Surgical Repair of Abdominal Aortic and Renal Artery Aneurysms in Takayasu's Arteritis

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Abstract

Takayasu's arteritis is a large vessel vasculitis that can be a challenging diagnosis to make and has a varied clinical presentation. Management largely depends on affected vessel disease severity and individual patient considerations. The diagnosis must be considered in a young patient with large vessel aneurysms. We present a case of a 30 year-old woman of Pacific Islander descent who presented to the Tripler Army medical Center Vascular Surgery Department in Honolulu, Hawai'i seeking repair of her abdominal aortic and renal artery aneurysms prior to conception.

A 30 year-old Pacific Islander woman with a history of a saccular abdominal aortic aneurysm and renal artery aneurysms presented to our clinic seeking vascular surgery consultation prior to a planned pregnancy. She had a renal artery stent placed at an outside institution for hypertension. She met the diagnosis of Takayasu's arteritis by Sharma's criteria. Physical exam was significant for a palpable, pulsatile, abdominal mass and CT angiography revealed a saccular irregular-appearing infra-renal abdominal aortic aneurysm, extending to the aortic bifurcation, with a maximum diameter of 3.3 cm. A right renal artery aneurysm was also identified proximally, contiguous with the aorta, with a maximal transverse diameter of 1.7 cm. The patient underwent a supraceliac bypass to the right renal artery with a 7mm Dacron graft, as well as excision of the right renal artery aneurysm. The abdominal aortic aneurysm was replaced using a Hemashield Dacron bifurcated 14mm x 7mm bypass graft. Intraoperative measurements of the renal artery aneurysm were 1.5 x 1.5 cm and the saccular appearing distal abdominal aortic aneurysm measured 3.6 x 3.3cm. The patient was discharged from the hospital 7 days post-operatively. At 1-year follow up, CT scan of the abdominal aorta revealed the repair was without any evidence of aneurysm formation, anastomotic pseudoaneurysm formation, or areas of stenosis. She has remained normotensive with a normal serum creatinine 18 months after her repair. She has since delivered her second child.

It is rare for Takayasu's arteritis to present with aneurysmal disease. It is much more common to present with stenosis or occlusion. It has yet to be proven that Takayasu's truly has a higher incidence in those of Asian descent. Takayasu's can be a difficult diagnosis to make but can be aided with the use of Sharma's criteria. Our particular patient posed unique considerations on the effects of the physiology of pregnancy on her aneurysms and repair.

Managing the active phases of disease is imperative, and though medical management is first line, surgical intervention may be necessary. Surgical intervention should be performed in a quiescent period of disease if possible given that biological inflammation at the time of intervention increases the complication rate. Repair of aneurysmal disease in a young female should also be considered prior to pregnancy.

Keywords

Arterial Aneurysms, Pregnancy, Abdominal Aortic Aneurysm, Renal Artery Aneurysm

Introduction

Takayasu's arteritis (TA) is a large vessel vasculitis. The current body of knowledge is varied, from the histopathology, diagnosis, and clinical manifestations. The histopathological features vary from an extensive acute inflammatory process to extensive post-inflammatory mural fibrosis.[1] The most severe manifestations of Takayasu's artertitis are Takayasu retinopathy, secondary hypertension, aortic regurgitation, and aneurysm formation. The diagnosis is made clinically according to Sharma's criterion for diagnosing Takayasu Arteritis.[2] The gold standard for diagnosis is angiography exhibiting stenosis particularly of midsubclavian artery lesions.[3]

We report the case of a young Pacific Islander woman who presented with aneurysmal disease and was successfully managed with open surgical repair. This case is significant to the heath care field in that our patient had an unusual presentation with aneurysmal disease and a complicated surgical history, including a prior renal artery stent placed across an inflammatory aneurysm that was freely floating. We also provide a review of the literature to aide clinicians in diagnosis and medical management and show that data supporting a racial predisposition is actually lacking. The surgical management of a young patient with complicated inflammatory aneurysms with a previously placed renal artery stent is unique and we hope to provide one method for consideration.

Case Report

A 30 year-old Pacific Islander woman (as defined as the areas within the Pacific Ocean encompassing Micronesia, Polynesia, and Melanesia) with a history of a saccular abdominal aortic aneurysm presented to our clinic seeking vascular surgery consultation prior to a planned pregnancy. She also had a proximal saccular renal artery aneurysm associated with a presumed distal renal artery stenosis, previously stented in Italy. The patient initially had her abdominal aortic aneurysm detected during her first pregnancy 4 years prior. Her first pregnancy was unremarkable. She delivered at term under high risk obstetric care. However, it was recommended by her subspecialist obstetrician at a US academic center, that if she were to consider pregnancy again she should seek repair of both aneurysms prior to conceiving. In the interim she developed refractory hypertension secondary to renal artery stenosis necessitating stent placement.

She reported having a history of fevers, chills, and rigors in her youth. By Sharma's diagnostic criterion for Takayasu's arteritis our patient met at least two major and three minor criteria. Her major criteria included characteristic signs and symptoms that were unexplained in her adolescence, and a documented discrepancy in systolic blood pressure in her upper extremities associated with a subclavian murmur (R 170/84 and L 151/99).

Her minor criteria included an abdominal aortic lesion, elevated ESR (28mm/hr at age 29), and hypertension. Physical exam was significant for a palpable, pulsatile, abdominal mass and CT angiography revealed a saccular irregular-appearing infra-renal abdominal aortic aneurysm, extending to the aortic bifurcation, with a maximum diameter of 3.3 cm (Figure 1). A right renal artery aneurysm was also identified proximally, contiguous with the aorta, with a maximal transverse diameter of 1.7 cm (Figure 2). Given the saccular nature of the aortic aneurysm, as well as the patient's desire to become pregnant, the decision was made to proceed with an open repair. The patient underwent a supraceliac bypass to the right renal artery with a 7 mm Dacron graft, as well as excision of the right renal artery aneurysm. The abdominal aortic aneurysm was replaced using a Hemashield Dacron bifurcated 14mm x 7mm bypass graft. Intraoperative measurements of the renal artery aneurysm were 1.5 x 1.5 cm and the saccular appearing distal abdominal aortic aneurysm measured 3.6 x 3.3cm. There were no intra-operative or post-operative complications, and the patient was discharged from the hospital 7 days post-operatively. On attempted sectioning of her removed renal aneurysm, the tissue was extensively calcified without active inflammation. At 1-year follow up, CT scan of the abdominal aorta (Figure 3 and 4) revealed the repair was without any evidence of aneurysm formation, anastomotic pseudoaneurysm formation, or areas of stenosis. She has remained normotensive with a normal serum creatinine 18 months after her repair. She has recently conceived and will be monitored closely throughout her pregnancy.

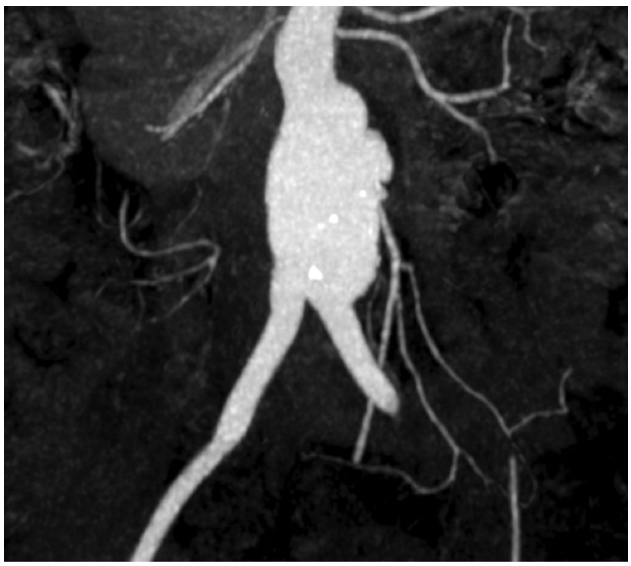

Figure 1. Saccular irregular-appearing infra-renal abdominal aortic aneurysm, extending to the aortic bifurcation with a maximum diameter of 3.3 cm.

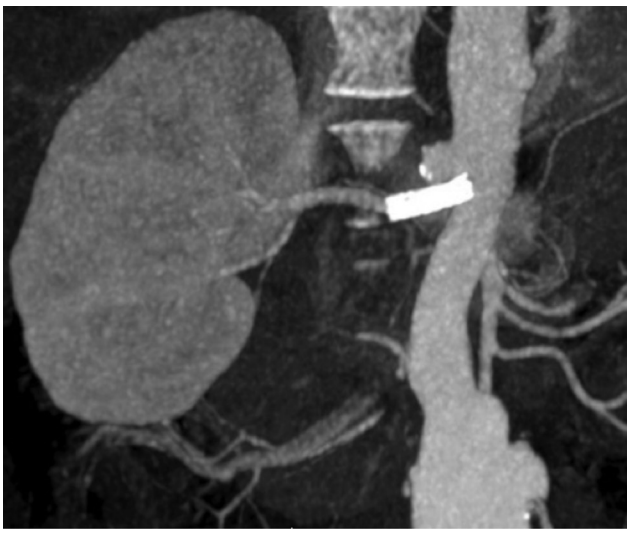

Figure 2. Right renal artery aneurysm contiguous with the aorta with a maximal transverse diameter of 1.7 cm. Stent is seen protruding into the proximal aneurysm cavity.

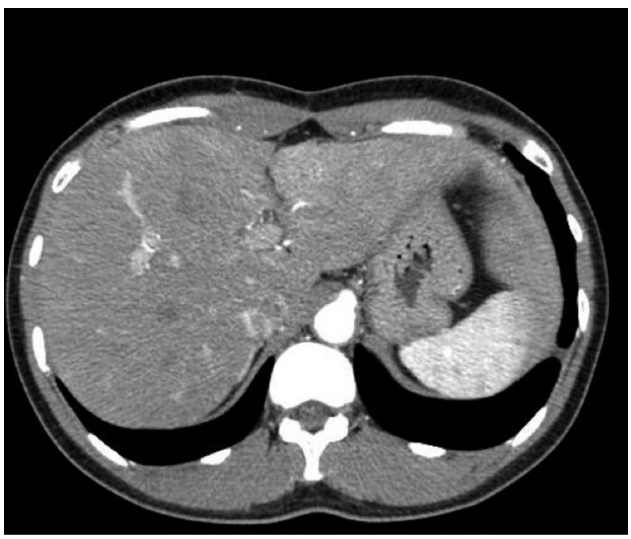

Figure 3. Supraceliac origin of right renal artery bypass graft.

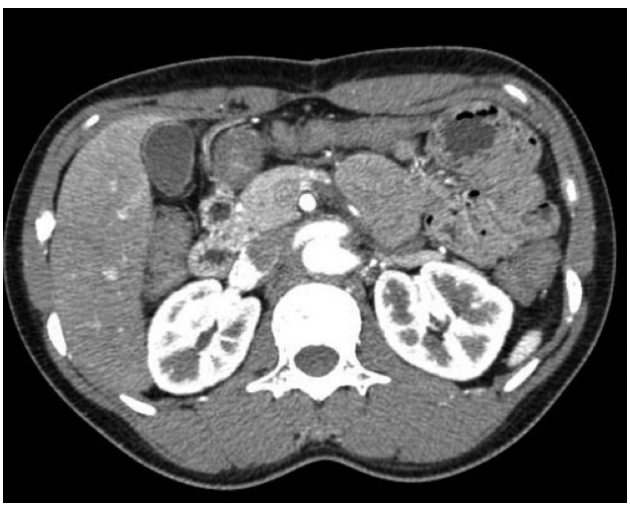

Figure 4. Right renal artery aneurysm exclusion with thrombosis in the original renal artery aneurysm.

Discussion

In young patients with aneurysmal disease connective tissue disorders should be ruled out. TA more commonly presents with stenosis or occlusion than aneurysmal disease, and should also be included in the differential. Our patient had both stenosis and aneurysmal disease, requiring a stent placement for refractory hypertension. Aneurysmal disease is more common depending on the geographic location, with 71% of TA patients in South Africa having aneurysmal disease, versus only 32% in Japan.[4] The aneurysms associated with TA are frequently multiple, saccular aneurysms that are associated with stenotic lesions.

TA is a large vessel vasculitis known for its propensity to affect women from childhood until the age of forty. TA is rare, with an annual incidence of 1.2-2.6 cases/1 million.[5] Most series reveal that 85% of cases are women, although the gender association has been shown to vary with geographic location.[6] While classically thought to be a disease with increased prevalence in Asians, no racial predisposition has been proven.[6] Vessels most commonly involved are the aorta and its immediate branch vessels. TA presents pathologically as fibrous transmural thickening following necrotizing inflammation of the aorta and major vessels, similar to giant cell arteritis. The transmural intimal thickening results in weakening of the vessel wall, predisposing the patient to aneurysm formation.[7] Clinical manifestations are dependent on the vessels involved; however, nonspecific and systemic complaints such as fever, malaise, weight loss, arthralgia, myalgia, and night sweats are well described.[7] The transmural thickening often results in severe stenosis of major branch vessels, hence the term "pulseless disease." Certain HLA haplotypes have been identified as being associated with TA, however, the pathogenesis is unknown.[8]

Diagnosis may be made with histology, revealing panarteritis of all three layers of the vessel wall, with skip areas along the length of the vessel.[9] However, histology is usually not available, so historically the most widely accepted clinical diagnostic criteria were Ishikawa'a criteria (1988), which have since been modified by Sharma, et al, improving sensitivity to 92.5% while maintaining a 95% specificity when applied to their population.[2] Sharma's criteria include three major and ten minor criteria (Table 1).[2] The presence of two major, or one major and two minor, or four minor criteria imply a high clinical suspicion for TA.

Treatment of TA is initially medical. However, approximately 20% of patients are resistant to medical treatment, which justifies an aggressive medical regimen and possible surgical intervention.[10,11] Corticosteroids are used as first-line therapy to control the acute arteritis. If a patient is resistant to corticosteroids then treatment with cyclophosphamide, azathioprine, or methotrexate is started. Other proposed treatment modalities include mycophenolate mofetil and anti-tumor necrosis factor-α biological agents for those unresponsive to the other immunosuppressors.[11,12]

It is important to control the acute inflammation and acute arteritis prior to surgery. A multicenter retrospective analysis of 79 patients with TA who underwent surgical or endovascular intervention revealed that biological inflammation at the time of revascularization was associated with the occurrence of arterial complications regardless of the method of repair, with biological inflammation defined as ESR >30mm/hr and CRP >6 mg/L.[10] The most commonly encountered complication after endovascular intervention reported in this series was renal artery re-stenosis. Other post-intervention complications include bleeding, stroke, thrombosis, infection, and anastomotic failure.[9,10]

Table 1. Sharma's Criteria. Characteristic signs and symptoms include limb claudication, pulse differences or pulselessness in limbs, difference of >10 mm Hg in systolic blood pressure between limbs, fever, pain, transient amaurosis, blurred vision, syncope, dyspnea, or palpitations.

10 Minor Criteria	3 Major Criteria
Persistently high ESR	Left mid-subclavian artery lesion
Carotid artery tenderness	Right mid-subclavian artery lesion
Hypertension	Characteristic signs and symptoms for at least one month's duration
Aortic regurgitation or Annuloaortic ectasia	
Pulmonary artery lesion	
Left mid-common carotid lesion	
Distal brachiocephalic trunk lesion	
Descending thoracic aorta lesion	
Abdominal aorta lesion	
Coronary artery lesion in the absence of risk factors	

The periarterial adventitial thickening of aneurysms with TA results in a lower rate of rupture compared to degenerative aneurysms.[13] The life expectancy of these patients is expected to be nearly the same as healthy individuals. Over a longer time period, patients with long standing TA and aneurysm formation do have an increasing risk of aneurysm rupture. These patients may not suffer from the same medical comorbidities as patients with degenerative aneurysms and would be suitable candidates for surgical repair.

Hypertension with aneurysmal formation in TA is present in a significant portion of patients. In a paper by Sharma et al, of the 88 patients diagnosed to have TA, eight patients had aneurysms. Those eight patients all had long standing uncontrolled hypertension.[14] The high prevalence of systemic hypertension and TA suggests the causative factor for aneurysm formation is high blood pressure. Stenotic disease is associated with aneurysm formation in TA patients. Isolated aneurysm formation occurs in only 2% of patients with TA.[15] Surgically correctable stenotic lesions would prevent future aneurysm formation and progression.

TA with aneurysm formation in pregnant patients presents a unique challenge. The effects of hypertension during pregnancy on fetal demise are well known. The other concern is further

aneurysm growth during pregnancy. Under the age of 40, half of ruptured arterial aneurysms occur in pregnant women.[16] Pregnancy related circulatory and endocrine changes may cause new aneurysm and further expansion of aneurysms already present.[16] A ruptured aortic aneurysm during pregnancy could mimic benign disease processes leading to high mortality rates. Early repair of TA related aneurysms will potentially eliminate the potential for rupture and allow future pregnancies.

Conclusion

There is often a delay in diagnosis of TA given its varied presentation and nonspecific symptoms. Managing the active phases of disease is imperative, and though medical management is first line, surgical intervention may be necessary. Surgical intervention should be performed in a quiescent period of disease if possible given that biological inflammation at the time of intervention increases the complication rate. Repair of aneurysmal disease in a young women should also be considered prior to pregnancy. After endovascular treatment of renal artery stenosis, our patient successfully underwent open aortic and renal artery aneurysm repair and has delivered a second child by an uncomplicated repeat low transverse Caesarean section.

The views expressed in this manuscript are those of the authors and do not reflect the official policy or position of the Department of the Army, Department of Defense, or the US Government.

Conflict of Interest

None of the authors identify any conflict of interest.

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