Takayasu disease and cardiac manifestations, presentation of a case with Takayasu and coronary artery disease

Sadeghpour Tabaee Ali (MD)*, Rostami Alireza (MD)*,Baghaei Tehrani Ramin (MD)*
Deris Bahador (MD)*, Nourizadeh Eskandar (MD)*, Pourabasi Mohamadsadegh (MD)*

Abstract:
A 48 years old male who was reffered to us for coronary artery bypass graft,was scheduled for surgery and during the operation it was found that the patient is suffering from Takayasu arteritis. Patient had no suitable left internal thoracic artery for revascularizaton, so ,saphenous veins and off-pump coronary artery by pass graft were used for revascularization .The case is presented below and the cardiac manifestations and challenges in these patients is discussed.

Introduction: Takayasu disease or Takayasu arteritis is a chronic inflammation of the large blood vessels.It is most common in women (90%) of Asian descent. It usually begins between 10 and 30 years of age. The cause is unknown(1). The first case of Takayasu's arteritis was described in 1908 by Dr. Mikito Takayasu(2). Japanese studies support 90-95% survival rates in these patients. Morbidities in persons with TA are related to ischemia and hypertension and include congestive heart failure, transient ischemic attacks, stroke, and visual disturbances. Chronic low-grade dissection of the aorta may cause recurrent chest pain for years. At autopsy, children with TA who have died from acute rupture of the aorta often are found to have evidence of multiple prior small dissections that did not progress. Females comprise 80-90% of patients with TA. Pediatric studies are more varied. Sex distribution usually mirrors the 80-90% female preponderance observed in adults. Series of studies of TA in childhood from India and South Africa report a 2:1 female-to-male ratio. However, these are countries in which TA is associated strongly with tuberculosis, and additional etiologic and pathophysiologic factors may be present. TA is the most common large vessel vasculitis of adolescence. The youngest patient reported was aged 6 months. Systemic symptoms include the following: Fever, night sweats, Fatigue, Weight loss, Myalgia and/or arthralgia and/or arthritis, Skin rash (eg, erythema nodosum, pyoderma gangrenosum), Headaches and/or dizziness and/or syncope, Congestive heart failure, palpitations, angina, Hypertension (may be paroxysmal), Symptoms related to ischemia include the following: Ischemic stroke and/or transient ischemic attack,Visual disturbances (eg, blurred vision, diplopia, amaurosis), Carotidynia, Abdominal pain, Claudications (vary due to the development of collateral circulations; symptom is rare in children). The findings on physical examination are as follow: Blood pressure difference greater than 30 mm Hg between arms, Asymmetric pulses, Diminished or absent pulses (midaortic lesions found in children may not affect pulses), Asymmetric pulses (common) and absent pulses (rare), even in the later stages of the disease (awareness of this is critical), Poststenotic dilatations producing what appear to be bounding pulses (often present), Hypertension (may be paroxysmal): Since this typically results from renovascular compromise, this is a high-renin hypertension, Bruits, especially over subclavian arteries or aorta, on Funduscopic examination, Retinal hemorrhages, Cotton-wool exudates, Venous dilatation and beading, Microaneurysms of peripheral retina, Optic atrophy, Vitreous hemorrhage and Classic wreathlike peripapillary arteriovenous anastomoses (extremely rare) may be seen.Reported skin lesions include erythema nodosum-like lesions,

***Cardiovascular surgery Department, Rajaee Heart Center,Valiasr Ave,park mellat,Tehran,Iran ,postal code:1996911151.**
Corresponding author:Alireza Rostami,Rajaee heart center,Tehran,Iran Tel:098-21-23922589
Email:dr_ar_rostami@yahoo.com

pyoderma gangrenosum, leukocytoclastic vasculitis, and panniculitis . Takayasu disease is divided into early and late phases. The early phase is inflammatory and has been called the prepulseless phase, and the late phase is an occlusive one called the pulseless phase (4).

Case presentation:

A 48 year old smoker male, with history of diabet(NIDDM) , hyperlipidemy and hypertension with exertional dyspnea ,undervent coronary angiography and significant stenosis at the origins of Left main ,LAD and RCA was found.Angiography of carotid arteries showed nonsignificcant proximal right internal carotid artery stenosis , total occlusion of proximal left internal carotid artery,nonsignificant bilateral common carotid artery stenosis,and significant bilateral external carotid artery stenosis.Echocardiography showed:EFof 45-50%,normal LV size,mild LV dysfunction,mild LVH,mild diastolic dysfunction, moderate eccentric MR,AV sclerosis with peak PG=41mmHg,small size ascending aorta and aortic arch,normal RV size and function,mild PAH. CT angio of aorta and carotid arteries was performed and the results were as follow:dense calcified plaque at aortic arch, dense calcified plaque at the origine of supra aortic branches,significant stenosis in proximal part of both subclavian arteries,50-60% stenosis in distal part of both common carotid arteries,50-60% stenosis in the left internal carotid artery origin,remarkable stenosis in right external carotid artery origin, and stenosis at the origine of left vertebral artery.EKG:NSR/LBBB.positive findings on physical examination were as follow:bruit on auscultation of both carotid arteries ,right brachial BP:170/63 mmHg,left brachial artery BP:100/64 mmHg,an aortic 3/6 systolic murmur , and absent left radial pulse.pateint was reffered for CABG by cardiologist .there were no preoperative diagnosis of takayasu by cardiologist.During the operation, we found no palpable LIMA pulsation and severe calcification of ascending aorta and arch(Fig.1). Ascending aorta dimension was about 1.5-2 cm.Further examination of aorta showed a very small area lacking calcification just over RCA and in aortic root,measuring about 0.5 cm* 0.5 cm.canulation of aorta was impossible because of sever calcification of aorta. LIMA was harvested for free graft despite being pulseless,but after harvesting we found it thick,inflamed and calcified , and did not use it .we used saphenous vein grafts for revascularization of PDA and LAD off-pump,and made proximal anastomosis of PDA graft to aortic root in the small area over RCA which was suitable for anastomosis because of lacking calcification (Fig1)..

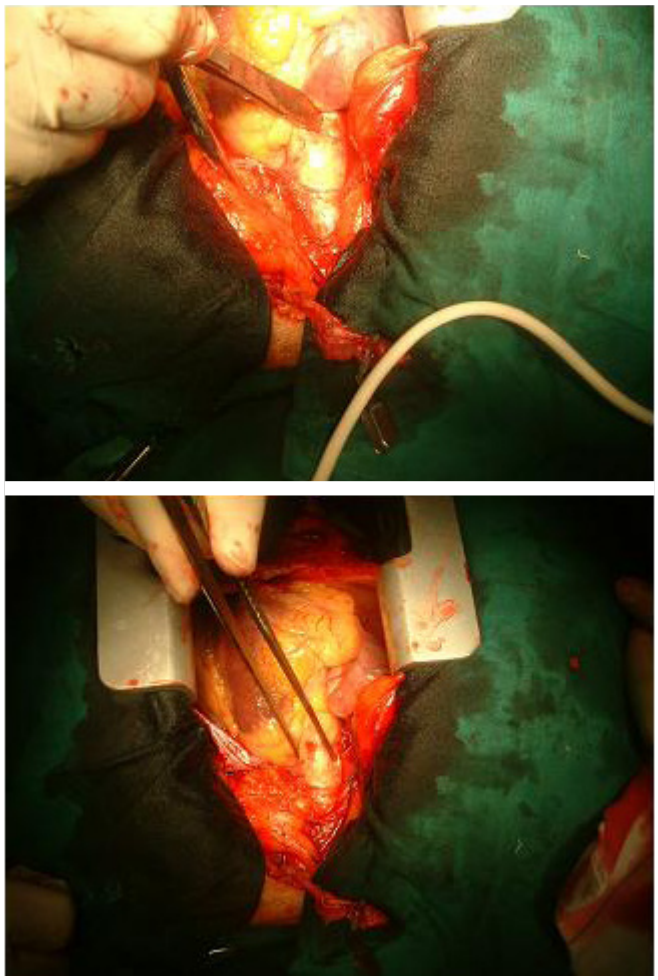

Fig.1. *Picture showing the only area over aortic root that was grafable(upper),and rigidity and small diameter of aortic root(lower)*

OM was not graftable.Aortic side clamp was used for a limited period of time and with difficulty but successfully. LAD graft was anastomosed over PDA graft in a manner most suitable for prevention of kinking .patient could be got off the pump without difficulty and underwent a normal, uneventful post operative course.rheumatologic counsultation was performed and corticosteroid was begun by rheumatologist with the diagnosis of takayasu. Pathologic report of aortic biopsy from the punched portion of aorta for proximal anastomosis showed inflammation and arteritis.CT Angiography was performed before the

patient was discharged form hospital,showing multiple stenotic lesions in different parts of large blood vessels that distribute blood from aorta and our grafts looked to be patent (Fig.2) .

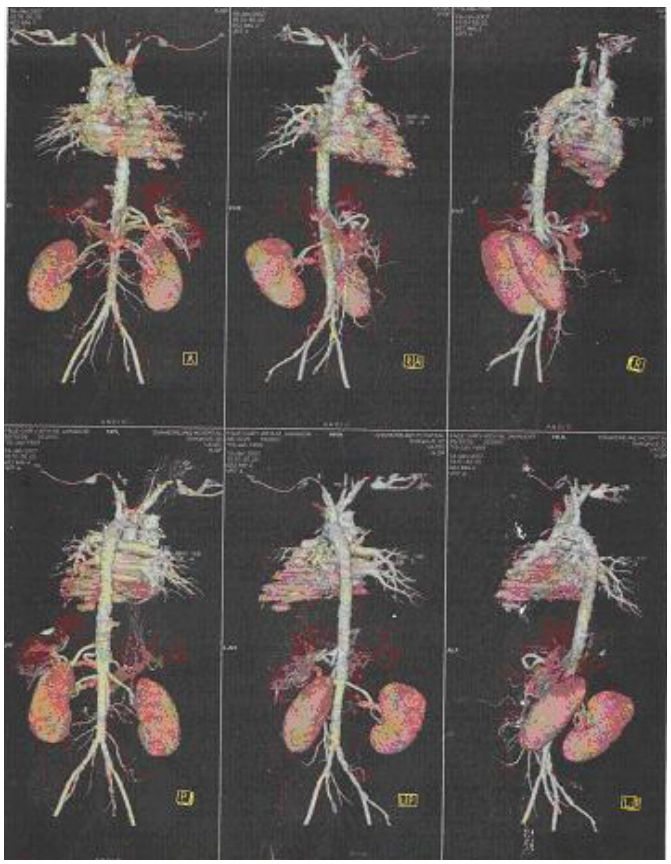

Fig.2.CT angiographic image of the patient at the time of discharge

Diagnosis:

Six criteria were selected for the traditional format classification: onset at age less than or equal to 40 years, claudication of an extremity, decreased brachial artery pulse, greater than 10 mm Hg difference in systolic blood pressure between arms, a bruit over the subclavian arteries or the aorta, and arteriographic evidence of narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities. The presence of 3 or more of these 6 criteria demonstrated a sensitivity of 90.5% and a specificity of 97.8%(5).From the above mentioned criteria, decreased brachial artery pulse, greater than 10 mm Hg difference in systolic blood pressure between arms, a bruit over the subclavian arteries or the aorta, and arteriographic evidence of narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities,were found in our patient. Arteriography is the standard criterion for assisting in making the diagnosis of TA. However, the use of MRA is increasing rapidly.Peripheral blood pressure monitoring frequently is inaccurate in persons with TA; pressure readings during angiography alone may reveal aortic root hypertension.Drawbacks to arteriography, including morbidity from use of contrast dye in patients with renal disease and cumulative radiation exposure over time, can be avoided by using MRA.Arteriography often demonstrates long, smooth, tapered narrowings or occlusions. Stenoses occur in 90-100% of patients with TA and aneurysm formation in only 27%. Three-dimensional MRA imaging of the aorta and its branches are providing exciting new data that may improve the understanding of the disease.Some authors recommend arteriography of the entire aorta.Magnetic resonance imaging, magnetic resonance angiography,computed tomography.These examinations are useful for serial examinations and diagnosis in the early phase of TA.CT scan and MRI may demonstrate mural thickening of the aorta and luminal narrowing. The delayed phase of the contrast-enhanced CT shows a double-ring configuration of the thick thoracic aortic wall, which is similar to CT findings reported for syphilitic arteritis(7). Use of contrast may reveal inflammatory lesions prior to the development of stenoses; these lesions may be missed by angiography. Aortic lesions including stenosis, dilatation, wall thickening, and mural thrombi are well visualized on MRI, which is less adequate in visualizing distal lesions of the subclavian vessels and common carotids.Noncontrast T2-weighted STIR images may be used to monitor edema in the aortic wall, which may be a surrogate for inflammation; edema was found in 94% of patients with clinically active disease. Both MRA and color Doppler flow imaging (6) may be used to evaluate and monitor disease in the common carotids and subclavian arteries; however, this imaging study is not useful in evaluating the aorta.Carotid evaluation reveals a homogenous circumferential thickening of the vessel wall that is distinguishable from atherosclerotic thickening. Chest radiographs may reveal widening of the ascending aorta, irregular descending aorta, aortic calcifications, and rib notching (late findings). Echocardiography is performed to evaluate the aortic valve and follow aortic insufficiency. From the above mentioned imaging studies

,angiography,echocardiography,Ct angiography and chest xray were performed and the results are presented above. Histologic findings are : Mononuclear infiltration of the adventitia with perivascular cuffing of the vasa vasorum may occur early in the disease. Granulomatous changes may be observed in the tunica media with Langerhans cells and central necrosis of elastic fibers and smooth muscle cells. Later, fibrosis of the media and acellular thickening of the intima may compromise the vessel lumen. Grossly, wrinkling of the intima is found. Nonspeciefic arteritis was the finding in the pathologic examination of the part we punched out of aortic root for construction of proximal anastomosis.Following the acute phase, patients with fibrotic changes require surgical treatment of symptomatic stenotic or occlusive disease. This can be achieved by percutaneous angioplasty or stenting or, in severe cases, by resection and placement of a manmade graft. Children with TA rarely require bypass surgery or carotid stenting(4). Our patient had a very narrow ,fibrotic and calcific aortic root,though at this time it seemd there was no necessity for any intervention. TA has no specific markers.Complete blood count reveals a normochromic normocytic anemia in 50% of patients with TA. Acute phase reactants are elevated, with leukocytosis and thrombocytosis. Westergren erythrocyte sedimentation rate is elevated. Comprehensive metabolic profile may indicate elevated transaminases and hypoalbuminemia.The von Willebrand factor–related antigen (factor VIII–related antigen) may be elevated.Antiendothelial antibodies are present. Antinuclear antibody usually is negative.Rheumatoid factor is elevated in 15% of individuals with TA.Increased levels of immunoglobulins G, M, and A are present(4). There were no abnormal finding in the laboratory tests of our patient to be worth of mentioning here.

Internationally,TA is a common affliction in third world countries, where the disease is associated closely with tuberculosis. The nature of this association is unclear because most patients with TA in the US do not have tuberculosis. In contrast, many third world physicians assume tuberculosis in every patient with TA(4).If we have such association in our country we don't know ,but after discharge we referred our patient to a specialist in infectious disease ,and nothing in favour of tuberculosis was found.TA more frequently is found in Asian populations but has been described in patients of all races. Japanese patients with TA have a higher incidence of aortic arch involvement. In contrast, series from India report higher incidences of thoracic and abdominal involvement. In US patients with TA, the most commonly involved vessels are the left subclavian, superior mesenteric, and abdominal aorta. In US children with TA, lesions of the thoracic and abdominal aorta, rather than lesions of the aortic arch, are found most commonly. However, all patterns of vascular involvement have been observed in every country(4). The most commonly involved vessels include the left subclavian artery (50%), left common carotid artery (20%), brachiocephalic trunk, renal arteries, celiac trunk, superior mesenteric artery, and pulmonary arteries (50%). Infrequently, the axillary, brachial, vertebral, coronary, and iliac arteries are involved. (3). In our patient ,aortic root and arch,and its major branches were involved but the thoracic and abdominal aorta were spared.

Takayasu arteritis is associated with a low incidence of coronary artery involvement(in 3-5% of patients)(16) such as stenosis, obstruction, aneurysm, and coronary steal syndrome, but coronary ischemia can be fatal, then, when indicated ,surgical treatment is recommended for patients with coronary ostial stenoses, because coronary ischemia can be one of the major causes of death (8).proximal parts of coronary arteries are usually involved(17).The timing preferred for surgical intervention is during an inactive phase. Two procedures are commonly chosen for surgical intervention, either transaortic endarterectomy or coronary revascularization with vein grafts. Postoperative steroid therapy is strongly recommended to those patients who are operated in the clinically or histologically active stage(9). Sometimes Because of Poor flow in bilateral internal thoracic arteries,they are not suitable for revascularization. the calcification of the ascending aortic wall may be too severe to be able to anastomose saphenous vein grafts to it. Therefore, the proper hepatic artery ,gastroepiploic artery or other donor arteries may be used for anastomosing saphenous vein grafts to them(10) . other than coronary artery disease that is a rare ocurrance in TA,other Cardiac involvements in Takayasu's disease are well documented and often the result of severe hypertension. However, severe clinical manifestations of aortic regurgitation and coronary insufficiency are much less common(11). Surgical treatment of aortic root and coronary artery lesions due to Takayasu arteritis has many potential difficulties due to its inflammatory nature and some times are a challenge for the surgeon and demand special considerations. Steroid

therapy is recommended in cases diagnosed as being in an active stage until the inflammatory signs disappear (12) (13). The use of off-pump coronary artery bypass grafting techniques should be considered in surgical coronary revascularization of patients with Takayasu's disease, thus leading to wide-spreading indication for the surgery(14). xenopericardial patch or aortic connector for anastomosis of saphenous vein grafts to aortic wall have been reported in literature(15).

Comment:

The problems a cardiac surgeon may encounter in confrontation with Takayasu's disease are:calcification of aortic root making on pump surgery impossible and finding a donor site for proximal anastomosis a challenge,lack of flow in internal thoracic arteries and other treatments necessary in collaboration with internists,vascular surgeons,interventional cardiologists,radiologists and other specialists . Wether takayasu is a prevalent disease in iran or not,we do not know.we presented this case in order to show the challenges and problems a surgeon may encounter in confrontation with unusual circumstances and cases,and the solutions and innovations that may help him in such circumstances.

References:

1. *William C. Shiel Jr., MD, FACP, FACR , Takayasu Disease (Takayasu Arteritis) ttp://www.medicinenet.com/takayasu_disease/article.htm*

2. *David Hellmannn, M.D. (F.A.C.P.), Types of vasculitis,takayasu's Arteritis , A discussion of Takayasu's Arteritis written in medical terms by David Hellmannn, M.D. (F.A.C.P.), The Johns Hopkins Vasculitis Center, for the Rheumatology Section of the Medical Knowledge Self-Assessment Program published and copyrighted by the American College of Physicians (Edition 11, 1998). http://vasculitis.med.jhu.edu/typesof/takayasu.html*

3. *Robert L Cirillo, Jr, MD, MBA, Arteritis, Takayasu,Last Updated: June 10, 2005 http://www.emedicine.com/radio/topic51.htm*

4. *Christine Hom, MD , Takayasu Arteritis,Last Updated: March 27, 2006 ,http://www.emedicine.com/ped/topic1956.htm*

5. *Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, Fauci AS, Leavitt RY, Lie JT,Lightfoot RW Jr, et al, The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. : Arthritis Rheum. 1990 Aug;33(8):1129-34.*

6. *Cantu C,Pineda C, Barinagarrementeria F, Salgado P, Gurza A, Paola de Pablo, Espinosa R, Martinez-Lavin Mr, et ,Noninvasive cerebrovascular assessment of Takayasu arteritis, : Stroke. 2000 Sep;31(9):2197-202.*

7. *Kimura F, Satoh H, Sakai F,Nishii N, Tohda J, Fujimura M, Haruta S, Yamazaki K, Endo M, Sakomura Y, Kurosama H, Kasanuki H,Computed tomographic findings of syphilitic aortitis. Cardiovasc Intervent Radiol. 2004 Mar-Apr;27(2):179-81.*

8. *Endo M, Tomizawa Y, Nishida H, Aomi S, Nakazawa M, Tsurumi Y, Kawana M, Kasanuki H, Angiographic findings and surgical treatments of coronary artery involvement in Takayasu arteritis. J Thorac Cardiovasc Surg. 2003 Mar;125(3):570-7*

9. *Amano J, Suzuki A,Coronary artery involvement in Takayasu's arteritis. Collective review and guideline for surgical treatment.J Thorac Cardiovasc Surg. 1991 Oct;102(4):554-60*

10. *Nishiyama A, Matsubara S, Toyama J. Takayasu arteritis with multiple cardiovascular complications. Heart Vessels. 2001 Dec;16(1):23-7*

11. *Favre JP ,[Morelon P,Obadia JF, David M. Heart valvular and coronary manifestations of Takayasu disease. Apropos of a surgically-treated case. Arch Mal Coeur Vaiss. 1992 Dec;85(12):1865-7*

12. *Amano J, Suzuki A. Surgical treatment of cardiac involvement in Takayasu arteritis. Heart Vessels Suppl. 1992;7:168-78*

13. *Suzuki A, Amano J, Tanaka H, Sakamoto T, Sunamori M. Surgical consideration of aortitis involving the aortic root.Circulation. 1989 Sep;80(3 Pt 1):I222-32*

14. *Yamaguchi A, Endo H, Adachi H, Kawahito K, Ino T.Off-pump coronary artery bypass in patients with Takayasu's disease. Ann Thorac Surg. 2004 Jun;77(6):2186-8*

15. *Tanaka M, Abe T, Takeuchi E,Watanabe T, Tamaki S. Revascularization for coronary ostial stenosis in Takayasu's disease with calcified aorta. Ann Thorac Surg. 1992 May;53(5):894-5*

16. *Hamdan A, Porter A, Georghiou GP, Mulad Y, Raanani E, Hasdai D, Battler A, Assali AR. Unusual presentation of takayasu arteritis. Int J Cardiol. 2006 Oct 23; Epub ahead of print*

17. *Matsumiya G, Ohtake S, Sawa Y, Ishizaka T, Yoshimine T, Matsuda H.Simultaneous repair of stenosis in coronary and vertebral arteries and aortic regurgitation secondary to Takayasu's aortitis.Jpn J Thorac Cardiovasc Surg. 2002 Feb;50(2):88-91*