# Intracranial Vessel Affection in Takayasu Arteritis- Exception, or Rule

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**ABSTRACT**

**Introduction:** Takayasu Arteritis is a large vessel vasculitis affecting mostly young women. It was thought to affect extracranial arteries only resulting in stenosis and / or aneurysm but recently it has been found to affect intracranial arteries too.

**Case report:** We present a case of a young lady who was diagnosed as Takayasu Arteritis, had high inflammatory parameters and Interleukin-6 levels and was treated with Tocilizumab. She subsequently developed central nervous system symptoms and imaging studies were revealed affection of intracranial arteries. Remarkably, her symptoms developed after the inflammation was well controlled with therapy.

**Conclusion:** We conclude that intracranial affection may not be uncommon even in patients without stroke or hypertension, but are usually missed in absence of imaging studies of intracranial vessels. We also propose more studies to elucidate if intracranial affection is associated with high Interleukin-6 levels.

**Keywords:** Inflammatory Parameters, Intracranial Affection, Interleukin-6, Takayasu Arteritis, Tocilizumab

## INTRODUCTION

Takayasu Arteritis (TA) is an inflammatory disease of unknown etiology characterized by granulomatous vasculitis affecting the aorta, its main branches, and the pulmonary arteries, and occurs most often in women of childbearing age. Inflammation of the vessels may result in stenosis or in aneurysm formation. Central Nervous System (CNS) symptoms like transient ischemic attacks, stroke, dizziness, syncope, headache, or visual changes are thought to be due to occlusion in the extra cranial parts of the carotid and vertebral arteritis. We present a case where a patient of Takayasu arteritis had symptoms due to affection of intracranial arteritis.

## CASE

Ms X, a 19 year girl was already diagnosed as a case of TA by the department of cardiology based on the findings of asymmetrical pulses, and proved by computed tomogram (CT) angiogram which showed focal dilatation and narrowing with asymmetric circumferential intimal thickening of the arch of aorta, fusiform aneurysm of brachiocephalic artery, asymmetric circumferential intimal thickening and wall irregularity of descending and the abdominal aorta, and dilatation of the celiac axis. The right common carotid artery (CCA) showed circumferential intimal thickening and narrowing for 3.5 cm. The left subclavian artery (SCA) was had approximately 50% narrowing along its whole course [Figure 1]. She was planned for stent insertion, prior to which she was referred to our department in view of carotidynia. Her evaluation revealed Haemoglobin (Hb) of 9.2 g/dL, Erythrocyte Sedimentation Rate (ESR) of 120 mm in 1st hour by westergren's method, C-reactive protein (CRP) of 105.66 mg/dL (normal < 5 mg/dL). Her leucocyte counts, differential counts, platelet counts, hepatic and renal function tests were within normal range. She tested negative for Hepatitis B and C, and HIV. However, her Interleukin-6 (IL-6) levels were raised at 13.70 pg/mL with normal values being less than 6 pg/ml. Considering the clinical and laboratory parameters of high disease activity, she was put on prednisolone 1mg/Kg bodyweight, and after negative latent tuberculosis infection (LTBI) screening, Injection Tocilizumab 162 mg by pre-filled syringe (PFS) every two weeks. After 6 weeks, her ESR was 36 mm in 1st hour, and CRP was 0.41 mg/dL. She was again planned to be taken up for stenting.

However, a week prior to stenting, she developed recurrent attacks of vomiting, and throbbing pulsatile headache with photophobia and phonophobia, but without aura, fortification spectrum, or meningeal signs for which she presented to the Emergency. She had to be administered morphine to control the pain. The symptoms recurred again after another 3 days and a Rheumatology call was given. Migraine, high disease activity of TA, and intracranial vasospasm were kept as the differential diagnoses. Subsequent evaluation revealed no new loss of pulses, normal ESR and CRP, and normal scans of the brain. She was advised contrast enhanced MRI (CEMRI) and MR Angiogram (MRA). MRI of the brain revealed serpiginous FLAIR hyperintense structures along sulci of both cerebral convexities suggestive of leptomeningeal collaterals without any evidence of bleed or infarct [Figure 2]. MR Angiography of Circle of Willis showed irregularity and narrowing of distal petrous, supraclinoid, segments of right internal carotid arteries (ICA) with 50% stenosis, irregularity of the horizontal segments (M1) of both middle

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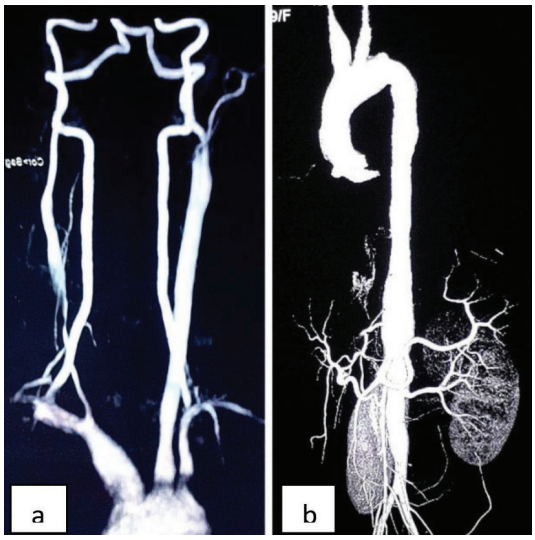


**Figure-1:** CT Angiogram of Aorta showing fusiform aneurysm of the brachiocephalic artery and proximal right subclavian artery and severe stenosis of the proximal right common carotid artery (a), and variable narrowing of the left subclavian artery, thoracic and abdominal aorta (b).

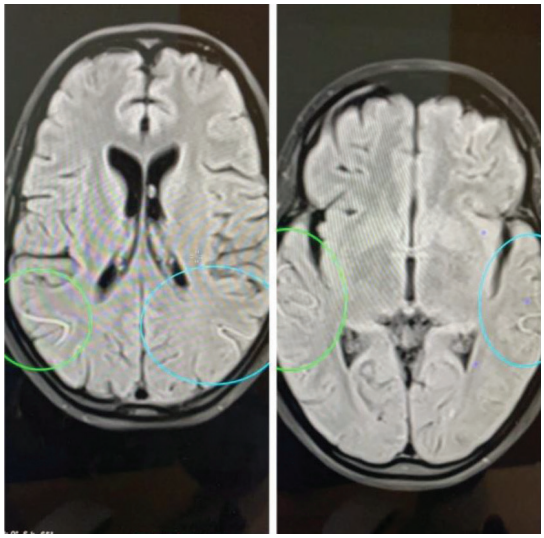

**Figure-2:** FLAIR axial image of the brain reveals multiple leptomeningeal collaterals seen as sulcal hyperintensity(circled in blue and green).

cerebral arteries, and irregular and attenuated caliber of proximal aspect of A1 segment of the left anterior cerebral artery with 60% stenosis [Figure 3]. MR Angiogram of the neck revealed asymmetrical thickening of the aortic arch with mild dilatation, fusiform dilatation of the brachiocephalic and right subclavian artery, and circumferential thickening of the right common carotid artery from origin till 44 mm causing >90% luminal stenosis, and short segment asymmetrical circumferential thickening of the wall of the proximal left subclavian artery. Stenting of the carotid artery was carried out. She has not reported any recurrence of symptoms.

## DISCUSSION

TA is diagnosed incidentally in a fifth of cases, with the rest four-fifths having systemic or vascular symptoms with the

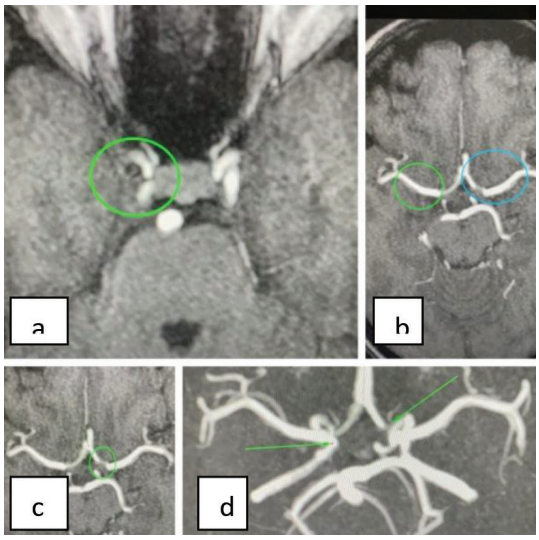


**Figure-3:** Magnetic resonance angiography of the intracranial vessels showing variable narrowing of the cavernous segment of right ICA (a), A1 segment of left ACA (b) and horizontal (M1) segments of both MCAs (c & d)

later being either due to active vasculitis or damage due to prior disease including dissection[1] and stenosis[2].Thought to be mostly extracranial, affection of carotids or vertebral arteries may have intracranial manifestations like transient ischemic attacks, stroke, dizziness, syncope, headache, visual changes, and even epilepsy [2, 3].

Direct involvement of intracranial arteries was first demonstrated in 1984 by Molnar P and Hegedus K, who showed intense inflammation with lymphocytes and plasma cells in vessel walls of all major intracranial arteries in the pathological examination of a 20 year old man who had presented with headache and rapidly evolving spastic quadriparesis [4].

A study by Bond KM, Nasr D, et al found intracranial affection in 7.8% of patients, with aneurysms in 3.9% of patients and occlusions in 3.9% of patients, while 7.6% had intracranial vasculitis and 1.3% having reversible vasoconstriction[5]. Another study found intracranial affection in 20% of patients[6]. Thus intracranial TA may be more common than is thought to be. Patients of intracranial TA are thought to be older, have stroke and/or hypertension, coronary artery lesions, and extracranial affection of ICA[7].

Our case is that of a young lady who likely had TA for quite some time as evidenced by presence of intracranial collaterals. Intracranial radioimaging showed irregularity of vessel walls, stenosis, as well as aneurysm, with affection of both the anterior and middle cerebral arteries and the Circle of Willis. Extracranial affection in our patient has been described previously. It is interesting to note that the migraine like symptoms of vomiting, and throbbing pulsatile headache with photophobia and phonophobia occurred when the disease activity of TA was already under control as evidenced by normal values of ESR and CRP. Apart from the young age, other aspects of our case which needs highlighting is the high levels of IL-6 which is associated with high disease activity of TA and monitoring of which

can help in making decisions about treatment modalities[8] and normalization of ESR and CRP after 3 doses of IL-6 antagonist Tocilizumab, which is an accepted modality of treatment[9]. In contrast to the study by Guo Y et al, our patient was younger and did not have stroke or hypertension.

Thus we conclude that intracranial TA is not uncommon but can present with innocuous symptoms like migraine and has to be thought of by the clinician even in a young patient. High disease activity in TA may have devastating consequences and this underlines the need for early detection and treatment. Further studies will be required to explore if high levels of IL-6 are associated with intracranial affection in TA.

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