The term woolly hair refers to an abnormal variant of fine, tightly curled hair with 180° longitudinal twisting and an increased tendency to fracture. Transverse sections of hair shafts show varying ovoid shapes of different morphology and hair may be sparse and hypopigmented. Lichen planopilaris (LPP) is an inflammatory, cicatricial alopecia with several different patterns of hair loss. Woolly hair and LPP have not been previously reported together. We believe that the accumulation of these rare associations in the literature will be useful in understanding etiopathogenesis of the diseases.

**Keywords:** Alopecia; lichen planus; cyclosporine

**ÖZET** Yünsü saç, negroid kökenli olmayan bir bireyde meydana gelen saçlı derinin bir kısmını ya da tamamını etkileyen sıkı sarılmış saçların karakterine, saçlı deri sayısını nadir konjenital bir anormallikidir. 1974’te Hutchinson ve ark. yünsü saç lokal alt tip (yünsü saç nevü) ile otozomal dominant herediter yünsü saç ve otozomal rezesif (OR) ailesel yünsü saç içeren iki jeneratize alt tip olmak üzere üç farklı varyanta ayrıldılar. Lichen planopilaris ise nedeni tam olarak bilinmemeyen primer skatrisel alopesinin en sık görülen formudur. Burada daha önce bildirilmemiş OR ailesel yünsü saç ve lichen planopilars birlikteliği bulunur bir hastayi tərəf ediyoruz. Nadir görülen bu birlikteliklerin literatürde birikmesinin hastalıkların etiyopatojenezini anlamada faydali olacağını düşünmekteyiz.

**Anahtar Kelimeler:** Alopesi; liken planus; siklosporin

**ABSTRACT** Woolly hair is a rare congenital abnormality of the structure of the scalp hair characterized by tightly coiled hair involving part or the entire scalp occurring in an individual of non-negroid origin. In 1974, Hutchinson et al. classified woolly hair into three variants: A localized variant (woolly hair nevus) and two generalized variants, including autosomal dominant hereditary woolly hair and autosomal recessive (AR) familial woolly hair. Lichen planopilaris is a most frequent presentation of primary cicatricial alopecia. We hereby describe a patient with AR familial woolly hair and lichen planopilaris who has not been previously reported. We believe that the accumulation of these rare associations in the literature will be useful in understanding etiopathogenesis of the diseases.

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**CASE REPORT**

A 24-year-old female presented with complaints of increasing local hair loss for tree years. Her alopecia was regarded as alopecia areata and intrale-
sional steroid therapy had been done two times about two years ago without improvement by another dermatologist. She had also short, curly since birth. There was no history of physical or mental retardation and no history of systemic involvement. Similar history was also present in her little nephew and her mother’s-grand father. There was no similar complaints in the parents and the two other sisters. Dermatological examination revealed the presence of fine, coiled hairs over the scalp (Figure 1). There was three cicatricial alopecia patch on bilateral parietal and right occipital region (Figure 2). There were no abnormality of body, pubic, axillary hair and eyebrow. Nail, teeth and systemic examination was normal without micrognathia. Routine hematological investigations including complete blood counts, liver and renal function tests, chest radiograph and urinalysis were all normal. Trichoscopy showed “crawling snake appearance” cicatricial areas with white and milky-red areas lacking follicular openings (Figure 3). Skin punch biopsy taken from the periphery of cicatricial area demonstrated slight lymphocytic infiltration around infundibulum of some hair follicles, and fibrous tract with destroyed hair follicles and sebaceous glands (Figure 4). Detailed cardiac evaluation including electrocardiogram and echocardiography was normal. Genetic analysis to determine the gene loci could not be carried out. Based on history, clinical features, trichoscopy and histopathologic findings a diagnosis of autosomal recessive/ familial woolly hair and liken planopilaris was made. We chose cyclosporin treatment with the dosage of 250 mg/day for LPP. At six months follow up lesions and scar progression stopped and improvement was seen at active sides of LPP. Informed Consent was received from the patient for use of patient data.
DISCUSSION

Woolly hair is a rare congenital abnormality of the structure of the scalp hair characterized by tightly coiled hair involving part or the entire scalp occurring in an individual of non-negroid origin. In 1907, Gossage was the first to describe a case of woolly hair in a European family, comparing this hair anomaly with the characteristic curly hair of black people. In 1974, Hutchinson et al. classified woolly hair into three variants; a localized variant: Woolly hair nevus and two generalized variants: Autosomal dominant/hereditary woolly hair and autosomal recessive/familial woolly hair. In addition to this, diffuse partial woolly hair has also been described. There is a marked reduction in the diameter of hair shafts, which may be poorly pigmented. It is present at birth with tightly coiled, thin-caliber hair, which usually grows only to 2-3 cm due to a truncated anagen phase. Very few cases have been reported in non-negroid races and these patients are typical cases of familial woolly hair. In some patients, woolly hair may become darker and less curly with time, but there is no effective treatment. Our patient color of hair is dark and also hairs are not too short. Various other anomalies have been reported to be associated with different types of woolly hair. These include Noonan syndrome, Cardiofaciocutaneous syndrome, Naxos disease, Carjaval disease and keratosis pilaris. AR variant has been associated with palmoplantar keratoderma and cardiac anomalies. Naxos disease, as a result of mutation in the plakoglobin gene, is characterized by right ventricular cardiomyopathy and nonepidermolytic diffuse palmoplantar keratoderma in addition to features of woolly hair. Another similar variant is Carvajal syndrome, which is due to mutation in the desmoplakin gene. In addition, a family with woolly hair has been reported with features of ectodermal dysplasia in the form of nail dystrophies, acral hyperkeratosis, and changes in peridontium. Our case did not fit into any of the syndromes described above.

The pathogenesis of LPP remains to be fully elucidated but like other cicatricial alopecias involves the irreversible destruction of hair follicle stem cells and loss of a hair follicle’s capacity to regenerate itself. Overgrowth of microorganisms with hyperkeratosis results in factors that disrupt the immune privilege of the hair follicle, leading to an immunologic reaction that is limited to the follicle and spares surrounding eccrine structures. A scalp biopsy shows a lymphocytic infiltrate involving the isthmus and infundibulum. Treatment is prescribed with the goal to alleviate patient symptoms and to halt the progression of hair loss. Treatment involves use of potent topical corticosteroids and/or intralesional corticosteroids. Options for systemic treatment include anti-inflammatory agents such as hydroxychloroquine, tetracyclines, pioglitazones, and immunosuppressive medications such as cyclosporine, mycophenolate mofetil, or systemic corticosteroids. In our case the cicatricial alopecia was due to late lichen planopilaris and there was also new active lesions. Cyclosporin has begun with the dosage of 250mg/day and used for six months. Any side effect was not seen during therapy and activity of the disease ceased. Treatment for woolly hair is not currently available, although in some patients the hair may become darker and less curly with time as in our case.

Our case was an autosomal recessive woolly hair associated with lichen planopilaris which is a rare association and no case was reported up to date. In our patient, the increase in microorganism caused by the woolly hair structure and hyperkeratosis may have caused immunological damage in the hair follicle.
Conflict of Interest
Authors declared no conflict of interest or financial support.

Authorship Contributions
Idea/Concept: Sümeyye Altıntaş Kakşi; Design: Sümeyye Altıntaş Kakşi; Control/Supervision: Sümeyye Altıntaş Kakşi.

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