A 74-year-old woman with no relevant past medical history presented with lesions of 4 years’ duration that had started in the submammary region and extended to the presternal and periorbital region. The patient had been administered corticosteroids but there had been no improvement. She also reported chronic asthenia and exercise intolerance. The physical examination showed well-circumscribed, yellowish-orange, xanthomatous plaques with palpable, erythematous edges. The central area of these lesions showed marked atrophy and telangiectasias (Fig. 1). The general blood analysis with serology, autoimmunity tests, and protein profile showed an increase in immunoglobulin G (3060) with free κ chains (4.79). Positron emission tomography and bone marrow studies were normal. The skin biopsy results confirmed a diagnosis of necrobiotic xanthogranuloma associated with monoclonal gammopathy of undetermined significance.

Necrobiotic xanthogranuloma is an uncommon entity classified within the histiocytoses. Clinically, it tends to present as indurated red-violaceous papules and plaques that are variable in location although they are mostly found in the periorbital region. Prognosis depends on the severity of the lesions and level of extracutaneous involvement, because, as seen with our patient, necrobiotic xanthogranuloma sometimes occurs in association with gammapathies or lymphoproliferative disorders.