

Case Report

Takayasu's arteritis: A case report

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ABSTRACT:

Takayasu's arteritis is a chronic nonspecific inflammatory and stenotic disease of medium and large sized arteries mainly involving aortic arch. It is an uncommon disease prevalent in adolescent girls and young women and is a common cause of renovascular hypertension in Asian females. Aortic dissection is one of the rare complications of Takayasu's arteritis. We hereby discuss an interesting case of middle aged female who presented with easy fatigability and pain of left upper limb.

Key words: Takayasu's arteritis

Received: 12 December, 2020

Revised: 24 December, 2020

Accepted: 29 December, 2020

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This article may be cited as: Gudka S, Shetty V, Chandrakar S, Moulick ND, Peshimam U. Takayasu's arteritis: A case report. J Adv Med Dent Scie Res 2021;9(2): 15-17.

INTRODUCTION

Takayasu's arteritis is a chronic nonspecific inflammatory disease mainly involving aorta and its branches mainly including brachiocephalic, carotid, subclavian, vertebral, renal, coronary and pulmonary arteries. Involvement is more marked proximally than distally. Histologically presents as pan arteritis with inflammatory cell infiltration, marked intima proliferation, fibrosis and scarring of media along with involvement of vasa vasorum, narrowing of arterial lumen with or without thrombosis. Pathological changes in various organs reflect the compromise of blood flow through the involved vessels.¹⁻³ We hereby discuss an interesting case of middle aged female who presented with easy fatigability and pain of left upper limb.

CASE REPORT

A 37 year old female presented to our institution with complaints of fatigue since 1 year, pain (non-radiating) in the left upper limb since 1 year, and upper abdominal pain since 1 year. There was no history of chest pain, syncope, palpitations, wheezing, cough, altered bowel habits or menstrual irregularities. Past medical history and family history were insignificant. On general physical examination, mild pallor was seen with bilateral lower extremity edema. Brachial and radial pulses were

absent on the left side. Pulse rate was 100 beats per min whereas carotid bruit was present on left side. There was a remarkable blood pressure discrepancy between the right and the left arm. Systemic blood pressure was 160/100mm hg in right arm in supine position and 146/90 mm hg in left arm in supine position. Abdominal examination revealed tender hepatomegaly 5cm below the costal margin but there was no abdominal bruit. Preliminary blood examination showed presence of dimorphic anaemia and elevated ESR. No significant abnormalities were seen on chest X-ray and abdominal USG. Doppler ultrasound of renal arteries was also normal. However; abdominal Doppler revealed mild stenosis at origin of celiac artery, diffuse thickening of wall of aorta which was suggestive of aorto-arteritis. Colour Doppler of both upper limbs showed left proximal subclavian artery stenosis with consequent dampened flow in left distal subclavian artery with collaterals. Monophasic and dampened flow was present in left upper limb arteries and there was mild dilatation of right brachiocephalic trunk. Carotid Doppler showed diffuse wall thickening of bilateral carotid arteries more on left side along with short segment moderate stenosis in the left distal common carotid artery and external carotid artery. Presence of multiple manifestations supported the diagnosis of Takayasu's arteritis. Treatment therapy

which was administered included: methotrexate, azathioprine, folic acid, metoprolol and atorvastatin. Inflammatory parameters normalised within 3 weeks and she was shifted on maintenance regimen. At 6 months follow up, the patient was asymptomatic.

DISCUSSION

Takayasu's disease is a chronic inflammatory disease of large- and medium-sized arteries, involving the aorta and its main branches, the pulmonary arteries, and the coronary tree. Since the original report of Takayasu's disease in 1908, the estimated worldwide incidence is 2.6 cases per million per year, with women more commonly affected than men. Peak onset is in individuals in their 30s. The disease has been mainly studied in Japan but Western studies have also been published. Cardiac features are present in up to 40% of cases. Patients usually have no risk factors for atherosclerosis and yet have atheromatous aorta, suggesting the importance of inflammation in atherosclerosis.¹⁻⁵

In the present case report, we have discussed the case of middle aged female who presented with easy fatigability and pain of left upper limb. A 37 year old female presented with complaints of fatigue since 1 year, pain (non-radiating) in the left upper limb since 1 year, and upper abdominal pain since 1 year. There was a remarkable blood pressure discrepancy between the right and the left arm. Abdominal Doppler revealed mild stenosis at origin of celiac artery, diffuse thickening of wall of aorta which was suggestive of aorto-arteritis. Wang et al found that high resolution sonography was able to discern tiny dissected intimae and provide precise images for evaluation of TA. Many factors, such as arteriosclerosis, long-term hypertension, dyslipidemia, and connective tissue disorders, have been identified that can damage the aortic wall and lead to dissection. Autoimmune diseases, including TA, can cause persistent inflammation of the aorta, but rarely dissection because of the dense adventitial fibrosis and intimal scarring. Some researchers have speculated that decreased wall elasticity and aortic mobility due to fibrous adhesion between fibrous adventitia and surrounding tissue may cause dissection in TA cases.⁶⁻⁸

Manfrini O et al presented a case report which focused on a Caucasian middle-aged woman which complained of weakness, malaise, and fatigue for as many as 19 years. Delayed diagnosis and lack of specific treatment could explain the extent and the clinical severity of the disease at time of hospital admission. Angiography showed focal narrowings of the abdominal and thoracic aorta and occlusion of both the subclavian arteries, of the right coronary artery and severe stenosis of the first marginal obtuse. Takayasu's arteritis is not limited to women of Japanese origin but is present worldwide. Early diagnosis and treatment is warranted. Outcome appears to be favorable when the disease is quiescent.⁹

The diagnosis of TA is primarily based on clinical and radiological findings, as the results of biopsy are nonspecific as the histopathology may imitate other types of vasculitis. Suspected TA always warrants prompt

vascular imaging, enabling earlier diagnosis and further decreasing the risk to the patient. Although angiography was considered to be the standard method for diagnosis of TA, it has been replaced by computed tomography angiography or angiography or magnetic resonance angiography. Furthermore, literature has shown that ultrasound with color Doppler flow imaging and angiography are highly useful for detecting and determining the severity of the disease (except for right brachiocephalic artery).¹⁰⁻¹²

In the present case report, treatment therapy which was administered included: methotrexate, azathioprine, folic acid, metoprolol and atorvastatin. Inflammatory parameters normalised within 3 weeks and she was shifted on maintenance regimen. At 6 months follow up, the patient was asymptomatic. Two phenotypic variants are recognized; pyridoxine (vitamin B6) responsive and pyridoxine non-responsive homocystinuria. Pyridoxine is a cofactor of the enzyme cystathionine β -synthase and supplementation leads to prompt metabolic normalization in about 50% of the patients. Folate and vitamin B12 optimize the conversion of homocystine to methionine by methionine synthase, thus helping to decrease the plasma homocystine concentration. Diagnostic tests include cyanide nitroprusside test in urine, elevated levels of homocystine and its metabolites in blood and measurement of cystathionine β -synthase enzyme activity in tissue extracts. Treatment includes pyridoxine, folate and vitamin B12 supplementation in responsive subgroups and methionine restriction in diet.¹³⁻¹⁵ MW Winiecki et al presented a case report of a 33-year-old female who was diagnosed with Takayasu's arteritis. The patient underwent a laparoscopic tubal ligation under general anesthesia without complication. Takayasu's arteritis is a chronic inflammatory disease seen primarily in women. It can involve the aorta and its major branches, as well as the coronary, hepatic, mesenteric, pulmonary, and renal arteries. Takayasu's syndrome is also known as the "pulseless disease," because of the characteristic lack of peripheral pulses. A wide variety of presentations are possible with this syndrome. Patients range from asymptomatic to severely compromised, and their medical condition and previous therapy can offer an interesting set of challenges. Monitoring patients with Takayasu's syndrome can give rise to several unusual problems.¹⁶

From the more typical features of Takayasu's arteritis, the American College of Rheumatology (ACR) defined specific diagnostic criteria for this disorder in 1990. Angiography remains the gold standard for diagnosis. Assessment of pulmonary vasculature by angiography is not universally recommended, being reserved for patients with symptoms of pulmonary hypertension. Doppler ultrasound is a useful non-invasive procedure for the assessment of vessel wall inflammation. In view of the vessels involved, histological diagnosis is usually impractical and histological assessment is limited to those cases undergoing revascularisation procedures.¹⁷ Steroids have formed the mainstay of treatment for Takayasu arteritis and reports of efficacy vary. It is now

accepted that approximately half of patients treated with steroids will respond. This lack of universal success and the side effects associated with steroid use have led to a search for a more effective treatment. Immunosuppressive agents including cyclophosphamide, azathioprine, and methotrexate have all been tried. Kerr et al studied 25 steroid unresponsive patients receiving cytotoxic medications including cyclophosphamide, azathioprine, or methotrexate, although not concurrently. The overall remission rate was 33%. Twenty three per cent of all treated patients in their study never achieved remission.¹⁸

CONCLUSION

Takayasu's arteritis is an uncommon presentation accompanied by life threatening complications. Such unusual presentation underscores the importance of complete and thorough evaluation of patients including utilisation of non-invasive investigations. Patients with Takayasu's arteritis such as this unique one should be managed according to standard guidelines, appropriate medical management and surgical intervention when required.

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