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CLINICAL RECORD

# Extracranial internal carotid artery pseudoaneurysm in a two-year-old child: case report

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## Abstract

*Background*: Extracranial internal carotid artery pseudoaneurysm is very rare in children.

*Method*: This paper discusses the case of a boy, aged two years and six months, who presented with an enlarging neck mass and unilateral bloody otorrhoea. Special investigations revealed an extracranial internal carotid artery pseudoaneurysm.

*Results*: The patient made a full recovery after endovascular occlusion of the internal carotid artery and pseudoaneurysm using coils. At six months' follow up, the internal carotid artery and pseudoaneurysm remained excluded from the circulation. The patient did not display any neurological deficits during hospital stay or follow up.

*Conclusion*: This paper reports on one of the youngest patients documented to date who presented with an internal carotid artery pseudoaneurysm, possibly secondary to ear infection. Although rare, this condition should be excluded in children presenting with a mass of the neck or pharynx because of the dire consequences if left undiagnosed and untreated.

**Key words:** Carotid Pseudoaneurysm; Child; Endovascular Procedures; Infection; Ear

## Introduction

Extracranial internal carotid artery (ICA) pseudoaneurysms in children are extremely rare, with few reported cases in the literature. A pseudoaneurysm or false aneurysm represents a blood-filled cavity in contact with an artery, with a persistent connection with the arterial lumen.[1,2] This occurs as a result of a localised disruption of the arterial wall.[2] The aneurysm wall is formed by haematoma and reactive connective tissue.[1] It has a high mortality rate if left untreated because of the great potential for thromboembolic events and massive life-threatening haemorrhage.[1,3,4] Therefore, a high index of suspicion and timely diagnosis remain very important in order to manage these children before the development of life-threatening complications.[3,4]

We report on the case of a boy aged two years and six months with an extracranial ICA pseudoaneurysm, who presented with a large parapharyngeal space mass and bloody otorrhoea secondary to ear infection. A successful endovascular occlusion procedure of the ICA and pseudoaneurysm was performed without any neurological sequelae.

## Case report

A previously healthy boy, aged two years and six months, with a one-month history of intermittent bloody otorrhoea from the right ear and a right-sided neck mass, was seen at the ENT Clinic of Universitas Hospital, Bloemfontein, South Africa.

On examination, the patient appeared acutely ill and irritable. A hard, tender, right-sided neck mass was noted in the infra-auricular area, which was non-fluctuant and non-pulsatile. Examination of the right ear revealed an inflamed, granulomatous mass on the inferior aspect of the right external auditory canal, with an intact tympanic membrane. Neurological examination findings were normal. On inspection of the oral cavity, the patient was noted to have mild trismus and narrowing of the oropharynx due to a large right-sided parapharyngeal mass. The airway was not imminently threatened. He had persistent hypertension, observed since admission.

A neck ultrasound scan showed a poorly circumscribed mass on the side right, suspected to be of parotid gland origin. Computed tomography (CT) scans of the neck revealed a pseudoaneurysm of the right proximal internal carotid artery (ICA) measuring $4.8 \times 3.6 \times 4.4$ cm, with mass effect on the pharynx and narrowing of the airway (Figures 1 and 2). Opacification of the right mastoid air cells and middle ear were observed, with an intact circle of Willis.

With regard to laboratory results, white cell count was $7.69 \times 10^9$/l, erythrocyte sedimentation rate was 62 mm/hour, C-reactive protein was 18 mg/l, enzyme-linked immunosorbent assay findings for human immunodeficiency virus (HIV) were negative, and renal functions and electrolytes were normal.

A biopsy of the mass in the right external auditory canal revealed ulcerated squamous epithelium and underlying inflamed granulation tissue, suggestive of chronic otitis externa. Auramine-O stain and tuberculosis cultures of the

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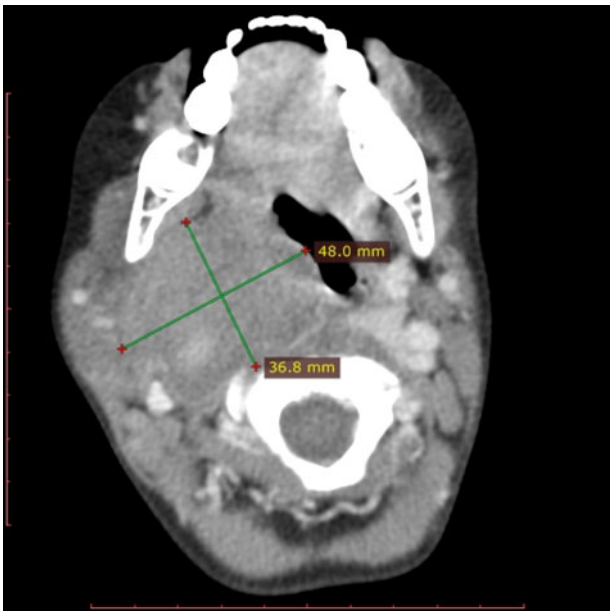


FIG. 1
Axial computed tomography scan demonstrating internal carotid artery pseudoaneurysm.

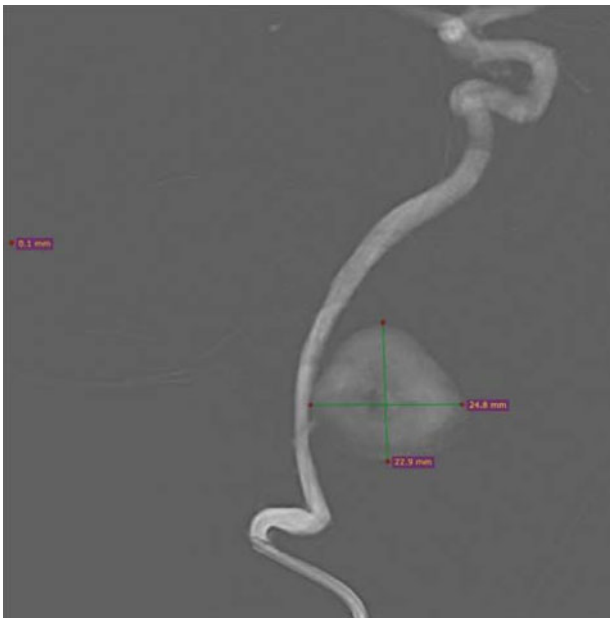

FIG. 3
Arteriogram demonstrating pseudoaneurysm of right internal carotid artery.

biopsy were negative, while a pus swab of the right ear cultured *Klebsiella pneumoniae* and beta-lactamase negative *Staphylococcus aureus*. Blood cultures were negative. A renal ultrasound scan was normal and autoimmune investigation findings were negative.

The patient underwent uneventful intubation, followed by endovascular occlusion of the ICA and pseudoaneurysm by coiling (Figures 3 and 4). Intubation was carried out by experienced personnel because of the potential for rupture during intubation. An endovascular approach was used because direct surgical repair would have required wide exposure of the neck in order to obtain proximal and distal control of the aneurysm, leading to higher morbidity and possible mortality.[1]

The patient was extubated 4 days after the procedure. He made a full recovery without any neurological sequelae. Post-procedural Doppler scans did not demonstrate any flow in the ICA or pseudoaneurysm. No anticoagulation was prescribed. The parapharyngeal mass gradually resolved and the right ear was successfully treated with systemic antibiotics and ofloxacin drops. Blood pressure gradually returned to normal after the resolution of the neck mass, and the assumption was made that the hypertension was

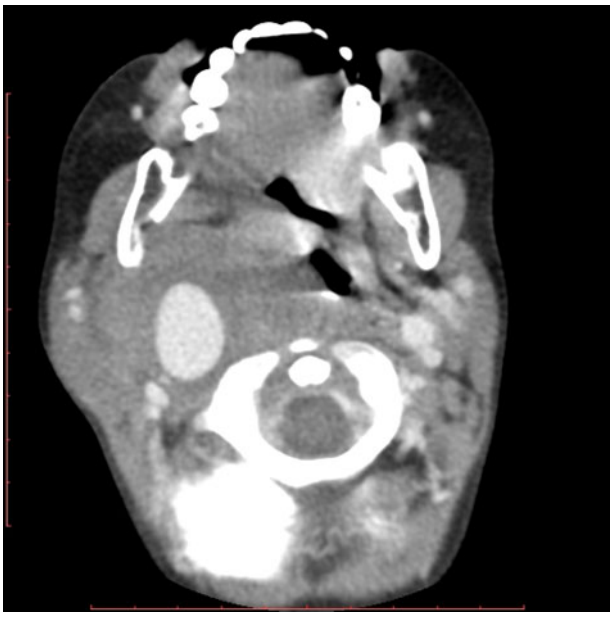

FIG. 2
Axial computed tomography scan with post-contrast enhancement in pseudoaneurysm.

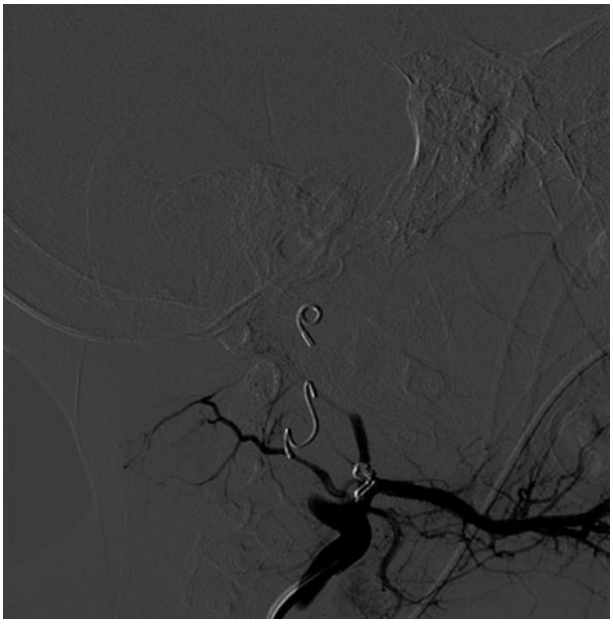

FIG. 4
Arteriogram post-coiling, demonstrating exclusion of internal carotid artery and pseudoaneurysm from circulation.

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secondary to the pressure on the carotid body caused by the pseudoaneurysm. At six months' follow up, the ICA and pseudoaneurysm remained excluded from the circulation.

## Discussion

Risk factors for extracranial internal carotid artery (ICA) pseudoaneurysm include prior carotid surgery or intervention, blunt and open trauma, primary or contiguous infection, spontaneous carotid dissection, connective tissue diseases, and radiation therapy.[1,2,5] Contiguous infection, typically from meningitis, cervical lymphadenitis and ear infection, remains the most common cause of carotid artery aneurysm in children.[2] The infection typically spreads to involve periarterial lymphatics and vasa vasorum.[2] The incidence of infected aneurysms due to peritonsillar and middle-ear infections has declined with the use of antibiotics.[2,3] The main reason for involvement of the vessels with infection of the pharynx and deep neck spaces is the close proximity of the ICA to these structures.[1,4] A case study published in 2012, on a five-year-old child, reported on an ICA pseudoaneurysm secondary to a throat infection, which was treated successfully with stent placement after thrombin injection failure.[3]

*S aureus,* salmonella, *Streptococcus pyogenes, Escherichia coli* and klebsiella are the most common pathogens associated with infected extracranial ICA pseudoaneurysm.[1,2] The high incidence of HIV in our environment has led to an increase in HIV-associated carotid aneurysms.[6] Carotid artery aneurysms represent 25 per cent of all HIV-related aneurysms and have been thought to occur as a result of HIV-related vasculopathy.[6]

Congenital aneurysms have also been described in children, and occur as a result of structural defects of the arterial wall secondary to developmental defects of the arterial muscular layer.[4]

Symptomatology of extracranial ICA pseudoaneurysm may be due to the local mass effect, embolism, thrombotic occlusion or aneurysm rupture.[2] Common symptoms include: a painful or painless mass, stroke or transient ischaemic attacks, visual disturbance, a protruding pharyngeal mass, epistaxis, and Horner's syndrome. Hoarseness, dysphagia, auricular pain and tongue weakness may occur as a result of cranial nerve palsies.[1,2,3] The condition may also be asymptomatic and discovered incidentally.[2] Rupture is a potentially life-threatening complication that may occur in as many as one in three children, and may present as pharyngeal haemorrhage, epistaxis or bloody otorrhoea.[1,2,4] Pourhassan *et al*. reported the highest risk of rupture to occur with traumatic and mycotic aneurysms.[4]

Diagnosis relies on the demonstration of an aneurysm on imaging studies. Ultrasound is the initial study of choice to evaluate a neck mass, but CT angiography and magnetic resonance angiography are useful confirmatory imaging studies that can determine the extent of dilatation and thrombus formation of the aneurysm, and demonstrate the relationship of the aneurysm with the surrounding structures.[2]

Intervention is recommended for symptomatic aneurysms, aneurysms with intraluminal thrombus and small asymptomatic aneurysms that demonstrate expansion on imaging.[2,4] Management was traditionally with open surgical repair, although it has recently been replaced by endovascular techniques, which seem to be a feasible and safe option.[3,5] Open surgical repair is reported to have higher morbidity and mortality rates, and a higher risk of cranial nerve injury.[5] With free or impending rupture, it may be necessary to convert to an open surgical technique if there is continuous bleeding from the sac during endovascular procedures.[5]

Options for open surgical repair include ligation of the carotid artery and excision of the aneurysm, with reconstruction using a saphenous vein graft or prosthetic material.[2,4] In children younger than four years, the greater saphenous vein graft may be too small for supra-aortal interposition,[4] with the theoretical risk of stenosis. In these cases, the use of the profunda femoral vein graft for interposition has been described.[4] In children, the delicate anatomy of the small vessels makes open surgery challenging.[4]

Endovascular repair techniques include stent placement, trans-stent coil embolisation, stent-graft exclusion of the aneurysm or endovascular carotid artery occlusion.[2] In a reported case of a four-year-old child with ICA pseudoaneurysm secondary to pharyngitis, the use of a covered stent was reported to be an effective therapeutic option to exclude the pseudoaneurysm while keeping the parent vessel patent.[1] It was reported that long-term patency, outcomes and best antithrombotic therapy still need to be evaluated in children with covered stents.[1] There are also concerns regarding the adaptability of the growing carotid artery to the covered stent.[3]

- **Extracranial internal carotid artery (ICA) pseudoaneurysms in children are rare**
- **Endovascular intervention remains the choice of treatment for this condition**
- **It has a high mortality rate if left untreated given the potential for development of life-threatening complications**
- **A high index of suspicion and timely diagnosis remain important**
- **This paper reports on a child successfully treated with endovascular occlusion of the ICA and pseudoaneurysm by coiling**

Carotid sacrifice may be performed via an open ligation or via an endovascular approach.[2] Ligation of the ICA is associated with a substantial risk for neurological morbidity in the short and long term.[7] Stroke and even death may result if the collateral circulation is not intact.[4] McCann (as cited in Rosset *et al.*)[7] reported a stroke risk of 25 per cent and mortality rate of 20 per cent with ICA ligation. Although endovascular exclusion techniques are usually safe and curative, there is a small risk for aneurysmal disease in the contralateral ICA due to chronic increase in flow.[1,3] Given the fragile nature of the pseudoaneurysm wall, selective embolisation of the pseudoaneurysm may be hazardous.[3]

## Conclusion

Extracranial internal carotid artery pseudoaneurysm is rare in children, but has a high mortality rate if left untreated. The diagnosis should be kept in mind in a child presenting with a neck mass, especially in a patient with a concomitant protruding pharynx mass. This entity may rarely be caused by recent ear infection. Endovascular intervention remains the treatment of choice in children because of lower associated mortality rates compared to an open surgical approach.[2,5]

### References

1 Requejo F, Sierre S, Lipsich J, Zuccaro G. Endovascular treatment of post-pharyngitis internal carotid artery pseudoaneurysm

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with a covered stent in a child: a case report. *Childs Nerv Syst* 2013;**29**:1369–73
2 Extracranial carotid artery aneurysm. In: http://www.uptodate.com/contents/extracranial-carotid-artery-aneurysm [10 June 2015]
3 Garcia-Monaco RD, Kohan AA, Martinez-Corvalan MP, Cacchiarelli N, Peralta O, Wahren CG. Thrombin injection failure with subsequent successful stent-graft placement for the treatment of an extracranial internal carotid pseudoaneurysm in a 5-year-old child. *Cardiovasc Intervent Radiol* 2012;**35**:704–8
4 Pourhassan S, Grotemeyer D, Fokou M, Heinen W, Balzer K, Ramp U *et al*. Extracranial carotid arteries aneurysms in children: single-center experiences in 4 patients and review of the literature. *J Pediatr Surg* 2007;**42**:1961–8
5 Pulli R, Dorigo W, Pratesi G, Fargion A, Pratesi C. Single-center experience on endovascular repair of noninfected extracranial internal carotid artery pseudoaneurysms. *Ann Vasc Surg* 2013; **27**:672.e13–17
6 Padayachy V, Robbs JV. Carotid artery aneurysms in patients with human immunodeficiency virus. *J Vasc Surg* 2012;**55**: 331–7
7 Rosset E, Albertini J, Magnan PE, Ede B, Thomassin JM, Branchereau A. Surgical treatment of extracranial internal carotid artery aneurysms. *J Vasc Surg* 2000;**31**:713–23

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