

cannot be used, monitoring of serum and urinary silver levels is recommended.<sup>9</sup>

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Key words: argyria, epidermolysis bullosa

Conflicts of interest: none declared.

## Rapid diagnosis of monilethrix using dermoscopy

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SIR, Monilethrix (from the Latin for 'necklace' and the Greek for 'hair') is a rare autosomal dominant disease of the hair shaft characterized by small elliptical nodes of normal thickness that are regularly separated by dystrophic constrictions. The lanugo hair is usually clinically normal, and thus an affected newborn will present normal-appearing scalp hair. Only when it is shed during the first few weeks of life does the regrown mature hair show the anomaly.<sup>1</sup> In the mildest form the disease involves only the occiput and the nape of the neck, but in its severe form the entire scalp, secondary hairs, eyebrows and eyelashes may also be involved. Occasionally, regrowth of apparently normal hair may occur at the time of puberty or during pregnancy.<sup>2</sup> Ultrastructurally, vacuolation and altera-

tions in the fibrillar structures of lower cortex cells have been described.<sup>3</sup> Clinical features evident to the naked eye may be limited, especially in the milder form. In order to avoid misdiagnosis, several investigations have been made in this hair disorder using light microscopy, scanning electron microscopy, transmission electron microscopy and autoradiography. Dermoscopy without immersion gel was referred to as 'dry dermoscopy', which was reportedly utilized to examine dandruff adherence to hair,<sup>4</sup> skin weathering and ashiness,<sup>5</sup> and skin surface patterns of xerosis.<sup>6</sup> Despite this main application, it has also been described as an aid to diagnosis and treatment follow-up of hair and scalp disorders, such as androgenetic alopecia, alopecia areata, lipoedematous alopecia or pediculosis.<sup>7,8</sup> Studies on a few entities suggest that clinical accuracy may be better with dermoscopy than with the unaided eye. In addition, novel features of the disease have been discovered.

A girl aged 1 year and 10 months visited us with diffuse hair loss which was noted by her family since a few months after birth. Physical examination showed sparse, fine, black, brittle scalp hair and an erythematous patch (Fig. 1). Her eyebrows and eyelashes appeared normal. Dermoscopy (DermLite II PRO HR; 3Gen, LLC, San Juan Capistrano, CA, U.S.A.) of the hair revealed uniform elliptical nodes and multiple constrictions (Fig. 2a,b). The nails and teeth appeared normal. A diagnosis of monilethrix was made and was confirmed by further examination with light microscopy which showed characteristic alternating nodes and cracked areas on the thin part of the hair shaft, as well as a tendency of hair shafts to fracture at the sites of these constrictions (Fig. 3). The girl was born by uneventful normal spontaneous delivery. Both parents had normal hair and the personal and family history were noncontributory. No evidence of developmental delay, mental retardation, epilepsy or systemic diseases has been noted to date.

Pseudomonilethrix should be ruled out before establishing the diagnosis of monilethrix. Some authors suggest that pseudomonilethrix is an autosomal dominant inherited nodal



Fig 1. A girl aged 1 year and 10 months having sparse, fine, black, brittle scalp hair and an erythematous patch. Diffuse hair loss, particularly at the occipital scalp, was noted.

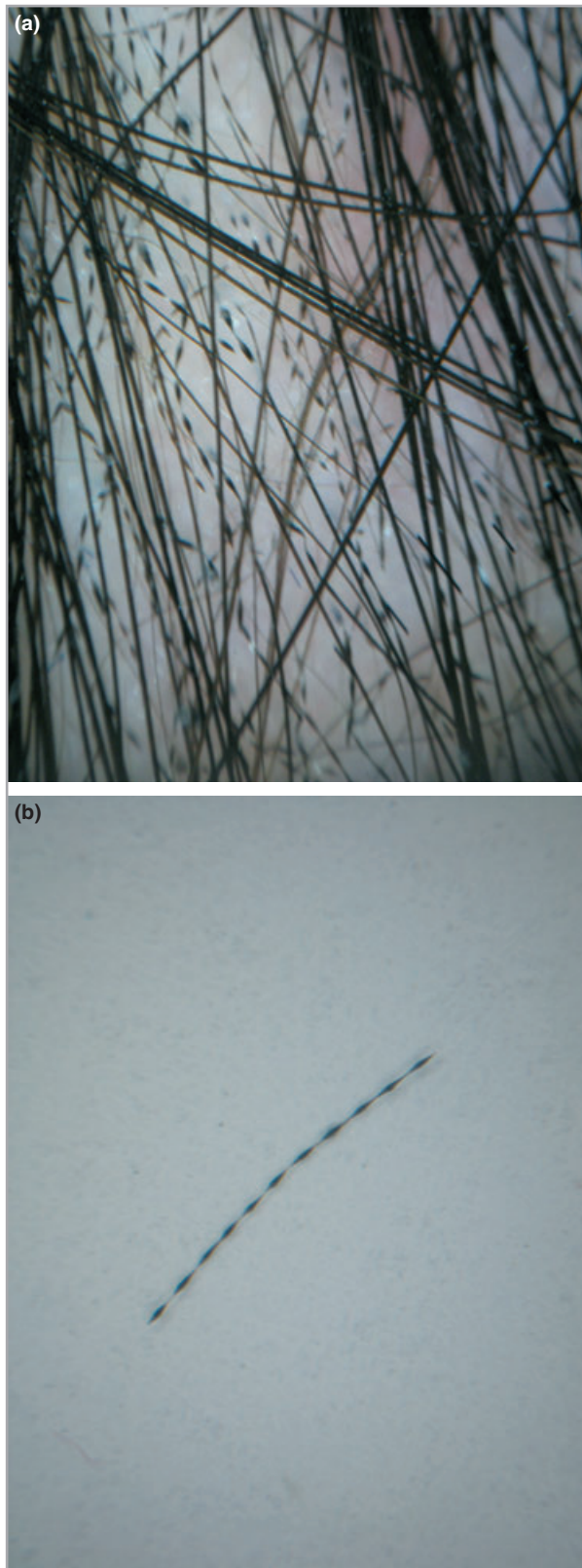


Fig 2. (a) Dermoscopy of the hair revealed uniform elliptical nodes and multiple constrictions. (b) A closer dermoscopic view showed a hair shaft characterized by small elliptical nodes of normal thickness, regularly separated by dystrophic constrictions.



Fig 3. Light microscopy showed characteristic alternating nodes and cracked areas on the thin part of the hair shaft, as well as a tendency of hair shafts to fracture at the sites of these constrictions.

trichodystrophy with late onset,<sup>9</sup> while others suggest iatrogenic pseudomonilethrix as a condition produced by the procedure of preparing hairs for microscopic examination which occurs when hairs are pressed together between glass slides. It appears as irregularly constricted hair shafts, depending on the pressure applied or on the presence of cosmetic haircare products at the visualized site under dermoscopy. Irregularity of intermittent constrictions and lack of tendency to break at constricted sites is consistent with light microscopy findings in pseudomonilethrix.<sup>10</sup>

The diagnosis of monilethrix is based on demonstrating beaded hairs with periodic narrowing under light microscopy. However, numerous hairs have to be removed for microscopic evaluation and most dermatologists will not perform microscopic examination on site in their practice. Besides, despite characteristic microscopic findings, the disease is easily overlooked in a dermatological practice, especially in milder cases. Dermoscopy was much easier and less time consuming than *ex vivo* microscopic examination, and the characteristic dermoscopic features enabled us to establish a rapid diagnosis. Further, misdiagnosis as iatrogenic pseudomonilethrix, which is caused by overlapping hairs pressed under glass slides prepared for microscopic examination,<sup>10</sup> could be prevented by *in vivo* dermoscopy. Therefore, dermoscopy may provide an alternative to standard light microscopy for examination of affected hairs.

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## Lower serum dehydroepiandrosterone sulphate concentration in chronic idiopathic urticaria: a secondary transient phenomenon?

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SIR, In some cases urticaria is associated with hormonal changes, including thyroid dysfunction<sup>1</sup> and menstrual hormonal alterations.<sup>2</sup> In addition, we have demonstrated for the first time that chronic urticaria may be accompanied by lower serum dehydroepiandrosterone sulphate (DHEA-S) concentration, yet its significance and underlying mechanisms are unclear.<sup>3–5</sup> However, it seems that the decline in DHEA-S observed in patients with chronic urticaria may be a secondary

phenomenon resulting from psychological distress.<sup>5</sup> DHEA-S is the major adrenal androgen converted into active DHEA hormone which may exert multiple immunomodulating effects.<sup>6</sup> Decreased serum concentration of DHEA-S can be observed in chronic inflammatory and immune-mediated diseases as well as in stress.<sup>6,7</sup>

Natural clinical remission is frequently observed in chronic urticaria.<sup>8</sup> As chronic urticaria may be associated with lower serum DHEA-S concentration in the active period of the disease, we wanted to determine whether such a phenomenon is present when the disease reaches the clinical spontaneous remission phase. Therefore, serum DHEA-S concentrations were analysed in patients with symptomatic chronic idiopathic urticaria (CIU) and compared with levels in the same patients with CIU upon remission as well as in healthy controls.

The study group consisted of 32 patients with CIU (15 men, median age 34 years, range 18–52; and 17 women, median age 34 years, range 19–44), all identified causes of urticaria having been excluded by appropriate investigation. All patients showed a negative response to an autologous serum skin test (ASST). The ASST was performed according to the method described by Sabroe et al.<sup>9</sup> Patients were examined twice: first, during the symptomatic period, and next, during complete spontaneous remission lasting for at least 4 months (range 4–6 months) without any medication.

The control group consisted of 40 healthy subjects (20 men, median age 34.5 years, range 18–48; and 20 women, median age 33 years, range 18–44). Serum concentration of DHEA-S was measured by automatic electrochemiluminescence immunoassay (ECLIA; Roche Diagnostics, Mannheim, Germany). The Mann–Whitney test was employed to assess differences between patients and healthy controls. Wilcoxon's paired test was employed to compare the urticaria groups.

DHEA-S serum concentration was significantly lower in patients with active CIU as compared with those with CIU in remission and with the healthy subjects. Furthermore, DHEA-S serum concentration during the remission period was similar to that of the healthy controls (Table 1).

**Table 1** Serum dehydroepiandrosterone sulphate (DHEA-S) concentration ( $\mu\text{g dL}^{-1}$ ) in patients with chronic idiopathic urticaria (CIU) during symptomatic periods and during remission as well as in healthy controls

Analysed parameter (unit)	Healthy controls		CIU patients			
			Symptomatic		During remission	
	F (n = 20) Median	M (n = 20) Median	F (n = 17) Median	M (n = 15) Median	F (n = 17) Median	M (n = 15) Median
DHEA-S ( $\mu\text{g dL}^{-1}$ )	254.0*	320.8*	203.9	181.8	262.2**	337.3**
	Normal range [51.7–417]	Normal range [160–601]				

n, number of subjects; F, female; M, male; CIU, chronic idiopathic urticaria.

\*Significant differences between controls vs. symptomatic CIU patients  $P < 0.01$  (Mann–Whitney test); \*\*Significant differences between asymptomatic vs. symptomatic CIU patients  $P < 0.01$  (Wilcoxon paired test).

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