Sir

We present a case of adult urticaria pigmentosa: maculo-papular type- with temporary disappearance of the lesions during treatment with Enoxaparinum.

History
A 52-year-old female patient, with a 20 years history of asymptomatic, erythematous-to-brown macules and papules on the trunk, neck, buttocks and extremities, presented in our department a few months ago searching for a diagnosis (Fig. 1). Her medical problems were: an arterial hypertension (controled with Indapamidum) and osteoporosis (with no medication for).

Physical Examination
Scattered, erythematous, edematous papules and brown macules were present on the neck, chest, abdomen, back, extremities and buttocks. The face, palms, soles, and genitals were spared.

Lab
A complete blood count, basic metabolic profile, hepatic and lipid panels were within normal limits and we excluded systemic involvement. A bone density study showed osteoporosis. Serum tryptase levels, 24 hour urinary N-methylhistamine, N-methylimidazolacetic acid and prostaglandin D2 metabolites excretion were within normal limits.

Skin biopsy confirmed the diagnosis of generalized cutaneous mastocytosis (Urticaria pigmentosa) (Fig. 2A-D).

The patient left the Dermatology Unit with no medication, but she called us, a few weeks later for a new appointment. She described and we confirmed the disappearance of the cutaneous lesions during the last weeks, while she was hospitalised for a hip fracture and treated with Enoxaparinum 40mg s.c/daily for 14 days. The patient refused a new biopsy and we saw her again three months later, she again showed the characteristic brownish-red skin lesions of Urticaria pigmentosa, exactly as at the first appointment. The lesions had begun to appear very soon after she had stopped taking Enoxaparinum (Fig. 3).

Figure 1. Yellow-tan to reddish-brown macules and slightly raised papules scattered over the trunk and extremities.
Discussions

In the mast cell granules, tryptase is stored in complex with negatively charged heparin proteoglycans. Apart from the critical role of heparin proteoglycan in storage of tryptase in the secretory granules, heparin has been implicated in the autocatalytic processing of protryptase into mature tryptase monomer (Sakai). It has been known for a long time that heparin is required for stabilization of the mature tryptase tetramer (Schwartz). Small heparine molecules, as is Enoxaparinum, in excess, could block/interfere with H-receptors family in a way that would prevent further degranulation of mastocytes. This case report is the first observation in the literature regarding the transitory favorable effect of Heparine administration on the evolution of adult urticaria pigmentosa lesions. Further studies are needed to confirm or not our observation.

Figure 2. A. Sparse infiltrate with mast cells perivascular. (HE stain x200); B. Hyperpigmentation of the basal layer of the epidermis. (H&E stain x400); C. Mast cells in the papillary dermis. (Giemsa stain x100); D. Round and spindle-shaped mast cells. (Giemsa stain x400)

Figure 3. A slight hyperpigmentation scattered just in a few places on the trunk.