CASE REPORT

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An interesting case of Takayasu's arteritis

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ABSTRACT

Takayasu's arteritis is a chronic, inflammatory, granulomatous vasculitis, affecting predominantly large elastic arteries like the aorta and its main branches. We report a case of 29 year old female patient, who had newly detected hypertension and was ultimately diagnosed to have Takayasu arteritis based on clinical profile and angiography. Renovascular hypertension is very common in Indian population. However, there was no renal artery involvement in our patient which is an uncommon manifestation. The patient was treated successfully with immunosuppressants and blood pressure got reduced without the use of anti-hypertensives.

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CASE REPORT

A 29 year old female presented to the OP with a history of a severe headache on and off for 6 months. There was no fever, blurred vision, nausea, vomiting or any other history suggestive of raised intracranial pressure or sinusitis. There was no significant past medical history. The patient was married and had two children with uneventful pregnancies. On examination, the patient had a mild pallor. All the pulses were normal on the right side whereas all the pulses were weak on the left side. There was a bruit heard over the right carotid. BP was 180/80 mm Hg in the right upper limb, 190/90 mm Hg in the right lower limb and 180/80 mm Hg in left lower limb. BP was not recordable in the left upper limb. Fundus examination was normal.

Complete blood count and tests of renal function, liver function, and thyroid function were normal. CRP level was normal and ESR was 48 mm in the first hour. HIV, VDRL and APLA tests were negative. USG abdomen revealed normal sized kidneys. The echocardiogram was normal. Renal arterial Doppler was normal.

To rule out vasculitis as a cause of secondary hypertension, CT angiogram was done. CT angiography showed complete stenosis of origin of left common carotid and left subclavian artery with attenuated left internal and external carotid arteries. Intracranial arteries were normal. (Figure 1) There was involvement of descending thoracic aorta and abdominal aorta in the form of wall thickening. There was also ostial stenosis of the celiac artery and the superior mesenteric artery with collateral between the superior and inferior mesenteric artery (Figure 2). Bilateral renal arteries were normal (Figure 3). She was diagnosed to have Takayasu's arteritis by the American College of Rheumatology Classification Criteria for Takayasu's Arteritis. (Table 1) (Moriwaki R *et al.,* 1997) These features were

consistent with the diagnosis of Takayasu arteritis type V. (Table 2)

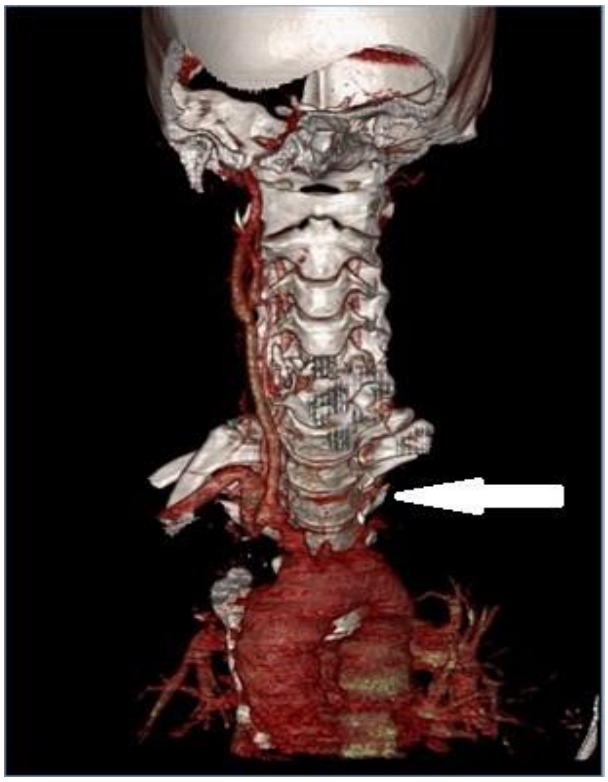

Figure 1: CT Angiogram of the arch of the aorta and carotid arteries (false colour picture with 3D reconstruction) showing complete stenosis of origin of left common carotid and left subclavian artery with attenuated left internal and external carotid arteries (white arrow)

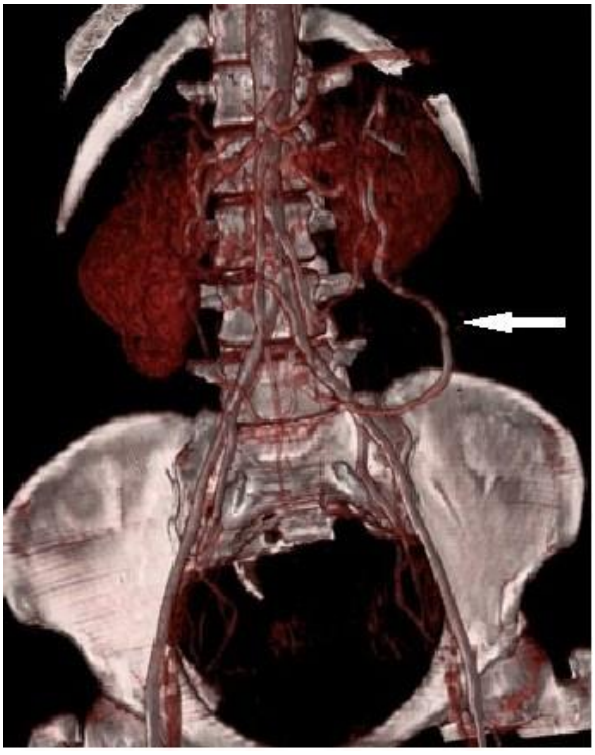

Figure 2: CT Angiogram of the abdominal aorta (false colour picture with 3D reconstruction) showing collateral between superior and inferior mesenteric arteries (white arrow)

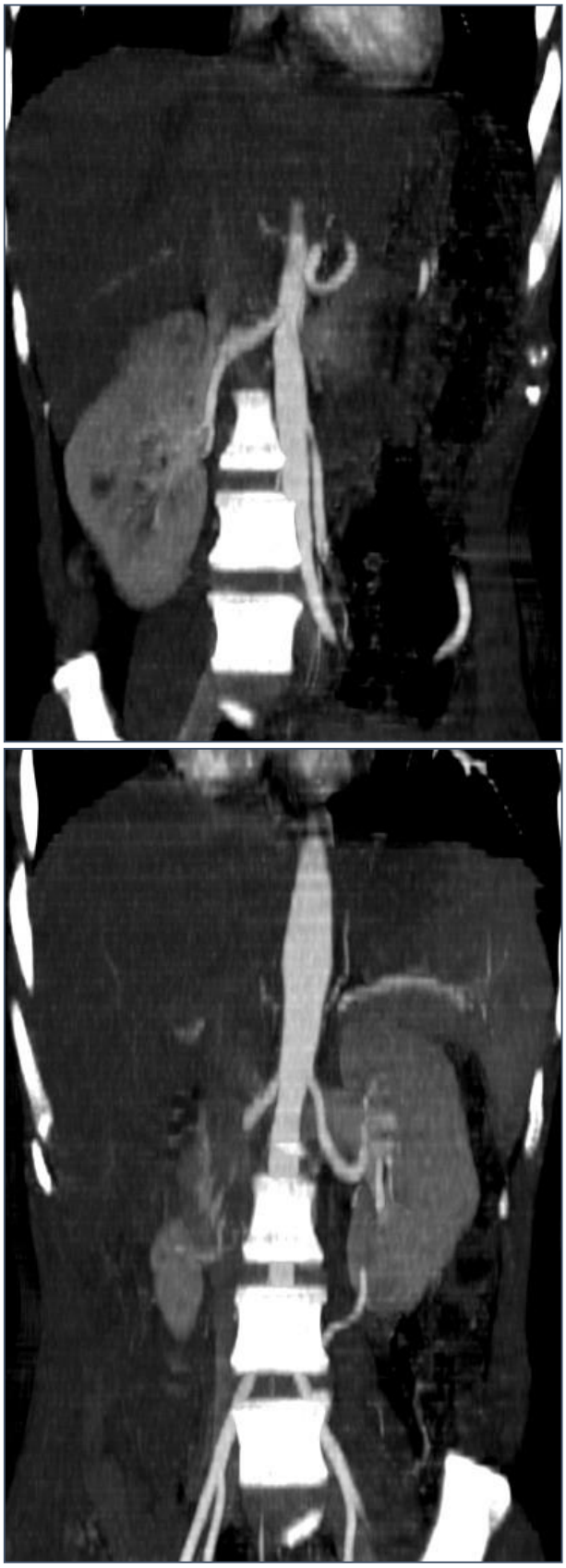

Figure 3: CT Angiogram of the renal arteries on the Right and Left sides

Table 1: American College of Rheumatology Classification Criteria for Takayasu's Arteritis (any 3 out of the 6 criteria)

The onset of the disease before the age of 40 years
Limb claudication
Decreased brachial artery pulse
Unequal arm blood pressure (> 10 mm Hg)
Subclavian or aortic bruit
Angiographic evidence of narrowing or occlusion of the aorta or its primary branches or large limb arteritis

Table 2: Angiographic classification of Takayasu's arteritis

Type	Vessel involvement
Type I	Branches from the aortic arch
Type IIa	Ascending aorta, aortic arch and its branches
Type IIb	Ascending aorta, aortic arch and its branches, descending thoracic aorta
Type III	Thoracic descending aorta, abdominal aorta, and/or renal arteries
Type IV	Abdominal aorta and/or renal arteries
Type V	Combined features of types IIb and IV

The patient has treated with Atorvastatin 10 mg, Aspirin 75 mg, Methotrexate 7.5 mg/week and Folic acid 5mg after consultation with the rheumatologist. Prednisolone 40 mg/day was initiated and titrated downwards over a period of 4 weeks.BP reduced and came to normal limits within one week without the use of anti-hypertensives (Lateef *et al.,* 2018).

DISCUSSION

Takayasu arteritis often referred to as "occlusive thromboaortopathy" or "pulseless disease", was first described in 1908 by Dr. Mikito Takayasu, a Japanese ophthalmologist in a 22 year old woman. (Terao C 2014) It affects women more commonly than men, with a ratio of 8:1. The median age of onset is around 25 years. Our patient was also a young female aged 29 years.

Clinical features results either from systemic inflammation or vascular insufficiency or both. (Park M-C *et al.,* 2005; Kerr GS *et al.,* 1994) Symptoms due to vascular insufficiency are asymmetric arm blood pressures (15%), carotidynia (20%), hypertension (20%), carotid bruit (20%), lightheadedness (20%), reduced or absent pulse (25%),), and claudication (35%). Common vessels involved are left subclavian artery (93%) and aorta (63%) as in our patient also. (Kerr GS *et al.,* 1994) Renal, carotid and vertebral arteries also can be affected. Lesions can be occlusion, stenosis, thrombosis or aneurysm formation in the affected vessels. Constitutional symptoms like fever, myalgia and weight loss can occur due to systemic inflammation. (Ishikawa K and Maetani S 1994). The diagnosis is made by American College of Rheumatology Classification Criteria for Takayasu's Arteritis (Figure 1 & Table 1) (American 1990). The presence of three or more of the six criteria is required for the diagnosis of the disease as in the present case.

Renal artery involvement can lead to hypertension or renal insufficiency. (Jain S *et al.,* 1996). It is an important cause of renovascular hypertension by causing renal artery stenosis. Our patient did not have renal arterial involvement in angiogram and renal function tests were normal throughout the illness. In Takayasu arteritis, hypertension can result from atypical coarctation of the aorta, loss of vascular compliance, aortic insufficiency, or renal artery stenosis. (Jain S *et al.,* 1996) In our patient, none of these was found, but the patient had hypertension. The cause of hypertension in our patient could be due to vasculitis. Renal arterial compliance was not measured separately but renal artery Doppler was normal. The patient fitted into Type 5 of angiographic classification of Takayasu's arteritis. She unusually did not have renal artery involvement which is commonly seen in 20 – 90% of Indian patients with the disease (Jain S *et al.,* 1996).

Treatment was started with oral Prednisolone 40 mg per day along with Methotrexate 7.5 mg per week along with atorvastatin and folic acid. The patient remained as an inpatient under constant monitoring. BP started to fall on the fourth day and within a week's time, BP returned to baseline. Over a period of six weeks, the patient became symptom free and BP remained under control without the need for additional medications. Follow up investigations showed no progression of disease activity and ESR returned to normal. Prednisolone was tapered and stopped over 3 weeks. Currently patient is on Methotrexate 2.5 mg per week, atorvastatin and folic acid. This case signifies an uncommon cause of secondary hypertension which was treated successfully without the use of anti-hypertensives.

CONCLUSION

Our patient was a young female who was diagnosed to have hypertension due to Takayasu's arteritis. Even though she was an Indian, she did not have renal artery involvement which is a rare presentation. Her BP came down without any antihypertensive drugs which was unique in our case.

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