

Pili Annulati: A Case Report in Young Siblings

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Abstract

Pili annulati is an uncommon inherited hair shaft disorder, predominantly recognised as an incidental finding typified by spangled appearance and alternate light and dark bands along the hair shaft. In this report we describe 2 sisters aged 6 and 3 years of age, who presented with 3-month history of silvery appearance of hair. The diagnosis was confirmed by the identification of periodic light and dark bands on polariscopic examination.

Keywords: Autosomal inherited hair shaft abnormality; Spangled hair; Ringed hair; Polarized microscopy

Introduction

Pili annulati is an uncommon disorder classified under hair shaft abnormalities clinically presenting as. This genetic condition is inherited in an autosomal dominant fashion, although sporadic cases have been reported. The pathognomonic feature is the alternating transverse light and dark bands along the hair shaft on polariscopic examination.

Case Report

We report two sisters, born of nonconsanguineous marriage aged 6 and 3 years old with a complaints of change of hair colour to a silvery hue over 3 months duration. There was no history suggestive of retarded hair growth or increased fragility. On examination, the hairs appeared dark, soft, straight, and silky with a silvery appearance under bright light (Figure 1 and 2). The family history and medical history were unremarkable. General and systemic examination was normal. The hair thickness and tensile strength were clinically normal. There were no other associated abnormalities; the skin nails and mucosal surfaces were normal. The hair was plucked and sent for light microscopy and polarized microscopic examination. Although the light microscopic examination was normal, polarized microscopic examination revealed alternate dark and light bands along the length of the hair shaft (Figure 3). The hair shaft was otherwise normal in density and tensile strength. Based on clinical examination and polarized microscopic examination findings, a diagnosis of Pili annulati was made.

Discussion

Pili annulati is a rare hair shaft abnormality with characteristic light and dark bands, primarily affecting the scalp hairs. The diagnosis pili annulati is synonymous with ringed hair or spangled hair [1].



Figure 1: Spangled Appearance with reflected light.



Figure 2: Spangled Appearance with reflected light.



Figure 3: Polarized Microscopy showing alternate banding.

The disease is predominantly inherited in an autosomal dominant fashion; however sporadic cases have also been reported. Genetic mapping in families with pili annulati has narrowed the target locus to the telomeric region of long arm of chromosome 12 but the exact gene responsible is still elusive [2]. Abnormal ribosomal synthesis of microfibril –matrix complex has been regarded to cause intracellular defect leading to potential spaces within the cortex [3]. These empty spaces are filled with air due to the relative permeability of the hair

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fibres and form the air cavities that appear as light bands along the hair shaft against reflected light. Further insight of the underlying defect is revealed by transmission electron microscopy of the hair root, which shows reduplicated lamina densa [4]. Immunohistochemical studies, shows a wavy basement membrane zone. An altered amino acid level with elevated lysine and decreased cysteine by amino acid analysis suggests a structural defect [5].

The clinical features are variable as the expression and penetrance of this genetic defect is variable [6]. Pili annulati may appear during infancy or later. The hair appears shiny and attractive and reveals the typical alternative light and dark bands with illumination. The number of white bands has been noted to decrease distally. Pili annulati is usually an incidental finding in dark hair as the banding is less prominent with the additional pigment [7]. In most cases the manifestation is limited to the scalp hairs however the beard, axillary and pubic areas have also been affected. A variable range of 20-80% of scalp hairs may be affected in a given patient. Despite the structural defect, classically the hair diameter and tensile strength are within normal limits. Nonetheless there have been reports of increased risk of weathering and fragility [8].

The diagnosis is confirmed by further microscopic examination of the hair shaft. Trichoscopic and polariscopic examination demonstrates regular alternating white bands along the hair shaft, which corresponds to the air filled cavities [9]. In light microscopic examination alternating light and dark bands are reversed wherein the dark bands represent the white bands seen by naked eye.

Pili annulati has been reported along with atopic dermatitis, alopecia areata, blue nevi, anhidrotic ectodermal dysplasia, syndactyly, polydactyly and woolly hair [5,10-12]. Despite the pathognomonic clinical features of pili annulati, diagnostic dilemma may occur with the following; intermediate medullation, pseudo pili annulati, bubble hairs, pili torti and trichothiodystrophy [5].

Intermittent medullation is a normal variant seen with individuals with thick hair shafts. Trichoscopy reveals light coloured medulla, which constitutes less than 50% of the hair shaft width. Pseudo pili annulati is also a variant of normal characterized by abnormal twisting of hair without any banding under light microscopy [5,13]. Bubble hairs occurs when wet hair is exposed to extreme temperatures causing gas formation within the hair shaft. Light microscopy reveals irregular white oval vacuoles resembling Swiss cheese structures without the dark bands seen in pili annulati [14]. Pili torti represents twisting of flattened hairs at 180 degrees at irregular intervals. At low magnification, sharp bending at irregular intervals is seen. Trichothiodystrophy is a hair shaft disorder distinguished by the tiger tail pattern seen in polariscopic examination. Here the alternating light and dark bands are perpendicular to the long axis of the hair shaft [15].

We report two sisters with childhood onset of shiny hair and diagnosis confirmed by polariscopic examination, which showed the characteristic light and dark bands. Generally no treatment is required for this condition when diagnosed in isolation. Protective measures from weathering may be offered. Although multiple cases of pili

annulati have been reported in the past, there is a paucity of reports in the literature in ethnic groups, owing to the difficulties in recognising this condition. We hereby, emphasize the need for clinicians to be aware of its presence in dark hair.

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