Abstract
Systemic sclerosis (SSc) is an autoimmune connective tissue disease characterized by cutaneous and visceral fibrosis and widespread vascular pathology. Raynaud's phenomenon is one of the most common manifestations of SSc. It may lead to complications such as digital ulceration and infarction and its treatment remains elusive. We present the case of a female patient with severe digital ischemia secondary to SSc. Treatment using vasodilators and prostacyclin analogues was ineffective, but the patient experienced a remarkable response to bosentan, a dual endothelin receptor antagonist. This report suggests that endothelin antagonists may benefit patients with SSc and vasculopathy as a major feature of the disease, including limb ischemia.

Keywords
Systemic sclerosis, digital ischemia, bosentan.