

Endothelin receptor antagonists and scleroderma related digital ulcers

Wha-Yong Lee¹, Shoaib Faruqi¹, Alyn H Morice¹

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An 84-year-old woman with scleroderma presented with recurrent non-healing digital ulcers despite treatment with iloprost. A year later she developed pulmonary hypertension and treatment with bosentan was initiated. Although there was mild improvement in her symptoms related to pulmonary hypertension, the digital ulcers did not improve at all (Figures 1A, 1B). Another year later, in view of lack of sustained response to bosentan in symptoms related to pulmonary hypertension, she was commenced on sitaxentan 100 mg once daily. In 2 months, remarkably her digital ulcers had almost healed completely (Figures 2A, 2B) and her symptoms of pulmonary hypertension also improved. She tolerated her treatment well and had no further recurrence of digital ulcers.

Digital ulcers (DUs) in scleroderma result from recurrent Raynaud's phenomenon and microtrauma. Vasodilatory agents such as calcium channel blockers and prostaglandin derivatives have been used to treat scleroderma related DUs, but with the advent of selective and non-selective endothelin receptor antagonists (ETRA) new therapeutic options are available. Bosentan, a non-selective ETRA has been shown to reduce the number of new ulcers and improve ulcer healing long term¹ but this was not observed in our case. Sitaxentan is a highly selective ETRA, and has been shown in only two previous reports to improve refractory digital ulcer healing, similar to our patient^{2,3}. Our case is unique as non-selective ETRA did not result in improvement earlier. In December 2010, sitaxentan was withdrawn from the market due to two cases of fatal liver injury and is currently not available for prescription. Other selective ERTAs such as ambrisentan could be considered as an alternative to sitaxentan, and in cases where non-selective ETRA are ineffective. However, further randomised control trials are necessary to bet-



FIGURE 1A, 1B. Right index finger of the patient with digital ulcers before treatment with sitaxentan



FIGURE 2A, 2B. Improvement of the digital ulcers after treatment with sitaxentan

1. Division of Cardiovascular and Respiratory Studies, Hull York Medical School, University of Hull, Castle Hill Hospital, Cottingham, United Kingdom

ter assess the role of selective and non-selective ETRAs for the treatment DUs in scleroderma.

CORRESPONDENCE TO

Wha-Yong Lee
Department of Respiratory Medicine,
Castle Hill Hospital, Cottingham,
HU16 5JQ, United Kingdom
E-mail: Wha-Yong.Lee@hey.nhs.uk

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