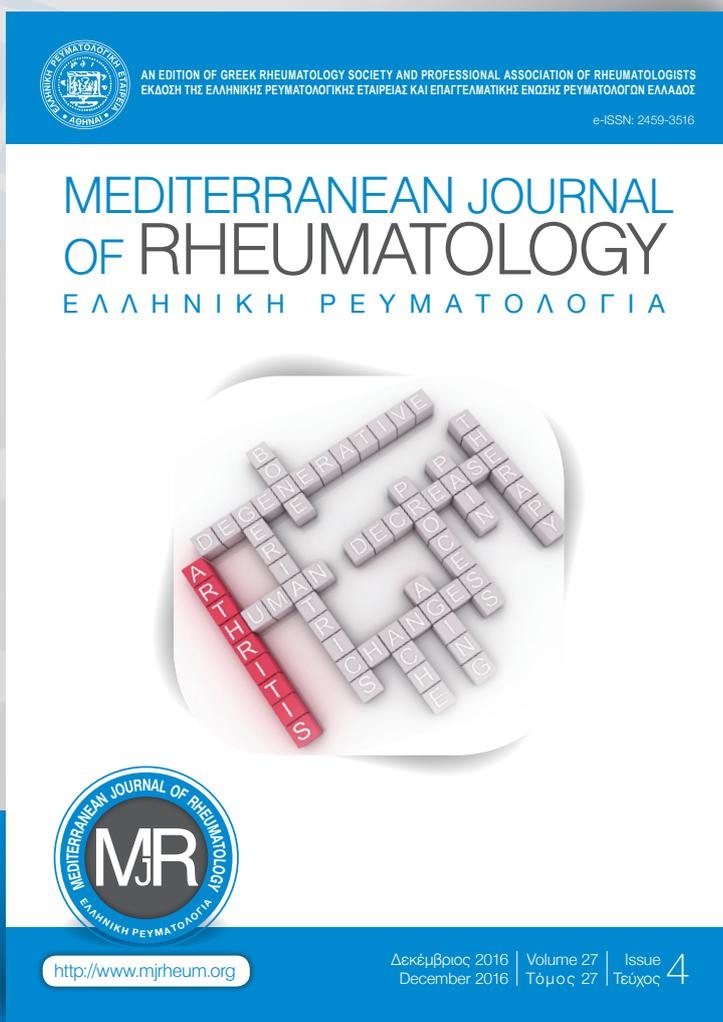


58-year-old patient with IgG4-related thoracic aortitis

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58-year-old patient with IgG4-related thoracic aortitis

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ABSTRACT

We report a 58-year-old male with a one-month of fever up to 38.5° C and back pain with reflection in the anterior chest wall. Chest CT and magnetic resonance angiography (MRA) revealed thickening of the descending thoracic aorta throughout its length and laboratory tests showed elevated serum immunoglobulin IgG4. The patient was diagnosed with noninfectious IgG4-related thoracic aortitis. He was treated with corticosteroids and Azathioprine, and 3 months later, the patient was asymptomatic with normal inflammatory markers, reduction of IgG4 serum levels and with no findings on imaging with MRA.

Cases of IgG4-related thoracic aortitis have been currently described in literature. This rather new entity accounts for around 9% of noninfectious aortitis. As in our case, IgG4-related disease should be considered in any patient found to have aortitis or periaortitis. An early diagnosis and treatment is essential for the survival of such patients.

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CASE

A 58-year-old male was presented with one-month of fever up to 38.5° C and back pain with reflection in the anterior chest wall. Before admission to our hospital, the patient had received, via private physician, oral antibiotics and antiviral treatment without remission of symptoms.

The patient had a history of hypertension and diabetes mellitus, but was not taking any medication. He was a smoker and social drinker. He had no family history of any rheumatic disorder and was of Greek origin.

The physical examination revealed an overweight patient, afebrile, with vital signs within normal limits. In the examination, signs for cardiovascular, pulmonary, abdominal, joint, and neurologic disease were insignificant. Peripheral arteries were uniformly palpable.

Initial laboratory tests showed leukocytosis (WBC 12.240 x 10⁹/L) with neutrophilia, mild normocytic anemia, and elevated inflammatory markers (ESR 64mm/h, CRP 29.1 mg/dl).

Chest Computed Tomography (CT) showed thickening of the descending thoracic aorta throughout its length (**Figure 1a**) and a magnetic resonance angiography (MRA) of the thoracic aorta confirmed those findings (**Figure 1b**).

Further laboratory examinations ruled out the presence of any infectious agent (Salmonella, Staphylococcal species, Streptococcus, etc.). Immunologic work up declared elevated serum immunoglobulin IgG4 (540 mg/dl - normal value <135 mg/dl).

TREATMENT AND FOLLOW-UP

The patient was diagnosed with noninfectious IgG4-related thoracic aortitis.

He was initially treated with intravenous methylprednisolone pulses for 3 days, followed by oral corticosteroids, and Azathioprine as a corticosteroid-sparing agent, leading to direct remission of symptoms and improvement of inflammatory markers.

Three months later, the patient was asymptomatic with normal inflammatory markers, reduction of IgG4 serum levels and with no findings on MRA (**Figure 2**).

DISCUSSION

Thoracic aortitis is a very rare condition that involves a broad array of infectious and noninfectious causes. In nowadays, in the antibiotic era, the occurrence of thoracic aortitis due to an infectious agent is extremely rare.¹ Isolated reports present cases of patients with prior atherosclerotic aorta disease and/or associated infective endocarditis.^{2,3}

The most common causes of noninfectious thoracic aortitis are large-vessel vasculitis giant cell arteritis

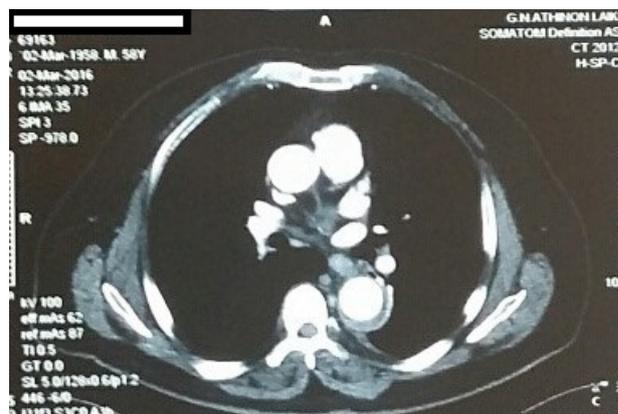


Figure 1a. Chest Computing Tomography (CT) showing thickening of the descending thoracic aorta throughout its length.

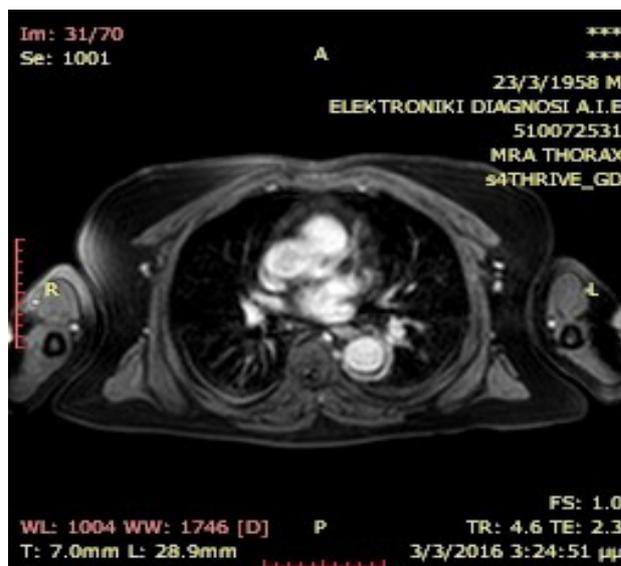


Figure 1b. Initial magnetic resonance angiography (MRA) of thoracic aorta.

(GCA) and Takayasu arteritis.⁴ In the literature, cases of IgG4-related thoracic aortitis have been currently described.⁵ This rather new entity accounts for around 9% of noninfectious aortitis.⁶ The diagnosis is based on the clinical presentation, laboratory data, and especially the presence of infiltration with IgG4-positive cells in histological samples.⁷ Due to the difficulty of surgical approach for tissue samples from the aorta, imaging findings along with serum IgG4 levels are essential for diagnosis.

IgG4-related diseases are newly defined multi-faceted diseases, and have been described in virtually every organ system; including the salivary glands, lacrimal glands, lungs, thyroid, lymph nodes, central nervous

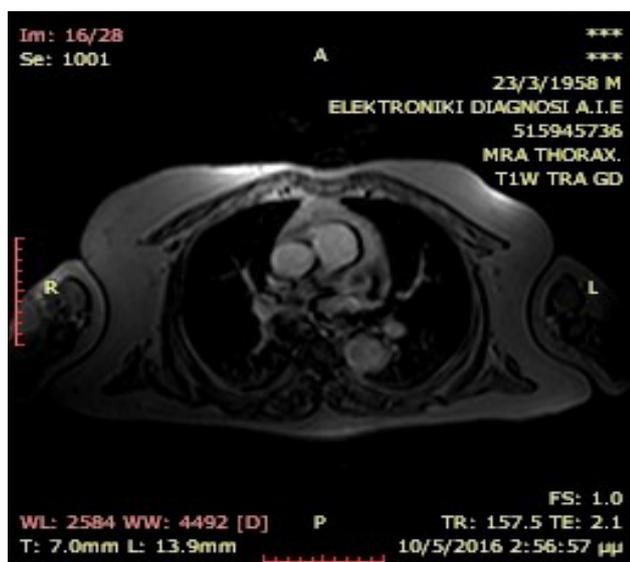


Figure 2. Magnetic resonance angiography (MRA) of thoracic aorta 3 months after therapy.

system, pituitary body, aorta, pancreas, liver, gall bladder, bile ducts, kidney, and prostate.⁸ Predominant histologic findings are lymphocytic infiltration and fibrosis of the affected organs. The aorta is involved in the form of aortitis, abdominal aneurysm or periaortitis and retroperitoneal fibrosis. As in our case, IgG4-related disease should be considered in any patient found to have aortitis or periaortitis. An early diagnosis and treatment is essential for the survival of such patients.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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