

CASE REPORT

RARE PRESENTATION OF TAKAYASU'S ARTERITIS

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INTRODUCTION: Takayasu's arteritis is a rare form of large vessel granulomatous arteritis affecting young or middle aged women of Asian origin. It leads to intimal fibrosis and vascular narrowing. It mainly presents as pulseless extremities, so it is also called as pulseless disease. Treatment is mainly with steroids and immunosuppressants but TNF- α blockers gives promising results. Surgical options may need to be explored for patients who don't respond to steroids.

KEYWORDS: Takayasu's arteritis, steroid.

CASE REPORT: A 30 year old female presented with 10 months duration of pain, discoloration and disfigurement in digits of both hands and feet. There is history of increase in pain during night. There is no variation in pain on exposure to cold. She was treated for the same complaints by a Dermatologist, but the symptoms did not subside even after treatment. There is history of chest pain for 15 days. No history of Retrosternal chest pain/tearing/constricting type of pain/radiation of pain to left upper limb/jaw. No history of diaphoresis/vomiting. There is history of difficulty during swallowing for 10 days. There is no variation in swallowing for liquids or solids and swallowing difficulty subsided by itself in 3 days. There is history of low grade, intermittent fever and malaise for 3 days. There are no localizing features for fever.

History of night sweats occasionally present. History of shortness of breath on exertion for 3 days. There is no history of symptoms suggestive of angina/ respiratory tract infections. History is not suggestive of any habits or addictions. The patient is suffering with pain in digits of both hands and feet for the last 10 months. She was treated for the same complaints, but the symptoms persisted even after treatment. No history of any co morbidities.

On examination patient is conscious/coherent/oriented. She is moderately built and nourished. She is anemic. There is no clubbing/cyanosis/icterus/ pedal edema/ Lymphadenopathy. Pulse: 100 per min. Rate/Rhythm/volume/character/condition of the vessel wall are normal. Pulses are feeble in both brachial arteries. All other pulses felt equally bilaterally. Blood pressure: 180/ 120 mm Hg in left upper limb in supine position and 160/110mm Hg in right upper limb in supine position. There is no bruit over carotid/ subclavian/aorta/renal artery. Cardiovascular system/respiratory system/gastro intestinal system/central nervous system examination are within normal limits.

Local Examination:

1. Discoloration and disfigurement of left tip of index finger. (Fig. 1)
2. Swelling at the tip of left ring finger. (Fig. 2)
3. Swelling at the tip of right middle and right index fingers. (Fig. 3a, 3b)
4. Discoloration of right great toe.

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Figure 1



Figure 2



Figure 3a



Figure 3b

INVESTIGATIONS:

Hb: 8.9 gms/ dl

Normocytic, Normochromic anemia is present.

T.C.: 9,700 cells/mm³

ESR: 110 mm/ Hour

CRP: 14 mg/ dl

ANA/ds DNA: Negative. Auto antibodies for other connective tissue disorders are also negative.

Fasting blood sugar: 118 mg/dl

Na+: 130/k+: 4.2/Creat: 1.4 mg/ dl. Other biochemical investigations are within normal limits.

Urine analysis normal.

2D Echo: Normal

Doppler Ultrasonography: Normal.

TREATMENT: She was treated with prednisolone 1mg/ kg/ day, Azathioprine 100mg two times a day, Pentoxifylline 400mg two times a day, Metoprolol 25mg two times a day, prazosin 5mg once a day, Telmisartan 40 mg once a day, Atorvastatin 20mg once a day, Tramadol 50mg once a day. Cyclophosphamide, Methotrexate or TNF blockers like Infliximab, Etanercept can also be used in the

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treatment. Auto amputation of gangrenous parts of fingers occurred after treatment. Her symptoms subsided after three months of treatment.

DISCUSSION: Takayasu's Arteritis is a rare, idiopathic, noninfectious, chronic granulomatous, large vessel vasculitis of young adults affecting the aorta^{1, 2} and its branches such as the pulmonary³ arteries. It is also called as Aorto arteritis or pulse less disease or Aortic arch⁴ syndrome because of its predilection for brachio cephalic vessels. Takayasu's Arteritis with digital gangrene is a rare manifestation.

It was named after the Japanese Ophthalmologist^{5, 6} Mikito Takayasu who first described the disease in 1905⁷. Shikhare reported the first case report in India in 1921.⁸

The incidence of the disease is 1.26 to 2.6/million/year. It is 10 times more common in women than in men. Age of onset of the disease is in between second and third decades. All races are equally affected.

Deposition of Mononuclear cell infiltrates (macrophages, T cells, gamma, delta, cytotoxic and NK cells) in the arterial intima is the underlying pathogenic mechanism of the disease. Cytokines, IL-6, TNF in these lesions prompted therapeutic approaches using cytokine targeted biologic agent.

Clinical features;-Claudication is the most commonest complaint (due to involvement of subclavian artery) followed by Bruit, (approx. 80%) Blood Pressure or Pulse asymmetry, (60-80%) Hypertension due to Renal Artery Stenosis or a chronically damaged rigid aorta^{9, 10}. Cardiovascular¹¹, Renal¹², Vascular¹³ complications cause severe morbidity and mortality. Mortality range is 3-35%.

Patients with active disease presents as increased extremity or visceral pain, malaise, myalgias, arthralgia, night sweats, fever and laboratory evidence of elevated ESR.

INVESTIGATIONS: Besides routine investigations, Acute phase reactants like ESR/ CRP are elevated. Normocytic normochromic anemia may be present. Auto antibodies are negative. Arteriography is the gold standard test. Doppler Ultrasonography helps in assessing occlusive changes, dilation and can measure post-stenosis flow velocity, arterial wall thickening. CT angiography helps in early diagnosis of wall thickening. MRI angiography may be useful for follow up and to assess vessel wall. FDG-PET is a noninvasive metabolic imaging test. It has high sensitivity and specificity for the presence of inflammation.

DIAGNOSIS: Diagnostic criteria for Takayasu's arteritis as per 1990 American college of Rheumatology (ACR):

1. Age at onset < 40 years
2. Limb claudication
3. Diminished brachial pulsations
4. Difference of > 10 mm Hg systolic pressure between arms
5. Bruit over subclavian or aorta
6. Abnormal angiogram

A patient has Takayasu' arteritis if > 3 criteria are present. (Sensitivity: 90.5%, Specificity: 97.8%).

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There is poor correlation between Clinical, Laboratory, Radiologic and Histologic data in Takayasu's arteritis. Arteriography is the Gold standard test in Takayasu's arteritis. MRI detects qualitative abnormalities in the vessel wall that imply inflammatory change. FDG –PET scan has high sensitivity and specificity in inflammation. No specific diagnostic test for Takayasu's arteritis. Diagnosis depends on clinical features in conjunction with vascular imaging abnormalities

DIFFERENTIAL DIAGNOSIS: The following are the differential diagnosis for Takayasu's arteritis

1. Marfans syndrome 2. Ehler Danlos syndrome 3. Inborn genetic errors 4. Large vessel aneurysms caused by Bacteria, Syphilitic, Mycobacteria, Fungal infections. 5. SLE 6. Cogan syndrome 7. Behcets disease 8. Spondyloarthropathies 9. Kawasaki disease 10. Giant cell arteritis 11. Sarcoidosis.

TREATMENT: Corticosteroids are the drug of choice for treatment of the disease. Prednisone 1 mg/kg/day is the dose of steroid for 3 months. Relapses occur during steroid tapering. Steroid resistant and relapsing disease responds to cyclophosphamide 2 mg/kg/day or methotrexate 20 mg/week. Dramatic improvement occurs with TNF blockers. Patients who do not respond to medical management should go for anatomic correction of lesions. Angioplasty,¹⁴ stenting,¹⁵ valve replacements and bypass surgeries can be done for anatomic correction of lesions.¹⁶

REFERENCES:

1. Braunwald's heart disease. A textbook of cardiovascular medicine -9th edition. Editors: Robert O. Bonow, Douglas L. Mann, Douglas P. Zipes, Peter Libby. Page no 1876-1878, Published by Saunders, An imprint of Elsevier, 3251, River Port Lane, St.Louis, Missouri, 63043, Published by Elsevier Inc.
2. Sen PK, Kinare SG, Kulkarni TP, Parulkar GB. Stenosingaortitis of unknown etiology- A study of four cases. *Surgery* 1962; 51:317.
3. Lupi-Herrera E, Sanchez Torres C, Marcushamer J et al. Takayasu Arteritis, clinical studies of 107 cases. *Am Heart J* 1977; 93:94.
4. Ueno A, Awane G, Wakahayaci A. Successfully operated obliterative brachiocephalic arteritis (Takayasu) associated with elongated coarctation. *Jap Heart J* 1967; 8:538.
5. Hurst's The heart text book 13th edition.vol 2 page no 2285-2286.Editors: Valentin Fuster, Richard A. Walsh, Robert A. Harrington, Hunt, King III, Nash, Prytowsky, Roberts, Rose. Published by The McGraw Hill Companies Inc, United States of America.
6. Takayasu M. A case with unusual changes of the central vessels in the retina. *Acta Soc Ophth Jap* 1908; 12:554.
7. Harrison's principles of internal medicine 18th edition. vol 2, page no: 2796-2797. Editors: Dan L. Longo, Anthony S. Fauci, Dennis L. Kasper, Stephen L. Hauser, J. Larry Jameson, Joseph Loscalzo. Published by The McGraw Hill Companies Inc. United States of America.
8. Shikhare PV. Note on a remarkable case of absence of pulsation in the arteries of upper part of the body. *Ind J Med* 1921; 2:326.
9. Panja M, Sarkar C, Kumar S, Kar AK, Mazumder B, Roy S, Sinha DP, Sarkar NC. Coronary artery lesions in Takayasu's arteritis: clinical and angiographic study. *J Assoc Phys Ind* 1998; 46; 67881

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10. Di Giacomo V. A case of Takayasu's disease occurred over two hundred years ago. *Angiology* 1984; 35:750.
11. McKusick VA. A form of vascular disease relatively frequent in the Orient. *Am Heart J* 1962; 63:57.
12. Kieffer E, Piquois A, Bertal A, Blétry O, Godeau P. Reconstructive surgery of the renal arteries in Takayasu's disease. *Ann Vasc Surg* 1990; 4:156-165.
13. Sharma S, Gupta H, Saxena A, Kothari SS, Taneja K, Guleria S, Rajani M. Results of renal angioplasty in nonspecific aortoarteritis (Takayasu disease). *J Vasc Interv Radiol* 1998; 9:429-435.
14. Tyagi S, Kaul UA, Nair M, Sethi KK, Arora R, Khalilullah M. Balloon angioplasty of the aorta in Takayasu's arteritis: initial and long-term results. *Am Heart J* 1992; 124:876-882.
15. Taketani T, Miyata T, Morota T, Takamoto S. Surgical treatment of atypical aortic coarctation complicating Takayasu's arteritis- experience with 33 cases over 44 years. *J VascSurg* 2005; 41:597- 601.
16. Miyata T, Sato O, Deguchi J, Kimura H, Namba T, Kondo K, Makuuchi M, Hamada C, Takagi A, Tada Y. Anastomotic aneurysms after surgical treatment of Takayasu arteritis: a 40-year experience. *J VascSurg* 1998; 27:438-445.

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