Giant cell arteritis as the cause of a chronic fever of unknown origin

Authors: Agnieszka Ciba-Stemplewska, Dorota Krzos, Magdalena Kal, Ewa Pater, Mariola Kleist, Beata Wożakowska-Kapłon

Article type: Clinical image

Received: August 7, 2020.

Accepted: August 10, 2020.

Published online: August 11, 2020.

ISSN: 1897-9483
Giant cell arteritis as the cause of a chronic fever of unknown origin

Agnieszka Ciba-Stemplewska¹, Dorota Krzos², Magdalena Kal³, Ewa Pater⁴, Mariola Kleist⁵, Beata Wożakowska-Kapłon⁶

1 Department of Internal Medicine, Regional Hospital, Kielce, Poland
2 Healthcare Center, Święta Katarzyna, Poland
3 Department of Ophthalmology, Regional Hospital, Kielce, Poland
4 Rheumatology Unit, Regional Hospital, Włoszczowa, Poland
5 Department of Nuclear Medicine with Positron Emission Tomography (PET) Unit, Holy Cross Cancer Centre, Kielce, Poland.
6 First Department of Cardiology and Electrotherapy, Świętokrzyskie Cardiology Centre, Faculty of Medicine and Health Sciences, Jan Kochanowski University, Kielce, Poland

Short title: Giant cell arteritis as the cause of fever

Corresponding author: Dorota Krzos, MD, Healthcare Center, ul. Żeromskiego 5, 26-010 Święta Katarzyna, Poland, phone: +48 41 311 21 18, email: krzosd@wp.pl

Conflict of interest: none declared
Giant cell arteritis (GCA) is an inflammatory disease affecting medium and large arteries. Its incidence is 15 to 25 per 100,000 population, and it usually affects individuals aged above 50 years (typically in their seventh or eighth decade of life).[1,2] The etiopathogenesis involves proinflammatory and autoimmune mechanisms.[3] Patients present with symptoms related to an ongoing inflammatory process (fever, malaise) and ischemia of tissues supplied by the affected vessels. These symptoms may not occur simultaneously. Some patients may display one predominant symptom (e.g., fever), which complicates the diagnosis. A common comorbidity in GCA is polymyalgia rheumatica. The diagnosis is based on ultrasonography (which reveals the characteristic hypoechoic halo sign around the artery lumen), computed tomography (CT), magnetic resonance imaging (MRI), and positron-emission tomography/computed tomography (PET-CT).[4,5]

A 64-year-old man was admitted to the Department of Internal Medicine after 2-month empiric antibiotic therapy due to sustained subfebrile temperature. He reported headache, depressive disorders, and weight loss (6 kg over 3 months). On admission, enlarged neck and axillary lymph nodes with ultrasound features of reactive lymph node enlargement, tooth decay, and left knee bursitis were noted. The patient did not consent to lymph node biopsy. We did not observe arthritis or symptoms of polymyalgia rheumatica. Laboratory tests showed high levels of inflammatory markers (C-reactive protein, 114 ng/ml; erythrocyte sedimentation rate, 120 mm/h), anemia of chronic disease, and thrombocytosis. Multiple blood and urine culture tests were negative. Protein electrophoresis did not show increased γ-globulin and monoclonal protein levels. Typical infections were excluded (hepatitis B and C, HIV, cytomegalovirus, mononucleosis, toxoplasmosis, Lyme disease, and influenza). Antinuclear, proteinase 3, myeloperoxidase, β2-microglobulin, and antiphospholipid antibodies were negative. Chest X-ray showed no inflammatory lesions. Abdominal ultrasound revealed kidney stones, gallbladder polyp, and hepatic cyst. Endoscopy showed no
abnormalities. Due to suspicion of malignancy, imaging studies were performed, including chest, abdominal, and pelvic CT. They revealed enlarged lymph nodes (up to 9 mm) in the mediastinum and lesser curvature (up to 12 × 18 mm), aortic atherosclerosis, kidney stones, hepatic cyst, and a metallic foreign body in the left eye (which precluded head MRI). Chest CT did not reveal features of sarcoidosis. Following a dental consultation, 6 teeth were extracted as a possible source of systemic inflammation. Broad-spectrum antibiotic therapy was started (metronidazole, trimethoprim/sulfamethoxazole, clindamycin). However, subfebrile temperature and the levels of inflammatory markers were not reduced. The patient was referred for PET-CT to a nuclear medicine unit. The examination showed increased glucose metabolism in the aorta and its branches (Figure 1A and 1B). Based on the clinical presentation and extensive workup, GCA was diagnosed. Treatment with intravenous methylprednisolone (0.5 g for 3 days) was administered. Then, oral glucocorticoids were continued at the initial prednisone dose of 60 mg. Additionally, methotrexate at a titrated dose of up to 25 mg/wk was administered. The patient’s general condition improved, with a reduction in the body temperature and levels of inflammatory markers.

We presented a complex case of fever and malaise due to vasculitis. The wide differential diagnosis and numerous recommended imaging studies were inconclusive [3,5]. The cause of nonspecific symptoms and persistently elevated levels of inflammatory markers was ultimately identified by PET-CT, which has high sensitivity and specificity for early GCA diagnosis [4].

References


2 Jennette JC, Falk RJ, Bacon PA, et al. 2012 revised international chapel hill consensus

3 Milchert M, Brzosko M. Pathogenesis of large vessel vasculitis as an example of innate and adaptive immunity relations – case report and literature review [in Polish]. Allergy Asthma Immunology – clinical review. 2018; 23: 29-34.


Figure 1 A and 1B

Positron emission tomography/computed tomography showing abnormal $^{18}$F-fluorodeoxyglucose uptake in the aorta and its branches.