

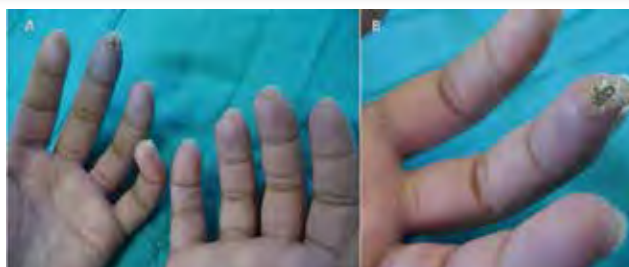
## Úlceras digitais numa doente com esclerose sistémica

### *Digital ulcers in a systemic sclerosis patient*

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Female patient, 22 years- old, followed in the Internal Medicine Outpatient clinic with a diagnosis of Limited Cutaneous Systemic Sclerosis (LCSS). At first she had frequent episodes of Raynaud's phenomenon. Capillaroscopy has detected the presence of dilated capillary vessels and areas of devascularization, and the lab analyses showed positive anticentromere antibodies. She was medicated with angiotensin receptors antagonists (ARA) and platelet antiaggregants. In December 2008 she started showing edema in the hands and digital cutaneous ulcers on the 3<sup>rd</sup> finger of the left hand, and on the 2<sup>nd</sup> and 3<sup>rd</sup> fingers of the right hand (Fig. 1). These were refractory to treatment with ARA and calcium channels antagonists in the maximum doses the patient could tolerate. She had complaints of intense pain in the fingers pulps using often painkillers with NSAIDs. In February she started therapy with bosentan, an endothelin receptor antagonist (ERA). By the end of the first two weeks, the pain was slowing down and by the end of 8 weeks a total healing of digital ulcers occurred (Fig. 2). After 12 months of treatment, no recurrence happened, nor new ulcers. The hands edema was kept stable and the patient was kept in therapy with ERA. No side effects were seen.

System Sclerosis (SS) is an autoimmune disease of the connective tissue characterized by diffuse vasculopathy, cutaneous and visceral fibrosis. Digital cutaneous ulcers are one the main complications of peripheral microvasculopathy in SS. The etiopathogeny of vasculopathy in SS is unknown, but it is thought to be related with a lesion in the endothelial



A) Digital cutaneous ulcers: 3<sup>rd</sup> finger of the left hand, 2<sup>nd</sup> and 3<sup>rd</sup> fingers of the right hand. B) Cutaneous ulcer on the 3<sup>rd</sup> finger of the left hand

FIG. 1



Total healing of the digital ulcers after 8 weeks of having started treatment with bosentan.

FIG. 2

cell,<sup>1,2</sup> with a loss of vasodilation mediators including prostacyclin and nitric oxide<sup>3</sup> and an increase on vasoconstrictors production, as endothelin<sup>4</sup> binding to two kinds of receptors (ET<sub>A</sub> and ET<sub>B</sub>). Bosentan is an ERA, with a preferential action at the level of ET<sub>A</sub> receptors, reducing vasoconstriction mediated by endogenous endothelin.<sup>5</sup> It is used in the treatment of pulmonary arterial hypertension both idiopathic

Internal Medicine Service of Coimbra University Hospitals  
Received for publication on the 31st March 2011  
Accepted for publication on the 10th March 2011

as secondary to scleroderma since 2007, its use in the prevention of SS digital ulcers has been approved by INFARMED. Such approval was based in the result of two studies (RAPIDS-1 and RAPIDS-2) which have revealed that bosentan was associated with a 48% reduction in the average number of new ulcers<sup>5</sup> and such effect was higher in patients presenting more than three multiple ulcers.<sup>6</sup> The authors take the view the case presented cannot prove that bosentan has helped the treatment of digital ulcers in this patient, although it helped with the healing whilst avoiding the occurrence of new ulcers. ■

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