

Correspondence

Chronic autoimmune urticaria in a patient with multiple piloleiomyomas

doi: 10.1111/j.1365-2230.2007.02400.x

A 67-year-old woman was referred to our dermatological unit for recurring urticaria–angio-oedema episodes that had begun in 1986, and had been unsuccessfully treated with antihistamines.

The patient's medical history revealed hypertension under management with beta-blocking agents. She had no history of urticaria-triggering drugs. She had undergone laparohysterectomy for uterine leiomyomas in 1986.

Upon physical examination, multiple, firm, red-brownish grouped nodules, a few millimetres in diameter, were evident on the shoulders, presternal region, back and lumbosacral surface (Fig. 1). The nodules had gradually developed since 1986, concomitantly with the beginning of the urticaria, and had persisted unchanged.

The only abnormality revealed by laboratory assays was the titre of antinuclear autoantibodies (ANA 1 : 80). In order to gain further information about the aetiopathogenesis of the urticaria, we performed an autologous serum

skin test and *in vitro* histamine release using basophils from healthy donors, which gave a positive result.

Histopathological examination of a cutaneous nodule of the shoulder showed circumscribed, nonencapsulated dermal bundles of smooth muscle arranged in an interlacing and whorled pattern. The cells showed elongated nuclei with blunt ends. Mitosis was not seen. The histopathological features were consistent with cutaneous piloleiomyoma (Fig. 2). A diagnosis of Reed's syndrome associated with chronic autoimmune urticaria (CAU) was made.

Chronic idiopathic urticaria (CIU) is a form of long-lasting urticaria, the pathogenesis of which is still poorly understood. The ability of an intradermal injection of autologous serum to induce a weal-and-flare response in about two-thirds of CIU patients, attributed to the activity of different types of functional circulating autoantibodies, led to the hypothesis that at least some forms of CIU could be sustained by an autoimmune process.¹ Fagiolo² suggested that skin reactivity to autologous serum could be due to an as yet unidentified non-Ig serum factor present in the sera of patients with CIU, which might be related to the coagulation–fibrinolytic system. We reported the case of a middle-aged woman affected by CAU associated with multiple piloleiomyomas.

Cutaneous leiomyoma is a rare benign tumour derived from smooth muscle, which can develop in the erector pili



Figure 1 Multiple grouped piloleiomyomas of the left shoulder.

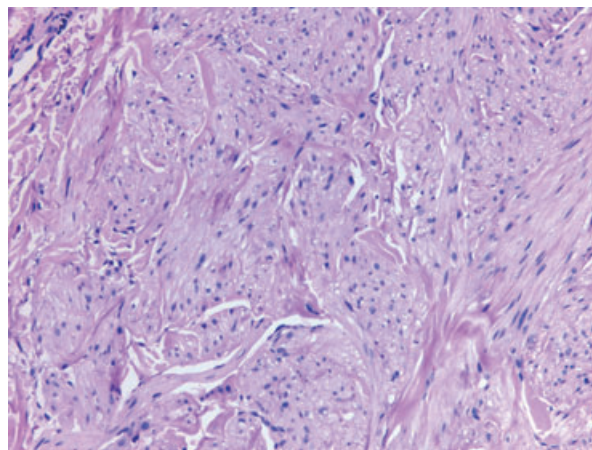


Figure 2 The reticular dermis is replaced by interlacing bundles of smooth muscle cells (haematoxylin and eosin, original magnification $\times 20$).

muscles, in the specialized muscles of the genital skin (including the scrotum, vulva and nipples) and in the walls of blood vessels.³ It can be inherited as an autosomal dominant trait, or can occur sporadically.⁴ The genetic locus for multiple piloleiomyomas has been mapped to chromosome 1q42.3–43, and germline mutations in the fumarate hydratase gene have been identified.⁵

The tumour can occur in association with polycythaemia (as the tumour may produce erythropoietin), papillary renal carcinoma, myeloid leukaemia and uterine leiomyomas.⁴ The latter association is known as Reed's syndrome.⁴

Although piloleiomyoma represents a benign condition, it may suggest eventual visceral involvement. Patients should be warned about the risks of gynaecological complications that may require an early hysterectomy.

Although the occurrence of CAU with piloleiomyomas is probably unrelated, it could be speculated that tumour activity might lead to the production of an unidentified circulating mediator able to provoke a typical urticarial response, in keeping with Fagiolo's proposal, and analogous to what happens in polycythaemia induced by piloleiomyoma-released erythropoietin.²

L. Melani, E. Schincaglia, E. Antiga, M. Caproni, D. Massi,* and P. Fabbri

Departments of Dermatological Sciences, and *Human Pathology and Oncology, University of Florence, Via della Pergola 58, 50121, Florence, Italy

E-mail: fabbri@unifi.it

Conflict of interest: none declared.

Accepted for publication 2 November 2006

References

- 1 Sabroe RA, Fiebiger E, Francis DM *et al.* Classification of anti-FcepsilonRI and anti-IgE autoantibodies in chronic idiopathic urticaria and correlation with disease severity. *J Allergy Clin Immunol* 2002; **110**: 492–9.
- 2 Fagiolo U, Kricek F, Peserico A *et al.* Effects of complement inactivation and IgG depletion on skin reactivity to autologous serum in chronic idiopathic urticaria. *J Allergy Clin Immunol* 2000; **106**: 567–72.
- 3 Alper M, Parlak AH, Kavak A *et al.* Bilateral multiple piloleiomyomas on the breast. *Breast* 2004; **13**: 146–8.
- 4 Carey-Remonnay S, Salard D, Algros MP, Laurent R. Multiple familial cutaneous leiomyoma. *Ann Dermatol Venereol* 2003; **130**: 1017–20.
- 5 Chuang GS, Martinez-Mir A, Engler DE *et al.* Multiple cutaneous and uterine leiomyomata resulting from missense mutations in the fumarate hydratase gene. *Clin Exp Dermatol* 2006; **31**: 118–21.