

Defying the Standard: Tackling Refractory Takayasu Arteritis with Novel Approach.

Dr Kausalyaa Radha¹, Dr Thaneswari Tharumalingam¹, Dr Suhaida Ahmad Maulana¹,
Dr Eashwary Mageswaran¹, Dr Emilia Illa²

1. Department of Rheumatology, Hospital Tengku Ampuan Rahimah, Klang.

2. Department of Radiology, Hospital Tengku Ampuan Rahimah, Klang.

INTRODUCTION:

Large vessel vasculitis, such as Takayasu arteritis, is a rare but serious inflammatory disease affecting the aorta and its major branches. Standard immunosuppressive therapy may be insufficient in refractory cases, necessitating alternative treatments.

CASE SERIES:

1. 16-year-old Burmese male presented with loose stools, blood pressure discrepancy in the upper limbs, and a feeble left radial pulse.

Lab results

ESR 89 mm/hr, CRP 74 mg/dL, negative infectious and autoimmune screens, including ANCA.

CT angiography (Figure 1a,b).

He was treated with IV Methylprednisolone, tapering Prednisolone, and 8 cycles of IV Cyclophosphamide. Despite this, patient remained symptomatic with unrecordable upper limb blood pressures and a carotid bruit. Inflammatory markers remained elevated. The patient was subsequently switched to IV Tocilizumab, resulting in normalization of inflammatory markers, resolution of blood pressure discrepancies, palpable carotid pulses without bruits and improved arterial findings on repeat CT angiography (Figure 2a,b).

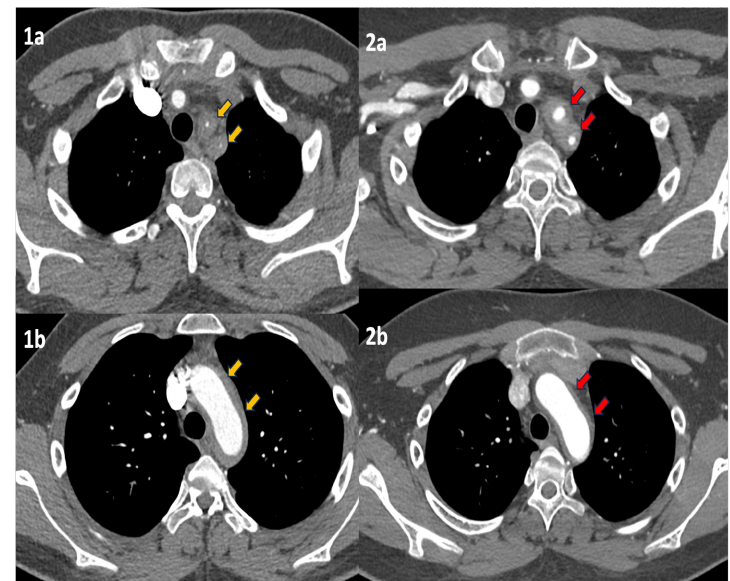


Figure 1a and 1b- shows diffuse wall thickening of proximal left Common Carotid and proximal left Subclavian Artery causing near total luminal occlusion (1a) diffuse thickening of Aortic Arch (1b).

Figure 2a and 2b- CT (13-months interval) shows reduced severity of arterial wall thickening (2a, 2b, red arrows) indicating treatment response.

2. A 29-year-old Malay man presented with right lower limb cellulitis following a motor vehicle accident, presented with unequal blood pressures in both upper and lower limbs, with diminished and absent pulses.

Lab results:

ESR 83 mm/hr, CRP 36 mg/dL, negative autoimmune screen including ANCA and APLS.

CT angiography (Figure 3a,b)

He was treated with IV Methylprednisolone, tapering Prednisolone, and 6 cycles of IV Cyclophosphamide, with initial improvement.

However, the disease relapsed on Methotrexate, with absent bilateral Dorsalis Pedis pulses and raised inflammatory markers. Switching to IV Tocilizumab led to marked clinical and biochemical improvement after four cycles. His care was later transferred for logistical reasons.

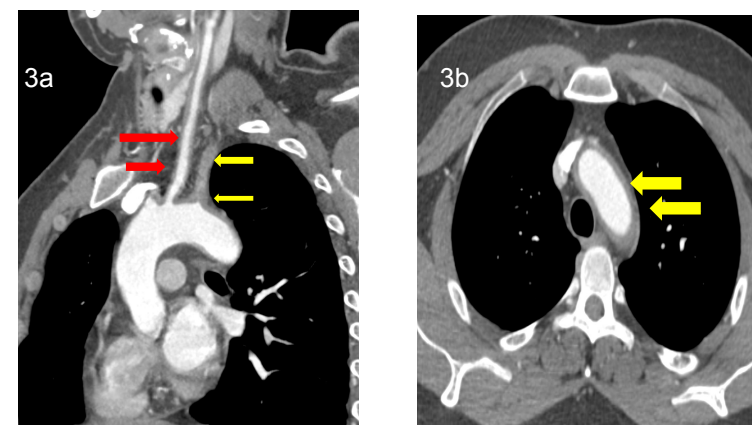


Figure 3a- Diffused thickening and enhanced left common carotid arterial wall (red arrow), thickened wall with severe luminal stenosis of the left subclavian artery (yellow arrow).

Figure 3b- Diffused aortic wall thickening (yellow arrow).

CONCLUSION

Both patients with refractory large vessel vasculitis failed to respond to high-dose glucocorticoids and Cyclophosphamide. Introduction of Tocilizumab, an Interleukin-6 receptor antagonist, led to normalization of inflammatory markers, improved vascular imaging, and clinical remission, highlighting its efficacy for young patients with rapidly progressive, refractory disease.