REFRACTORY LIMB-THREATENING ISCHEMIA IN BEHÇET'S DISEASE TRIGGERED BY CHIKUNGUNYA INFECTION

Victoria Dornelas¹, Maria Cecília Dias Corrêa¹, Pedro Laurindo Fiorio Junior¹, Ana Cláudia Guimarães Carvalho¹, Guilherme Bicalho Civinelli de Almeida¹, Ronald Kleinsorge Ronald¹, Viviane Angelina Souza¹, Gustavo Guimarães Moreira Balbi^{1,*}

¹Universidade Federal de Juiz de Fora, Juiz de Fora (MG), Brazil.

*Corresponding author: ggmbalbi@gmail.com

BACKGROUND

Behçet's disease (BD) is a systemic autoimmune disease that may cause variable vessel vasculitis. Even though medium vessel vasculitis may occur, limb ischemia is seldom reported. In cases with limb-threatening ongoing ischemia, high-grade immunosuppression is the cornerstone treatment. The use of hyperbaric oxygen therapy (HOT) has never been reported in this context. We hereby report the case of a patient previously diagnosed with BD evolving with refractory limb ischemia after chikungunya infection that was treated with HOT.

CASE REPORT

A 22-year-old female patient was referred to our outpatient vasculitis clinic in 2019, due to oral and genital ulcers, pseudofolliculitis, erythema nodosum and abdominal pain. She was diagnosed with BD and treated with prednisone, colchicine and azathioprine, with good response. In 2021, she was admitted with high-grade fever (up to 40 °C), diffuse myalgia and fixed cyanosis in hands and feet, associated with oral ulcers and abdominal pain. Laboratory workup revealed markedly increased levels of ESR, CRP, direct bilirubin, CPK and leukocytes. At this time, both hemorrhagic fever and infective endocarditis were suspected, and empirical antibiotic treatment was started. Also, prednisone dose was increased to 60 mg once daily. Specific workup confirmed chikungunya infection (reagent IgM and nonreactive IgG); transesophageal echocardiogram and serology for dengue, Zika virus, leptospiral and rickettsia infection were unremarkable. During inpatient care, limb ischemia has worsened despite treatment, with progressive cyanosis of the left foot, and necrosis of the right first and second finger phalanges, left first and fifth finger phalanges and right forefoot. Pulse therapy with methylprednisolone and cyclophosphamide, intravenous vasodilation with alprostadil and therapeutic anticoagulation were started, with poor therapeutic response. Diagnostic angiogram showed occlusion of all tibial artery and absent right plantar arch. After multidisciplinary discussion with the vascular surgery team, HOT (initially 20 sessions of 90 min – 2.5 ATM, with 100% oxygen concentration) was initiated as an attempt to allow a more conservative amputation (necrotic region vs. infrapatellar amputation of the right lower limb). After that, perfusion, cyanosis and ischemic pain improved in both feet, and she underwent amputation of the right forefoot. After 4 weeks of outpatient care, she was still on HOT, with a satisfactory outcome.

CONCLUSION

To the best of our knowledge, this is the first description of chikungunya infection triggering BD activity. Also, this is the first report of HOT successfully used to avoid major amputation in limb-threatening ischemia in BD.

KEYWORDS

Behçet's disease, Behçet's syndrome, Systemic vasculitides, Chikungunya, Hyperbaric oxygen therapy.