CASE REPORT

UNUSUAL PRESENTATION OF A CASE OF TAKAYASU ARTERITIS

Adnan Shabbir, Adil Mohyudin, Khushbakht Javeria Rao*, Ghias Un Nabi Tayyab, Abu Fahad Abbasi*****

Department of Medicine and Gastroenterology, *Department of Radiology, Lahore General Hospital, Lahore, **Sheikh Zayed Medical College, Rahim Yar Khan-Pakistan, ***Saint James School of Medicine, Anguilla, BWI-United States

Takayasu arteritis is a rare disease of medium size and large vessels causing granulomatous inflammation of the involved territory. The most usual area involved is the arch of the aorta and its major branches. We came across an atypical presentation of this rare disease in our medicine department at Lahore General Hospital. A 52-year-old male presented with complaints of vertigo, bilateral upper limb cramps on physical activity. On examination, pulses were absent bilaterally in upper limbs. On CT angiography brachiocephalic and left subclavian arteries were not visualized. Inflammatory markers were not raised to the extent indicating a very aggressive disease. The patient was not meeting the age limit as described by Ishikawa diagnostic criteria. The patient was referred to the cardiovascular surgery department for revascularization.

Keywords: Takayasu arteritis; Ishikawa criteria; Aortic arch syndrome

Citation: Shabbir A, Mohyudin A, Rao KJ, Tayyab GN, Abbasi AF. Unusual presentation of a case of Takayasu arteritis. J Ayub Med Coll Abbottabad 2019;31(4 Suppl 1):683–5.

INTRODUCTION

Takayasu arteritis is a rare disease of medium size and large vessels causing granulomatous inflammation of the involved territory.[1] It has a strong predilection for the arch of the aorta and its major branches. When it comes to the epidemiology of disease 80–90 percent affectees are women and the usual age of onset in between 10 and 40 years of age.[2,3] The disease is more common in Asia but the geographical or racial restriction is observed.[4–6]

The pathogenesis of Takayasu arteritis (TAK) is not well understood. The proposed mechanism signifies the role of cell-mediated immunity as seen in giant cell arteritis (GCA).[7] Analysis of aortic tissue by Immuno-histopathological means shows that primary cells involved are cytotoxic cells especially gamma delta T lymphocytes.[8] Vascular injury is thought to be a consequence of perforin (cytolytic protein) released by these cells. MRA and CTA of patients with Takayasu arteritis mostly show occlusion of the vessels with luminal narrowing and tapering. Sometimes vessel wall thickening is also observed.[9–14]

Researchers and clinicians use ACR (The American College of Rheumatology) and Ishikawa criteria for diagnosing Takayasu arteritis. But the perfect diagnostic criteria is still a future goal.

CASE REPORT

Fifty-two years old male presented with complaints of vertigo for the last 6 months and cramps in arms on prolonged manual work. He was a farmer by profession. He has no history of smoking, diabetes and any chronic illness. There is no history of fever, malaise, joint pain, rash or other neurological complaints except vertigo. The patient didn't have complains of shortness of breath, chest pain, palpitations or any previous vascular event. There was no family history of ischemic heart disease or stroke. On physical examination, he had absent peripheral arterial pulses in both upper limbs. Blood pressure was not recordable by both auscultatory and palpatory methods. No murmur was found on cardiac auscultation. Pulses were feeble in lower limbs. On the basis of history and clinical examination, our provisional diagnosis was Takayasu arteritis. We proceeded with the second basic component of diagnosis, radiological evidence. Brachiocephalic trunk and left subclavian artery were not visualized on CT aortogram. Left common carotid artery showed narrowing at origin (ostial narrowing). Figure (1a) and (1b).

MR angiogram of the brain showed no flow in the right carotid artery; however, right anterior and middle cerebral arteries were normal and supplied by the circle of Willis. Left anterior, middle and posterior cerebral arteries were normal. Vertebral, basilar, anterior communicating, posterior communicating and left common carotid artery were normal.

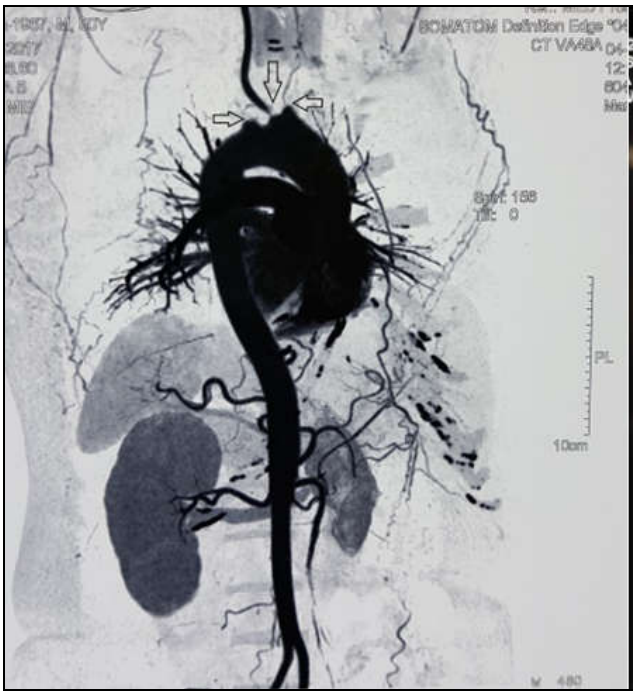

Figure (1a) CT aortogram

Figure (1b) CT aortogram

On laboratory reports, markers of inflammation were found to be normal. ESR was borderline raised 18mm/hr and CRP was normal. But inflammatory markers did not correlate well with disease activity. Fasting lipid profile, chest x-ray and echocardiography showed no abnormality. This case was unusual in many regards. According to Ishikawa criteria, age must be less than 40 years for the diagnosis of Takayasu arteritis.[1] But our patient was 52 years old at presentation. The disease was insidious in onset without any constitutional symptoms. It was very unusual to have such an extensive disease with involvement of major branches of the aorta but with very late presentation. Our patient had developed a good amount of collateral circulation to compensate for arterial insufficiency. Now extensive disease resulted in intermittent claudication in upper limbs and intracranial arterial insufficiency resulted in vertigo on walking and standing up.

DISCUSSSION

Takayasu arteritis is the large vessel vasculitis causing granulomatous inflammation of the involved territory and is less commonly experienced in clinical practice. Mainly it targets the arch of the aorta and its major branches but other territories can also be affected.[1,2] India, Japan, Southeast Asia, and Mexico contribute to the major disease burden. Annual incidence is reported to be 2.6 per million people in North America.[6] No epidemiological data is available for the Pakistani population. There is no specific test to diagnose Takayasu arteritis. Historically Ishikawa criteria was used to diagnose Takayasu arteritis. According to this criteria, it was mandatory that the age of the patient must be below 40 years.[6] In our case, the age of the patient was 52 years which was way above the age limitation set by Ishikawa. Age below 40 years at diagnosis has also been proposed in the ACR (The American College of Rheumatology) criteria but is not mandatory.[15] Our case is a rare presentation of Takayasu arteritis at this extreme of age which is unusual for both diagnostic criteria but still fulfils ACR diagnostic approach. But the perfect diagnostic criteria is still a future goal. As the pathophysiology of different types of vasculitis is a subject of extensive research, and modern laboratory investigations are at disposal, EULAR (European League against Rheumatism) and ACR are striving to reach an international consensus regarding accurate diagnostic and classification criteria.[15,16] Due to the extensive involvement of vessels, our patient needed extensive revascularization for the brachiocephalic trunk and left subclavian arteries. He refused to undergo extensive life-risking surgery and chose to live with symptoms. The patient was discharged on steroid on standard dose.

CONCLUSION

Takayasu arteritis can rarely present above 50 years of age which is unusual for both diagnostic criteria. We need to review our diagnostic approach towards Takayasu arteritis. EULAR and ACR are striving to

reach an international consensus regarding accurate diagnostic and classification criteria.

REFERENCES

1. Ishikawa K. Diagnostic approach and proposed criteria for the clinical diagnosis of Takayasu's arteriopathy. J Am Coll Cardiol 1988;12(4):964–72.
2. Lupi-Herrera E, Sánchez-Torres G, Marcushamer J, Mispireta J, Horwitz S, Vela JE. Takayasu's arteritis. Clinical study of 107 cases. Am Heart J 1977;93(1):94–103.
3. Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, *et al*. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 1990;33(8):1129–34.
4. Dabague J, Reyes PA. Takayasu arteritis in Mexico: A 38-year clinical perspective through literature review. Int J Cardiol 1996;54(Suppl):S103–9.
5. Hall S, Barr W, Lie JT, Stanson AW, Kazmier FJ, Hunder GG, *et al*. Takayasu arteritis. A study of 32 North American patients. Medicine (Baltimore) 1985;64(2):89–99.
6. Ishikawa K. Natural history and classification of occlusive thromboaortopathy (Takayasu's disease). Circulation 1978;57(1):27–35.
7. Weyand CM, Goronzy JJ. Medium- and large-vessel vasculitis. N Engl J Med 2003;349(2):160–9.
8. Seko Y, Minota S, Kawasaki A, Shinkai Y, Maeda K, Yagita H, *et al*. Perforin-secreting killer cell infiltration and expression of a 65-kD heat-shock protein in aortic tissue of patients with Takayasu's arteritis. J Clin Invest 1994;93(2):750–8.
9. Hata A, Numano F. Magnetic resonance imaging of vascular changes in Takayasu arteritis. Int J Cardiol 1995;52(1):45–52.
10. Yamada I, Numano F, Suzuki S. Takayasu arteritis: evaluation with MR imaging. Radiology 1993;188(1):89–94.
11. Yamada I, Nakagawa T, Himeno Y, Numano F, Shibuya H. Takayasu arteritis: evaluation of the thoracic aorta with CT angiography. Radiology 1998;209(1):103–9.
12. Paul JF, Hernigou A, Lefebvre C, Bletry O, Piette JC, Gaux JC, *et al*. Electron beam CT features of the pulmonary artery in Takayasu's arteritis. AJR Am J Roentgenol 1999;173(1):89–93.
13. Kissin EY, Merkel PA. Diagnostic imaging in Takayasu arteritis. Curr Opin Rheumatol 2004;16(1):31–7.
14. Keenan NG, Mason JC, Maceira A, Assomull R, O'Hanlon R, Chan C, *et al*. Integrated cardiac and vascular assessment in Takayasu arteritis by cardiovascular magnetic resonance. Arthritis Rheum 2009;60(11):3501–9.
15. Craven A, Robson J, Ponte C, Grayson PC, Suppiah R, Judge A, *et al*. ACR/EULAR-endorsed study to develop Diagnostic and Classification Criteria for Vasculitis (DCVAS). Clin Exp Nephrol 2013;17(5):619–21.
16. Luqmani RA, Suppiah R, Grayson PC, Merkel PA, Watts R. Nomenclature and classification of vasculitis - update on the ACR/EULAR diagnosis and classification of vasculitis study (DCVAS). Clin Exp Immunol 2011;164(Suppl 1):11–3.

Submitted: 12 July, 2019	*Revised: 4 October, 2019*	*Accepted: 4 October, 2019*

Address for Correspondence:

Adil Mohyudin, House No. 498/499 Block C, Gulberg Colony, Kasur 55050.
Cell: +92 322 658 6009
Email: adilmohyudin@gmail.com