CASE REPORT

Hybrid Thoracic Endovascular Repair of a Large, Saccular Aortic Arch Aneurysm with Coil Embolization of the Left Subclavian Artery in a 31 Year Old Filipino Female with Takayasu Arteritis and Multiple Intracranial Aneurysms: A First in the Philippines

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Abstract

BACKGROUND: Takayasu Arteritis (TA) is a rare, primary large-vessel vasculitis frequently leading to stenosis and less commonly, aneurysm formation. Saccular aneurysms of the aortic arch in patients with TA are fatal, have rarely been reported and represent a significant technical challenge due to the difficult anatomical location and need for protection of the cerebral circulation. Concomitant intracerebral aneurysms in patients with TA are extremely uncommon and have mostly been documented in very few case reports in literature.

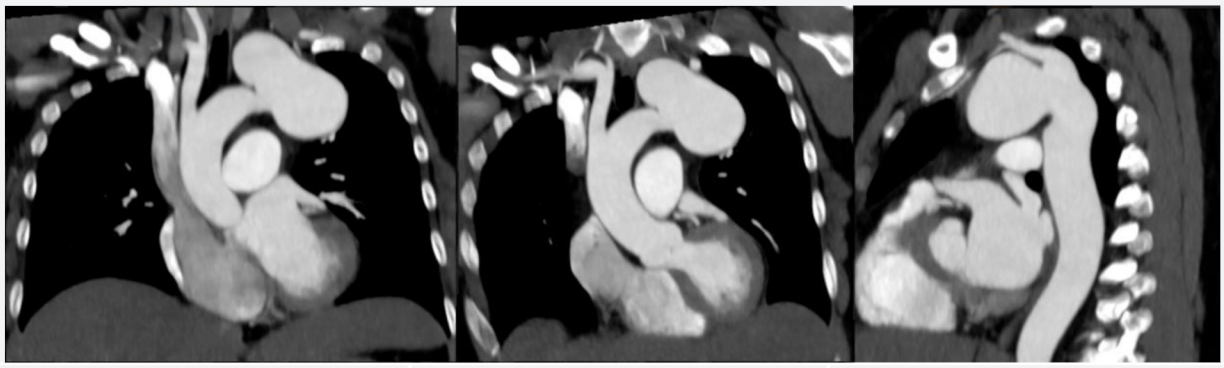
CASE DISCUSSION: We present a case of a 31 year-old Filipino female with recurrent chest and neck pain radiating to the upper back. Computed tomographic (CT) angiography demonstrated a large saccular aortic arch aneurysm without branch stenosis. CTA of the cerebral circulation likewise demonstrated multiple, saccular, intra-cerebral aneurysms. She underwent hybrid thoracic arch repair with supra-aortic debranching via mini-sternotomy and proximal ligation of the left common carotid artery and staged endovascular aortic arch replacement with coil embolization of the ostial-to-proximal left subclavian artery segment. Post-operative aortogram showed optimal repair with thrombosed aneurysmal sac, optimal graft position, no endoleaks and preservation of cerebral circulation. Patient improved symptomatically post-procedure and remained symptom-free during follow-up after six months. Careful review of local literature suggests that this is the first Philippine TA case with a saccular aortic arch aneurysm successfully managed in this manner.

CONCLUSION: Saccular aortic arch aneurysms in patients with Takayasu are unusual and presence of concomitant multiple cerebral saccular aneurysms have rarely been reported in literature. This case highlighted that hybrid endovascular arch repair in patients with TA is feasible, minimally invasive and effective.

KEYWORDS: Takayasu arteritis, aortic arch aneurysm, cerebral aneurysm, hybrid endovascular arch repair, aortic de-branching

INTRODUCTION

Takayasu Arteritis (TA) is a rare, chronic inflammatory large vessel vasculitis primarily affecting adolescent girls and young females[1] with a worldwide incidence of 0.3 to 3.3/million/year and prevalence rate of 4.7 to 360 cases/million.[2] It is a systemic granulomatous inflammatory process which most often results in stenosis, occlusion, dilatation, and aneurysms of the arterial wall.[3] Aneurysmal involvement of the aorta in the absence of classic stenotic lesions have been reported in 2.8% to 31.9%.[4,5] Isolated saccular aneurysms of the aortic arch have been rarely reported.[4,5] Comorbid aneurysmal involvement of the intracerebral circulation is unusual and



A B C

Figure 1A - B. CTA demonstrating the large saccular, lobulated, aortic arch aneurysm arising between the left common carotid and left subclavian artery. **Figure 1C.** Image showing the saccular lobulated aneurysm just before the takeoff of the left subclavian artery (LSA) which appears widely patent and tortuous.

probably underreported in the literature. In a local retrospective study done by Afos et. al in 2018[6], arterial stenoses are seen in 90% of patients with TA and associated with mostly fusiform aortic aneurysmal involvement in about 31.8%.

The diagnosis of TA is challenging as it often presents with nonspecific symptoms that delays the diagnosis and the treatment.[1,2] Intracranial aneurysmal involvement in patients with TA is rare and based on our review, it has been reported in 25 cases worldwide.[7-13] However, intracranial vascular abnormalities in patients with TA may actually be underestimated given limited studies and lack of evidence regarding the value of routine imaging of the intracranial vessels in this disease.[13-15]

CASE PRESENTATION

A 31 year old Filipino female, obese, pre-diabetic, hypertensive for two years on combination anti-hypertensive agents came in for an incidental finding of widened mediastinum on chest x-ray. Four years prior, a chest x-ray from another institution showed a suspicious mediastinal density, however, follow-up chest CT scan was unremarkable. She remained asymptomatic until six months prior when she experienced recurrent, intermittent, 5/10, stabbing chest pain lasting for a few minutes radiating to the upper back and left scapular area, spontaneous resolving. She is a previous smoker, occasionally drinks alcohol without a history of illicit drug use. She is nulligravid with a regular menstrual cycle. Family history was unremarkable.

She appears obese, with right upper extremity blood pressure of 120/80 mmHg and 130/80 mmHg on the left, audible systolic bruits over the left carotid and left subclavian artery. Pulses were full and equal. Cardiopulmonary exam was normal. There was no abdominal bruit. Chest x-ray demonstrated a widened superior mediastinum with left-sided convex prominence**.** A follow-up thoraco-abdominal CTA showed a large, lobulated, saccular aneurysm originating between the takeoff of the left common carotid artery and left subclavian artery and directed laterally and superiorly (Figure 1A and 1B). It measured 3.9 x 7.4 x 4.3 cm (APxCCxT) with a neck of 4.2 x 2.3 cm (APxCC). The left subclavian artery was markedly tortuous and angulated as a result of compression by the aneurysmal dilatation. It appeared dilated proximally measuring 1.8 x 2.1x 1.3 cm (APxTxCC) (Figure 1C). There were no stenotic lesions in the abdominal visceral branches nor the aorta. Carotid duplex showed bilateral carotids with smooth blood vessel intima and mid-systolic velocity deceleration in the left vertebral artery indicative of pre-subclavian steal phenomenon. CRP, p-ANCA, c-ANCA, ANA, C3, RF, anti-CCP, total Vitamin D total, RPR, quantiferon TB, procalcitonin, hepatitis screen and renal profile were normal. ESR was 65 mm/hr. A transthoracic echocardiography was unremarkable.

She was referred to Rheumatology service for medical management and HEART team for possible surgical aortic arch replacement. However, she refused open repair and opted for hybrid arch repair with staged fully percutaneous endovascular repair of the aortic arch and coil embolization of the proximal left subclavian artery.

She was started on steroid pulse therapy, methotrexate and folic acid. She subsequently underwent debranching of the supra aortic vessels with innominate to left common carotid and subclavian bypasses and proximal ligation of the left common carotid artery via mini-sternotomy. The LSA was not ligated due to difficult access and enhanced risk of aortic rupture with manipulation. Staged fully percutaneous endovascular arch repair with coiling of the ostial to proximal LSA was carried out two days after the de-branching procedure.

Endograft repair was done using a thoracic stent grafts within the arch of the aorta (Zone 1) ensuring its optimal position just distal to the innominate artery. Another stent graft was deployed within overlap of the two grafts in order to ensure optimal overlap and improved radial force followed by balloon expansion. Coil embolization was done in order to prevent retrograde filling of the aneurysmal sac. Follow up angiographic images demonstrated optimal placement of the coils within the

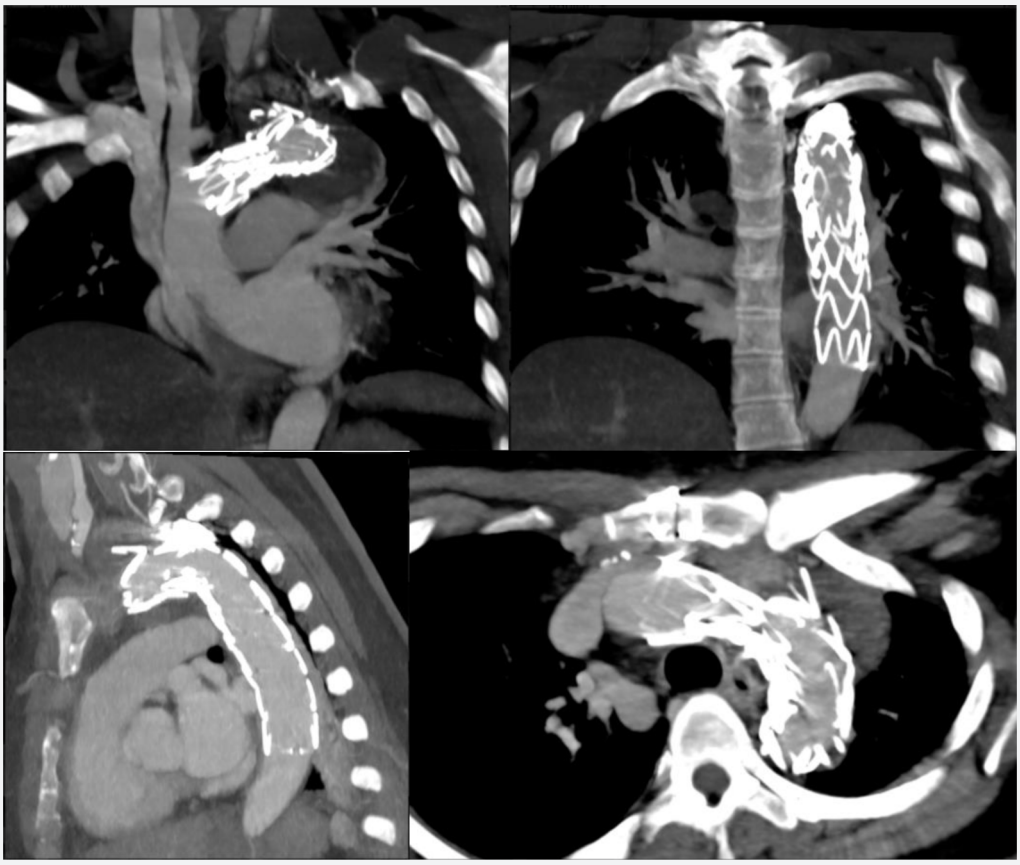


Figure 2. Follow-up CTA after one week demonstrating optimal repair with thrombosed aneurysmal sac, optimal graft position and no endoleaks and preservation of the cerebral circulation. Coils within the LSA are likewise stable.

LSA without retrograde flow into the aneurysmal sac (Figure 2). The patient tolerated the procedure well and was subsequently started on cardiac rehabilitation. The patient was continued on Aspirin 80 mg and Clopidogrel 75 mg once a day post-operatively.

Following repair, she developed recurrent left temporal headaches, generalized fatigue, diaphoresis, and abdominal discomfort. A positron emission tomography (PET) scan with 18F-FDG showed no hypermetabolic lesions in the aorta and arterial walls however suggested the presence of a saccular aneurysm at the left anterior cerebral circulation. CTA demonstrated incidental findings of pulmonary emboli at the right interlobar pulmonary artery, superior and anterior segmental branches of the right lower lobe. A wedge shaped hypodensity on the spleen suggested a splenic infarct. Workup for hypercoagulable state which included antiphospholipid antibody syndrome panel, Protein C, Protein S, Factor V Leiden and Jak2V617F all tested negative. She was started on a therapeutic enoxaparin dose. Increasing ESR trends prompted escalation of immunosuppression with pulse therapy with addition of Tocilizumab. Brain CTA demonstrated multiple saccular aneurysms (Figure 3). She refused cerebral angiogram and further invasive assessment of her intracerebral disease. She was thus managed with aggressive risk factor modification, optimized control of hypertension, avoidance of cigarette smoking as well as optimized medical therapy. She was discharged on tapering doses of steroids and oral anticoagulation with serial monitoring of ESR. On follow up after six months, she remains asymptomatic and fully functional.

CASE DISCUSSION

Majority of TA cases can be treated medically but due to diagnostic delays, arterial injury accumulates and surgical intervention may already be necessary.[2] Aneurysms involving the aortic arch are considered one of the major complications related to prognosis in TA and once present usually require surgical repair.[3] This poses a technical problem due to difficulty in resection and replacement of these arteries with a high incidence of stroke but the advent of hybrid repair offers a promising approach.[17-18] Currently, there are no existing guidelines to direct the choice between performing an open surgery versus endovascular approaches.

In the case of this patient, the location of the aneurysm poses a technical difficulty in that the endograft needs at least two centimeters landing zone which would mean occlusion of the

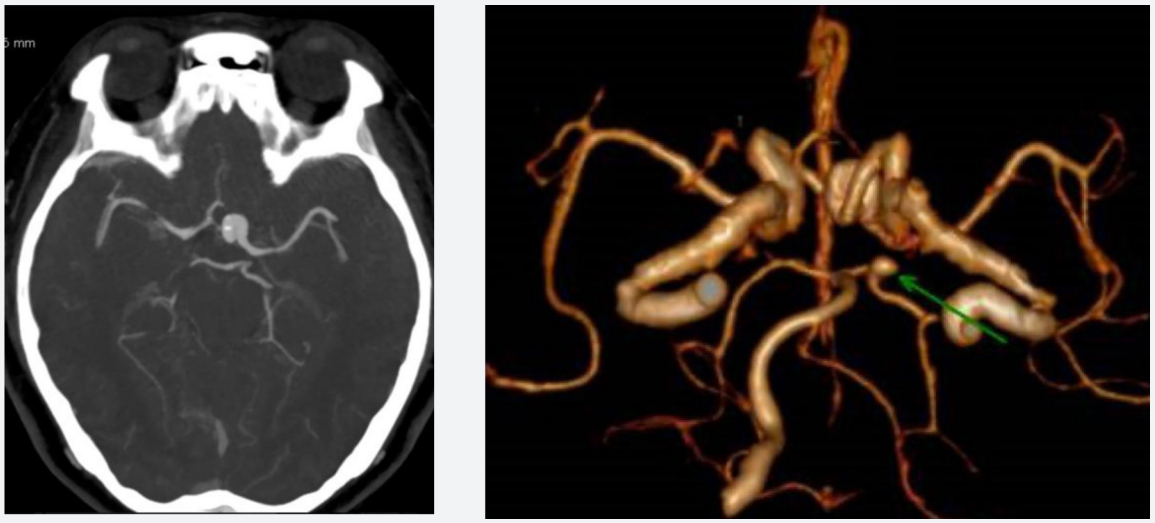


Figure 3. Inferiorly directed saccular aneurysm at the junction of the left posterior communicating artery and left P1 segment of the posterior cerebral artery.

arch vessels and compromising blood flow to the head and neck. The patient also has a left subclavian artery aneurysm where aortic arch revascularization is necessary to avoid posterior circulation stroke. Also considering a young female, a median sternotomy approach to repair aortic arch aneurysm would raise aesthetic concerns. A hybrid approach would minimize invasiveness and insult to this patient by minimizing cross clamping and circulatory arrest time. Anatomic management dilemma in this patient includes compromising the left common carotid and left subclavian artery with an endograft hence extrathoracic debranching was done with bypass graft connecting the brachiocephalic and left common carotid artery. This ensures that vital perfusion of the aortic side branches is maintained.

Studies show that periprocedural immunosuppression has significantly improved outcomes in those that undergo aortic repair.[4] This patient receive periprocedural immunosuppression with methylprednisolone, methotrexate, and post operative tocilizumab which is an effective therapy for TA that when added to steroids may haste steroid discontinuation.[19] Determining disease activity and pattern of arterial involvement in TA remain a challenge since hallmarks of active disease are yet to be established[19].

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