# Takayasu arteritis: showed by a hypertensive crisis

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## INTRODUCTION

Takayasu Arteritis (TA) is a primary granulomatous large vessel vasculitis, affecting predominantly young women[1]. It mainly affects the aorta and its major branches. TA and the required immunosuppressive therapy cause considerable morbidity and mortality[2]. Early in the disease course, symptoms can be non-specific, leading to a difficult diagnosis[3]. Consequences of vascular stenosis, occlusions, and, less commonly, vascular dilation, account for the typical clinical presentation; the latter can sometimes lead to aneurysmal rupture or dissection. Though the importance of a comprehensive history and a thorough physical examination cannot be over-emphasized, clinical assessment is frequently inaccurate when evaluating disease activity, which may sometimes progress silently. Also the diagnostic modalities currently used are unsatisfactory. X-Ray angiography, the "gold standard" for TA diagnosis, can image luminal defects but does not detect changes of the vessel wall[4]. The diagnosis of TA can be confirmed with histopathological examination; however, tissue from blood vessels is obtained only in the minority of patients requiring a surgical intervention. There are currently no specific biomarkers for diagnosing TA. Identifying disease activity in TA is challenging[3].

Glucocorticosteroids are anchor drugs for this disease, like other vasculitis. Most cases in Japan respond with 0.3–0.5 mg/kg/day prednisolone, but we frequently found that some patients revealed flare-ups during tapering of glucocorticosteroids. Since TAK mainly affects young women, side-effects of glucocorticosteroids, especially moon face, severely damage their quality of life. Immunosuppressive agents, including methotrexate, cyclosporine, cyclophosphamide, mycophenolate mofetil and TACROLIMUS have been used for patients with TAK. Biological agents targeting tumor necrosis factor (TNF) have also been used for patients with TAK. Since IL-6 is highly expressed within inflamed arteries and serum levels correlate with disease activity, blocking IL-6 showed effectiveness in TA. Tocilizumab is a humanized monoclonal antibody against the IL-6 receptor, and the first report of successful use of tocilizumab in a patient with refractory TA was published in 2008. Later, nine additional cases of TA treated with tocilizumab 8 mg/kg every 4 weeks were reported. In the majority of the cases, disease activity improved and CS doses were discontinued or tapered. Abatacept is another promising biologic agent inhibiting the co-stimulation of T cells, and is currently being investigated in the first randomized, placebo-controlled trial of LVV patients including TA[7].

This article aims the description of a clinical case of a serious appearance form of an uncommon disease, Takayasu's arteritis.

**Keywords:** Takayasu Arteritis, Large vessel vasculitis, renal artery stenosis

**Palabras clave:** Arteritis de Takayasu. Vasculitis de grandes vasos. Estenosis de la arteria renal

## CLINICAL CASE

Female, 61 years old, caucasian with personal background of a Takayasu's arteritis (TA) without medication and without medical follow-up, Hypertension and Depressive syndrome, presented to the emergency department (ED) with fever, dyspnea with polypnea, productive cough with severe hypoxemia. The physical examination showed a pulmonary auscultation with loud crackles in both lung bases. Laboratory tests showed a marked increase of inflammatory parameters. Thorax X-Ray confirms evident condensations compatible with bilateral pneumonia. Patient was admitted with a diagnosis of Community Acquired Pneumonia. On the 2nd day at the ED, the patient had an acute lung edema associated with hypertensive crisis with respiratory failure and was transferred to the Intensive Care Unit. He was extubated after eight days and accomplished empirical antibiotic Clarithromycin and Ceftriaxone treatment. After eight days and after stabilization, patient was then transferred to the Internal Medicine Service.

Based on a long-term fever, a suspicion of relapsing Takayasu's arteritis was registered.

Inpatient made a CT angiography, examination towards the systemic arteries from the base of the skull to the distal portion of the lower limbs, showing the following aspects:

- Unchanged pulmonary arteries;
- In the circle of Willis there's an occlusion of the posterior communicating artery, the anterior cerebral artery and the right anterior communicating artery;
- Examination to the supra-aortic trunks shows the absence of significant changes in the permeability of the right carotid and right vertebral arteries. On the left there's an early carotid stenosis, chronic occlusion of the left subclavian artery, with this artery permeabilized with stolen blood by a vertebral artery. There's evident occlusion of the carotid artery along its entire length. The other visible portions of the left vertebral artery and carotid artery have a tapered diameter tapered in a general way;
- No changes to the diameter or the permeability of the thoracic and abdominal aorta. However, there are mural thrombi and extensive calcifications;
- In the distal branches of the abdominal aorta stands a stenosis of the right renal artery, which is uneven across its path and with a more pronounced stenotic segment with hemodynamic meaning. Consistent to that, there's a diffuse decrease in the thickness of the parenchyma, compared to the one observed on the left.
- Celiac trunk with stenosis but the irrigation is assured by the superior mesenteric artery. In the inferior mesenteric artery there's evident sub-occlusive stenosis of the proximal portion.

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- At the arteries of the lower limbs can be observed a diffuse stenosis of primitives and external iliac arteries, as well as in the common and superficial femoral arteries;
- Popliteal Arteries and infra-popliteal trunks without any modifications observed.

Pharmacological stent was implanted in the renal artery, whose stenosis probably caused the hypertensive crisis and consequently the Lung edema. The patient starts GC therapy during the internment with GC bolus of 250mg before oral steroids. Inpatient started double platelet anti-aggregation therapy.

The internment took place without further complications and patient was discharged with an indication of smoking cessation and steroid prednisolone 30mg to 10mg at breakfast and dinner every day, anti-hypertensives, platelet anti-aggregation and statin therapy. Patient started to be followed-up at regular autoimmune medical consultations and in the Imaging follow-up study: "Regular caliber of the remaining arterial structures, without any evidences of significant ectasia and/or stenosis. There weren't any abnormal vascular structures identified. A stent is evident at the right renal artery, which remains permeable".

With regard to the admission, recent exams noticed some improvements in stenosis and occlusions.

## DISCUSSION

Takayasu arteritis (TA) is a large vessel vasculitis (LVV) characterized by granulomatous inflammation of the vessel wall with an unknown etiopathogenesis, often resistant to treatment and associated with high morbidity and mortality[6]. TA predominantly affects young females during the second or third decades of life and mainly involves the aortic arch and its primary branches, ascending aorta, thoracic descending aorta and abdominal aorta[4]. Although there is considerable variability in disease expression, the initial vascular lesions frequently occur in the left middle or proximal subclavian artery[4].

Is one of the 2 main causes of large vessel vasculitis, giant cell arteritis being the other[2]. The pathogenesis of Takayasu's arteritis is poorly understood.

Vascular symptoms are rare at presentation, but evidence of vascular involvement and insufficiency becomes clinically apparent as the disease progresses due to dilation, narrowing, or occlusion of the proximal or distal branches of the aorta. 1 Clinical manifestations of TA are quite variable, ranging from tissue ischemia due to vascular stenosis and occlusion, and to aneurysm formation that may occasionally rupture or dissect. Systemic symptoms (fever, malaise, weight loss, night sweats, polyarthralgia or arthritis) may predominate at the onset of TA[2].

In addition to a careful history and physical examination, diagnosis of TA should be based on laboratory studies (acute phase reactants), and imaging studies. Rarely, histology of the resected vessel is also available, and may help establishing the diagnosis and assess its severity[7].

In most cases the diagnosis is based upon suggestive clinical features and imaging of the arterial tree by MRI, CT, or angiography that demonstrates smoothly tapered luminal narrowing or occlusion that is accompanied by thickening of the wall of the vessel that is best demonstrated by CT or MRI.

Contrast arteriography may be preferred to CT or MRI for diagnosis if there is life- or limb-threatening ischemia for which immediate revascularization is anticipated since this may be accomplished by angioplasty and/or stenting of the affected vessel or vessels.

Smoking in the context of an inflammatory vascular disease is strongly discouraged. Patients should be repeatedly reminded of this, and referred for appropriate counseling if necessary. Diet is important for bone health and cardiovascular health in general, and also for maintaining a healthy body weight while on glucocorticoids, but it has not been shown to have a clear role in affecting the disease course as such[2].

Low salt intake, calcium and vitamin D supplementation and regular exercise are essential to reduce the metabolic side effects of CS agents. Monitoring and control of blood pressure may be difficult in cases with absent or reduced pulses in some extremities. Blood pressure measurements should be made in the unaffected extremities[7].

Corticosteroids have been the mainstay of therapy for active TA[8].

The response to high dose prednisolone is generally favorable, but relapses may occur while gradually tapering the dose and adverse effects of long-term treatment can cause problems. Therefore, many physicians tend to start conventional IS agents together with the initial CS treatment or while tapering the CS dose. Since MTX is an inexpensive, easily available and relatively safe agent that is widely used in rheumatology, it is the first choice of many physicians[7].

Surgical treatment, which should preferably be avoided, is also needed at times based on specific circumstances. Results of surgery are often unsatisfactory if TA is active preoperatively. When TA is felt to be in remission, the short term results of revascularization procedures may be acceptable, but relapses occur quite often when patients are followed over several years. It is imperative that better biomarkers and better imaging modalities be devised for accurate evaluation of ongoing vascular inflammation, so that the effect of therapy can be precisely quantified. A more detailed understanding of the pathogenesis of TA is likely to provide new targets for therapy[2].

Takayasu's arteritis is a rare and complex disease that can complicate and eminently end fatally. Therefore, patients with Takayasu´s arteritis must be followed up carefully, with a regular follow-up imaging, not allowing medication errors[2].

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