Original Article

CT Evaluation of Takayasu Arteritis an Initial Experience at CPEIC, Multan

Takayasu Arteritis Among Teenagers & Adults

Maham Munir Awan[1], Muhammad Aftab Akbar[2], Muhammad Zubair Zaffar[3], Afshan Noreen[4], Syed Naseem Bukhari[3] and Faisel Yunus[5]

ABSTRACT

Objective: To identify the Takayasu Arteritis (TA) among teenagers & adults patients of South Punjab (Pakistan).
Study Design: Prospective Study
Place and Duration of Study: This study was conducted at the at radiology department of Chaudry Pervaiz Ellahi Institute of Cardiology, Multan (Pak) for a period of one year from April 2016 to April 2017.
Materials and Methods: All patients underwent color doppler ultrasound study for limb ischemia, reno vascular hypertension and carotid doppler ultrasound for stroke like symptoms followed by CT angiography using Toshiba Aquilon 128 slice CT scan and low-osmolar contrast media. Inclusion criteria to establish diagnosis was based on the American College of Rheumatology criteria. SPSS 20 was used to compile and consolidate the data findings.
Results: A total of 4012 patients were studied from them 56% were females and 44% were males, mean age of these patients were 26.34±5.12 years. Amongst them six patients were diagnosed with the disease (TA) from them(83.3%) patients were females and one was male (16.7%).All the six patients radiologically diagnosed as Takayasu Arteritis were further pathologically and clinically evaluated. Mean C-reactive protein (CRP) was 3.60±2.60mg/L while mean ESR 44.33±29.78mm/hr and mean wall thickness was 4.46±1.86mm.
Conclusion: CT Angiography is a very useful and reliable method of diagnosing Takayasu Arteritis, assessing disease activity and a guide to treatment & follow-up.
Key Words: 128 Slice CT Angiography, Computed Tomography Angiography, Takayasuarteritis, Pulseless Disease, South Punjab, CPEIC, Pakistan

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INTRODUCTION

Takayasu arteritis alternatively known as pulseless disease and aortic arch syndrome named after Dr. Mikito Takayasu in 1908; is an inflammatory and stenotic disease of medium and large sized arteries characterized by a strong predilection for the aorta and its major branches[1]. Vessels of arms, kidneys and arteries to the brain are exposed to frequent effects whilst coronary and pulmonary arteries retain less frequent effect.

[1.] Department of Radiology / Medicine[2], Nishtar Medical University, Multan.
[3.] Department of Cardiology, Chaudry Pervaiz Ellahi Institute of Cardiology, Multan.
[4.] Department of Radiology, Children Hospital & Institute of child health (CHC), Multan.
[5.] Department of Director & Consultant Public Health Medicine, Luton, UK.

Correspondence: Dr Maham Munir Awan, Assistant Professor Radiology. Nishtar Medical University Multan.
Cell No.: 03317276872
Email: maham.amj@hotmail.com

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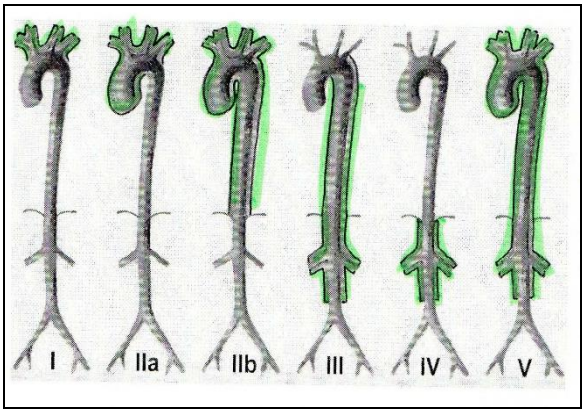
Although causation of Takayasu Arteritis is yet to be discovered but it is assumed to be an auto-immune disorder where immune defense instead of protecting body cells start attacking them. Extensive research has revealed prevalence of Takayasu Arteritis in Asia among teenagers and young adults resulting in consistent high blood pressure[2]. On the contrary, it is very rare in North America and Europe[3]. Emergence of Takayasu Arteritis is commonly experienced among women below the age bar of 40 and in contrast of emergence with males; the ratio of one male to nine females of thousand patients is collectively witnessed across the globe[4,5]. Distribution of lesion in the aorta is localized in 37.5% with abdominal aorta involvement in adults and thoracic + abdominal involvement in children. Diffuse involvement occurs in 62.5% withthoraco-abdominalpredilection[6].

Descending thoracic Aorta is maximally affected area. Aortic arch has more distal involvement than proximal. Patients present with constitutional symptoms of Headache (50%-70%), Malaise (35%-65%), Arthralgias (28%-75%), Fever (9%-35%) & Weight loss (10%-18%)[7]. Pan-arteritis with marked intimal proliferation, fibrosis, scarring and vascularization of media, disruption, degeneration of elastic lamina, and narrowing of lumen with or without thrombus formation resulting in segmental stenosis or aneurysm. Complications occur mainly due to arterial occlusion

leading to limb ischemia, renal failure, cardiac ischemia or stroke[8].

Types of TA

Type I	Branches of the aortic arch(Classic Type)
Type IIa	Ascending aorta, aortic arch, and its branches
Type IIb	Type IIa region plus thoracic descending aorta
Type III	Thoracic descending aorta, abdominal aorta, renal arteries, or a combination
Type IV	Abdominal aorta, renal arteries, or both
Type V	Entire aorta and its branches



Sharma Criteria for Diagnosis of TA[8]	
Major criteria	• Left mid-subclavian artery lesion • Right mid-subclavian artery lesion • Characteristic signs and symptoms of at least one-month duration
Minor criteria	• High ESR • Carotid artery tenderness • Hypertension • Aortic regurgitation or annuloaorticectasis • Pulmonary artery lesion

Takayasu Arteritis is a very rare disease and sofar, no specific study regarding its prevalence in South Punjab region of Pakistan has ever been conducted. Chaudhry Pervaiz Elahi Institute of Cardiology, Multan (CPEIC) being the hub of vascular diseases receives a large number ofyoung patients with limb ischemia, high blood pressure and cardiac issues which encouraged us to probe and found TA as a rare cause. In this study selected patients with prevalence of TA suspected on color doppler ultrasound study were further evaluated with CT angiography using Toshiba Aquilon 128 slice CT scan[10]. This study is scheduled as research methods, lab &radiological investigations, imaging findings, discussion and finally conclusion will be stated at the end of this paper.

MATERIALS AND METHODS

This paper is a prospective study and the study population consisted of 4012 patients who were referred to our Radiology department of Chaudhry Pervaiz Elahi Institute of Cardiology (CPEIC), Multan between April, 2016 and April, 2017. All patients underwent color doppler ultrasound study for limb ischemia, reno vascular hypertension and carotid doppler ultrasound for stroke like symptoms followed by CT angiography using Toshiba Aquilon 128 slice CT scan and low-osmolar contrast media. MIP, MPR and 3Dreconstruction of images data done with slice thickness 0.5mm including curve planar reformation (CPR) to display tortuous vessels along its long axis, multiplanar reconstruction (MPR) for anatomical information of vessels, volume rendered images to see the extension of luminal lesions and following the collaterals after arterial occlusion. Bolus of contrast was given at the rate of 4 to 6ml per second with automated injector, total volume of contrast was 80 to 100ml and patients were scanned in 15 to 20 seconds using breath hold technique.

Inclusion criteria to establish diagnosis was based on The American College of Rheumatology criteria. Referred patients from Cardiology Department between ages 15 to 40 years and either sex, with raised CRP and ESR levels, intermittent claudication, blood pressure difference of more than 10mmhg in both arms, decreased brachial artery pulse, reno vascular hypertension, bruit over subclavian, carotids or aorta, and shortness of breath or chest discomfort[11].

Exclusion criteria includes patients with S. creatinine more than 1.5mg/dl, history of iodine allergy, pregnancy and those who refused for consent. SPSS 20 was used to compile and consolidate the data findings.

RESULTS

Amongst the total 4012 patients referred to our Radiology department, 56% of the patients (2,247) were females whereas 44% of the patients (1,765) were males. Based on the inclusion criteria between ages 15 to 40;mean age of these patients 26.34$\pm$5.12 while 69 percent of the patients (2,768)were aged from 15 to 30 years whereas 31% of the patients (1,244) were from ages 31 to 40 years.

Imaging Findings: All the patients reported with Takayasu Arteritis were young with age ranging from 18 to 36 years. Mean age of these patients was 25.33$\pm$6.02 years (ranges from 18 to 36 years)Five out of six (83.3%) patients were females and one was male (16.7%). All patients showed the presence of stenosis involving different primary branch vessels. The degree of stenosis also varied from patient to patient. Stenosis being the hallmark of Takayasu Arteritis and its presence among all patients encouraged further evaluation. Three patients (50%) showed hyper-enhancement of vessel wall in delayed arterial phase representing active phase of the disease process.

Table No.1: Clinical and Radiologic Data on Patients with Takayasu's Arteritis

Clinical and Radiologic Data on Patients with Takayasu's Arteritis							
Patient	Age (yrs)	Sex	Stenosis	CRP (mg/L)	ESR (mm/hr)	MWT (mm)	Delayed Hyperenhancement
A	22	F	Present	5.0	100	4.5	Present
B	18	F	Present	0.3	20	2.3	Absent
C	26	F	Present	5.5	40	4.5	Present
D	36	M	Present	0.3	18	2.5	Absent
E	24	F	Present	6.0	48	7.0	Present
F	26	F	Present	4.5	40	6.0	Absent

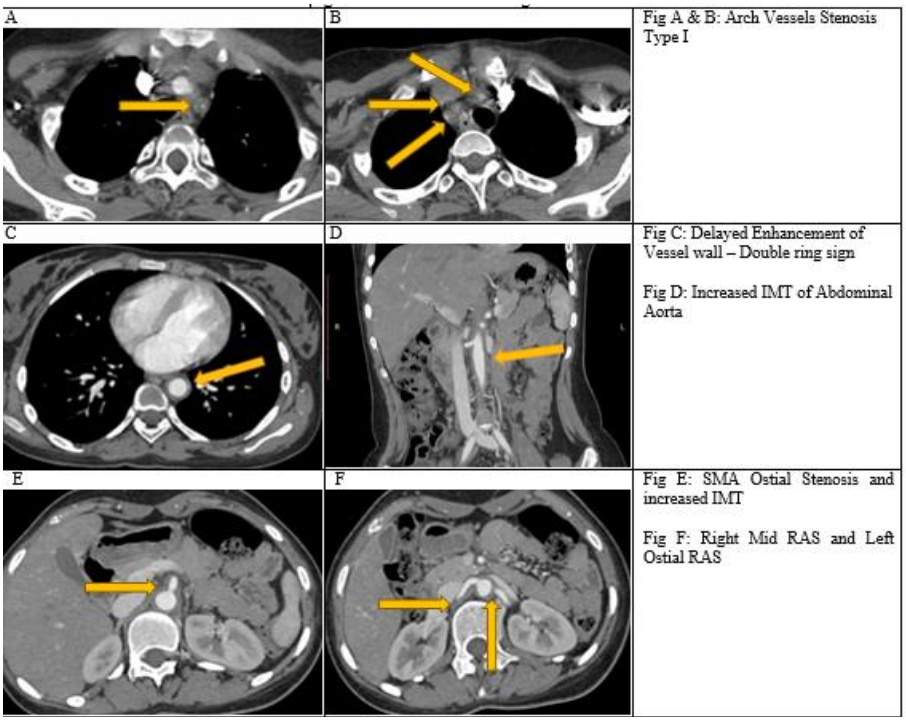


Figure No.1: CTA Pictorial Findings of Patient A

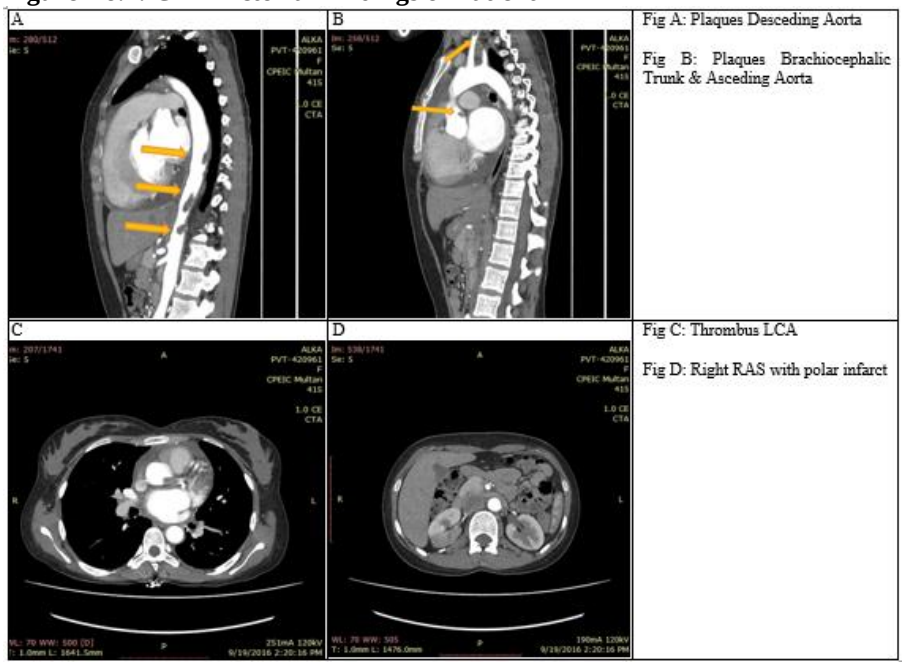


Figure No.2: CTA Pictorial Findings of Patient B

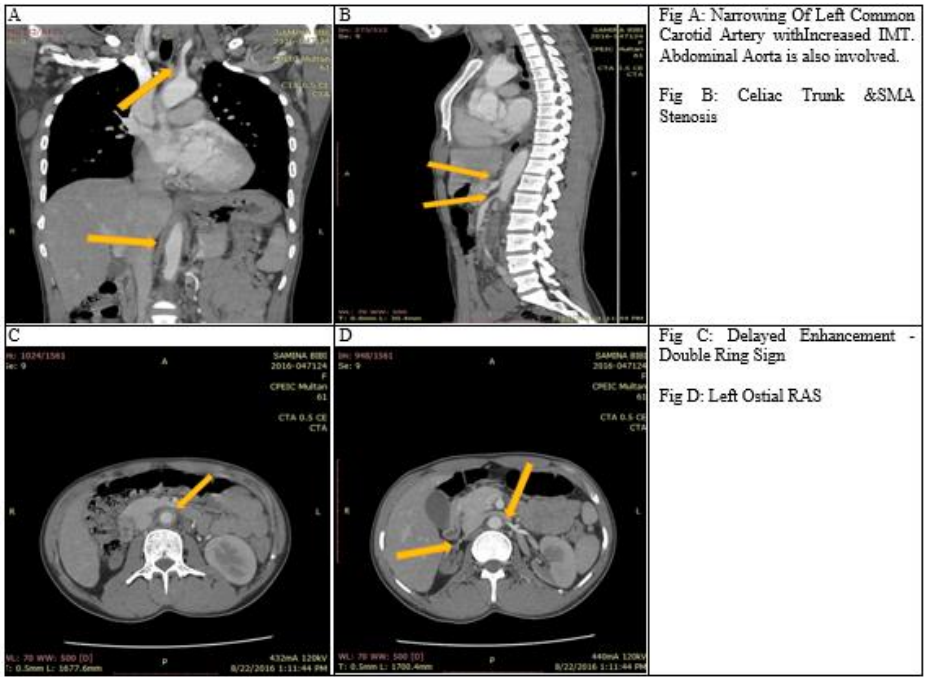


Fig A: Narrowing Of Left Common Carotid Artery withIncreased IMT. Abdominal Aorta is also involved.

Fig B: Celiac Trunk &SMA Stenosis

Fig C: Delayed Enhancement - Double Ring Sign

Fig D: Left Ostial RAS

Figure No.3: : CTA Pictorial Findings of Patient C

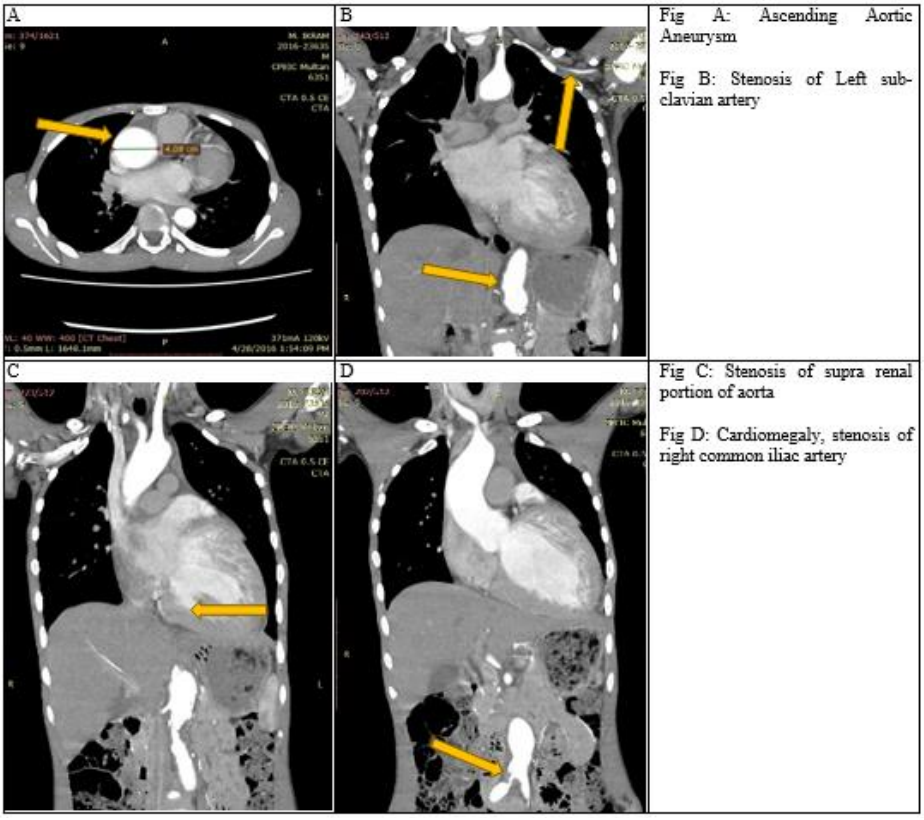


Fig A: Ascending Aortic Aneurysm

Fig B: Stenosis of Left sub-clavian artery

Fig C: Stenosis of supra renal portion of aorta

Fig D: Cardiomegaly, stenosis of right common iliac artery

Figure No.4: CTA Pictorial Findings of Patient D

All the six patients radiologically diagnosed as Takayasu Arteritis were further pathologically and clinically evaluated. Mean C-reactive protein (CRP) was 3.60$\pm$2.60mg/L while mean ESR 44.33$\pm$29.78 mm/hr and mean wall thickness was 4.46$\pm$1.86mm. It was found that those patients which showed delayed hyper-enhancements of the vessel walls also had increased C-reactive proteins levels. ESR was also found to be higher in patients with delayed hyper-enhancement of vessel walls (Figure 1-4). This showed that three patients were going through active phase of disease process.

DISCUSSION

Takaysau Arteritis also known as aorto-arteritis is a type of vasculitis of unknown etiology of the vessel walls. It has higher incidents in young females and more common among Asians. Takayasu Arteritis is pan-arteritis, inflammatory mononuclear infiltrates with intimal proliferation, fibrosis, scarring and vasculization of media. This study describes our initial experience of diagnosing Takayasu Arteritis and defining its types according to involvement of major aortic branches using CT angiography protocol in delayed arterial phase[12]. This method showed arterial wall hyper-enhancement which represented active phase of the disease.

Although all the six patients had characteristic stenosis. Three of them showed delayed hyper-enhancement of the vessel wall with slightly increased mean wall thickness none more than 5.0mm[13-15].

Patient A, young female aged 22 presented with complaints of hyper tension and bilateral cold upper extremities, showed segmental irregular stenosis/ occlusion of major aortic arch vessels at their origin including right brachio-cephalic trunk, left common carotid and left sub-clavian artery. Diffused circumferential wall thickening and delayed vessel wall hyper-enhancement of thoracic[16] and abdominal descending aorta up to iliac bifurcation with involvement of SMA and right renal artery. She was diagnosed as Takayasu Arteritis type I classic with type III in acute active phase. ESR was reported at 100mm at first hour with CRP at 5.0 mg/L.

Patient B, young unmarried female aged 18 years presented with constitutional symptoms on examination; patient had hypertension. She was referred to our department for ultrasound abdomen and renal artery doppler which showed multiple thrombi along the posterior wall of abdominal aorta adjacent to the diaphragm, parvustardus pattern in both intra renal arteries and wedge shaped lower polar hypoechoic area with no vascular supply seen in right kidney. Patient was advised for abdominothoracic CT angiography which showed left ostial renal artery stenosis with collaterals formation. Right renal artery stenosis with lower polar infarct. Thrombus visualized in left main coronary artery with left ventricular hypertrophy and thrombus in LV apex. Multiple plaques and thrombi were observed in aortic root, descending thoracic and upper abdominal aorta and braciocephalic trunk[14-16].

Patient C was a young female aged 26, presented with light headedness, hyper tension and pain abdomen. She was referred for carotid Doppler which showed significantly reduced Doppler flow in left common carotid artery and renal artery Doppler was also done which showed parvustardus pattern of flow in bilateral intra renal arteries. Right kidney was also smaller in size. CTA findings reported as narrowing of left common carotid artery at its origin with narrow caliber throughout its course in neck. Intima and media thickening and wall irregularity with delayed hyper-enhancement giving a double ring sign were seen in the abdominal aorta. Significant narrowing from the origin of right renal artery throughout its course up to right kidney with a streak of contrast passing through it. Right kidney was smaller in size with a cranio-caudal length of 4.9cm. Ostial narrowing seen in left renal artery with normally outlined course and caliber of distal vessel. Multiple collateral channels were seen extending from left external iliac vessel supplying the mid portion of left renal artery. Left kidney was normal in size. Partial stenosis of major abdominal aortic branches involving origins of celiac trunk superior mesenteric arteries. The origin of inferior mesenteric artery also showed significant narrowing with multiple collateral channels arising from lumbar arteries. Patient was diagnosed Takayasu arteritis type IV with involvement of left common carotid artery (CCA)[14, 16].

Patient D, middle aged male at 36 years presented with numbness of both arms. Doppler study of both upper limbs showed parvustardus pattern of flow in both limbs arterial systems up to distal sub-clavian arteries. CT aortogram was suggested to rule out proximal disease. CTA of Abdomino Thoracic aorta showed short segments of significant narrowing identified in both proximal sub-clavian arteries. Area of significant stenosis identified in proximal abdominal aorta with post stenotic fusiform aneurysm involving suprarenal part of distal aorta. The segment of narrowing is suprarenal below the origin of SMA. Aneurysmal dilatation of ascending aorta also seen. Short segment stenosis is also noted in proximal right common iliac artery. Cardiac size was also enlarged[7, 15-16].

Patient E, young female aged 24 years presented with hypertension, numbness/ pain in right arm. Patient was referred to us for abdominal and carotid doppler. Right kidney was small echogenic with right renal artery stenosis. Carotid doppler showed dilated right common carotid artery, bilateral increase intimal medial thickening of 22mm on right and 16mm on left side. Patient was advised for abdominothoracic CT aortogram which showed increased mean wall thickness in arch of aorta (7mm), descending thoracic and abdominal aorta with calcification. Bilateral common carotid arteries also showed increased mean wall thickness. Focal stenosis at the origin of left ECA with multiple collaterals visualized filling distill portion of ECA and its branches. Right subclavian artery showed focal stenosis more than 70% just after crossing the first rib. However, contrast was trickling through it. Left subclavian artery is stenosed after origin of costo-cervical trunk with multiple collaterals recons truing blood in axillary artery. The length of stenos segment was 6.2cm.Left CCA had narrow caliber. MWT was

also increased in SMA and celiac trunk at their origin. Narrow right renal artery with small sized right kidney.

Patient F, young female aged 26 years was diagnosed earlier as a case of Takayasu Arteritis. She was hypertensive at presentation and was taking treatment. CT Aortogram was carried out which showed aneurysmal dilatation seem involving ascending aorta, lower portion of descending thoracic aorta which measured 3.2cm in sagittal section, IMT was increased involving major arch vessels, descending thoracic aorta, abdominal aorta, and origin of celiac trunk and SMA causing their narrowing & significant stenosis at celiac trunk with MWT of 6.0mm in descending aorta[15-16]. Main pulmonary trunk was also involved with increased IMT and narrowing of right and left pulmonary arteries measuring 6.4mm and 10mm respectively[14]. Stenosis seen at the origin of right renal artery**.**

CONCLUSION

CTA is very useful and reliable method of diagnosing TA and assessing disease activity and a guide to treatment/follow-up. Findings of the procedure include vascular stenosis, occlusion, aneurysm as well as mural wall thickenings and plaques formation. Delayed contrast enhancement of the vessel wall giving a double ring sign correlated with raised serum markers suggesting active disease. In our view all patients with suspected vascular stenosis should undergo MSCTA as a standard diagnostic workup protocol in furnishing the diagnosis of TA.

Author's Contribution:

Concept & Design of Study:	Maham Munir Awan
Drafting:	Muhammad Aftab Akbar, Muhammad Zubair Zaffar
Data Analysis:	Afshan Noreen, Syed Naseem Bukhari and Faisel Yunus
Revisiting Critically:	Maham Munir Awan, Muhammad Aftab Akbar
Final Approval of version:	Maham Munir Awan

Conflict of Interest: The study has no conflict of interest to declare by any author.

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