HHS Public Access

Author manuscript

Scand J Rheumatol. Author manuscript; available in PMC 2024 April 29.

Published in final edited form as:

Scand J Rheumatol. 2020 July ; 49(4): 341. doi:10.1080/03009742.2020.1744713.

Anti-neutrophil cytoplasmic antibody-positive vasculitis presenting with periaortitis and muscle vasculitis in a patient with chronic Chagas disease: comment on the letter by Garcia-Bustos et al.

Ester Roffe[1], **Philip Murphy**[1]

[1]Molecular Signaling Section, Laboratory of Molecular Immunology, National Institute of Allergy and Infectious Diseases (NIAID), National Institutes of Health (NIH), Bethesda, MD, USA

To the Editor:

In their interesting Letter entitled **"Anti-neutrophil cytoplasmic antibody-positive vasculitis presenting with periaortitis and muscle vasculitis in a patient with chronic Chagas disease"**, published by the Journal on 11 Feb 2020, Garcia-Bustos et al. describe a previously healthy 53-year-old Bolivian male who had resided in Spain for 15 years, but had traveled to Bolivia 3 months before presenting with acute onset of severe myalgia and fever. In addition to vasculitis, the diagnosis of chronic Chagas disease was made at that time based on a strongly positive IgG serology for *T. cruzi* and the presence of asymptomatic megacolon. This case raises two interesting issues, one related to when the patient became infected, and one relating to the natural history of the disease.

On the first point, we suspect the patient was most likely not initially infected during his recent trip to Bolivia but instead when he was resident there at least 15 years earlier. This aligns with the known natural history of the disease in which the chronic symptomatic forms, cardiomyopathy, megaesophagus and megacolon, all take 10 to 30 years to develop. Moreover, approximately 70% of *Trypanosoma cruzi* seropositive individuals remain asymptomatic throughout life, and most patients, especially emigrants from Latin America, are unaware that they are infected. Of all Latin American countries, Bolivia has had the highest prevalence of Chagas disease for the past four decades (Moncayo, 1999; OPS, 2006; WHO, 2015; Lidani et al., 2019). Nevertheless, since the patient presented with fever, it's possible that he was reinfected with T. cruzi during the recent trip to Bolivia. In support of this, chronically infected mice have been reported to present with exacerbated electrocardiographic symptoms, enhanced myocarditis and myositis and increased mortality after experimental reinfection (Bustamante et al., 2002; Bustamante et al., 2004; Andrade et al., 2006; Reis Machado et al., 2014).

On the second point, the patient's presentation with vasculitis is a particularly interesting feature of the case and, to our knowledge, represents the first report of vasculitis in a Chagas disease patient. Whether *T. cruzi* caused the patient's vasculitis and whether vasculitis may occur subclinically in other patients with Chagas disease cannot be determined from single case report. Nevertheless, we have found that chronic experimental infection with T. cruzi can cause paralyzing systemic necrotizing vasculitis in mice by a mechanism involving

pathogen-specific type I immunity (Roffê et al., 2016). The skeletal muscle lesions are very similar in appearance to those of the patient, although antibody deposits and neutrophilic inflammation were not features of the mouse lesions. Nevertheless, based on the obvious similarities, we propose that Chagas disease should be considered as part of the differential diagnosis for patients presenting with vasculitis, and vasculitis should be considered as a potential cause of myalgia in patients with Chagas disease.

References:

1. Garcia-Bustos V, Calabuig E, López-Aldeguer J, Moral Moral P. Anti-neutrophil cytoplasmic antibody-positive vasculitis presenting with periaortitis and muscle vasculitis in a patient with chronic Chagas disease. Scand J Rheumatol. 2020 Feb 11:1–2.
2. Moncayo A. Progress towards interruption of transmission of Chagas disease. Mem Inst Oswaldo Cruz. 1999;94 Suppl 1:401–4. [PubMed: 10677765]
3. Pan American Health Organization. Estimación cuantitativa de la enfermedad de Chagas en las Américas. [Quantitative estimation of Chagas Disease in the Americas]. Report No.: OPS/HDM/CD/425-06. Montevideo: PAHO (2006).
4. Chagas disease in Latin America: an epidemiological update based on 2010 estimates. Wkly Epidemiol Rec 2015; 90: 33-43. [PubMed: 25671846]
5. Lidani KCF, Andrade FA, Bavia L, Damasceno FS, Beltrame MH, Messias-Reason IJ, Sandri TL. Chagas Disease: From Discovery to a Worldwide Health Problem. Front Public Health. 2019 Jul 2;7:166. [PubMed: 31312626]
6. Bustamante JM, Rivarola HW, Fernández AR, Enders JE, Fretes R, Palma JA, Paglini-Oliva PA. Trypanosoma cruzi reinfections in mice determine the severity of cardiac damage. Int J Parasitol. 2002 Jun 15;32(7):889–96. [PubMed: 12062560]
7. Bustamante JM, Rivarola HW, Palma JA, Paglini-Oliva PA. Electrocardiographic characterization in Trypanosoma cruzi reinfected mice. Parasitology. 2004 Apr;128(Pt 4):415–9. [PubMed: 15151147]
8. Andrade SG, Campos RF, Sobral KS, Magalhães JB, Guedes RS, Guerreiro ML. Reinfections with strains of Trypanosoma cruzi, of different biodemes as a factor of aggravation of myocarditis and myositis in mice. Rev Soc Bras Med Trop. 2006 Jan-Feb;39(1):1–8. [PubMed: 16501758]
9. Reis Machado J, Silva MV, Borges DC, da Silva CA, Ramirez LE, dos Reis MA, Castellano LR, Rodrigues V, Rodrigues DB. Immunopathological aspects of experimental Trypanosoma cruzi reinfections. Biomed Res Int. 2014:648715. [PubMed: 25050370]
10. Roffê E, Marino APMP, Weaver J, Wan W, de Araújo FF, Hoffman V, Santiago HC, Murphy PM. Trypanosoma cruzi Causes Paralyzing Systemic Necrotizing Vasculitis Driven by Pathogen-Specific Type I Immunity in Mice. Infect Immun. 2016 Mar 24;84(4):1123–1136. [PubMed: 26857570]