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Case Report

Takayasu Arteritis Presenting with Hypertensive Urgency in a Young Male

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A B S T R A C T

Here we present a 31-year-old male with a one-month history of persistent generalized headache and significantly elevated blood pressure (220/130 mmHg). Initial evaluation revealed no specific symptoms indicative of a hypertensive emergency, prompting further investigation. Imaging studies, including 2D echocardiography and MRI aortogram, revealed hypertensive changes with left ventricular hypertrophy and diastolic dysfunction, as well as complete stenosis of the right renal artery and mild stenosis of other arterial territories respectively. Subsequent clinical assessment supported a diagnosis of Takayasu arteritis (TAK) based on the 2022 ACR/EULAR classification criteria despite normal inflammatory markers.

Treatment involved a multidisciplinary approach, including systemic glucocorticoids, methotrexate, and antihypertensive medications, resulting in blood pressure control and specific disease management.

Keywords: Hypertensive urgency; Treatment, Young hypertension, Large-vessel vasculitis, Takayasu arteritis

Introduction

Takayasu arteritis (TAK) is a rare large-vessel vasculitis primarily affecting the aorta and its branches, commonly presenting with constitutional symptoms and vascular damage manifestations[1]. Diagnosis relies on clinical findings and imaging criteria, with treatment involving systemic glucocorticoids and immunosuppressive agents to control inflammation and prevent complications. Despite advancements in diagnosis and management, challenges persist due to the disease's chronic and relapsing nature, emphasizing the need for lifelong follow-up and individualized treatment strategies.

Case Presentation

A 31-year-old male presented with a history of persistent generalized headache of one month duration. He did not have early morning headaches, nausea vomiting or any aura preceding the headache. Following that he consulted his GP and found out that his BP was 220/130mmHg. He was conscious and otherwise well. No symptoms such as chest pain, shortness of breath and hematuria to suggest a hypertensive emergency at the time of the presentation. But it was suggested that he should undergo extensive investigations to find the underlying cause for his high BP and headache. He consumes alcohol occasionally and does not smoke. Denies of consuming any recreational drugs. No history of recent weight loss. There were no signs suggesting heat intolerance, proximal muscle weakness. He is also not a known diabetic. No episodic palpitations or anxiety. He does not snore while sleeping. No history of renal stones or frothy urine.

He also did not have any history of oral and genital ulcers. He denied any painful joint swelling, or skin rashes.

On examination, He is an average built man with a BMI of 20.5. He was not pale. No signs of peripheral or periorbital edema. He did not have rash or swelling of joints. His PR is 80bpm with good volume. All peripheral pulses were felt but the left radial and brachial pulse was weaker than the right side, the rest of the peripheral pulses were normal. No femoral or radio-radio delay. His Blood pressure at the time of examination on the right side was 230/120 mmHg and left side 210/110, no significant difference noted in the lower limbs. His apex beat was felt at 5th ICS and not heaving in nature. Auscultation was normal. There was a bruit heard over the right renal artery, no bruit heard over carotid and subclavian artery. The rest of the examination including musculoskeletal examination was normal. There were no hypertensive changes seen on fundoscopy.

Based on the investigation (**Table 1**) a clinical diagnosis of Takayasu arteritis was made based on 2022 ACR/EULAR criteria. His inflammatory markers were normal. Immunological markers including antinuclear antibody (ANA), Double stranded DNA (DsDNA), antineutrophil cytoplasmic antibodies(ANCA) were all negative. His MRI aortogram showed complete stenosis of the right renal artery, moderate stenosis of left proximal subclavian artery and stenosis of proximal part of superior mesenteric artery. He was initially admitted and managed as hypertensive emergency and his Blood pressure was controlled with Cilnidipine 10mg twice daily, Oral prazosin 1mg three times a day, losartan 50mg twice a day and oral spironolactone 50mg daily. Then he was pulsed with IV methylprednisolone 1 g for three days and discharged on 1mg/kg/body weight oral prednisolone with a plan to slowly taper it down and started on Oral Methotrexate 10mg on Sundays with folic acid 5mg on Wednesdays.

Table 1: Investigations.

Investigations		Values	Reference range
Full blood count	White cell count (*10^3)	6.38	4–11
	Neutrophils (%)	66	50–70
	Lymphocytes (%)	35	20–40
	Eosinophils (%)	4	
	Hemoglobin (g/dL)	14	12–14
	Platelets (*10^3/µL)	161	150–400
InflammatoryMarkers	Erythrocyte sedimentation rate (mm in 1st hour)	5	<20
Serum electrolytes	Serum sodium (mmol/L)	133	135–145
	Serum potassium (mmol/L)	4	3.5–5.5
Renal function tests	Serum creatinine (µmol/L)	1.34	0.9-1.3
	Serum Urea	25	15-45
	EGFR	70.09	>90
24-hour urine for metanephrines		143.52	<350
Aldosterone: Renin Ratio		1.25	<30
Micro-albumin in spot urine	Albumin/creatinine ratio	254	<30
Antineutrophilic cytoplasmic antibody (ANCA)		Negative	
Anti-Double strandedDNA antibodies(DSDNA)		Negative	
Anti-Nuclear Antibody (ANA)		Negative	
Thyroid profile	Serum TSH (µIU/mL)	3.679	0.35–5.5
Mri Aortogram and Renal artery		Evidence of complete stenosis of right renal artery,Moderate narrowing at the subclavian artery. Proximal part of superior mesentericartery is also narrowed. No evidence of aneurysms.	

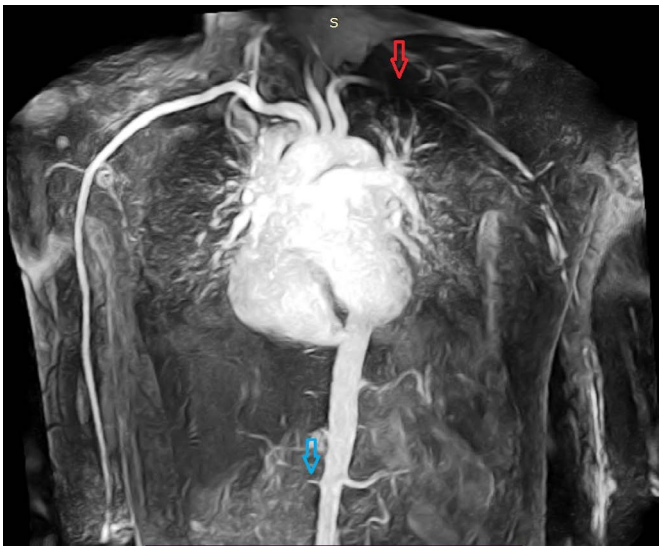

Figure 1. Aortogram

Red Arrow - showing moderate narrowing of left subclavian artery.

Blue arrow-Showing complete narrowing of right renal artery

Discussion

Takayasu arteritis is classed as a large-vessel vasculitis affecting mainly aorta and its main branches[1]. It has a similar histological picture with giant cell arteritis[2]. Latter predominantly involves only large arteries, involvement of renal arteries is rare and it mainly affects the elderly population. The main presentation is nonspecificconstitutional symptoms and symptoms evolve once significant damage is caused by vascular damage thus a greater suspicious is required to make diagnosis early[2]. It has a high prevalence in Japan and Asian population with majority being females in their thirties[3,4].

The pathogenesis of Takayasu arteritis (TAK) is poorly understood. The main proposed mechanism is cell

mediated damage to the blood vessel. Immunohistopathologic examination is characterized by the evidence infiltration of cytotoxic lymphocytes, especially gamma delta T lymphocytes causing granulomatousinflammation[5]. Involvement may be localized to a portion of a large vessel or may involve the entire vessel. There is a great variability in disease expression which causes wide range of clinical presentation[6], There is a prediction to involve mainly proximal or middle subclavianartery, carotid artery and vertebral arteries[7]. The onset has a very insidious and may sometimes be asymptomatic for couple of years. Young hypertension is a common presentation involving the renal artery, but diagnosis sometimes is delayed unless there is a huge suspicion.

So Constitutional symptoms like weight loss, low grade fever are common in the early phase of Takayasu arteritis and should never be overlooked[8]. Arthralgia, tenderness over the carotid artery, limb claudication, features of ischemic type chest pain due to coronary artery ostium association are also common presentations[8]. Involvement of carotid and vertebral arteries can lead to features of reduced blood supply and present with lightheadedness, vertigo, syncope and rarely seizures or strokes[9]. In a very minority the presentation may be dermatological manifestations like erythematosus or pyoderma gangrenosum.

Targetted examination is crucial including the palpation of all major arteries and Blood pressue. Measurement of blood pressure should be done in all 4 limbs. Most common involvement is the sublclavian artery Obstruction giving rise to erroneously low-pressure readings in the ipsilateral arm. Bruits should be listened for over the bilateral carotid, subclavian, axillary, renal artery[10]. A clinical diagnosis of Takayasu arteritis (TAK) can be made in a patient with both indicative of clinical findings and imaging showing narrowing of the aorta and/or its primary branches. The 2022 ACR/European Alliance of Associations for Rheumatology (EULAR) classification criteria for TAK (**Table 2**) use a weighted algorithm that includes clinical and imaging criteria[11].

Table 2: 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology/European Alliance for Rheumatology classification criteria for Takayasu arteritis.

Considerations when applying for these criteria	
These classification criteria should be applied to classify the patient as having Takayasu arteritis when a diagnosis of medium vessel or large vessel vasculitis has been made	
Alternate diagnosis mimicking vasculitis should be excluded prior to applying the criteria	
Absolute requirements	
Age <60 years at time of diagnosis	
Evidence of vasculitis on imaging	
Additional Clinical criteria	
Female sex	+1
Angina or ischemia cardiac pain	+2
Arm or leg claudication	+2
Vascular bruit	+2
Reduced pulse in upper extremity	+2
Carotid artery abnormality	+2
Systolic blood pressure difference in arms >20 mmhg	+1
Additional Imaging criteria	
Number of affected arterial territories (select one)	
One arterial territory	+1
Two arterial territories	+2
Three or more arterial territories	+3
Symmetric involvement of Paired arteries	+1
Abdominal aorta involvement with renal or mesenteric involvement	+3

These criteria were composed in part to reflect the developing role of noninvasive imaging for the evaluation of patients with TAK. Sum the scores for 11 items, if present. A score of ≥5 points is needed for the classification of Takayasu arteritis.

Our patient had the absolute necessity, with lowered pulse in the upper extremity, significant blood pressure discrepancy renal bruit, with three arterial territories involved in imaging with renal artery association giving a total score[11]. A total score more than is required for the classification of Takayasu arteritis. There was evidence of renal involvement with mild renal impairment with microscopic proteinurisa.

The main differential diagnosis to consider is Giant cell arteritis which is commonly seen in the elderly and involves external carotid artery and branches[12]. Association of renal artery is extremely uncommon, the next is Fibromuscular dysplasia. However, this syndrome often has characteristic radiographic

findings including "strings of beads appearances" is usually more focal in its involvement and is not associated with the systemic symptoms of Takayasu arteritis[13]. Atherosclerosis and Behçet syndrome also need to be entertained.

The mainstay of treatment for Takayasu arteritis is systemic glucocorticoids[14]. As it's a chronic disease with relapsing nature its paramount important to add a "steroid-sparing" benefit and longer-term disease control. For most patients with active TAK, we suggest the addition of either methotrexate (20mg once weekly) or azathioprine (2 mg/kg daily) to therapy with glucocorticoids Leflunomide and Mycophenolate mofetil are also recommended[14-16].

There are limited published data on the use of cyclophosphamide in Takayasu arteritis[17]. Biologic disease modifying anti rheumatic drugs (DMARDs) such as TNF inhibitor namely infliximab or other biologics like tocilizumab, abatacept, Ustekinumab and rituximab can be tried[18].

Treatment of Takayasu which is resistant to initial management, combination with an oral nonbiologic agent with a biologic agent, is recommended with oral corticosteroid whenever needed. Commonly used combinations are a TNF inhibitor to either methotrexate, azathioprine, or leflunomide. Combination of tocilizumab with eithermethotrexate, azathioprine, or leflunomide is also a reasonable approach.

Some patients maybe benefitted by vascular intervention if there is ongoing organ ischaemia, resistant hypertension or aneurismal disease[19]. Anerusymal disease are at risk for dissection or rupture, severe aortic regurgitation (AR), and aortic coarctation also demands surgery. Revascularization procedures, however, should be delayed during the active phase of the disease[20]. In our patient, we have started the high dose steroids with methotrexate but need serial monitoring including blood pressure and at least yearly MRI if stable disease to see the interval changes and early if clinically indicated.

Conclusions

Takayasu arteritis poses diagnostic and therapeutic challenges due to its chronic and variable course. Early recognition despite normal inflammatory markers and aggressive treatment are crucial to mitigate morbidity and mortality associated with the disease. Lifelong follow-up and close monitoring are essential for optimal management of TAK, highlighting the importance of further research to enhance understanding and management of this rare condition.

Refserences

1. Jennette J, Falk R, Bacon P, et al. Revised international chapel hill consensus conference nomencalture of vasculitidis. Arthritis Rheum 2013;65(1):11.
2. Weyand C, Goronzy J. Medium- and large-vessel vasculitis. N Engl J Med 2003;349(2):160.
3. Lupi-Herrera E, Sánchez-Torres G, Marcushamer JM. Takayasu's arteritis. Clinical study of 107 cases. Am Heart J 1977;93(1):94.
4. Arend W, Michel B, Bloch D, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 1990;33(8):1129.
5. Seko y, Minota S, Kawasaki A, 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.
6. Cid M, Font C, Coll-Vinent B, Grau J. Large vessel vasculitides. Curr Opin Rheumatol 1998; 10:18. Curr Opin Rheumatol 1998;10(1):18.
7. Goel R, Gribbons K, Carette S, et al. GribboDerivation of anangiographically based classification system in Takayasu's arteritis: an observational study from India and North America," Rheumatology (Oxford) 2020;59(5):1118.
8. Kerr GS, Hallahan C, Giordano J, et al. akayasu arteritis. Ann Intern Med 1994;120:919.
9. Rodríguez-Pla A, de Migue G, López-Contreras J, et al. Bilateral blindness inTakayasu's disease. Scand J Rheumatol 1996;25(6):394
10. Grayson PC, Tomasson G, Cuthbertson D, et al. Association of vascular physical examination findings and arteriographic lesions in large vessel vasculitis. J Rheumatol 2012;39:303.
11. Grayson C, Ponte R, Suppiah and et al. American College of Rheumatology/EULAR Classification Criteria for Takayasu Arteritis. Arthritis Rheumatol 2022;74:1872.
12. Michel B, Arend W, Hunder G. Clinical differentiation between giant cell (temporal) arteritis and Takayasu's arteritis. J Rheumatol, 1996;23:106.
13. D'Souza D, Liao A, Yap Jand et al. Fibromuscular dysplasia. Reference article: Radiopaedia.org.
14. Maz M, Chung S, Abril A, et al. American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Giant Cell Arteritis and Takayasu Arteritis. Arthritis Rheumatol 2021;73:1349.
15. Maz M, Chung S, Abril A, et al. American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Giant Cell Arteritis and Takayasu Arteritis. Arthritis Care Res (Hoboken) 2021;73:1071.
16. Hellmich B, Agueda A, Monti S, et al. Update of the EULAR recommendations for the management of large vessel vasculitis Ann Rheum Dis 2020;79:19.
17. Rodríguez-Hurtado F, Sabio J, Lucena J, Jimenze-Alonoso J. Ocular involvement in Takayasu's arteritis: response to cyclophosphamide therapy. Eur J Med Res 2002;7:128.
18. Hoffman G, Merkel P, Brasington R, et al. Anti-tumor necrosis factor therapy in patients with difficult to treat Takayasu arteritis. Arthritis Rheum 2004;50:2296.
19. Ogino H, Matsuda H, Minatoya K, et al. Overview of late outcome of medical and surgical treatment for Takayasu arteritis. Circulation 2008;118:2738.
20. Fields C, Bower T, Cooper L, et al. Takayasu's arteritis: operative results andinfluence of disease activity. J Vasc Surg 2006;43:64