imaging Findings:

PPHA usually originates from the cervical ICA distal to the carotid bifurcation between C1 and C3 level. The PPHA then enters the hypoglossal canal before anastomosing with the basilar artery [1, 2]. When enlargement of the hypoglossal canal is identified, the presence of a PPHA should be strongly suspected. On computed angiogram, the PPHA is usually the main supply to the posterior circulation and may be associated with miniscule vertebral or basilar arteries.

A carotid mycotic aneurysm is usually large and saccular in appearance with surrounding soft tissue infection or phlegmone, sometimes it can be surrounded by an enhancing rim of collection.

Treatment & Prognosis:

A multidisciplinary approach with combined medical and surgical treatment is the mainstay therapy for mycotic aneurysms. In a review by Soravia-Dunand et al, the mortality rate of Salmonella-induced mycotic aneurysms was 40% for patients who received combined surgical and medical treatment compared to 96% for those patients who received medical therapy alone [10].

Early surgical intervention for mycotic aneurysm usually consists of wide debridement of the infected tissues, extra-anatomic revascularization or in-situ graft replacement [7]. In most cases of PPHA, the vertebral arteries are either hypoplastic or aplastic with absence of the posterior communicating arteries, hence the main supply to the posterior circulation comes from the PPHA (1).

In our patient, the mycotic aneurysm was situated between the carotid bifurcation and the origin of the PPHA along with heavily calcified atherosclerotic plaques, rendering surgical repair difficult and hazardous with increased operative risks of thromboembolisms.

Resection of the mycotic aneurysm with graft replacement was subsequently complicated by inevitable distal embolism in the ipsilateral carotid and vertebrobasilar systems which resulted in right MCA and posterior circulation infarctions with an ultimately fatal outcome.

Endovascular stent graft repair is an alternative treatment option; however, this procedure was deemed unsuitable in our patient as it would not allow debridement or removal of the infected tissues. This procedure predisposed the patient to high risks of delayed complications such as septic embolism, ischemia and hemorrhage [7]. Besides, there were concerns raised over the durability of an endovascular stent grafts in pre-existing "infected" surgical bed with Salmonella-induced bacteremia. Long-term follow-up data is therefore needed to help define the role and durability of conventional surgery versus endovascular repair in the treatment of mycotic aneurysms.

Different antimicrobial regimens with varying success rates have been described for the treatment of Salmonella bacteremia, including combinations of ampicillin or cefotaxime with gentamycin, ceftriaxone alone or fluoroquinolones [4]. A case series found that among 34 Salmonella isolated from people with infective aortic aneurysms, 65% were ampicillin resistant and 41% were resistant to ciprofloxacin, but none were resistant to cefotaxime [6]. Third-generation cephalosporins are therefore recommended as the initial drugs of choice for Salmonella-induced aneurysms [4]. There is no consensus regarding the ideal duration of systemic antimicrobial therapy. As a general rule, postoperative antimicrobial therapy for 6 to 8 weeks may

be adequate for preventing a relapse [4]. Biochemical parameters of inflammation (white cell count, CRP, and ESR) may help with disease monitoring and to guide the duration of antibiotic treatment. Lifelong suppressive antibiotic therapy has been suggested in some cases because of unacceptable operative risks of mycotic aneurysm and the concern of late recurrent infections of in-situ prosthetic material [10].

In conclusion, it is imperative to promptly diagnose a carotid mycotic aneurysm. Surgical resection of the aneurysm was challenging due to presence of an embryonic carotid-vertebrobasilar anastomosis, in this case a PPHA. Despite a multidisciplinary approach with combined medical and surgical treatment, the morbidity and mortality rates of carotid mycotic aneurysms have remained extremely high.

Differential Diagnosis of carotid mycotic aneurysms:

a. Carotid body tumour (CBT) which is synonym to carotid glomus, chemodectoma and paraganglioma.

Patients with CBT may present with a pulsatile mass anteromedial to the sternomastoid muscle near the angle of the mandible at the level of the hyoid bone. It is usually located at the carotid bifurcation, splaying the internal and external carotid arteries (Lyre sign). CT demonstrates a soft tissue mass with rapid and avid enhancement following contrast administration. On T1w MRI sequence, the lesion demonstrates a "salt and pepper" appearance due to a combination of slow and high flow signal within it. Surgical resection is the treatment modality of choice [8]. Radiotherapy is reserved for the elderly, patients unfit for surgery or those with multiple paragangliomas in whom resection may be highly morbid [8].

b. Glomus Vagale tumour (GVT)

Patients with GVT may present with a painless mass behind the carotid artery and frequently with vocal cord paralysis. The GVT is located along the course of the vagal nerve between the carotid artery and internal jugular vein without widening the carotid bifurcation. Besides, it is usually more rostral in location to the carotid body tumours. Ultrasound shows a solid heterogeneously hypoechoic lesion comprising of small vascular structures. MRI demonstrates a "salt and pepper" appearance on non-contrast T1W sequences and avid enhancement following contrast administration. Surgical resection is the treatment of choice.

c. Vagal nerve schwannoma (VS)

Patients with VS may present with hoarseness of voice and a smooth palpable neck lump. On examination, a paroxysmal cough may be elicited on palpation. It is located along the course of the vagal nerve and within the carotid sheath, displacing the carotid artery medially. Ultrasound demonstrates a hypoechoic nodule and Doppler shows flow signal with increased vascularity at the periphery of the lesion. MRI demonstrates T1w iso- (similar to muscle) and T2w hyperintensity, and heterogeneous enhancement following contrast administration. Surgical resection is the best treatment of choice.

d. Carotid “blow-out” syndrome (CBS)

CBS is defined as rupture of the carotid artery and its branches with serious complication following the treatment of advanced head and neck cancer. Potential causes of CBS include radical resection, radiation therapy and radiation necrosis, carotid exposure, wound infection, pharyngocutaneous fistula, and recurrent or persistent carcinoma [9]. Patients usually present with sudden onset oral bleeding and hypotension. Axial contrast-enhanced CT shows a mass encasing the carotid artery with contrast extravasation. Catheter angiogram demonstrates contrast “blush” and extravasation from the carotid artery or its branches. Urgent treatment consists of surgical ligation or permanent arterial occlusion of the artery. The incidence of immediate or delayed complications of cerebral ischemia can be as high as 15%–20% [9]. Stent-graft deployment, with or without coiling, is an endovascular treatment for CBS which can preserve the affected carotid flow with lower complication rates [9].

TEACHING POINT

Carotid mycotic aneurysms are extremely rare and even more unusual when associated with a persistent primitive hypoglossal artery, requiring a multi-disciplinary approach with combined medical and surgical therapy in view of the high associated mortality rate. Radiologists and surgeons should be familiar with the vascular imaging features of a persistent carotid-vertebrobasilar anastomosis during aneurysm resection as the embryonic artery could be the main supply to the posterior circulation; compromising it could lead to ischemic complications with potentially fatal outcome.

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FIGURES

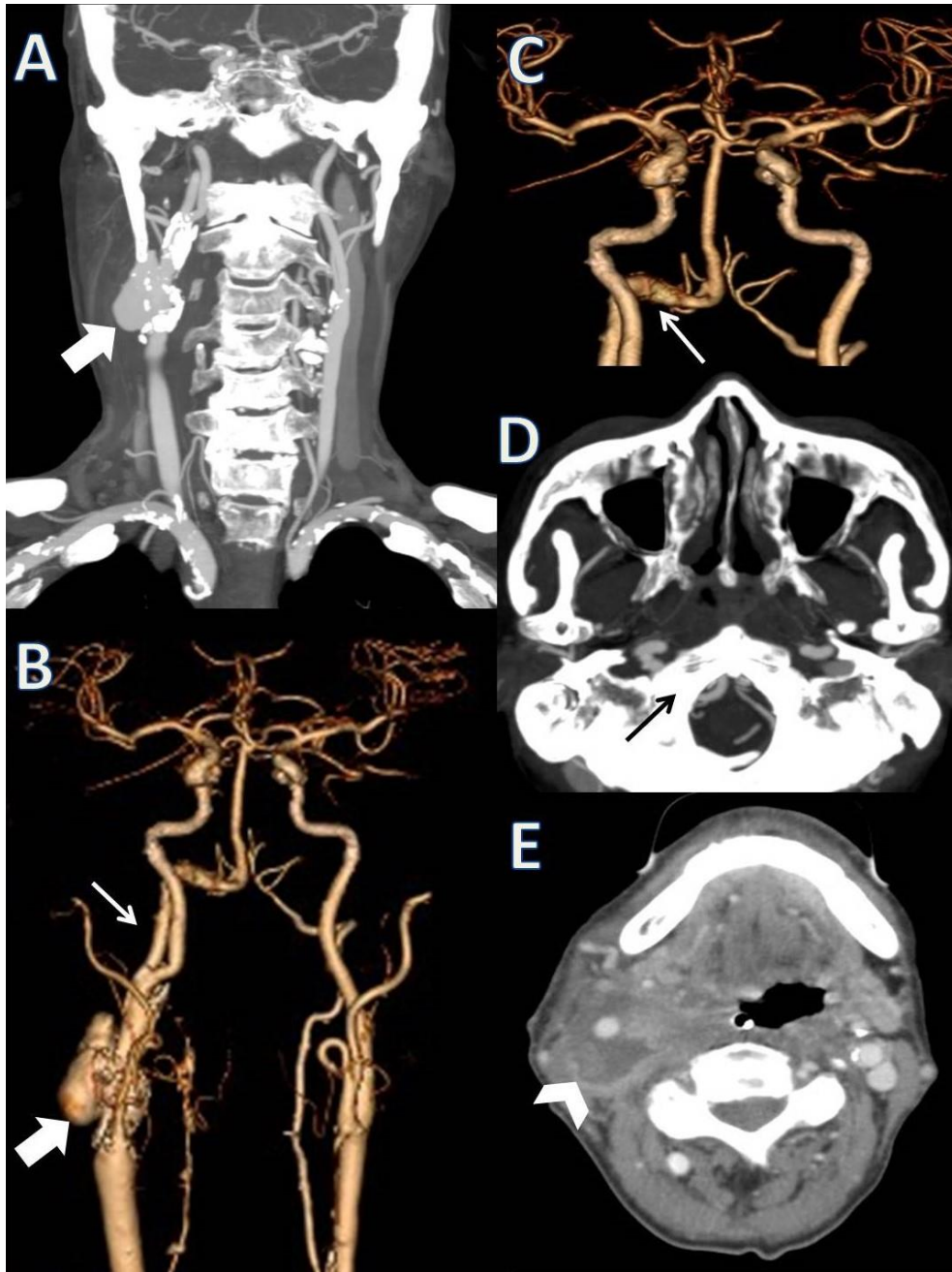


Figure 1: A 74-year-old man with a carotid mycotic aneurysm associated with persistent primitive hypoglossal artery (PPHA).

FINDINGS: Computed tomography angiogram (CTA) of the neck reveals a 2.8 x 2.5 cm large saccular mycotic aneurysm arising from the proximal extra-cranial right internal carotid artery (ICA) just distal to the carotid bifurcation (A-B, white block arrow). The PPHA (B-C, white arrow) arises from the right cervical ICA distal to the aneurysm as it anastomoses with the basilar artery after it courses through the right hypoglossal canal (D, black arrow). The proximal cervical ICA is surrounded by a thick rim of hypodense soft tissue indicating phlegmone formation (E, white arrowhead).

TECHNIQUE:

Philips iCT 256 CT scanner. Contrast-enhanced CT circle of Willis. Tube voltage = 120 kV. Tube current = 399 mA. A total of 60 mls of Omnipaque 350 was administered intravenously.

A: Coronal CT in the arterial phase with maximum intensity projection (MIP). Slice thickness = 15 mm.

B-C: 3D reconstruction CT angiogram. Slice thickness = 15 mm.

D: Axial CT in the arterial phase with MIP. Slice thickness = 15 mm.

E: Axial CT in the venous phase. Slice thickness = 5 mm.

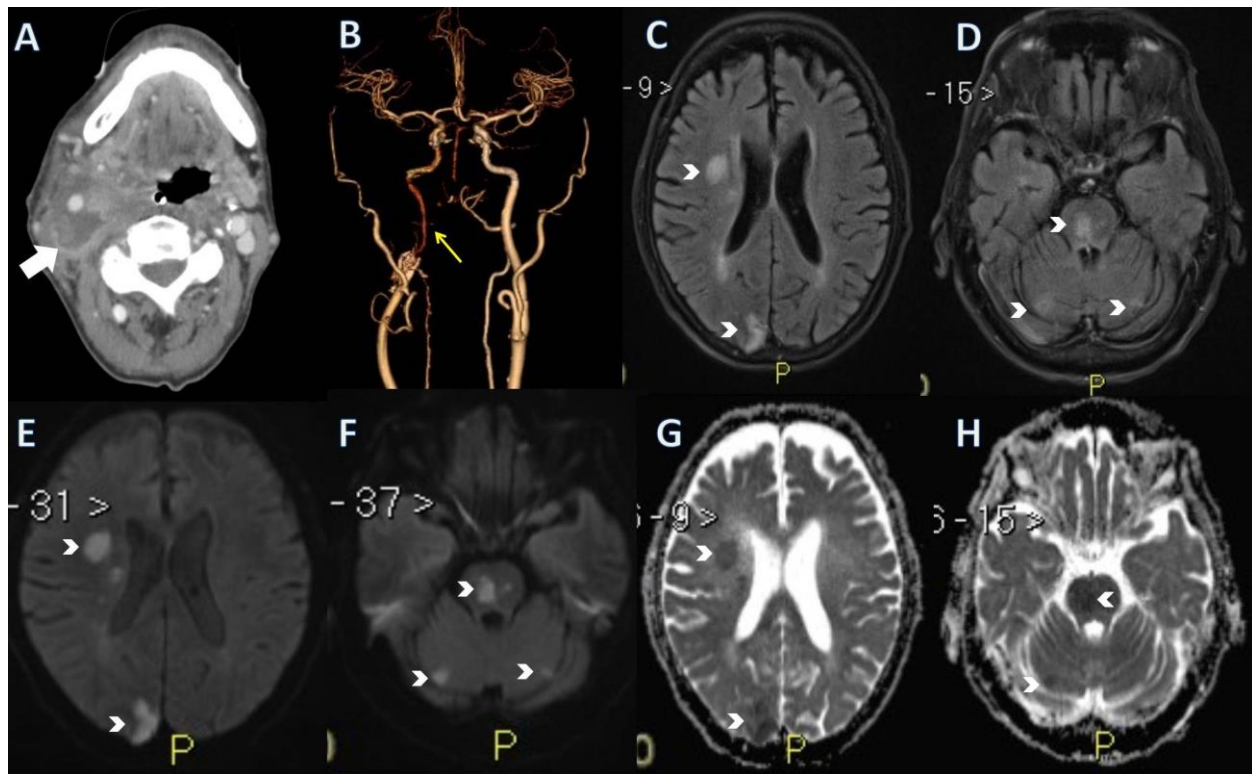


Figure 2: A 74-year-old man with a persistent primitive hypoglossal artery (PPHA) who underwent excision of the right carotid mycotic aneurysm with an end-to-end anastomosis using an inter-positional homograft.

FINDINGS: Computed tomography angiogram (CTA) of the neck showing a thin rim of hypodense collection surrounding the right carotid artery at the surgical anastomosis (A, white block arrow). 3D reconstructed CTA reveals narrowing of the right carotid graft repair (yellow arrow) following resection of the mycotic aneurysm (B). There is diminution of flow in the PPHA, now less well visualized (B). Axial FLAIR (C-D), DWI (E-F) and ADC (G-H) MRI reveals FLAIR hyper-intense foci in the pons, right corona radiata, right occipital lobe and bilateral cerebellar hemispheres with corresponding restricted diffusion (white arrowheads). These are compatible with acute infarctions in the territories supplied by the right middle cerebral artery and posterior circulation.

TECHNIQUES:

Philips iCT 256 CT scanner. Tube voltage 120kV. Tube current 402 mA. A total of 60 mls of Omnipaque 350 was administered intravenously.

A: Axial CT in the arterial phase. Slice thickness = 5 mm.

B: 3D reconstruction CT angiogram. Slice thickness = 15 mm.

Siemens Avanto MRI scanner. Magnetic strength = 1.5 Tesla. No intravenous contrast was administered.

C-D: Axial T2W FLAIR. TR = 6000 ms. TE = 2020 ms TI = 134 ms. Slice thickness = 5 mm.

E-F: Axial DWI. TR = 3500 ms. TE = 89 ms. Slice thickness = 5 mm.

G-H: Axial ADC. TR = 3500 ms. TE = 90 ms. Slice thickness = 5 mm.

ETIOLOGY	Acquired; Bacteremia, spread from a distant infectious source (e.g. endocarditis)
INCIDENCE	Extremely rare, accounted for 0.03% of all arterial aneurysms and 5% of all mycotic aneurysms
GENDER	Male:Female 6:4
AGE PREDILECTION	6th-7th decades of life
RISK FACTORS	Immunocompromised, drug abuse, dental extraction, prior head and neck surgery, advanced age, diabetes mellitus, hypercholesterolemia
IMAGING FINDINGS	Saccular aneurysm with surrounding soft tissue infection and collection on CT
PATHOGENS	Staphylococcus aureus followed by Salmonella, Streptococcus, Klebsiella and Escherichia coli
TREATMENT	Combined medical therapy (appropriate antibiotics) and surgical excision
PROGNOSIS	40-96% mortality

Table 1: Summary table for carotid mycotic aneurysm.

Differential diagnosis	Presentation	Location	CT or Ultrasound	MRI or conventional angiogram	Best treatment option
Carotid mycotic aneurysm	Sepsis from associated bacteremia	Carotid artery and bifurcation	Saccular aneurysm with surrounding rim-enhancing collection/inflammatory phlegmon	Similar to CT	Appropriate intravenous antibiotics and surgical excision.
Carotid body tumour synonyms: carotid glomus, chemodectoma, paraganglioma	Pulsatile mass anteromedial to the sternomastoid muscle near the angle of the mandible at the level of hyoid bone.	At carotid bifurcation, splaying the internal and external carotid arteries (Lyre sign)	Non-contrast CT demonstrates soft tissue with rapid and avid enhancement following contrast administration.	T1w MRI demonstrates "salt and pepper" appearance representing punctate regions of areas of slow flow and flow voids.	Surgical excision.
Glomus vagale	Painless mass behind carotid artery and frequently vocal cord paralysis.	Along the course of vagal nerve; more rostral in location to carotid body tumours and between the carotid artery and internal jugular vein.	Ultrasound shows solid heterogeneously hypoechoic lesion comprising of small vascular structures.	T1w MRI demonstrates "salt and pepper" appearance. Avid enhancement following contrast administration.	Surgical excision.
Vagal nerve schwannoma	Palpable smooth lump in the neck. Hoarseness. Paroxysmal cough maybe elicited on palpation.	Displacing the carotid artery medially within the carotid sheath.	Ultrasound demonstrates a hypoechoic nodule. Duplex Doppler shows flow signal with increased vascularity in the periphery of the lesion.	MRI demonstrates low T1 (similar to muscles) and high T2w signal. Heterogeneous enhancement following contrast.	Surgical excision.
Carotid "blow-out" syndrome (CBS)	Sudden onset of oral bleeding and hypotension. History of advanced head and neck cancer treatment with either surgery, radiation and/or chemo-therapy	Carotid artery and its branches.	Axial contrast-enhanced CT shows a mass-like contrast extravasation at the carotid artery.	Catheter angiogram demonstrates contrast extravasation at the carotid artery.	Deployment of stent graft with or without coil embolization.

Table 2: Differential diagnoses table for carotid mycotic aneurysm.

ABBREVIATIONS

CBS = carotid "blow-out" syndrome
 CBT = carotid body tumour
 CTA = Computed tomography angiogram
 GVT = glomus vagale tumour
 ICA = Internal carotid artery
 MCA = Middle cerebral artery
 MRI = Magnetic resonance imaging
 PIA = Persistent proatlantal intersegmental artery
 POA = Persistent otic (acoustic) artery
 PPHA = Persistent primitive hypoglossal artery
 PPTA = Persistent primitive trigeminal artery
 VS = vagal nerve schwannoma

KEYWORDS

Bacteremia; mycotic aneurysm; persistent primitive hypoglossal artery; Salmonella-induced infection

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