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Case report

Association between rheumatic fever and Takayasu's arteritis – Case report



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ABSTRACT

Takayasu's arteritis (TA) and rheumatic fever are diseases that can start with cardiac features, making the diagnosis difficult. There are reports of association of RF with Takayasu's arteritis beginning with cardiac involvement in pediatric patients. The aim of this study is to report the possible association of RF and TA in patients with cardiac abnormalities. We describe the case of an adolescent initially diagnosed with RF who progressed with changes that allowed making the diagnosis of TA. TA and RF are two important causes of valve involvement that may have systemic manifestations.

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Associação entre febre reumática e arterite de Takayasu – Relato de caso

RESUMO

A arterite de Takayasu (AT) e a febre reumática (FR) são doenças que podem ter início com manifestação cardíaca, o que dificulta o diagnóstico. Há relatos de associação de FR com AT que se inicia com comprometimento cardíaco na faixa etária pediátrica. O objetivo deste estudo é relatar a possibilidade da associação de FR e AT em paciente com alteração cardíaca. Descrevemos o caso de uma adolescente diagnosticada inicialmente como FR que apresentou na evolução alterações que permitiram o diagnóstico de AT. A AT e a FR são duas causas importantes de envolvimento valvular que podem apresentar manifestações sistêmicas.

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Introduction

Takayasu's arteritis (TA) is a chronic granulomatous inflammatory disease that can affect the aorta, its branches and pulmonary artery.^{1,2}

In children, it affects women at a ratio of 2:1, similarly to that found in a Brazilian multicentric study of 71 children and adolescents (2.6:1).³ Mean age of onset is 11.4 years, with 20% before 19 years and 2% before 10 years of age.⁴

The initial manifestations may be insidious and non-specific, such as asthenia, headache, fever, night sweats, arthralgia, muscle pain and weight loss, starting at variable times before vascular disease, or may be acute with high blood pressure, seizures and congestive heart failure.⁵

Cardiac impairment occurs in about 50% of patients in the course of the disease, and any cardiac structure can be affected. When presented with valve regurgitation, the patient may initially be diagnosed with rheumatic fever.

We reported a case of TA in an adolescent with cardiac valve impairment with early diagnosis of rheumatic fever.

Case report

A 15-year-old adolescent, female, white, was admitted with a history of palpitations associated with adynamia and 7-kg weight loss, 10 months ago. On physical examination, the patient was pale, tachycardic, and afebrile with blood pressure (BP) of 130 × 70 mmHg (90-percentile). On cardiac auscultation, loud P2 and systolic murmur in mitral area radiating to the back present symmetrical pulses.

Laboratory tests showed anemia, with mild leukocytosis and thrombocytosis, elevated inflammatory activity and titration of anti-streptolysin O antibody (ASLO) (Table 1).

Chest radiography showed cardiac area at the upper limit, the echocardiogram showed moderate left ventricular dilatation with thickening of the mitral valve and mitral (mild) and aortic (mild to moderate) regurgitation, with an ejection fraction of 51%, and inflammation on myocardium scintigraphy. With these data, she was diagnosed with rheumatic fever (RF). Treatment with prednisone, digoxin, and hydrochlorothiazide was initiated, with prophylaxis with benzathine penicillin, with normalization of inflammatory activity. In outpatient follow-up, corticosteroids were gradually reduced and

discontinued after three months of its initiation, maintaining adequate secondary prophylaxis.

After discontinuation of corticosteroids, she showed exertional dyspnea, tachycardia, lower limb edema, auscultation of epigastric murmurs and increased blood pressure. On laboratory investigation, inflammatory activity tests were high and PPD was negative. Echocardiography revealed normal mitral valve and mild aortic regurgitation and an ejection fraction of 63%. Chest tomography detected parietal thickening of the aorta and its branches, and presence of dilation (2.6 cm) in the thoracoabdominal transition that is suggestive of arteritis. The hypothesis of TA was confirmed by the changes evidenced in arteriography. (Fig. 1) Methotrexate and prednisone were associated with clinical and laboratory control of inflammation.

Currently, the patient is being followed on diagnosis of TA and RF, with prednisone, methotrexate, folic acid, digoxin, anti-hypertensives and prophylaxis with benzathine penicillin.

Discussion

TA is uncommon, especially in the pediatric population, although it is a major cause of renovascular hypertension.

Genetic factors and infectious and immunological processes are described in etiopathogenesis.

In the case described, as Jones criteria were met, it allowed the diagnosis of RF, but the nonspecific manifestations that preceded FR symptoms suggest the presence of a systemic inflammatory vasculitis of insidious onset.⁶ In the course of the disease, the presence of epigastric murmur, hypertension and abnormal angiography allowed the diagnosis of TA based on the revised criteria EULAR/PreS.⁷ The association of RF and TA is uncommon.⁸ Tejada et al.⁹ reported 2 cases of rheumatic heart disease in a series of 125 patients with TA. At our service, within 11 cases of TA, only this patient had heart changes consistent with RF.

Cardiac involvement occurs in up to 50% of patients with TA, and may impair any heart structure.^{10,11} Coronary arteritis, heart valve lesion, and ventricular aneurysm are the main manifestations reported in children with TA.

The aortic valve involvement is the most common, secondary to aortitis with annular dilation or inflammatory valve retraction.¹⁰ There are reports of involvement of mitral and

Table 1 – Progress of laboratory alterations.

	Hospital admission	12 days after the use of PRED	Postsuspension of PRED	18 days after reintroduction of PRED	3 months after PRED	6 months after PRED	Current
Hemoglobin	9.04	12.8	11.6	14.6	11.5		11.5
Leucocytes	13,000	24,000		12,600	5300		19,230
Platelets	635,000	523,000	630,000	543,000	530,000		444,000
ESR/CRP	120/7.8	5	37/19.5	20/<0.3	40/4.4	26	18/<0.4
Alpha-1-acid glycoprotein	195	188	246	–	129	125	–
ASLO	1379	1560	–	–	129	125	–
PPD	–	–	–	–	–	Negative	–
Creatinine/serum urea	–	–	–	–	–	0.5/14	–



Fig. 1 – Arteriographic alterations.

tricuspid valves in approximately 20% of TA patients, often with regurgitation, with no evidence of valve thickening.¹¹ In autopsy studies, there are reports of aortic valve thickening secondary to arteritis of the aorta extending to valve and mural endocardium.¹²

The presence of mitral valve thickening, associated with streptococcus, with resolution after treatment with corticosteroids does not allow ruling out TA-associated RF. According to Vale et al.¹³, the combination of RF and TA raises the possibility of a common immunologic basis in the pathogenesis of both disorders.

TA and RF are two major causes of valve involvement, the presence of systemic manifestations, echocardiographic and arteriographic findings are helpful in the diagnosis of cardiac involvement.

Conflicts of interest

The authors declare no conflicts of interest.

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