Systemic aortoarteritis: a case review

Aortoarterite Sistémica: apresentação de um caso

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Abstract

Systemic aortoarteritis of adults is an uncommon inflammatory disease of uncertain aetiopathogenesis, involving the aorta, pulmonary and temporal arteries as well as their branches.This article analyses the case of a 71-year-old man, a native of India, with symptoms suggestive of an inflammatory or consumptive disease. Positron emission tomography led to the diagnosis, demonstrating diffuse abnormal fluorine-18 fluorodeoxyglucose arterial uptake in the thoracic aorta and in both subclavian arteries. Magnetic resonance imaging confirmed the diagnosis, presenting furthermore inflammation of the pulmonary trunk. Differentiation between Takayasu arteritis and giant cell arteritis was not definitively possible, considering international established criteria. Prednisolone therapy resulted in clinical and laboratory remission. Positron emission tomography control showed total regression of the disease process. The data of this rare disease may be helpful for an early diagnosis and treatment as well as preventing severe vascular complications.

Key words: arteritis, aortoarteritis, Takayasu arteritis, giant cell arteritis.

Resumo

A aortoarterite sistémica dos adultos é uma doenca inflamatória rara, de etiopatogenia indeterminada, envolvendo a aorta, a artéria pulmonar, a artéria temporal, e os seus ramos.

Este artigo analisa o caso de um indivíduo de sexo masculino, indiano, de 71 anos de idade, com sintomatologia sugerindo um processo inflamatório ou uma doença consumptiva. Após a tomografia de emissão positrónica, que mostrou uma absorção anormal e difusa de fluorine-18 fluorodeoxyglucose na aorta torácica e nas artérias subclávias, chegou-se ao diagnóstico. A tomografia de ressonância magnética confirmou o diagnóstico, mostrando ainda uma inflamação do tronco pulmonar. O diagnóstico diferencial entre a arterite de Takayasu e a arterite de células gigantes não era possível, considerando os critérios internacionalmente estabelecidos. O tratamento com prednisolona deu lugar à remissão clínica e laboratorial. A tomografia de emissão positrónica de controle, revelou resultados normais. Os dados deste caso raro poderão ser úteis para um diagnóstico e tratamento precoces, evitando assim as complicações vasculares graves.

Palavras chave: arterite, aortoarterite, arterite de Takayasu, arterite de células gigantes.

Introduction

The systemic aortoarteritis (SA)[1,2,3] is mainly an inflammation of the aorta of unknown etiology. A relationship between SA and infectious agents has been suggested. A genetic predisposition to SA is supported by associations with human leucocyte antigens (HLA), evidence of the involvement of other immune response controlling genes, and familial clustering. SA, particularly affecting adults, is a rare disease. It includes Takayasu arteritis (TA) with a prevalence of 1-6/1000 and giant cell arteritis (GCA) with a frequency ranging from 10-50/100,000. TA is a granulomatous inflammation of the aorta, its major branches and the pulmonary artery; it appears predominantly in young females in their second and third decades; it has worldwide distribution with higher prevalence in the Far East and South America. GCA is a granulomatous arteritis with mononuclear and giant cell infiltration, involving the aorta and its major branches, especially the carotid and temporal arteries; it has a slight female preponderance, virtually never occurs before the age of 50 years, and it appears mostly in people of Scandinavian or Northern European heritage.

The initial clinical manifestations and laboratory findings are as such for severe inflammation or consuptive conditions. Further signs occur in advanced stages due to vascular complications.

The diagnosis and therapy delay leads to arterial occlusions or aneurysms.

Case report

A 71-year-old man, a native of India, presented with tiredness, night sweats, slight occipital and bilateral

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This work is dedicated to my family with love and gratitude

headache, occasionally, and weight loss of 7 kg during last 6 months. No fever. No jaw claudication and arthralgias. No visual troubles. The patient has type 2 diabetes, under good control with diet alone for 7 years; 40 years ago, he had acute hepatitis B, cured, no sequela. His weight was 63 kg and his height was 172 cm. On physical examination, the patient was afebrile and had normal blood pressure and pulse. No blood pressure difference between arms. No tenderness over the temporal arteries, no bruits over subclavian arteries, carotids or aorta. No any other clinical abnormalities. Tuberculin skin test slight positive.

Neurological, ophthalmological and otorhinolaryngological councils, electroencephalogram (EEG), funduscopic inspection and paranasal sinus x-ray were normal.

Laboratory data: Erythrocyte sedimentation rate (ESR) 88 mm/h. Hemoglobin 11.8 g/dl. Hematocrit 36.0%. Erythrocytes 4.41 x 10^6/µl. Thrombocytes 380 x 10^3/µl. Leucocytes 8.7 x 10^3/µl. Fibrinogen 799 mg/dl. C-reactive protein (CRP) 82.6 mg/l. P-iron 4.9 µmol/l. S-ferritin 281 µg/l. P-transferrin 191 mg/dl. Protein-electrophoresis: S-protein 7.1 g/dl, s-albumin 48%, s-a1-globulin 8%, s-a2-globulin 18%, s-ß-globulin 13%, s-g-globulin 13%. Blood glucose around 104 mg/dl. Hemoglobin A1c 5.7%. All other values, including complete blood count, urine-analysis, s-lipids, s-electrolytes, liver, kidney and thyroid function tests, hepatitis serology (HBsAg negative, anti-HBs positive), tumor marker, paraproteins, rheuma serology, immunoglobulins, bone-marrow examination, feces for occult blood and microbiological diagnostics, prostata specific antigen (PSA), lues, human immunodeficiency virus (HIV), borreliosis and malaria tests, and culture of gastric contents for tubercle bacillus were negative.

Electrocardiogram (ECG) within normal limits. Echocadiography: Normal dimensions and contractility, aortic and mitral insufficiency at first stage. Color-coded Doppler duplex sonography of extracranial and transcranial cerebral arteries, subclavian and temporal arteries without relevant abnormalities. Chest x-ray: normal. Sonography of abdomen and thyroid: a slight prostatic hypertrophie without residual urine, otherwise normal. Computer tomography (CT) of neck, thorax, abdomen and pelvis was normal up to a slight prostatic hypertophy. Cranial magnet resonance tomography (MRT) was normal. Total colonoscopy, oesophagogastroduodenoscopy, capsule-endoscopy of the small intestine, small intestine x-ray and contrast enema showed no substantial abnormalities. Positron emission tomography (PET) revealed massive aortitis of the thoracic aorta and inflammation of both subclavian arteries with diffuse abnormal fluorine-18 fluorodeoxyglucose ([F-18] FDG) arterial uptake. Magnetic resonance imaging (MRI) demonstrated inflammatory alterations of pulmonary trunk, in addition.

The initial therapy was conducted with prednisolone at a dose of 40 mg/d. Thereafter, the prednisolone dose was tapered gradually. Within 2 weeks, after corticosteroid medication, the patient has no troubles anymore, he gained weight and his laboratory parameters were all normal. PET control, after 18 months follow-up, under a prednisolone maintenance dose of 7.5 mg/d, showed total normalisation, correlating with clinical remission and normal laboratory findings. Body weight remained constant around 70 kg. After that, the maintenance dose of prednisolone was combined with azathioprine, 50 mg/d.

Discussion

The clinical signs in the beginning of SA are variable, depending on the location of the affected vessel and the severity of the vascular inflammation, and in absence of vascular features, they are also suggestive of a consumptive disease. After detailed diagnostics, this hypothesis could be excluded in our case.

SA was diagnosed by PET and MRI, demonstrating typical inflammatory findings of the thoracic aorta, pulmonary trunk and subclavian arteries. Pulmonary arteritis is a feature unique to TA and is not found in other forms of SA.[4]

Color Doppler duplex sonography did not reveal any relevant alterations of carotids and temporal arteries. For this reason, temporal artery biopsy was not performed, since this method has low predictive value[3] and the use of high-resolution ultrasonography may replace biopsy in the diagnosis of GCA [5]

PET and MRI are both effective techniques to assess early SA.[6] Ultrasonography reveals characteristic wall thickening of subclavian, carotid and temporal arteries.[5]

Since the mean delay from the onset of the symptoms to the time of diagnosis was relatively short (6 months), severe vascular complications were not found in our case, in contrast to other studies.[4,7]

The slight aortic and mitral insufficiency detected

by echocardiography has been described in TA; whether this valvular insufficiency is due to valve arteritis or valve ring dilatation remains controversial.[2,4,7]

Hypochromic anemia with sideropenia and elevated ERS, CRP, fibrinogen, thrombocytes, s-a1-globulin and s-a2-globulin are inflammatory in nature and have been mostly observed in SA.[3,4,7] These features serve as a useful guide to disease activity.

Hepatitis B infection, Borreliosis and tuberculosis has been postulated as possible etiological factors, but studies have not confirmed these relationships.[4,8] The literature review does not suggest any relationship between SA and type 2 diabetes.

Inflammatory cytokines play a key role in different pathological processes of the disease and may help to distinguish different forms of SA.[9]

The familial recurrence usually concerns the same form of SA and is supported by associations with HLA; the histocompatibility antigens of TA differ from those of GCA; there is also some geographical variation in the HLA constellations.[1,2,3]

Differentiation between both forms of SA, TA and GCA, was not definitely possible in our case, considering a set of 6 criteria selected by the American College of Rheumatology for the diagnosis of TA and GCA. Presence of 3 of these 6 criteria is required for the diagnosis.[3,10] However, these classification criteria were never meant to serve as diagnostic criteria and have some limitations; other authors have demonstrated that these criteria have a positive predictive value of only 29%.[11] Our patient met only 1 criterion, suggesting the diagnosis of TA, and 2 criteria for the diagnosis of GCA. The geographical origin of the patient and location of vascular process speak well for TA. The age of disease onset and elevated ERS tend to point to GCA.

First line therapy of SA is an immunosuppressive treatment,[2,3,4,12] primarily with corticosteroids in high doses. Prednisolone should be tapered gradually until a maintenance dose of 5 mg/d. During the taper, patients should be monitored for clinical relapse or an increase in the ERS and CRP. If either occurs, the taper is discontinued and the current dosage maintained. The low dose may last extended periods of time, usually 1-3 years. Patients with prednisolone adverse effects and steroid-resistant subjects respond in addition or alone to cytotoxic agents like azathioprine, methotrexate and cyclophosphamide. However, randomized studies have not proved that these drugs have a significant steroid-sparing effect. More recently, evidence is emerging that newer substances such as mycophenolate mofetil, rituximab and antitumor necrosis factor-alpha may be efficacious and act as steroid-sparing agents. Efficacy of antiaggregation and anticoagulation is not established. In some cases, iron substitution may be necessary for a short time. Our patient, treated with prednisolone, had clinical and laboratory remission after some weeks. After 18 months follow-up, PET control showed total normalisation of vascular findings and complete resolution of the abnormal FDG uptake. At present, he receives prednisolone, 7.5 mg/d, and azathioprine, 50 mg/d, before further reduction of prednisolone dose can be made.

Appropriate diagnostic and therapeutic measures should begin promptly to prevent severe vascular complications. This case report may be helpful in increasing physician's awareness. ■

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