

An Acquired Partial Hair Shaft Anomaly: Case Report

Edinsel Kısmi Saç Şaftı Anomalisi

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ABSTRACT A 7-year-old girl with a complaint of abnormality in her hair since 3 years old attended to our outpatient clinic. On her physical examination her complexion appeared rather pale. The hair was dull and different widths' of the effected hair and a tattered appearance was observed. First look of the patient's hair was similar with pili torti or wooly hair naevus. In the light microscopy, the atypical looking hair was flattened, in polarized light microscopy, the lateral sides of the hair shafts showed irregularity, confirming the irregular pattern on the light microscopy. No twisting of the hair shaft was observed. Besides, neither tapering nor breakage of the hair was observed at the tip of the hair. Scanning Electron Microscopy (SEM) examination showed that the hair surface structure was abnormal as well as cuticle pattern irregularity. X-Ray Fluorescent (XRF) study was performed and no abnormality was detected. The lead levels from both normal and affected hair were found to be high, while the serum lead and seruloplasmin levels were normal. Our findings were not consistent with any previously described hair deformity. We named our patient's deformity as "pili torti without torti" or "pseudopili torti". The different lead levels between the healthy and tattered hair may be explained by precipitation of the pollutants on the fringes caused by abnormal cuticle cells.

Key Words: Hair diseases; pediatrics

ÖZET 7 yaşında kız çocuğu saçlarının bir kısmının 3 yaşından itibaren taranamaması şikayeti ile başvurdu. Hastanın apikoposteriyör bölgesindeki saçların düzensiz, yolunmuş izlenimi veren yapıda olduğu izlendi. Hastanın saçı ilk bakışta "pili torti" ya da "yünsü saç" kliniğine benziyordu. Saç telinin ışık mikroskopik (SEM) incelemesinde saçın hafifçe yassılmış olduğu dikkati çekti. Polarize ışık mikroskopunda saç şaftının lateral duvarlarında irregularite izlendi. Saçın uç kısmında kırılma veya saçaklanma görülmedi. Taramalı elektron mikroskopik incelemede saç şaftının kutikul paterninde ışık mikroskopun benzer şekilde irregularite saptandı. X-ışını floresans (XRF) yöntemi ile eser elementler açısından incelenen hastanın, saç kurşun seviyesi dışında değerleri normaldi. Serum kurşun ve seruloplazmin seviyeleri ise normal sınırlardaydı. Hastanın klinik ve laboratuvar incelemeleri önceden tanımlanmış saç şaftı değişikliklerinin hiçbirine uymuyordu. Bu nedenle hastayı "psödopili torti" ya da "psödo-yünsü saç" olarak yorumladık. Saçında artmış olarak bulunan kurşun seviyesinin de anormal kutikul hücrelerinde yerleşebilen kurşuna bağlı olabileceğini düşündük.

Anahtar Kelimeler: Saç hastalıkları; pediatri

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CASE REPORT

A 7-year-old girl attended our outpatient clinic with a complaint of abnormality in her hair. Her history revealed that her complaint had started 3 years ago. Questioning of her bathing habits revealed no abnormality, she was using appropriate shampoos. Her mother denied vi-

gorous combing and brushing of her hair. On her physical examination her complexion appeared rather pale. Her weight and height were in the 25-50% and 75% percentiles respectively. No neurologic abnormality was observed. On dermatologic examination a bundle of hair on the apicoposterior of the scalp appeared uneven (Figure 1).

First look of the patient's hair was very similar with pili torti. However, on close examination, the hair was dull and different widths' of the effected hair and a tattered appearance was observed. In contrast, the hair on the frontotemporal region and the scalp skin seemed to be normal. Her routine laboratory tests were within normal range except for a decreased hemoglobin level (Hb: 13.4 g/dl). The anemia was consistent with iron deficiency anemia with MCV: 77 fL (normal ranges: 80-96 fL) and MCH: 26.9 pg (normal range: 27.5-33.2 pg), no basophilic stippling was observed in the peripheral blood smear. In the light microscopy, the atypical looking hair was slightly flattened (Figure 2). Besides, neither obvious tapering nor breakage of the hair was observed at the tip of the hair. In polarized light microscopy, the lateral sides of the hair shafts showed irregularity, confirming the irregular pattern on the light microscopy. Scanning Electron Microscopy (SEM) examination showed that the hair surface structure was abnormal and cuticle pattern was irregular (Figure 3).

Normal appearing hair localized on the frontotemporal region was also examined by all 3 methods and the results were consistent with normal hair shaft.

X-Ray Fluorescent (XRF) Study was performed in both abnormal and normal hair, nail, serum, and drinking water of the patient for trace elements. No abnormalities were detected from the samples of nail, serum and drinking water. In hair analysis, sulphur level was normal and the trace elements like Mn, Fe, Ca, Cu, Zn levels were found to be normal (Figure 4). The lead level from the normal hair was 45 ppm and 110 ppm from the affected hair. Examination of her mother's hair was negative for lead. The serum lead level of the patient was also performed in order to rule out chronic



FIGURE 1: The clinical appearance of the patient with tattered hair on the top, right side.

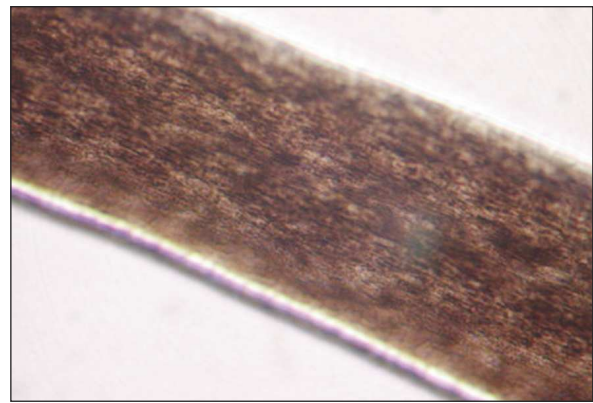


FIGURE 2: Light microscopy of the hair with x20 magnification.

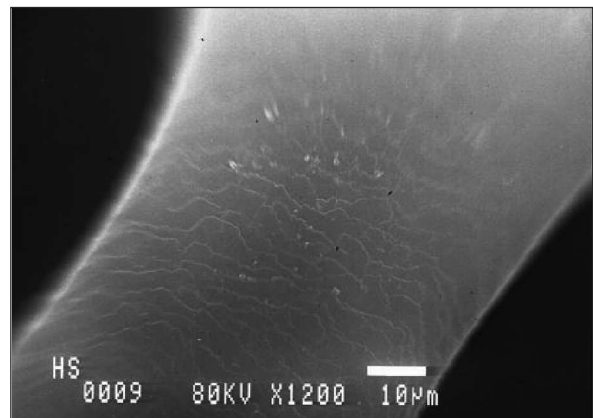


FIGURE 3: Scanning electron microscopy of the hair with x1200 magnification.

lead poisoning and the result was negative (<10 µg/dl).

Serum seruloplasmin level was also normal: 32.2 (normal range: 22-58). A written consent was taken from the patient's mother for publication.

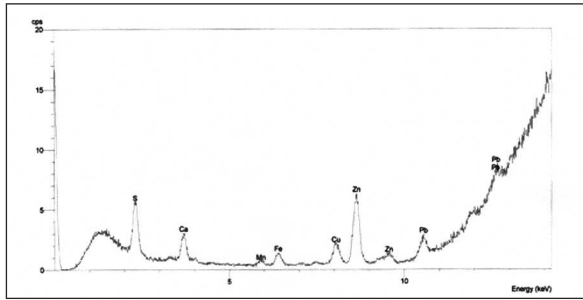


FIGURE 4: XRF analysis of the abnormal hair.

DISCUSSION

Many hair disorders are well described and grouped according to their underlying pathophysiologic mechanisms. Destruction of the hair follicle, abnormalities in hair cycling, and intrinsic defects of the hair itself are the major hair disorders met during daily practice.

The latter defect can be divided into hair shaft abnormalities associated with hair breakage, like trichorrexia nodosa, trichothiodystrophy, pili torti, trichorrexia invaginata and moniletrix, and hair shaft abnormalities associated with unruly hair, like uncombable hair syndrome and woolly hair.¹ Hair shaft abnormalities can either be congenital or acquired, generalized or localized.² Little data are found in the literature, especially if the hair shaft abnormality is isolated without any accompanying disorder.

In the review of the literature, Lalevic-Vasic et al reported cases showing common morphologic changes like torsions, angular kinks, weathering as well as grooved segments alternating with the normal ones, and described as a separate entity within the woolly hair group.³ Our patient's first appearance was very similar to pili torti. However, examination of the abnormal appearing hair of our patient both by light microscopy and scanning electron microscopy was not consistent with any previously described hair deformity, especially pili torti. Hence, we named our patient's deformity as

“pili torti without torti”. Our patient's dermatologic findings were also similar to woolly hair nevus, however, electron microscopy of the affected hair did not show the loss of cuticle, which is a prominent finding in woolly hair nevus. Besides, it is reported that, about half of the patients with woolly hair nevus show an epidermal nevus, in which we did not find such nevi in our patient.⁴

X-Ray Fluorescence (XRF) technique is widely applied for the analysis of inorganic constituents in light and heavy matrices, such as biological and geological samples. During the past three decades the investigation of trace element concentration in human scalp hair has become increasingly popular for monitoring environmental exposure, evaluating systemic intoxication, assessing nutritional status and diagnosing diseases.⁵ In our patient, lead levels from both normal and affected hair were found to be high, while the serum lead level was normal. This may be due to environmental factors of the urban area our patient accommodates. The different lead levels between the healthy and tattered hair may be explained by precipitation of the pollutants on the fringes caused by abnormal cuticle cells.

Despite the lack of vital function of hair, its psychological importance is significant. Hair disorders are frequently associated with lowered self-esteem and depression.¹

Unfortunately, the management of hair shaft abnormalities is limited. Trauma and drugs such as retinoid must be kept in mind as possible causes of acquired hair shaft disorders.⁶ As for congenital deformities, improvements of gene therapy in the future may offer promising treatment options for known genetic defects.⁷⁻¹⁰

As a conclusion, we would like to emphasize the fact that clinical appearance of the hair may be misleading and an examination with light microscopy is the essential in first step towards diagnosis.

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