Takayasu's disease : A case report
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Summary
Takayasu’s disease is an idiopathic inflammatory arteriopathy involving the aorta and its main branches. Symptoms are related to vascular insufficiency of affected organs, especially the brain, eye, heart and kidneys. Major ocular complications consist of ischaemic retinopathy, complicated cataract and neovascular glaucoma. The definite diagnosis can be established by carotid doppler study or aortic angiography. We here in reported a Bangladeshi old male patient with ocular ischaemia due to this disease.

Key words
Takayasu's disease, pulseless arteritis, color fundus photography, carotid doppler study.

Introduction
Takayasu's disease is an inflammatory disease of the aorta & its branches in aortic arch. This decreases the flow of blood to the areas supplied by these branches, which in turn lead to a lack of pulse in those areas- arms, legs, neck and head-producing symptoms. Symptoms consist of aphasia, transient hemiparesis, unilateral transient amblyopia or persistent blindness, headache, vertigo, syncopal attack and muscle wasting. Takayasu noted lack of pulse in the arteries of the eye, thus the name pulseless disease. Most common in the orient and showing predilection for young women, but also occurring in males throughout the world. It is a chronic relapsing disorder which may extend from childhood to late adult life and seldom occurs in persons over 40 years of age. The condition is progressive and the prognosis is poor, mostly in untreated cases. Death usually occurs after a few years, although survival for 20 years after onset of symptoms has been reported. It is probably of auto-immune origin.

This vascular disturbance was first described by Mikito Takayasu, a Japanese ophthalmologist at the 12th Annual meeting of the Japan Ophthalmology Society in 1908. His patient was a 28 years old Japanese girl with retinal neovascularization and absence of pulses in superior extremities. Subsequently 57 cases have been reported. It has been observed most frequently in young women. Major complications attributed to the disease are Takayasu's retinopathy, secondary hypertension, aortic regurgitation, aortic or arterial aneurysm etc. There are geographical variations in the clinical aspects of this disease.

Case report
A 55 years old man hailing from Faridgonj, Chandpur gradually developed pain and claudication in both hands and feet with visual disturbances on both eyes. He also gave the history of fatigue, weight loss, headache, dizziness, syncope, chest pain etc. Cardiovascular evaluation showed loss of pulses in upper limb. The lower limb pulses were normal. BP 90/62 mm of Hg in lower limb. Both carotid pulses were palpable with loud bruit. Ocular examination showed features of ischaemic retinopathy with early cataract. Neurological examinations were otherwise normal.

Haematological examination reveals normochromic, normocytic anemia, mild leukocytosis with raised ESR (80mm in 1st hour). Chest Radiography showed cardiomegaly with widening of aorta. ECG reveals features of anterior wall ischaemia.

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Carotid doppler study showed narrowing with atheromatous plaque at and near carotid bifurcation and its major two-external and internal carotid arteries.

Magnetic resonance imaging revealed thickening of the aortic arch extending into both carotid and subclavian arteries.

Arch aortography shows moderate stenosis in aortic arch and in its branches. It was diagnosed as a case of Takayasu's arteritis. Initially he was treated medically by high dose prednisolone (2mg/kg/day), methotrexate, cyclosporin, aspirin (150mg/daily) and then managed by surgical treatment- reconstructive vascular surgery & some carotid interventions. His symptoms were improved, the ESR returned to normal (5mm/hr). After proper counselling the patient was satisfied and remains asymptomatic with low dose prednisolone (5mg/daily).

Discussion

Takayasu's disease is a multisystem involvement disorder. Various categories are observed here:-

- Group-I : Uncomplicated Takayasu's disease, with or without involvement of the pulmonary artery.
- Group-II : TD with a single complication- ocular or cardiac.
- Group-III : TD with two or more complications.

There has been extensive research on this granulomatous vascular disorder throughout the whole world. There is often a long interval between the onset of symptoms, which usually begins at a young age and the establishment of the diagnosis, which is done at later age i.e. 35-60 years. We have diagnosed the case at the age of 55 years.

Most of the patients have general symptoms of malaise, headache, fever, fatiguability, dizziness, transient visual disturbances, neck pain, palpitation, dyspnoea, arthralgia, stiffness of shoulders and nausea. Syncopal attacks are not uncommon. Haemoptysis occurs rarely.

Our patient also presented with low grade fever, fatigue, weakness, headache, syncope, chest pain etc. Major symptomatology of Takayasu's disease was consistent with other Japanese and Scandanavian co-workers.

Though the aetiology of TD remains unknown, an autoimmune mechanism may attribute to the disease process. Attempts to demonstrate circulating antibodies against antigens of the arterial wall, group-A streptococcal infection, association with tuberculosis, hormonal imbalance, ethnic susceptibility and genetic predisposition may be pathogenetic factor.

The lesions in Takayasu's disease show a panarteritis of the aorta and its main branches and of the pulmonary artery. The lesions of the arterial wall begin with a mesoperiarteritis with subsequent fibrosis and are followed by fibrotic thickening of the adventitia and the vasa vasorum. These lesions lead to an intimal fibrosis, which progresses usually in marked thickening often with thrombi. The destruction of the arterial wall leads to both stenotic and ectatic changes of the lumen, especially occlusion. These affected portions are clearly demarcated from the adjacent normal sites & segmental 'skipped' lesions.

Figure 1: Carotid doppler study shows narrowing in common carotid artery

Figure 2: Carotid doppler study showing narrowing in right common carotid artery

Figure 3: Doppler study- narrowing in left subclavian artery

Figure 4: Ultrasound scan of the internal carotid artery demonstrating marked thickening of the arterial walls
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are observed. In our patient carotid doppler study showed gross thickening of the arterial walls with irregular narrowing in the carotid arteries¹¹.

Shimatsuku & co-workers found irregular narrowing with gross thickening of arterial walls in 71.81% cases¹².

Cardiac complications are common associations of Takayasu's disease. It is due to involvement of coronary artery and its branches. Patients usually present with varying degree of angina with or without arrhythmias and heart failure. Our patient also has got chest pain where ECG revealed anterior wall ischaemia. This cardiac event was supported by Yokatima & co-workers in 2004¹³.

In a large series of 161 patients, the 20 years overall survival rate after the diagnosis was established in nearly 83 percent. Recently the prognosis of patients with Takayasu's disease has improved among the major factors causing death or severe disability are congestive heart failure, cerebro-vascular accidents and blindness¹⁴.

Conclusion

Takayasu's disease is not uncommon in Bangladesh. Due to its varying symptomatology and multi-system involvement nature, it may be diagnosed incidentally. Proper cardiovascular evaluation is mandatory for each & every patient, and must be taken as a routine clinical evaluation tool. Simply examination of pulse can draw the attention of the attending physician to the diagnostic clue of Takayasu's disease.

So, our message to relevant doctors i.e. general practitioners, cardiologists, cardiac surgeons, ophthalmologists & other specialists is: "when you feel the pulse of a Bangladeshi patient please do not forget the name of a Japanese disease- Takayasu's arteritis".

References

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