

NO PULSE: A MEDICAL CONUNDRUM

Inês Pires Silva*, Carla Noronha**, António Panarra**, Nuno Riso**, Manuel Vaz Riscado**

Takayasu's arteritis (TA) is a chronic inflammatory disease that affects large vessels, predominantly the aorta and main branches, leading to vessel wall thickening, fibrosis, stenosis, aneurysms and thrombus formation¹.

TA presents a biphasic course: firstly, an inflammatory phase; and a later one, where vascular occlusion mainly occurs².

Diagnosis of TA is always challenging as the clinical presentation varies, biopsy of large-vessel lesions (gold standard) is rarely performed and because there are no serological or imaging specific diagnostic tests.

The treatment differs according to the phase of the disease. In the inflammatory phase the treatment is mainly immunosuppressive; however, in the latter, vaso-occlusive phase, surgical treatment is needed.

The authors describe the case of a 40 year-old caucasian woman referred to our Autoimmune Clinics due to a nine months duration of dizziness with episodic syncope, occipital headache, right cervicodinia and recurrent amaurosis fugax. Bilateral calf and right upper limb claudication, Raynaud's phenomenon, polyarthralgia, diffuse alopecia, oral ulcers, anorexia and weight loss of fifteen kilograms in two years were also reported. Her past history included: heavy smoking habits (28 pack-year); lumbar vertebral fracture surgery; G5P5A2 (2 first trimester spontaneous abortions) and intake of oral contraceptives. Physical examination was remarkable for a good general health status, but with anisophymia (BP RA: 60/30 mmHg; BP LA: 100/60 mmHg), asymmetry in arterial pulses (especially on the right) and bilateral carotid bruit (stronger on the left side).

Some diagnostic tests were performed: a) Lab workup (CBC, ESR, CRP, coagulation times, biochemistry profile, immunological profile, urinary-

sis), EKG, chest X-ray, head CT and ophtalmological exam were normal. b) nailfold capillaroscopy re-

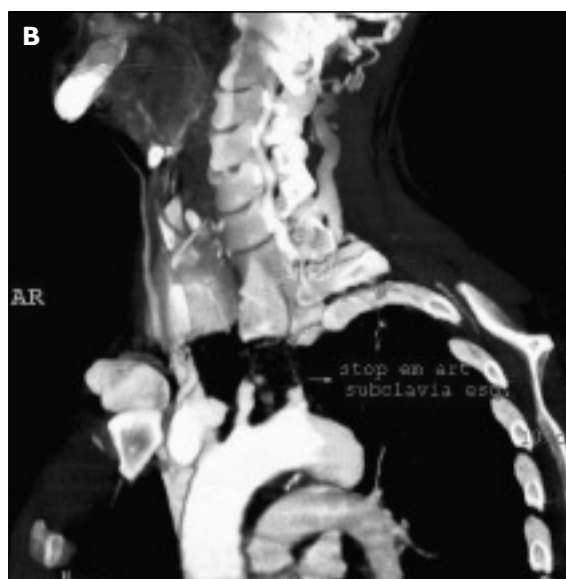
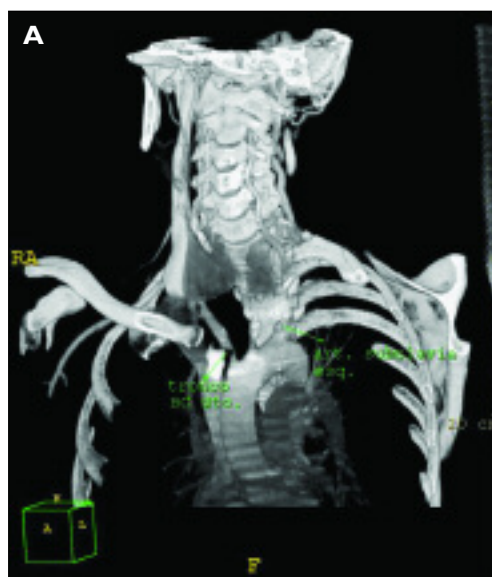


Figure 1a and 1b. Cervico-Thoracic revealing a complete occlusion of the left subclavian artery and a marked stenosis of the right brachiocephalic trunk

*Serviço de Oncologia Médica, Instituto Português de Oncologia de Lisboa Francisco Gentil (IPOLFG), Portugal, Programme for Advanced Medical Education

**Unidade de Doenças Auto-imunes, Serviço de Medicina 2, Hospital Curry Cabral, Lisboa, Portugal

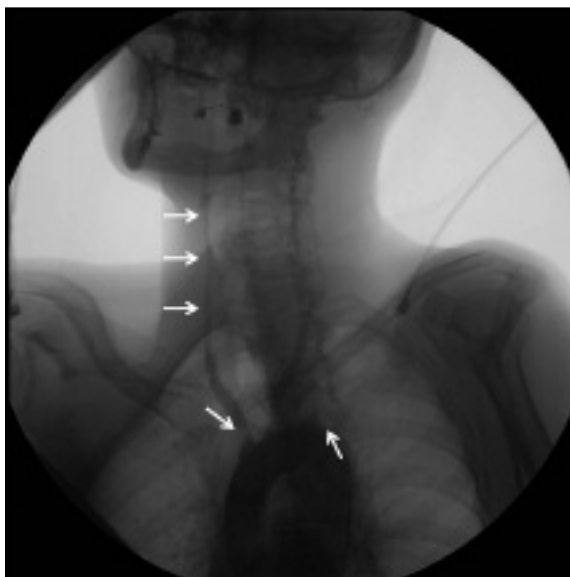


Figure 2. Supra-aortic Angiography confirming a complete occlusion of the left subclavian artery and a marked stenosis of the right brachiocephalic trunk



Figure 3. Lower limb Angiography revealing a stenosis of the right primitive iliac artery

vealed a secondary Raynaud's phenomenon; c) echocardiogram showed mitral regurgitation; c) 24h-Ambulatory Blood Pressure measurement documented a hypotensive profile; d) Arterial cervical doppler ultrasound revealed low peak systolic velocity and pulsability, marked high diastolic velocity in the brachiocephalic trunk, in bilateral carotid axes and the left subclavian artery; e) Cervico-thoracic angio-CT (Figures 1a and 1b) complemented by cervico-thoracic-abdominal angiography (Figures 2 and 3) revealed occlusion of the left subclavian artery, concentric stenosis of brachiocephalic trunk and mild stenosis of carotid axis and primitive right iliac artery.

A strong suspicion diagnosis of TA³ in the occlusive stage was established. A multidisciplinary treatment was adopted: smoking cessation, double antiplatelet therapy, statins, trimetazidine and endovascular surgery (stenting and brachiocephalic trunk and right primitive iliac angioplasty); immunosuppressive treatment was deferred due to surgical approach.

However, three months later, she complained of dysphonia, cough, upper dyspnea and bilateral cervical enlargement consistent with firm, painful polyadenopathy. An infiltrative and vegetant supraglottic mass was identified and consistent with a moderately differentiated squamous cell carcinoma of the larynx (T3N3M0). She was sub-

mitted to total laryngectomy, followed by radiotherapy. Presently, she is in oncological remission.

This case is particularly challenging because of the two concomitant diagnoses: TA and larynx neoplasia. TA was presumed due to clinico-radiological presentation, as vascular biopsy was not possible. The association to malignancy (Meig's Syndrome, Leukemias and oropharynx carcinoma)⁴⁻⁷ has been reported in literature, though still unclear.

Acknowledgments

The Programme for Advanced Medical Education is sponsored by Fundação Calouste Gulbenkian, Fundação Champalimaud, Ministério da Saúde e Fundação para a Ciência e Tecnologia, Portugal.

Correspondence to

Inês Pires da Silva
Rua Manuel Marques, nº 10, 9º B.
1750-171 Lisboa, Portugal
E-mail: inespda@gmail.com

References

1. Nishimoto N, Nakahara H, Yoshio-Hoshino N, Mima T. Successful treatment of a patient with Takayasu arteritis using a humanized anti-interleukin-6 receptor antibody. *Arthritis Rheum* 2008; 58: 1197-200.
2. Rodriguez-Orsorio X, Blanco M, Arias S, Castillo J. Biomarcadores y tomografía de emisión de positrones en la valoración de la actividad de la arteritis de takayasu. *Neurologia* 2008; 23: 329-332.
3. Arend WP, Michel BA, Bloch DA, et al: The American

- College of Rheumatology 1990 criteria for the classification of Takayasu's arthritis. *Arthritis Rheum* 1990; 33: 1129-1134.
4. Salman MC, Basaran A, Guler T, et al. Meigs' syndrome with highly elevated CA125 levels in a patient with Takayasu arteritis: a case report. *Arch Gynecol Obstet* 2005; 272:90-92.
 5. B'Chir Hamzaoui S, Abdallah M, Baili L, et al. Takayasu's arteritis associated with acute myeloblastic leukaemia. *J Mal Vasc* 2006; 31: 280-283.
 6. Amberger C, Denzlinger C, Janzen J, Müller-Schimpfle M, Mohren M, Kötter I. Takayasu's arteritis secondary to myelodysplasia as a predictor of poor outcome: two case reports. *Clin Exp Rheumatol* 2004; 22: 346-348.
 7. Kavanagh BD, Brizel DM, Leopold KA, Acker JC. Radiation Therapy for Head and Neck Cancer in a Patient with Takayasu's Arteritis. *Acta Oncologica* 1994; 33: 73-74.
-

XVII Jornadas Internacionais do Instituto Português de Reumatologia

**Lisboa, Portugal
9 a 10 de Dezembro 2010**