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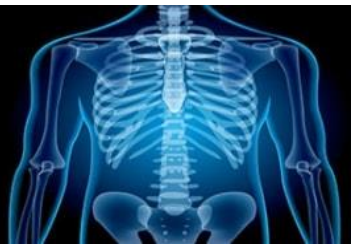

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A rare imaging case: Takayasu arteritis

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Abstract
Takayasu arteritis represents a rare, granulomatous inflammation of main arterial branches. Due to its low prevalence and polymorphic clinical aspects, often times it's difficult to make a diagnosis, requiring many investigations to be performed, with their associated costs. We present the case of a young-woman having the classic aspects reported in literature, however establishing the final diagnosis was tricky and it implied a range of multidisciplinary specialists.

Keywords: Large vessel, Takayasu, arteritis, Vasculitis

Introduction
Takayasu arteritis is a rare chronic granulomatous inflammation of large vessels that typically affects the aorta and its major branches [1]. Although imaging findings have a high sensitivity for depicting the disease through the observation of large vessel wall abnormalities, Takayasu arteritis is uncommon. Common imaging findings include stenosis, followed by occlusion, aneurysms, and in chronic cases evidence of increased collateral circulation [2].

Case report
We illustrate the case of a 23-year-old woman who presents to our hospital for prolonged fever, malaise, and dry cough for two months. She has a personal history of recurrent respiratory and urogenital infections. The physical exam revealed altered general condition, suffering facies, pale skin, poorly represented adipose connective tissue, right submandibular adenopathy, and painful axillary adenopathy on palpation. The laboratory results revealed microcytic hypochromic anemia, thrombocytosis, hyposideremia, hepatocytolysis syndrome, and non-specific biological inflammatory syndrome.We performed an echocardiogram, gastroscopy, colonoscopy, and non-contrast computer-tomography (CT) of the thorax, abdomen, and pelvis all of them with no pathological findings. Blood pressure, electrocardiogram were in normal range. Her urocultures, hemocultures, and sputum were negative with no trace of an infectious agent. Her anti-nuclear antibodies and rheumatoid factor were negative. She received symptomatic treatment with iron supplements, anti-fungic, and antibiotic therapy however her symptoms worsened and a month later she returns accusing of prolonged fever, cough and weight loss, a markedly pronounced biological inflammatory syndrome raised C-reactive proteine of 214,98 mg/L (normal range 0-5 mg/L), Fibrinogen 807 mg/dL (normal range 170-420 mg/dL) and erythrocite sedimentation rate >120 mm/h (normal range 0-20 mm/h) and positive fibrin monomers. On her non-contrast head CT scan, we found a left temporal arachnoidian cyst 35 mm in diameter, with no other pathological findings.
The cardiologist examines her again to exclude pulmonary thromboembolism and a CT angiography is ordered revealing no signs of pulmonary embolism, but a diffuse thickening of soft tissue around carotid and subclavian vessels with luminal narrowing in this regions (Figure 1. – A, B, C) and 3D VRT reconstruction images (Figure 2. – D, E) demonstrate imaging aspects in accordance with Takayasu Arteritis.
She received corticosteroids, antibiotics and antifungic therapy with the recommendation to report any adverse reaction and recall after 3 months. Patient comes back and now is doing well, her symptomes and biological inflammatory syndrome vanished.

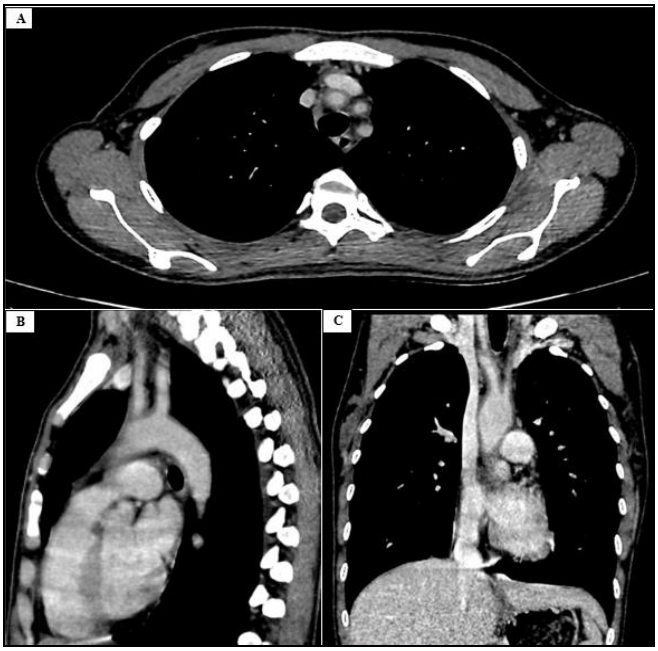

Fig 1: Contrast-enhanced CT scan axial (A), sagittal (B) and coronal (C) images markedly narrowing of the carotid and subclavian vessels lumen with dense perivascular tissue

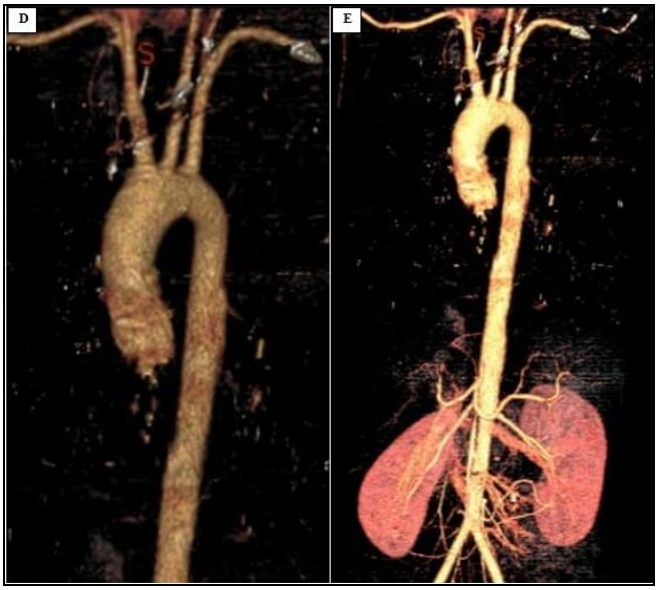

Fig 2: (D, E) - 3D VRT Reconstruction images reveal luminal narrowing of cervical vessels; Aorta and it's abdominal vessels having a normal course and anatomy

Discussion

Takayasu Arteritis (TA) is a granulomatous vasculitis of large vessels, usually occurring in patients younger than 50 years old, previously known as aortic arch syndrome or pulseless disease [2]. The disease was first described in 1908 after Mikito Takayasu after he saw multiple retinal vascular malformations in a patient [2].

Although the disease has a worldwide distribution, it is generally thought to be more common among Asian populations, with an incidence in Japan of 1-2 per million [3]. Epidemiologic studies suggest that TA is being increasingly reported in Europe with an incidence up to 1.5 per million [3]. Although the pathophysiological aspects are not well understood, the association of the gene with the HLA complex supports the genetic contribution to disease pathogenesis, the strongest causality involves HLA-B52 in Japan, patients in this category seem to carry a worse prognosis [2]. Patients from Mexico have also been found to have associations with HLA-B5, Arabs have had associations with HLA-B35, and North American patients have had associations with HLA-DR4 [2]. The majority of inflammatory lesions can be stenotic, occlusive, or seldom aneurysmal, panarteritis with intimal proliferation is seen in advanced lesions [2,4]. Wall thickening, thrombus formation, stenotic and occlusive lesions, as well as dilatation and aneurysms, are typically the results of inflammation and endothelial damage [2]. The lumen typically narrows in a patchy pattern that frequently affects multiple areas [4]. If the disease progresses rapidly, fibrosis may not be uniform and may lead to aneurysm formation [4].

The vasculitis can be divided base on microscopically findings into two phases: a healed fibrotic phase and an acute inflammatory phase [4]. In the acute phase, lymphocytes and occasionally giant cells with neovascularization infiltrate the adventitia and media [4]. Neoangiogenic vasa vasora, which most likely serve as the entry point to the arterial wall, are located close to inflammatory infiltrates [5]. Evidence suggests that vascular antigens are presented locally, with the arterial wall starting and maintaining the immune host response [5]. Lymphoid tissue forms in inflamed arteries, mainly in the adventitial layer [5]. In the chronic phase there is fibrosis with destruction of elastic tissue [4]. The infectious route has also been proposed, being reported cases of Takayasu arteritis in patients with HIV infection, Mycobacterium Tuberculosis and post-hepatitis B vaccination [2].

The majority of patients present with non-specific clinical general signs such as hypertension headaches, fever, dyspnea, weight loss, vomiting, myalgia or arthralgia [2].

A discrepancy in any of the four limbs blood pressures greater than 10 mmHg, diminished or absent pulses, limb claudication, dyspnea, valvular disease, and pericarditis are among the other cardiovascular findings that have been described most frequently in pediatric populations [2].

Pulmonary artery involvement typically manifests as cough, dyspnea, pleural effusion and pulmonary arterial hypertension [2,4]. In approximately a third of cases, prominent arterial vessels associated with oligaemic lung fields correlate with pulmonary vasculopathy [4]. Although there is little correlation between the systemic pattern of arterial involvement and pulmonary artery disease, pulmonary artery disease can be useful in the differential diagnosis by excluding pulmonary pulmonary embolism as it has been described in our case [4].

Headaches, stroke, cognitive impairment, seizures, and occasionally posterior reversible encephalopaty syndrome are all neurological manifestations that can be brought on by hypertension or ischemia [2].

Ocular features can be transient or persistent and progressive and are usually following stenosis of the carotid arteries [2]. Ischaemic changes may cause vitreous hemorrhage or retinal detachment [2]. Retinal involvement may lead to microaneurym and arterio-venous malformation [2].

IgA nephropathy and nephrotic syndrome have also been reported in the literature [2]. Renal involvement can result in glomerulopathy with hematuria and proteinuria, renal artery stenosis, and reno-vascular hypertension [2].

Gastrointestinal symptoms may be cause be arterial stenosic lesions causing arterial mesenteric ischemia with hematochezia or melena, vomiting and in severe cases

bowel perforation [2].

Laboratory findings are non-specific and most frequently may reveal an elevated level of erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) [2].

Concentrations of interleukin-6 are elevated in patients with active disease and corellate with the activity of the disease [4]. High serum concentrations of metalloproteinase-2 can suggest the presence of Takayasu's arteritis without any relation with the activity of the disease [4]. The connection between vascular progression and biomarkers and inflammation in the artery wall has been the subject of previous research [5]. When inflammation occurs, pentraxin-3 (PTX3) is released, regulating tissue repair and remodeling [5]. PTX3 levels have been shown to correlate with vascular progression [5].

Furthermore, imaging techniques are required to diagnosie and monitore the disease [2]. Most commonly used techniques are conventional angiography, magnetic resonance angiography (MRA), and computed tomography angiography (CTA) [2]. The best appraisal of the lumen can be obtained using the digital subtraction technique angiography (DSA), but this procedure is invasive and implies radiation [2]. MRA is equally accurate and sensitive as DSA in terms of providing information on artery wall architecture during inflammatory phases (edema, thickness and contrast enhancement) [2]. However, disadvantages of MRA include its inability to capture small vessels and the possibility of overestimating the degree of vascular stenosis [2]. It is a non-invasive imaging technique that doesn't imply radiation [2]. Stenosis, aortic wall thickening, mural thrombi, and a strong signal in T2-weighted images indicating inflammatory edema of the artery wall are most often described [2]. CTA, on the other hand, has a better resolution and produces images with outstanding anatomic information in 3D reconstruction [2]. It does, however, expose the patient to radiation and is not appropriate for recurrent follow-up examinations; yet, it is clearly useful when MRA cannot be performed [2].

Color Doppler Ultrasound offers information regarding vessel morphology detecting thrombosis and aneurysms, particularly in the carotid arteries [2]. It is a low-cost examination that does not involve radiation or contrast [2]. Ultrasonographic examinations have revealed a widespread thickening of the intima-media complex [2].

New nuclear imaging procedures, such as 18F-fluoro-deoxy-glucose positron emission tomography imaging (18F-FDG-PET), have recently been added to the arsenal of diagnostic procedures assesing the disease [2]. This technique incorporates the evaluation of the metabolic activity of the artery wall to offer information on the degree of disease activity and metabolic abnormalities well before morphologic changes take place on other imaging studies, with a sensitivity and specificity approaching to 100% [2].

Differential diagnosis includes: systemic infections like HIV, brucelosis, endocarditis; infectious aortitis – syphylitic aortitis and tuberculous aortitis; autoimmune systemic diseases – rheumatoid arthritis, polyarteritis nodosa, Kawasaki disease, systemic lupus erhythematosus, sarcoidosis, spondylarthropathies and Behcet's disease; atherosclerosis; Giant cell arteritis; Ig-G4 related disease and non-inflammatory conditions – Ehlers Danlos type IV and Marfan's syndrome [2, 6].

The treatment for TA focuses on controlling the inflammation that can lead to permanent damage to the organs and is divided in medical and surgical treatment [2,4].

Medical treatment involves using Prednisone, non-glucosteroidal therapies such as methotrexate, azathioprine, mycophenolate mofetil, cyclophosphamide; tumor necrosis factor inhibitors - etanercept, infliximab, and adalimumab; JAK-inhibitors such as Tofacitinib; Rituximab; Tocilizumab and Abatacept [7, 8]. Revascularization procedures are necessary for irreversible vascular lesions that have a significant influence on hemodynamics [2]. During the disease's inactive phases, these procedures should only be undertaken in institutions with expertise [2]. Cerebral artery disease, coronary artery disease, renovascular hypertension, end-organ ischemia, and gradual aneurysm enlargement with threat of dissection are all indications for revascularization [2].

Considering this case and the data available in literature, TA is a rare disease, affecting major arterial vessels and it implies a variety of signs and symptoms, representing a challenge for the medical team.

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