

Takayasu's Arteritis: Ascending Aortic Aneurysm and Coronary Artery Disease in a 19-year-old Young Adult

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Abstract

Takayasu's Arteritis (TA) is a disease of multiple presentation, which can involve many different systems and mainly affects women in the first decades of life. The present case reports on a 19-year-old patient with TA, with a diagnosis of ascending aortic aneurysm and severe coronary artery disease in the right coronary artery (RCA) and left coronary trunk (LCT), who underwent an implant of a supracoronary Dacron graft in the ascending aorta and myocardial revascularization with a double internal thoracic artery. The patient evolved satisfactorily in post-op and is today undergoing clinical follow-up in our medical services.

Introduction

Takayasu's Arteritis (TA) is a rare disease,¹ described for this first time in 1908 by Mikito Takayasu as a case of retinal vasculitis with the absence of a pulse in a young adult.² In an overall perspective, this disease is more common in women (80-90% of the cases), between the 2nd and 4th decades of life.¹ Its incidence varies and, according to statistical data from Japan, Europe, and the USA, around 1-3 new cases/one million inhabitants, are diagnosed each year.³

TA is defined as a progressive and granulomatous chronic inflammatory disease of the large blood vessels (aorta and its main supra-aortic branches, as well as the mid-sized pulmonary and coronary arteries), characterized by stenosis, occlusion, and/or dilation/aneurysm.^{4,5} Of unknown etiology and poorly understood pathogenesis, it is presumed that genetic factors and infectious agents can play key roles in the origin of the disease.⁶

There are no specific laboratory exams for the diagnosis and follow-up of the disease's activity. Angiotomography (Angio-CT) and the magnetic resonance angiography (MRA) represent the gold standards for the diagnosis of TA.^{7,8} According to angiographic findings, TA has been classified in 6 types of alterations, based on the affected region, which is useful for surgical planning.⁹

Keywords

Coronary Disease; Aneurysm; Takayasu Arteritis.

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The clinical treatment of this entity can be found in the guidelines, whose objective is the control of active inflammation and the prevention of new vascular damage. However, when in the presence of critical stenotic lesions, dilation, and/or aneurysm, percutaneous angioplasty and surgical treatment are recommended, on a case by case basis.⁸ In this context, this work aims to describe a rare clinical manifestation of TA

Case report

Our study reports on a 19-year-old female patient, weighing 57 kg, with TA diagnosed at 3 years of age, who was medicated with applications of pulse therapy and methotrexate until 12 years of age, at which time she ceased to continue her medical follow-up until she was 18 years of age. The patient resumed her follow-up treatment one year ago at the same medical institution, at which time she was diagnosed with ectasia of the ascending aorta, with 35 x 34 mm in the transthoracic echocardiogram (TEE). The patient was then sent to the medical reference service.

She underwent an MRA, which revealed a thickening of the aortic wall and dilation of the ascending aorta of 37x35 mm (Figure 1). Six months later, she returned to the outpatient clinic, reporting a medical condition of typical chest pain lasting 30 minutes, followed by syncope, but she did not go to the hospital on that occasion. An Angio-CT of the aorta and coronary arteries was requested, which revealed an aneurysm of the ascending aorta, 46 x 44 mm, with signs of circumferential parietal thickening and parietal calcification along its entire extension, as well as severe coronary artery involvement by extrinsic compression related to the parietal thickening of the aorta (Figure 2).

The coronary cineangiography showed severe lesions in the right coronary artery (RCA), 80% proximal and 80% proximal of the left coronary trunk (LCT). The case was discussed with the aorta Heart Team, and it was decided that the patient should be hospitalized for surgical treatment. The patient was asymptomatic at the time of hospitalization.

Upon physical exam, a systolic murmur appeared at a II/IV aortic focus. Laboratory data presented no significant alterations. During the pre-operative period, the patient's routine medication was maintained. Given the evidence of disease progression, the patient underwent a supracoronary Dacron graft implant in the ascending aorta and myocardial revascularization with double internal thoracic artery (ATIE-DA and ATID-CD, in Portuguese) (Figure 2). With no complications during hospitalization, the patient was released on the 8th post-operative day. She is currently undergoing optimized clinical treatment and is taking medicine to control the base disease, as advised by the healthcare team.

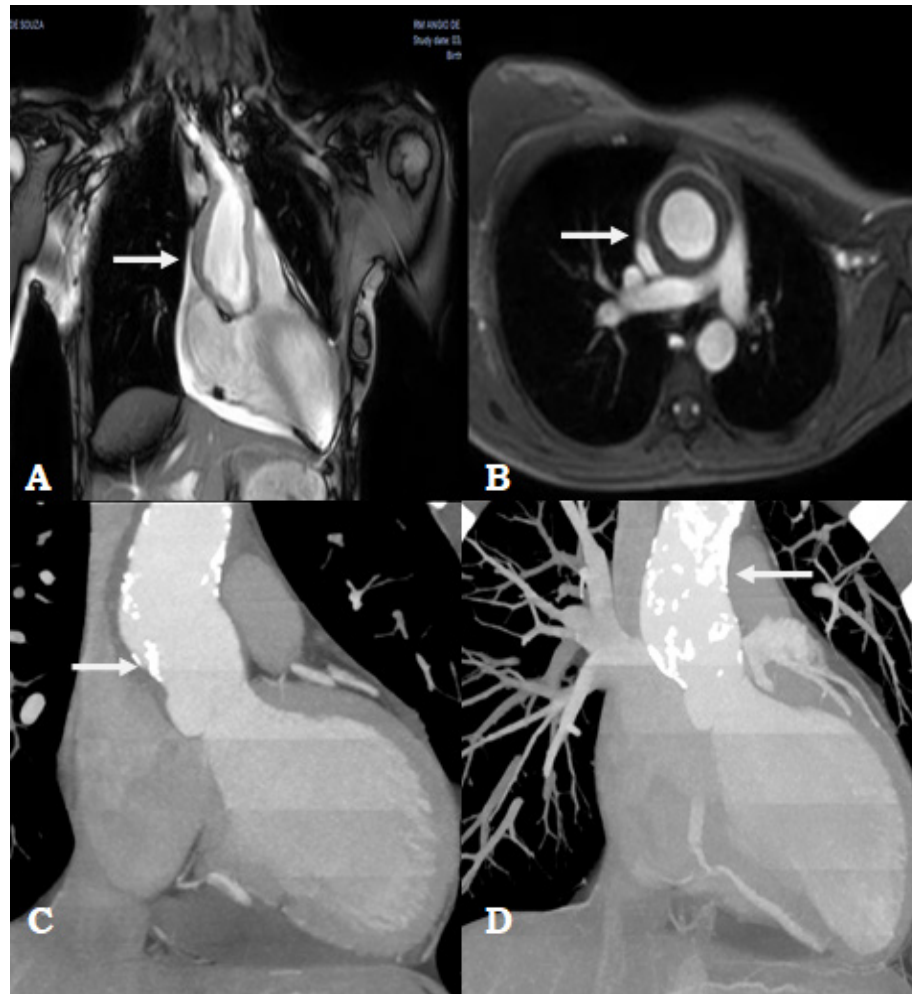


Figure 1 – A and B) CMR illustrating circumferential parietal thickening with dilation of the ascending aorta of 37x35 mm; C and D) Ascending aortic aneurysm by CT, measuring 46x44 mm, with signs of thickening and parietal calcification along its entire extension.

Discussion

The description of this case refers to the severity of TA, as well as to its complexity in the diagnostic approach and treatment. TA is a rare disease, most commonly found in women before 50 years of age, but many studies also show a high prevalence at more advanced ages.¹⁰

The diagnosis is based on diagnostic criteria from EULAR/PRINTO/PRES.¹¹ In this case, the clinical means of presentation was of coronary artery disease. Coronary artery involvement is uncommon, showing only a 9% incidence.¹² Although it is rare, myocardial ischemia is one of the main causes of death. The inflammatory process of the aorta has been identified as being responsible for these lesions, in which the occlusion of the ostium of the coronary arteries occurs more often, though there are many cases in the literature that describe coronary arteritis with no occlusion of the ostium.^{13,14} Coronary artery involvement in this patient is similar to that described by

Endo et al., in which 24 patients presented coronary artery stenosis, of whom 83.3% were women.¹²

According to angiographic findings, the TA was classified in six types of alterations based on the affected region, which is useful for surgical planning.^{7,8} The decision of when to use an endovascular approach or open surgery can be influenced by a series of factors.¹⁵ Percutaneous coronary interventions (PCI) have been used in the initial treatment of the obstructive lesions, presenting uncertain results and a long-term survival that is still not fully understood.¹⁶

Due to a serious chance of intra-stent restenosis, cardiac magnetic resonance (CMR) is the procedure of choice to treat coronary artery lesions.¹⁴ In a retrospective case series involving 106 Japanese patients, Miyata et al.¹⁷ observed a 15-year increase in the survival rate, rising from 43% (among those treated with medical therapy) to 67.5% after the addition of surgical intervention.

Case Report

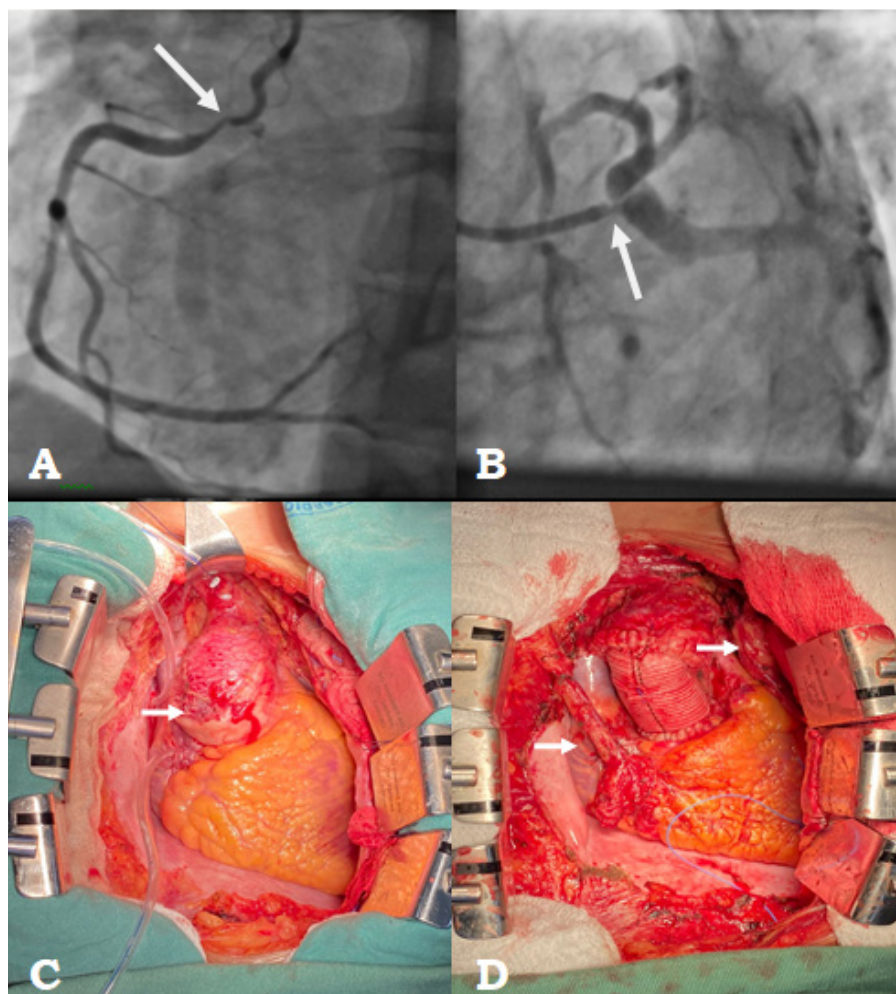


Figure 2 – A and B) Coronary angiography, showing severe lesions in the RCA and LCT; C and D) Final aspect of the surgery after myocardial revascularization and Dacron graft implant.

Yang et al.,¹⁸ in an analysis of 31 patients with TA who underwent revascularization by CMR (12) and PCI (19), in an average follow-up of 101 months, observed higher restenosis rates in the percutaneous group. The inflammatory process can affect different parts of the coronary artery, but with a greater prevalence of ostial/proximal lesions, in accordance with that reported in the present study. The authors also highlight that the myocardial ischemia resulting from coronary artery involvement constitutes the main cause of death among TA patients. In this sense, the importance of studying the coronary arteries of these patients when there is a clinical suspicion is crucial.

Huang et al.,¹⁹ in a study involving 90 patients comparing medicine vs. revascularization treatments in TA patients (CMR and PCI) were unable to identify any difference in mortality due to cardiovascular causes among the groups. Nevertheless, in the analysis of the subgroup through a revascularization strategy, the findings corroborated with those reported by Yang et al.¹⁸ with higher restenosis rates in the percutaneous

approach. Moreover, in this study, heart failure was an independent predictor of mortality.

Modalities of revascularization were also the focus of the study published by Wang et al.,²⁰ in an analysis of 46 patients with TA and coronary artery involvement. The authors found that the MACE rates (all-causes mortality, myocardial infarction, and revascularization) were greater in the group submitted to PCI. The multivariate analysis for the MACE outcome viewed the disease activity and the type of revascularization (CMR vs. PCI) as predictive factors. Such data reinforce the individualized indication and the importance of the Heart Team in these patients' medical evaluations.

Conclusion

Coronary artery lesions vary from patient to patient, especially proximal involvement, and therefore careful planning in all stages of the treatment through revascularization, be it percutaneous or surgical, is of utmost importance. The

data point to higher restenosis rates after the percutaneous approach, with the CMR being the preferred method when no contraindications were presented, such as the availability of grafts and the possibility of using the internal thoracic artery.

The adequate suppression of the inflammatory activity from the pre-operative stage and the choice of technique to be used guarantee better end results. Thus, the evaluation of the patient's stage of the disease is essential, given that it is a factor associated with higher MACE rates, as well as the shared decision on the case. The rigorous and specialized clinical follow-up in a reference center should be encouraged.

Author Contributions

Conception and design of the research, acquisition of data, analysis and interpretation of the data, statistical analysis, writing of the manuscript and critical revision of the manuscript for intellectual content: Antônio IBC, Maia AS, Rabelato J, Campos AAS, Menezes MP, Issa M.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This study was approved by the Ethics Committee of the Instituto Dante Pazzanese de Cardiologia, CAAE: 60291322.8.0000.5462, Protocol Number: 5.629.949. All the procedures in this study were in accordance with the 1975 Helsinki Declaration, updated in 2013. Informed consent was obtained from all participants included in the study.

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