

Pili Torti

Subjects: **Others**

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Pili torti is a rare condition characterized by the presence of the hair shaft, which is flattened at irregular intervals and twisted 180° along its long axis. It is a form of hair shaft disorder with increased fragility. The condition is classified into inherited and acquired. Inherited forms may be either isolated or associated with numerous genetic diseases or syndromes (e.g., Menkes disease, Björnstad syndrome, Netherton syndrome, and Bazex-Dupré-Christol syndrome). Moreover, pili torti may be a feature of various ectodermal dysplasias (such as Rapp-Hodgkin syndrome and Ankyloblepharon-ectodermal defects-cleft lip/palate syndrome). Acquired pili torti was described in numerous forms of alopecia (e.g., lichen planopilaris, discoid lupus erythematosus, dissecting cellulitis, folliculitis decalvans, alopecia areata) as well as neoplastic and systemic diseases (such as cutaneous T-cell lymphoma, scalp metastasis of breast cancer, anorexia nervosa, malnutrition, cataracts, and chronic graft-vs.-host disease). The condition may also be induced by several drugs (epidermal growth factor receptor inhibitors, oral retinoids, sodium valproate, and carbamide perhydrate). The diagnosis of pili torti is based on trichoscopic or microscopic examination. As pili torti is a marker of numerous congenital and acquired disorders, in every case, the search for the signs of underlying conditions is recommended.

pili torti

trichoscopy

hair shaft abnormalities

hair shaft disorder

hair disease

twisted hair

1. Introduction

Pili torti, also known as “twisted hair”, was first described by Galewsky, and, independently, by Ronchese in 1932 [1] [2]. It is characterized by the presence of the hair shaft, flattened at irregular intervals and twisted 180° along its long axis, with each twist being 0.4 to 0.9 mm wide and occurring in groups of 3 to 10 (**Figure 1**) [3].



Figure 1. Pili torti, characterized by the presence of the hair shaft flattened and twisted 180° along its long axis.

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Pili torti is a form of hair shaft disorder with increased fragility [4]. It is classified into inherited or acquired. A wide range of changes associated with pili torti suggest specific pathophysiological mechanisms [5]. In inherited forms, the twisting of the hair is caused by an unequal development of the outer root sheath cells. Cell vacuolation and the irregular thickness of the outer root sheath at the suprabulbar level induce an uneven molding of the inner root sheath and hair shaft [6]. In acquired forms, a perifollicular inflammation followed by fibrosis generates rotational forces and deforms the hair follicle [7]. The cross-sectional area of pili torti hair is significantly smaller than a normal hair sample (2210 ± 1090 vs. 3370 ± 821 (μm^2); $p < 0.001$) and the tensile strength of pili torti is 2.1 times lower than that of normal hair [8]. No abnormalities in the hair cortex keratin within pili torti axis are observed [5].

Clinically, patients with pili torti have fragile, brittle, dry, and coarse hair. Patchy alopecia may develop. The scalp hair, especially in the occipital and temporal areas, is most commonly affected [9]. However, the eyebrows, eyelashes, axillary, and pubic hair may also be involved [9]. Usually, not all hair is affected by pili torti, and only a part of hair length may be changed [10]. Isolated pili torti may be occasionally found in the normal scalp. However, it may be associated with numerous local and systemic conditions [11][12].

2. Diagnosis and Treatment

2.1. Diagnosis

The diagnosis of pili torti is based on trichoscopic and microscopic examination.

Trichoscopy, hair and scalp dermoscopy, is a rapid technique that is useful in the diagnosis of scalp and hair diseases as well as genetic disorders, including ectodermal dysplasias [10][13]. It can be performed with a manual dermoscope (10 magnification) or a videodermoscope (20–1000 magnification) [14]. This noninvasive method replaced light microscopy, which required pulling of multiple hairs for investigation. This is particularly burdensome in cases, where only few hairs might be affected [9]. In pili torti, low magnification trichoscopy reveals the hair shafts bent at different angles and at irregular intervals. Regular twists of the hair shaft along the long axis are observed at high magnification (**Figure 2**).

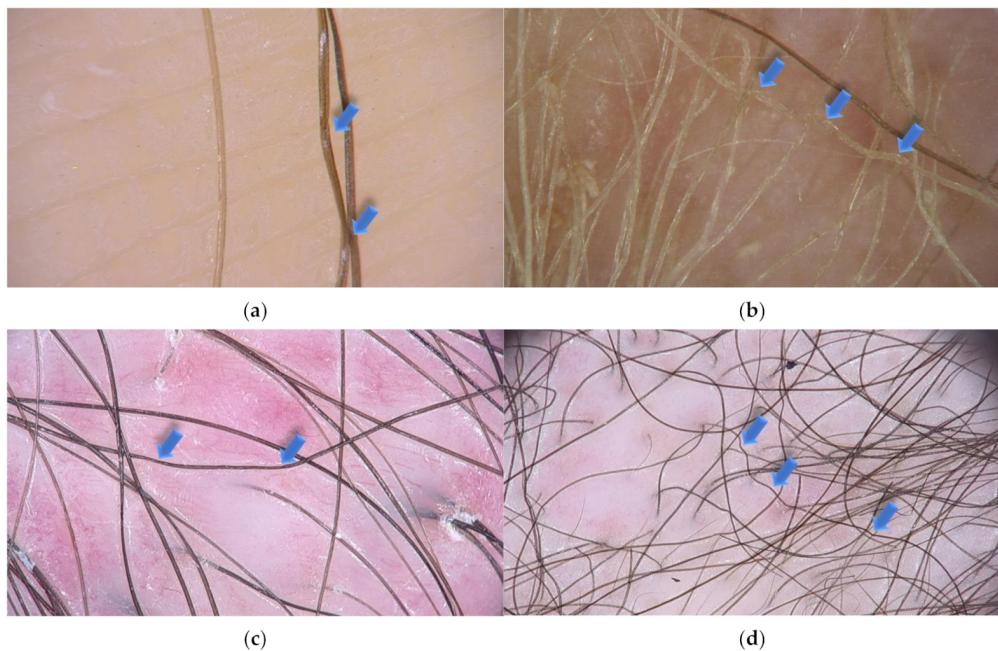


Figure 2. Trichoscopy shows pili torti (blue arrows) in various local and systemic conditions. (a) Pili torti in patient with late onset (Beare) type with single twists of hair ($\times 70$); (b) pili torti in patient with ectodermal dysplasia with the presence of multiple twist of hair ($\times 50$); (c) pili torti in patient with lichen planopilaris. Perifollicular scaling and milky-red areas are also presented ($\times 20$); (d) numerous pili torti in patient with mycosis fungoides ($\times 20$).

Microscopic examination shows groups of three or four regularly spaced twists at irregular intervals along the shaft [10].

Genetic diseases and syndromes should be excluded in every patient with pili torti. The search of other signs of underlying conditions should be performed in the case of acquired pili torti.

2.2. Treatment

There is no specific treatment of pili torti. The avoidance of trauma to the hair is recommended. Other forms of management include sleeping on a satin pillowcase, avoiding excessive grooming, braiding, heat treatments, and dying. Gentle shampoos may be beneficial [15][16].

Congenital pili torti may improve spontaneously after puberty. Drug-induced cases tend to resolve after the discontinuation of the offending agent [17][18]. In regard to acquired pili torti, the treatment of the underlying condition is most important.

Efficacy of pharmacological treatment in pili torti is limited [19]. Topical minoxidil has been suggested as a beneficial therapeutic option for patients with hair shaft abnormalities with increased fragility. However, it only has an impact on hair density and does not induce a causal treatment.

3. Conclusions

Pili torti is a rare condition, which may be associated with numerous congenital or acquired conditions. In every case of pili torti, the identification of the underlying disorder determines the therapeutic approach and prognosis.

References

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