Takayasu Arteritis as an Odd Cause of Chest Pain

Santiago Bernal-Macias, Maria A. Alzate, Adriana Rojas-Villarraga.

Center for Autoimmune Diseases Research (CREA), School of Medicine and Health Sciences, Universidad del Rosario, Bogota, Colombia.

Corresponding author:
Santiago Bernal-Macías, MD Center of Autoimmune Diseases Research (CREA)
School of Medicine and Health Sciences, Universidad del Rosario
Carrera 24 # 63 – c – 69, Bogotá, Colombia
Tel +57 349 9650.
Fax +57 349 9390
Email: bernal.santiago@urosario.edu.co

Highlights

We describe a case of Takayasu arteritis with an unusual presentation, having chest pain as the main symptom and a nasal septal perforation as a rare complication, without upper and lower extremities, abdominal, neck or cerebral compromise. The patient, a 34 year-old male, previously healthy, presented with acute onset of chest pain, dyspnea and severe compromise of his functional status. Severe stenosis of pulmonary arteries and coronaries, as well as signs of aortitis were found following CT angiography of the thorax, pulmonary angiography and right and left heart catheterization. Anti-neutrophil cytoplasmic antibodies were positive. The patient received 750 mg of Methylprednisolone and monthly bolus injections of Cyclophosphamide. Six months after the diagnosis, the patient developed a nasal septal perforation. This is the first reported case of Takayasu arteritis with rapidly progressing coronary disease as the only presentation, complicated by a nasal septal perforation.

Keywords: Takayasu arteritis, Vasculitis. Coronary artery disease

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Takayasu arteritis (TAK) is a chronic large vessel vasculitis mainly involving the aortic arch, and its primary branches. It predominantly affects young females during the second or third decades of life, with initial non-specific symptoms such as fever and weight loss, and in advance stages, extremity pain, claudication and abnormal pulses.[1,2]

We described the case of a young man, previously healthy, who was subjected to multiple (noninvasive and invasive) tests for the study of atypical chest pain following rapid progression of coronary disease. The final diagnosis was TAK.

A 34 year-old male patient, coalminer for 7 years, and smoker for 10 years, otherwise healthy, presented to the emergency department on multiple times for atypical chest pain and dyspnea, with important compromise of his functional status (NYHA IV/IV). First coronariography was normal, despite high troponins and electrocardiographic changes. As the patient kept deteriorating, six months later an CT angiography (CTA) of the thorax, pulmonary angiography and right and left heart catheterization were performed finding severe stenosis of pulmonary arteries and coronaries, as well as signs of aortitis (Figure 1A, B and C). An MRI of the heart was performed, showing signs of vasculitis compromising descending aorta and main pulmonary arteries. Standard autoimmune profile was negative except for Anti-neutrophil cytoplasmic antibodies by IFI (MPO ANCAS) positive 17 UI/ml and by ELISA positive 1:40. Infectious profile was negative. A diagnosis of Large Vessel Vasculitis – TAK was done. The patient received 750 mg of Methylprednisolone and monthly bolus of Cyclophosphamide (CYP) during 6 months. Additional tests were done to rule out abdominal, lower extremities, neck and cerebral compromise. All of them were negative. Despite there was no history of previous nasal trauma, surgery, smoke or any illicit drugs consumption, 6 months after the diagnosis of TAK, he developed a nasal septal perforation (Figure 2) that required otolaryngology study.

To our knowledge, this is the first reported case of TAK presenting with chest pain as the main symptom and a rare complication such as nasal septal perforation in a Colombian patient, without abdominal, lower extremities, neck and cerebral compromise, opposite to other publications of this rare disease in Colombia.[1,3] One similar case of rapid progression of coronary disease associated to TAK was reported in Portugal.[4]

There are two main problems that TAK patients have to deal with: A difficult diagnosis and a low level of evidence for their management.[2,5] There is not a gold standard for diagnosis and monitoring of disease. Radiological techniques have advanced in the assessment for diagnosis of TAK; especially CTA and magnetic resonance angiography which are less invasive than

* Corresponding author: bernal.santiago@urosario.edu.co

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conventional angiography and have similar sensitivity and specificity levels. Additionally, ultrasonography may have a role in detecting mural changes, particularly in early disease where CTA does not perform as well. [5,6]

The treatment for TAK has a low level of evidence because it is mainly based on case series, open studies and expert opinion.[2,5] As Keser G et al wrote there are four groups of therapeutic alternatives. The conventionally used are the steroids; however, they are used together with conventional immunosuppressive agents (i.e. methotrexate, azathioprine, or CYP). A novel alternative is the biological agents especially in patients refractory to all previous treatment. The last therapeutic alternatives are the invasive interventional treatment considered when patients are in chronic stages [2,5].

Our case supports the idea that invasive and non-invasive radiologic techniques are good alternatives for diagnosis of this disease, in combination of good assessment of clinical signs and symptoms, especially in odds manifestations of TAK. There is still a challenge in understanding the natural history of disease and the possible response to immunomodulatory treatment.

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The authors agree to abide by the requirements of the "Statement of publishing ethics of the International Cardiovascular Forum Journal".[6]

Declarations of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

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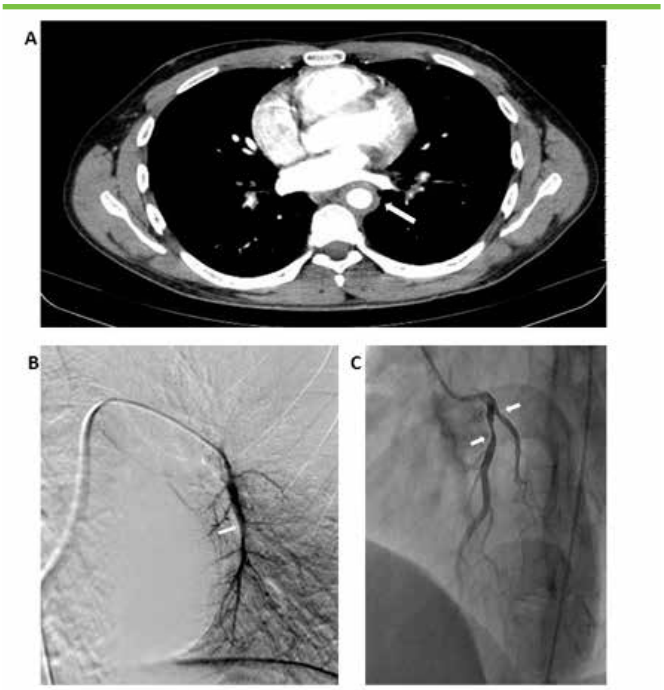

Figure 1.
A. Thorax CT angiography. Result was negative for signs of pulmonary embolism but showed mediastinal widening with changes suggestive of aortitis (arrow) and perivascular inflammation of main pulmonary arteries.
B. Pulmonary angiography. Moderate pulmonary hypertension (85/21/48 mmHg), multiple severe stenotic lessions in both main pulmonary arteries (arrow) with generalized hypoperfusion of upper lobes.
C. Right and left heart catheterization. The results of these tests were: 2 vessel coronary disease with 80% occlusion of Anterior descending artery and circumflex artery (arrows), anteroseptal hypokinesia with eyection fraction of 46%.

Figure 2. Subtotal nasal septal perforation on gross physical examination.