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Research Article

A CASE STUDY ON TAKAYASU ARTERITIS WITH DILATED CARDIOMYOPATHY: AN UNCOMMON SYSTEMIC INFLAMMATORY DISORDER OBSERVED IN TERTIARY CARE TEACHING HOSPITAL

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ABSTRACT

Takayasu arteritis is an uncommon, systemic granulomatous inflammatory large vessel vasculitis effecting the aorta and its major branches of unknown aetiology that most commonly affects women of ageless than 50. It is also known as aortic arch syndrome, pulse less disease, non-specific aortic arteritis. The disease progression can be controlled by systemic corticosteroids and Dilated Cardiomyopathy (DCM) is a disease of heart muscle in which the bottom chambers of heart (ventricles) stretches and thins and cannot pump blood normally. Here in, we present a case study of a 33 year old female patient admitted in the hospital due to weakness of right lower and upper limb (proximal and distal) and not progressive and is associated with fatigue, chest pain, dyspnea and was diagnosed as Takayasu Arteritis with Dilated Cardiomyopathy by basing on the physical examinations and clinical investigations. Treatment initiated with oral Tab. Prednisolone of dose 40 mg, Tab. Enalapril of dose 5 mg, Tab. Atorvastatin 40 mg and Tab. Clopidogrel of dose 75 mg and other drugs to achieve homeostasis. After close monitoring and management with medication for 10 days, the patient was discharged by performing discharge counselling and prescribing discharge medications.

Keywords: Large vessel vasculitis, Granulomatous inflammation, Aortic arteritis, treatment

INTRODUCTION

Arteritis refers to the inflammation of arteries (blood vessels that carry bloodfrom heart to rest of the body). Takayasu Arteritis (TA) is an uncommon condition in which the inflammation damages the large and medium sized arteries. It is also known as aortic arch syndrome, pulse less disease, non-specific aortic arteritis. Branches of aorta are the commonly affected arteries and then the arteries that supply blood to the arms, neck and then to the brain. Less commonly arteries are vessels of heart, intestines, kidneys and legs are involved. Takayasu arteritis can manifest as exceptional, abnormal and or catastrophic disease[1].

Takayasu's Arteritis was included in the Japanese government list of intractable diseases in 1990 and till date 5000 patients were diagnosed with Takayasu's Arteritis.

Criteria for diagnosing Takayasu's arteritis

1. Disease onset before 40 years - development of symptoms
2. Claudication of extremities – includes muscle fatigue and discomfort of at least 1 muscle or more extremity in use. Mainly in the upper limbs.
3. Decreased pulses in the brachial artery in most, carotid artery, radial artery.
4. Difference in the systolic blood pressure of 10 mmHg between both arms.
5. Auscultation over aorta (abdominal) or in sub-clavian arteries.
6. Arteriogram shows occlusion of aorta and hence narrowing or its branches or arteries in the upper or lower extremities.

2 or 3 criteria have to be met in order to diagnose the disorder as Takayasu's Arteritis. 1990 ACR (American Collage of

Rheumatology) criteria for the classification of Takayasu arteritis[2]

DCM is a progressive disease of heart muscle that is characterized by ventricular chamber enlargement and contractile dysfunction. The bottom right chamber of heart (ventricle) may also be dilated and dysfunctional[3].

In Dilated Cardiomyopathy, the myocardium is dilated, without any known cause. The systolic pump function of left or right ventricle is diminished, leading tohypertrophy, a process called remodelling. The presentation as DCM is uncommonly reported due to involvement of coronary artery, hypertension, and cardiac failure. DCM is the common cause for congestive cardiac failure in about one in three cases. About 30% of patients have genetical cause, with mutations affecting a gene that encodes cytoskeletal proteins and other proteins involved in constricting[4].

Continuing ventricular enlargement and dysfunction generally leads to developing heart failure with further decline in left ventricle contractile function. Sequela include ventricular and supra ventricular arrhythmias, conduction system abnormalities, thrombo embolism, and sudden death or heart failure–related death[5].

Angiography-based categories

Based on angiographic involvement, Takayasu Arteritis was of 6 types –

- TYPE-I: Aortic arch branches
- TYPE IIa: ascending aorta, its arch and branches
- TYPE IIb: ascending aorta, its arch and branches and descending aorta
- TYPE III: descending and abdominal aorta, arteries of kidney
- TYPE IV: abdominal aorta, arteries of kidney or both
- TYPE V: entire aorta and branches[6].

Inflammation of large blood vessels can cause segments of vessels to weaken and stretch causing aneurysm or may become narrowed i.e. causes and occlusion.

EPIDEMIOLOGY

Takayasu arteritis is rare, but most commonly seen in Japan, South East Asia and Mexico. In 1990, it was included in the list of intractable diseases and to date nearly 5000 patients have been registered. The incidence was about 1-2/million/year in Japan and 2.2 /million in Kuwait, 0.9/million in US. The symptoms of 106 patients over 16 years are documented. In India, the commonest mode of presentation of Takayasu arteritis cause of renovascular hypertension is reported in 28-75%, CHF in 76%, aortic regurgitation in 20-24%[7] and DCM is, however, reported to be seen in only 5-6% of cases of Takayasu arteritis[8].

AETIOLOGY

The cause of TA is unknown[9]. However, a hypothesis had been developed that a heat-shock protein in the aortic tissue called as 65k Dagets stimulated by unknown stimulus leading to an immune induction of major histocompatibility class I chain-related A (MICA) located on the vascular cells[10]. It mostly affects young women; it may also affect children and adults of all ages. At diagnosis, patients are often between the ages of 15 to 35. A link between the Takayasu's arteritis and an infection has not proven. However, pathogenesis of the disease induction is still not known. However, Tuberculosis, rheumatoid arthritis, collagen vascular related diseases or Streptococcal infections have been debated as its etiology in the past. Recently, more importance has been given on an immunopathological cause[11,12].

The role of genetics is unclear, however genetic susceptibility have been identified[13].

Clinical features

- Diminished or absent throb of patients associated with limb limping and blood pressure discrepancies
- Vascular bruits in 80–94% of patients, often involving several parts and particularly affecting the carotids, sub clavian and abdominal vessels[14].
- Hypertension in about 70% of patients[15], usually reflecting narrowing of arteries in the kidney, which is seen in about 60% of patients[16].
- Damage to the retina of eye due to TA was also includes in some patients[17].
- Congestive cardiac failure related with aortic regurgitation, hypertension and dilated cardiomyopathy[18].
- Neurological features including vertigo, convulsions, and amaurosis.
- Pulmonary artery inclusion in 14–100% of patients[19].

METHODOLOGY

The case was intervened at Maharajah's institute of Medical Sciences, Nellimarla, Vizianagaram, Andhra Pradesh, 535217. Before interacting with patient informed consent form was taken and the study is carried out as per International Conference of Harmonization- Good Clinical Practices Guidelines (ICH-GCP).

Case study

A 33 year old female patient was admitted in the general medicine department with complaints of weakness of both proximal and distal parts of right upper and lower limbs. There is no involvement of sensory disturbances or involuntary micturition or deviation of mouth. She is moderately built and nourished. She is not an alcoholic and smoker and takes mixed diet. Her bowel and bladder habits were regular. Similarly there is also no involvement of dyspnoea or neck rigidity in past. Co-morbidities such as left ventricular dysfunction were indicated which may be occurred due to dilated cardiomyopathy

Past medical history

The patient was known to possess hypertension and was on medication.
There is no record of tuberculosis, asthma or epilepsy.

Past medication history

The medication history indicated that she was on Tab. Enalapril 5 mg, Tab. Pantop 40 mg, Tab. Metoprolol 25 mg

RESULTS AND DISCUSSION

Table 1: Blood Pressure and Pulse Rate data

Days	D1	D2	D3	D4	D5	D6	D7
BP (mmHg)	130/80	140/80	110/70	110/70	100/60	100/60	100/60
PR (bpm)	100	92	80	82	76	82	80

Table 2: Complete Blood Profile data

Peripheral Blood Smear	
Hb (11-14gm/dl)	13.2
PCV	39%
Platelet count (1.5 – 4.5 lakhs/cumm)	3.4 lakhs /cumm
WBC (4000 - 11000)	12,400 CELLS/cumm
Differential Count	
Polymorphs (40 – 75%)	67%
Lymphocytes (20 – 45%)	31%
Eosinophils (01 – 06%)	2%
Erythrocyte Sedimentation Rate	21 mm/hour

Biochemical Investigations	
Blood urea (14 - 45 mg/dl)	44
Serum creatinine (0.6 – 1.4 mg/dl)	1.1

Liver Function Tests	
Total bilirubin (up to 1mg/dl)	0.3
Direct bilirubin (up to 0.25mg/dl)	0.1
Indirect bilirubin	0.2
Alkaline phosphatise (15- 116 IU/L)	95
SGOT (up to 37 IU/L)	24
SGPT (up to 65 IU/L)	15

Serum Electrolytes (mmoles/L)	
Serum sodium (135 - 155)	138
Serum potassium (3.6 – 5.5)	4.27

Vital signs were normal. As it was a pulse less syndrome, pulse was felt randomly on carotid, brachial and popliteal arteries of both the lower and upper limbs. The pulse as not felt on radial, post tibial, dorsalis pedis arteries.

MRI scan of brain revealed Lacunar infarct due to small vessel ischemia. Prominent sulcal spaces in left temporal and occipital regions due to ischemia. Based on all the presenting evidences and patients' condition a final diagnosis of Takayasu's arteritis was made.

Dilated cardiomyopathy (DCM) was diagnosed based on medical history and chest X-ray and MRI Chest X-ray shows enlarged left ventricle and atria with pulmonary edema associated pleural effusions seen.

MRI reveals dilated cardiomyopathy i.e. the left side of heart is markedly dilated and thinned and mid wall enhancement especially in the septum.

Table 3: Drug Chart

Trade name	**Generic name**	**Dose**	**Freq.**	**Route**	**D1**	**D2**	**D3**	**D4**	**D5**	**D6**	**D7**	**D8**	**D9**
T. Prednisolone	T. prednisolone	40 mg	OD	PO	√	√	√	√	√	√	√	√	√
T. enalapril	Enalapril	5 mg	OD	PO	√	√	√	√	√	√	√	√	√
T. Atenolol	Atenolol	50 mg	OD	PO	√	√	√	√	√	√	√	√	√
T BC	B complex		OD	PO	√								
T. Rantac	Ranitidine	150 mg	OD	PO	√	√	√	√	√	√	√	√	√
T. Aldactone	Spironolactone	25 mg	OD	PO	√	√	√	√					
T. Lasix	Furosemide	20 mg	OD	PO	√	√	√	√	√	√	√	√	√
T. Aspirin	Aspirin	75 mg	OD	PO		√	√	√	√	√	√	√	√
T. clopidogrel	Clopidogrel	75 mg	OD	PO				√	√	√	√	√	√
T. Atorvastain	Atorvastatin	40 mg	H/S	PO				√	√	√	√	√	√

DAY 1

Patient was coherent and conscious. She developed right sided weakness and was admitted in hospital. The therapy was initiated with corticosteroid like Prednisolone 40 mg to relieve from inflammation and anti-hypertensives like Enalapril (ACEI) 5 mg and Atenolol (β-blocker) 50 mg to reduce hypertension and for ventricular remodeling along with Spironolactone (potassium sparing diuretic) 25 mg and Furosemide (loop diuretic) was added to relieve hypertension. Ranitidine (H2 receptor blocker) of dose 150 mg is to reduce gastric secretion. Vitamin supplement of B complex was given. Patient's temperature was a febrile with normal heart sounds and no vascular breath sounds were felt in respiratory examination. Her motor examination includes tone and power on both the limbs was normal but mute in right lower limb. High mental function was seen with CNS examination. Peripheral pulses were felt except with posterior tibial artery.

DAY 2

Patient was conscious and coherent. Complete blood count, liver function tests, biochemical investigations and serum electrolytes test was advised to the patient. The same therapy was continued as of DAY 1 and Aspirin (Anti-platelet) 75 mg added in the therapy to prevent clots in the ventricle as pooling of blood takes place in the ventricle due to DCM. Her tone and power on both the limbs were normal but mute in right lower limb. The deep tendon reflexes were decreased in right limbs. Pustules were present in right elbow and face.

DAY 3

Patient was coherent and conscious. She developed right sided weakness of both lower and upper limbs in the morning and unable to walk. The same therapy was continued as of DAY 2. The difficulty in lifting the patients' hand and leg was observed. MRI scan of brain and MRI angiogram was advised to the patient.

DAY 4

Patient was coherent and conscious. Carotid and popliteal arteries pulse was felt on both left and right side. Brachial pulse was felt only on right side. Radial pulse was not felt. The therapy was continued same as DAY 3 along with those drugs Tab. Clopidogrel 75 mg as an adjunct to aspirin in anti-platelet therapy and Tab. Atorvastatin 40 mg for its cardiac remodeling and protective action were added.

DAY 5

Patient was coherent and conscious. Arterial pulse was felt for carotid, brachial, femoral, popliteal arteries and absent in radial and post tibial arteries. The deep tendon reflexes were very much reduced in right lower limb. The therapy was continued same as DAY 4. Tab. aldactone was removed from the therapy as it has serious interaction with furosemide if taken at a time. Co-administration of similar diuretics was not safe for the patient as it causes severe hypotension. Patient was experience symptoms like weakness, movement disorders, dysphonic which was concluded as functional neurological disorder (FND).

DAY 6

Patient was conscious and coherent with high mental function. Radial, post tibial, dorsalis pedis pulse was not felt. The patient difficulty in lifting the hand was observed. The same treatment was continued as of DAY 5.

DAY 7

Patient was conscious and coherent. Carotid, radial and brachial pulses were felt on right side. All her vitals were normal. The same therapy was continued as of DAY 6. Counseling to the patient was given regarding the disease and her medication in order to get relieved from FND.

DAY 8

Patient was conscious and coherent with carotid pulse, dorsalis pedis pulse was felt on right side. All her vital signs were normal. The patient ability to lift the hand was improved. Her mental function was normal.

DAY 9

Patient was conscious and coherent with high mental function. Radial, post tibial, dorsalis pedis pulse were randomly felt. The same treatments were continued as of DAY 8. The patient was able to lift the right limb without any restriction. Her walking ability was slightly increased.

DAY 10

Patient was conscious and asymptomatic and stable. She relieved from weakness and was able to walk and move her both right upper and lower limbs. Discharge medication was prescribed, and counseling was performed.

CONCLUSION

Takayasu's arteritis is an uncommon inflammatory disease occurring in about 1-2 million individuals in Japan, 2.2 million in Kuwait and 0.9 million in US. The common cause in India was hypertension, CHF, aortic regurgitation and DCM. Most commonly affects women of age less than 50. It is also known as pulse less disease, aortic arch syndrome, non-specific aortic arteritis. Based on the age on set of disease of less than 40 years-general symptoms of weakness of right lower leg and lower leg which was not progressive and decreased brachial pulses and claudication of extremities, it was diagnosed as Takayasu's arteritis. Patient was managed with corticosteroid like Tab. Prednisolone (40 mg) and antihypertensive agents like Tab. Enalapril (5 mg), Tab. Atenolol (50 mg), Tab. Spironolactone (25 mg), Tab. Furosemide (20 mg) and anti-platelet agents like Tab. Aspirin (75 mg), Tab. Clopidogrel (75 mg), HMG-CoA reductase enzyme inhibitor Tab. Atrovastatin (10 mg) and H2 receptor antagonist Tab. Ranitidine (150 mg). Patient was relieved from symptoms that are ease in lifting the limbs, increased pulses. Discharged medication was given, and discharge counseling was performed by clinical pharmacist

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