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#### Abstract

**Background:** Giant cerebral aneurysm occurring in the setting of right sided aortic arch (RAA) is exceedingly rare.

**Case Description:** A 56 years old female presented with acute headache. MRI of her brain showed subarachnoid haemorrhage (SAH) and a left internal carotid artery (LICA) paraclinoid segment giant aneurysm. During catheter angiography she was noted to have RAA. The left common carotid artery arose from the aortic arch with a very acute angle. Partial coiling of the LICA giant aneurysm was performed using a 6F guiding catheter with an inner 5F catheter with SIM curve in order to navigate the acute angle. Flow diverter was inserted three weeks later under double antiplatelet cover. Follow up angiogram at 6 months showed complete occlusion of the aneurysm.

**Conclusion:** cerebral aneurysms can be treated successfully with endovascular approach despite difficult and unpredictable anatomy in the rare situation of concurrence of RAA.

Keywords: right aortic arch, giant cerebral aneurysm

#### **INTRODUCTION**

RAA is a rare anomaly with estimated incidence of 0.01% to 0.1%. Among the cases with RAA, many anatomical variations of the origins of the common carotid arteries, in nominate arteries, subclavian as well as vertebral arteries have been documented. Occurrence of cerebral aneurysm in association with RAA is rare. In the presence of anatomical variations, endovascular access to treat the target intracranial aneurysm may be difficult or impossible, leaving surgical reconstruction as the only other viable option. Herein a case of ruptured giant LICA paraclinoid segment aneurysm in association with RAA treated with staged endovascular coiling followed by flow diversion is reported.

#### **CASE DESCRIPTION**

A 56 years old female with no past medical history presented with sudden onset of severe headache

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and neck pain. Clinically there was neck stiffness but no neurological deficit. MRI of her brain showed SAH and LICA paraclinoid segment giant aneurysm. During catheter angiography she was noted to have RAA. The aorta was on the right side. Arch injection in AP and multiple oblique views showed that the first branch was the left common carotid artery, followed by right innominate artery and then an aberrant left subclavian artery (Fig.1 and 2). The left common carotid artery arose from the proximal arch at a very acute angle. Attempts to cannulate it with a regular40degree multipurpose tipcatheter (4F, Ber II, Codman, USA) or Head-Hunter curve (5F, Radifocus, Terumo, Japan)was unsuccessful. It was eventually cannulated with a 5F diagnostic catheter with SIM 1 side winder curve (Radifocus, Terumo, Japan). At the origin of the left subclavian artery there was a Kommerell's diverticulum. The left vertebral artery arose from the left subclavian artery (Fig.3). The right innominate

artery gave rise to the right common carotid artery and the right subclavian artery. The right vertebral artery arose from the right subclavian artery (Fig.4). Selective injection of the left internal carotid artery showed a giant LICA aneurysm at the paraclinoid segment with a daughter sac (Fig.5). In view of the acute SAH, large size of the aneurysm as well as the presence of a daughter sac, the risk of rebleeding was assessed to be high. In the acute phase in view of the risk of rebleeding and the potential need for cerebrospinal fluid diversion procedures antiplatelet medication was deemed not advisable. Therefore, a strategy of staged coiling of the aneurysm to reduce the risk of rebleeding, followed by delayed flow diversion when double antiplatelet therapy could be administered was adopted. Surgical clipping of the aneurysm was also offered to the patient. She opted to have endovascular treatment rather than craniotomy. Partial coiling of the LICA

giant aneurysm was performed subsequently using a 6F Chaperone guiding catheter with SIM 1 curve (Microvention, Terumo Corporation, Japan) in order to navigate the acute angle. The inner 5F catheter of the Chaperone guiding catheter facilitated engagement and cannulation of the left common carotid artery. She tolerated the procedure well with no complication. Flow diverter was inserted three weeks later under double antiplatelet cover. A Pipeline Flex flow diverter (Medtronic Corporation, USA) was delivered with 6F Chaperone guiding catheter with SIM 1 curve inner 5F catheter, intermediate AXS Catalyst 5F catheter (Stryker Corporation, USA) and XT 27 microcatheter (Stryker Corporation, USA). Follow up angiogram at 6 months showed complete occlusion of the aneurysm (Fig.6). The patient made a full recovery and returned to work.



**Figure 1.** Angiogram arch injection chest AP view showed RAA with left common carotid artery arising as a first branch at an acute angle. Note the Kommerell's diverticulum at the origin of the left subclavian artery.



Figure 2. Angiogram arch injection chest right oblique view showing origin of the right innominate artery.



**Figure3.** Angiogram arch injection chest left oblique view showing Kommerell's diverticulum at the origin of the left subclavian artery. The left vertebral artery origin from the left subclavian artery was also demonstrated.



**Figure4.** Angiogram of the right innominate artery showing origins of the right common carotid and subclavian arteries. The origin of the right vertebral artery from the subclavian artery was also demonstrated.



**Figure5.** Left internal carotid artery angiogram oblique view demonstrating a paraclinoid segment giant aneurysm length 25mm with a daughter sac.



**Figure6.** Left internal carotid artery angiogram 6 months after staged coiling and flow diverter insertion showed complete obliteration of aneurysm.

# **DISCUSSION**

RAA is a rare congenital anomaly with estimated incidence of 0.01% to 0.1%.<sup>(2,4,7)</sup> Embryo logically it arose when the artery of the 4<sup>th</sup> branchial arch on the right side persists instead of the left side. It is known to be associated with other congenital heart anomalies such as tetralogy of Fallot or truncus arteriosus.<sup>(4,5)</sup> Those cases without severe congenital heart anomaly may remain asymptomatic well into adulthood. It is important to recognize RAA when endovascular or surgical treatment are considered for cerebral aneurysms. In the presence of RAA the origins and courses of common carotid, innominate, subclavian or vertebral arteries can be highly variable. <sup>(4,7)</sup> The presence or absence of either theinternal carotid or vertebral arteries must also be ascertained in case of the need to perform parent artery sacrifice with or without bypass for complex aneurysms. The occurrence of cerebral aneurysm in association with RAA is rare. PubMed search of published literature yielded two case reports of small cerebral aneurysms. In the case reported by Yamasaki et al, there was a small right internal carotid-anterior choroidal artery

internal carotid artery.<sup>(8)</sup> This aneurysm was treated with open surgical clipping. The choice of surgical clipping in this case was obvious because of the ability to reconstruct the parent artery or to perform a bypass when the need arises in view of agenesis of the contralateral carotid artery. The other case reported by Ichinose et alinvolved a small left middle cerebral artery bifurcation aneurysm that was treated with endovascular coiling without difficulty.<sup>(3)</sup> Thiscase reported herein differ from the other two in its internal carotid-paraclinoid location and the giant size. In view of the acute SAH, large size of the aneurysm as well as the presence of a daughter sac, the risk of rebleeding was assessed to be high. In the acute phase in view of the risk of rebleeding and the potential need for cerebrospinal fluid diversion procedures antiplatelet medication was not advisable. Without flow diversion or addition of a stent, the risk of recurrence will be high also. Therefore, a strategy of staged coiling of the aneurysm, followed by delayed flow diversion when double antiplatelet therapy could be administered was adopted.<sup>(1)</sup> The potential difficulty in this case

aneurysm in association with agenesis of the left

was the ability to navigate the tight acute angle of the left common carotid artery from the RAA to provide enough support for coiling of the aneurysm as well as delivery of a flow diverter with a 27 microcatheter. After failure with multipurpose tip and Head-Hunter curve, the SIM side winder curve successfully navigated the acute angle. This can be done with any tri-axial technique using any 0.035" wire combined with an inner 4 or 5F SIM with an outer 6F guide catheter. The alternative treatment option for this case is surgical clipping, with or without suction decompression.<sup>(6)</sup> The patient opted for endovascular approach. The excellent clinical recovery with the staged coiling followed by flow diversion, as well as angiographic complete obliteration of the aneurysm at 6 months was encouraging.

#### **CONCLUSION**

It is important to recognize the anatomical variations during angiography for SAH. Cerebral aneurysms, even with giant size, can be treated successfully with reconstructive endovascular approach despite difficult and unpredictable anatomy in the rare situation of concurrence of RAA.

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