Systemic Sclerosis after Bone Marrow Transplantation: A Case with Evidence of Donor-related Disease Transfer

To the Editor:

A 34-year-old man was diagnosed in January 2002 with acute B lymphoblastic leukemia. After induction chemotherapy with vincristine and daunorubicin, complete remission was achieved. In October 2011 a bone marrow relapse was detected. He was treated with clofarabine, attaining morphological remission with minimal residual disease, and in February 2012 he received a haplo-identical allogeneic bone marrow transplantation (BMT) from his mother, achieving a second complete remission. Graft versus host disease (GVHD) prophylaxis consisted of cyclophosphamide on days +3 and +4, short-term mycophenolate mofetil, and cyclosporine as maintenance therapy. Complete donor chimerism was seen from Day +30 and has been maintained since.

Ninety days after BMT, he developed a maculopapular rash with > 50% of the skin affected, diagnosed as acute GVHD grade II. He required prolonged corticosteroid treatment until clinical resolution. In March 2013 he presented with telangiectasia on his face, forearms, and abdomen, and dry eye and dry mouth that were attributed to chronic GVHD. Only artificial tears were added to the treatment with cyclosporine. In November 2016 a digital ulcer appeared on the index finger of his left hand. He was treated with nifedipine 5 mg twice a day and topical tacrolimus and betamethasone, without improvement.

On his first rheumatology visit in November 2017 he presented with 2 necrotic and 2 cicatricial ulcers (Figure 1), and telangiectasia on his face and neckline. Skin thickening was not observed but he complained about Raynaud phenomenon (RP) since the BMT.

He had positive antinuclear antibodies (ANA) at a titer of 1/640 with centromere pattern, anticientromere antibodies (ACA), rheumatoid factor (RF; 652 IU/ml), and a mild consumption of C3 (74.3 mg/dl). Prior to BMT, his ANA status was negative. On the nailfold videocapillaroscopy (NVC) we observed an active scleroderma pattern, with frequent giant capillaries, microhemorrhages and moderate loss of capillaries1 (Figure 2). He exhibited mild dysphagia, but his decision was for organ involvement not to be studied. He fulfilled the 2013 systemic sclerosis (SSc) American College of Rheumatology (ACR)/European League Against Rheumatism (EULAR) diagnostic criteria2 on the basis of digital ulcers, pitting scars, telangiectasia, abnormal nailfold capillaries, RP, and positive ACA.

He had no familial background of connective tissue diseases, but his mother had RP since her youth. On physical examination she had facial telangiectasia, sclerodactyly (modified Rodnan skin score of 4), and fingertip pitting scars, for which she had never sought treatment. She had positive ANA at titers of 1/320 with centromere pattern, ACA, RF (159 IU/ml), and complement consumption (C3 = 74.3 mg/dl, C4 = 7.6 mg/dl). On the NVC we observed a late scleroderma pattern, with severe loss of capillaries, disorganization of the normal capillary array, and ramified/busy capillaries3. She did not present symptoms of organ involvement. She was diagnosed with limited cutaneous SSc and fulfilled the 2013 ACR/EULAR diagnostic criteria2 on the basis of sclerodactyly, pitting scars, telangiectasia, abnormal nailfold capillaries, RP, and positive ACA.

Ethics board approval was obtained for publication and diffusion of this case report from the Ethical Committee of the Hospital General Universitario Gregorio Marañón, Madrid, Spain. Address correspondence to Dr. A. Silva Riveiro, Department of Rheumatology, Hospital General Universitario Gregorio Marañón, Calle del Doctor Esquerdo 46, 28009, Madrid, Spain. E-mail: alicia.silvariveiro@gmail.com

REFERENCES


Figure 1. Necrotic ulcers on the index finger of the left hand and on the middle finger of the right hand. Cicatricial ulcers on the middle finger of the left hand and on the index finger of the right hand.

Figure 2. Image of nailfold videocapillaroscopy. Moderate loss of capillaries and a giant capillary with remains of a microhemorrhage.