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Case Report

Magnetic resonance imaging in the diagnosis and follow-up of giant cell arteritis: case report and review of literature

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Abstract

A female patient with giant cell vasculitis of the abdominal aorta and its branches and strongly suspected of having extrapulmonary tuberculosis is presented. The diagnoses were based on the clinical picture, laboratory findings, and magnetic resonance imaging (MRI) findings. MRI is highly useful in cases where echosonography and/or vascular biopsy for histopathological analyses are not possible. A combination of giant cell vasculitis and extrapulmonary tuberculosis is extremely rare, and therefore, choosing the right treatment presents a considerable challenge. MRI performed after 6-month antituberculous therapy and 1-year glucocorticoid plus methotrexate therapy showed normal wall of the aorta and its branches, which was consistent with clinical and laboratory remission. Patients with large vessel vasculitis require regular follow-up by MRI.

Keywords: Giant cell vasculitis, magnetic resonance imaging, extrapulmonary tuberculosis, glucocorticoids, antituberculotics

Introduction

Giant cell vasculitis, which was first described by Horton, is a chronic granulomatous vasculitis of great blood vessels. Giant cell vasculitis involves the aorta and/or aortic branches, most frequently the extracranial branches of the carotid artery, including the temporal artery. Giant cell vasculitis is twice as common in women. Its clinical picture is predominated by general symptoms and affected artery symptoms. Giant cell arteritis is frequently associated with polymyalgia rheumatica. In giant cell vasculitis patients, laboratory tests usually reveal high erythrocyte sedimentation rate (ESR), elevated levels of C-reactive protein (CRP) and other acute phase proteins, anemia of chronic disease, and thrombocytosis. In clinical practice, the diagnosis of giant cell vasculitis is mostly based on history data, clinical examination, laboratory findings, and imaging studies, primarily Doppler ultrasonography of the temporal artery subsequently confirmed by temporal artery biopsy for histological analysis. Because inflammatory changes involve segments of the arterial wall, temporal artery biopsy is negative in 15% of patients with giant cell vasculitis (1). In giant cell vasculitis, inflammation may exclusively involve great blood vessels, which are unavailable to echosonography and biopsy. Therefore, magnetic resonance imaging (MRI) has a major role in the diagnosis of inflammatory lesions in major arterial walls as well as in the follow-up of therapeutic efficacy (2-4). MRI is also the method of choice in the follow-up of possible complications of inflammatory events in the aortic wall, e.g., aneurysms and dissection (4). Recent studies emphasize the role of fluorodeoxyglucose positron emission tomography (FDG PET) in the diagnosis of inflammatory lesions of great vessel walls, but it is not yet recommended in the follow-up of therapeutic efficacy or possible complications (4-8).

A female patient with giant cell vasculitis of the abdominal aorta and its branches is presented. Diagnosis was based on the clinical picture, laboratory findings, and images of vascular wall inflammatory lesions obtained by MRI. Positive purified protein derivative (PPD) test, positive family history, and lower leg skin lesions that histopathologically corresponded to erythema induratum of Bazin raised a strong suspicion of extrapulmonary tuberculosis. Therefore, treatment with antituberculous therapy was initiated and administered for 6 months. Glucocorticoids were introduced after 8 weeks of antituberculous therapy, followed by methotrexate after 8 weeks of glucocorticoid therapy. Complete clinical remission with normalization of inflammatory and other laboratory parameters was achieved after 1 year of treatment. Follow-up MRI performed after 1 year of treatment showed a normal aortic wall free from inflammatory changes.

Case Presentation

A 55-year-old Caucasian woman was admitted to Dubrava University because of febrile state of unknown etiology (37.5-39°C), fatigue, exhaustion, nocturnal sweating, weight loss (more than 20 kg in 6 months),

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painful erythematous lesions on the skin of the lower legs, migrating headache, all of which persisted for months. The family history revealed that the patient's mother and grandfather died from pneumonia, and her mother-in-law living in the same household was treated for pulmonary tuberculosis.

On admission, the patient was subfebrile (37.5°C) and pale. Her neck lymph nodes enlarged to up to 1.5 cm in diameter. The edema at the root of the nose was painful on palpation, with redness of the skin. The abdomen was diffusely sensitive on palpation, whereas red, painful, and demarcated skin edemas were found on the skin of both lower legs. Otherwise, the patient's status was normal.

Laboratory findings were as follows: ESR, 123 mm/h (normal 3-23 mm/h); CRP level, 245 mg/L (normal <5 mg/L); white blood cell count, 9.8×10^9/L (normal 4×10^9-10×10^9/L) with normal differential count; red blood cell count, 4.31×10^{12}/L (normal 3.86×10^{12}-5.08×10^{12}); hemoglobin level, 10.6 g/dL (normal 12-16 g/dL); platelet count, 990×10^9/L (normal 140×10^9-440×10^9/L); fibrinogen level, 9.3 g/L (normal <4.3 g/L); and ferritin level, 526 µg/L (normal 10-300 µg/L). Other biochemical findings were within the reference limits. Heart and lung X-rays, multislice computed tomography of the thorax, and abdominal echosonography showed normal findings. The cytology of enlarged lymph nodes of the neck biopsy specimens suggested reactive changes. The microbiological analysis of the urine, and urethral, cervical, laryngeal, and nasal swabs, and multiple blood samples yielded sterile findings. Sternal puncture finding was normal. Laboratory tests for systemic or malignant diseases were negative or within the normal range [latex rheumatoid factor, Waaler-Rose test, antibodies to nuclear antigen, anti-double stranded DNA antibodies, cardiolipin, cryoglobulin, complement levels, serum angiotensin-converting enzyme, and tumor markers]. The PPD test result was 25 mm (normal 5-15 mm). Microbiological analysis of the bronchial aspirate for Mycobacterium tuberculosis was negative. We were not able to perform the interferon-Gamma Release Assays test because of technical problems. The hepatitis B and C markers were negative. Echosonography of the heart indicated chronic pericarditis. The Doppler study of temporal arteries was normal. The histopathology of biopsy specimens obtained from the lower leg erythematous lesions corresponded to erythema induratum of Bazin.

MRI revealed enhanced imbibition on delayed post-contrast images of the aortic wall from the level of the crux of the diaphragm to the origin of renal arteries and asymmetric wall thickening at the origin of the celiac trunk, superior mesenteric artery, and both renal arteries (Figure 1). MR images were obtained using a 1.5 T MRI system (Avanto; Siemens, Erlangen, Germany). Contrast-enhanced T1-weighted imaging was performed with breath-hold sequences utilizing three-dimensional gradient-recalled echo and volumetric interpolated (3D GRE VIBE) technique and fat suppression (repetition time/echo time, 4.3/1.7; flip angle, 10°; slice thickness, 3.5 mm; gap, 0; mean number of slices, 72±15.2; range, 35-80 slices; field of view (FOV) 360 mm; matrix size, 144×320, phase×frequency; acquisition time, 19 s). Treatment was initiated with antituberculous therapy consisting of isoniazid+pyridoxine á 400 mg+25 mg (Eutizon B6, Pliva, Zagreb, Croatia), rifampin á 300 mg 2 caps and 1 caps (Arficin, Belupo, Koprivnica, Croatia) alternately, and pyrazinamide at a dose of 2×500 mg (Pyrazinamid, Krka, Novo Mesto, Slovenia) for 6 months. After this therapy, subfebrility, fatigue, and exhaustion persisted in the patient. After 8 weeks of antituberculous therapy, pulsed dosages of methylprednisolone (Medrol, Pliva, Zagreb, Hrvatska) were introduced (500 mg daily for 3 days), followed by 1-mg/kg body weight and acetylsalicylic acid (Andol, Pliva, Zagreb, Croatia) at a dosage of 100 mg daily. After 4 weeks of treatment, the patient showed marked clinical recovery accompanied by a decrease in acute phase reactants. The patient continued receiving methylprednisolone with dose tapering; after 8 weeks of methylprednisolone therapy, methotrexate (Metotreksat, Pliva, Zagreb, Croatia) was introduced at an initial dosage of 7.5 mg once a week. After 6 months of treatment, the dosage of methylprednisolone was 16 mg daily and that of methotrexate was 15 mg per week. Follow-up MRI performed after 1 year of treatment initiation showed normal wall of the aorta and its branches, free from aneurysmal dilation and signs of dissection (Figure 2). Clinically, the patient's condition was good, and her laboratory parameters were normal. She continued taking methylprednisolone at a dosage of 8 mg daily, methotrexate at a dosage of 25 mg per week, folic acid (Folacin, Jadran Galenski laboratorij, Zagreb, Croatia) and acetylsalicylic acid.

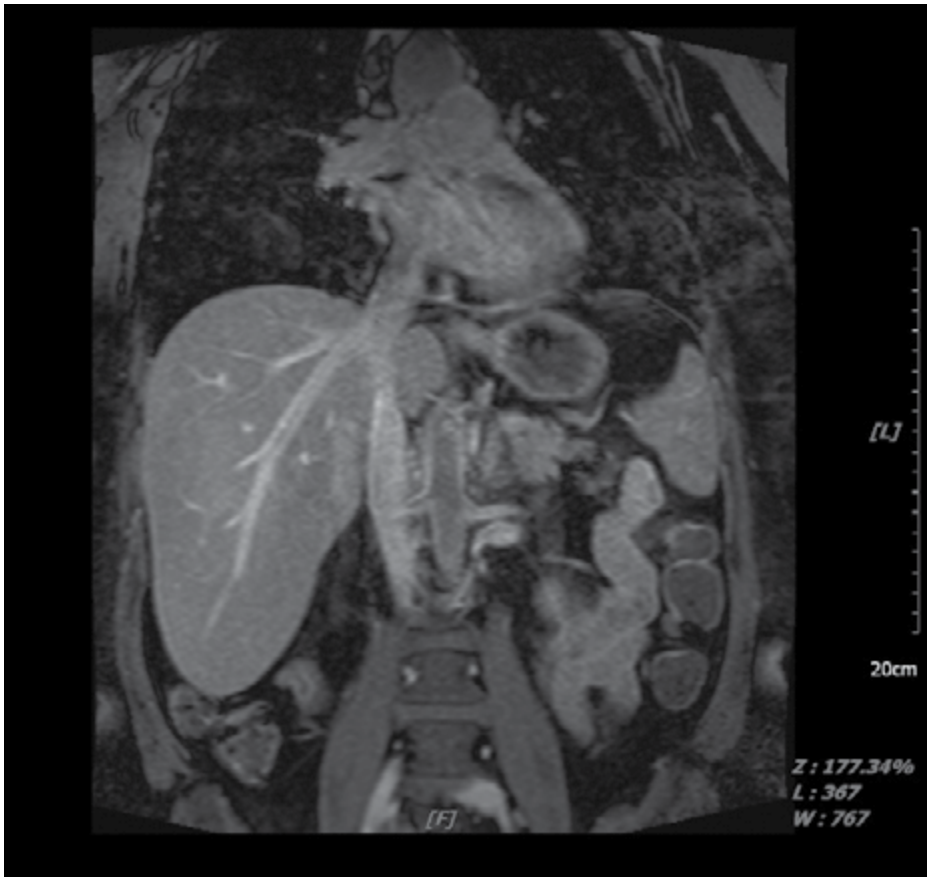

Figure 1. Coronal T1-weighted magnetic resonance image obtained after the administration of gadolinium contrast material showed significant wall thickening of the abdominal aorta and renal arteries

Discussion

Giant cell vasculitis is the vasculitis of great blood vessels, which involves the aorta and/or major aortic branches, mostly the extracranial branches of the carotid artery, including the temporal artery (5). The clinical picture is pre-

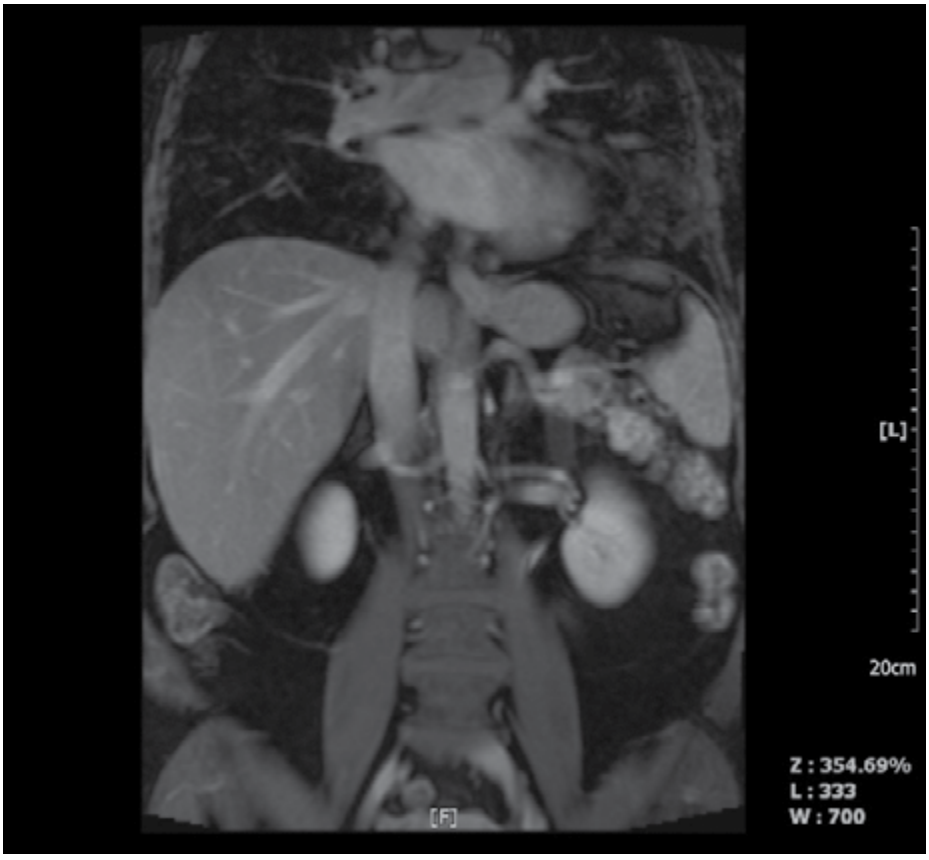

Figure 2. Post-contrast coronal T1-weighted magnetic resonance image of the same patient 1 year later showed normal findings

dominated by general symptoms and symptoms associated with the affected arteries. The diagnosis of giant cell vasculitis is based on clinical, laboratory, imaging, and histological findings and is confirmed by findings of temporal artery biopsy, suggesting vasculitis with mononuclear and giant cells with elements of granulomatous inflammation. Ultrasonography of temporal arteries is a noninvasive method which, in the hands of an experienced physician, can replace biopsy. It should be noted that inflammatory changes involve vascular wall segments and that temporal artery biopsy is negative in 15% of patients with giant cell vasculitis (1). Because giant cell vasculitis involves great blood vessels, the histological verification of giant cell vasculitis is not always possible because great blood vessels (i.e., the aorta and its branches) are unavailable for biopsy. In this situation, Doppler echosonografy is not useful. Recently, more studies point to the role of novel, noninvasive imaging methods in the diagnosis of inflammatory changes of the aorta and its branches. MRI can detect inflammatory changes of the cranial and extracranial arteries and the aorta and its other branches with high diagnostic accuracy (2-4). It is also very useful in the follow-up of therapeutic efficacy and in the detection of possible complications such as aneurysm and dissection (4).

The FDG PET scan also has high sensitivity in detecting inflammatory changes in the wall of great blood vessels in giant cell vasculitis, Takayasu's arteritis, and other arteritides (5-7). The PET scan may be even more efficient than MRI in the detection of initial inflammatory lesions of great vessel walls because of different 18-fluorodeoxyglucosae (18-FDG) accumulation in atherosclerosis compared that in arteritis (6). However, its role in the follow-up of patients with giant cell vasculitis and in detecting complications such as aneurysm and dissection is questionable, in particular following glucocorticoids application; thus, recent studies prefer MRI in patient follow-up (4).

In the case presented, giant cell vasculitis was diagnosed on the basis of history data, clinical examination, laboratory findings, and imaging methods. Because the Doppler ultrasonography of temporal arteries was normal, while large vessel vasculitis was suspected, MRI was performed to clearly visualize the inflammatory lesions of the wall of the abdominal aorta and its branches. However, extrapulmonary tuberculosis was also strongly suspected in our patient based on the information on a family member, who living in the same household, treated for pulmonary tuberculosis, presence of general symptoms in our patient (fatigue, febrility, nocturnal sweating, 20-kg weight loss in 6 months), positive PPD test, echosonographic elements of chronic pericarditis, and erythema induratum of Bazin. Painful erythematous skin lesions on the lower legs were first described by Bazin in 1861 in a young female patient with tuberculosis. These skin lesions histologically correspond to panniculitis and result from the cutaneous immune reaction to Mycobacterium tuberculosis antigen present in the blood (but not in skin lesions). Erythema induratum of Bazin is most frequently (but not exclusively) reported in association with tuberculosis. Although Mycobacterium tuberculosis was not isolated from the patient's sputum and blood, we immediately initiated (ex juvantibus) antituberculous therapy (isoniazid+pyridoxine, rifampin, and pyrazinamide) along with further diagnostic work-up and follow-up. After 4 weeks of antituberculous therapy, there was no clinical improvement; the patient was still subfebrile and exhausted. After 2 months of antituberculous therapy, intravenous administration of high dosage methylprednisolone (500 mg for 3 days) was introduced, followed by 1-mg/kg body weight, which led to considerable improvement. Antituberculous therapy was continued for up to 6 months. While the dosage of methylprednisolone was tapered, methotrexate was introduced after 8 weeks at a dosage of 7.5 mg once a week and was then gradually increased to 20 mg/week.

After 1 year of treatment with methylprednisolone and methotrexate, follow-up MRI showed a normal wall of the abdominal aorta, free from inflammatory changes and aneurysmal dilation. The patient was in clinical remission with normal laboratory parameters, confirming that treatment was efficacious and successful. Therapy with methylprednisolone at a dosage of 8 mg daily, methotrexate at a dosage of 20 mg once a week and folic acid at a dose of 5 mg was continued with regular follow-up.

MRI has a major role in the diagnosis of inflammatory changes of great blood vessels. Additional prospective studies are expected to provide clear data on the sensitivity as well as on the shortcomings of this noninvasive imaging method.

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Informed Consent: Written informed consent was obtained from the patients who participated in this study.

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