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Idiopathic Acquired True Leukonychia Totalis and Partialis- A case report with Review of literature

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Abstract

Leukonychia totalis and partialis are characterized by complete or partial whitening of nail plate. They can be inherited or acquired. Rarely, no cause or associations are found. Here, we report a case of a 17-year-old male with idiopathic acquired true leukonychia totalis and partialis.

Introduction

Leukonychia refers to white discoloration of the nails. Leuconychia can be true (involving nail plate or matrix) or apparent (due to nail bed pathology). [1] In true leuconychia, white color does not disappear on applying pressure on the nail plate. Leukonychia can also be classified as inherited or acquired.[2] Rarely, no cause or associations are found. [3] Here, we report a case of a 17-year-old male with idiopathic acquired true leukonychia totalis and partialis.

Case Report

17 years old male presented to our OPD with history of whitening of his finger and toenails. The nail changes started as ill-defined white discoloration on few finger and toenails which gradually and synchronously progressed to almost total whitening of all the twenty nails over 3 years. The growth of the nails was otherwise normal. He was born out of a non-consanguineous marriage. His developmental milestones were normal. He had no previous medical or surgical history. He denied history of any medications, exposure to chemicals or mechanical trauma to the nails. There was no history of leukonychia or other dermatologic diseases in family.

On physical examination, partial to total leukonychia of all finger and toenails was observed. Few nails had a totally white discoloration. Most of the nails had a pink transverse band about 2- 3 mm wide at the distal edge. The nail folds and cuticles were normal. The nail surface was smooth with no pitting, crumbling or nail thickening **(Figure 1).** On applying pressure on the nail plate, white color did not disappear. Thorough systemic examinations did not reveal any abnormalities.

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Figure 1: partial to total leukonychia of all finger and toenails

The routine laboratory investigations like complete blood count, liver and renal function tests were done and found to be within normal limits. Hemoglobin was 14.9 gm /dl, total leucocyte count 4800/ mm3, platelets 264000/mm3, total bilirubin -0.5mg/dl, conjugated bilirubin- 0.2 mg/dl, SGPT-15U/L, SGOT-11 U/L, ALP-81 U/L, urea- 12 mg/dl, creatinine - 0.4 mg/dl, Na- 142, K- 4.0). Further workup including serum albumin, serum calcium, urine analysis, thyroid function test, parathyroid function, chest X-rays, echocardiography, ultrasonography of abdomen and pelvis, iron studies, vitamin assays, antinuclear antibodies did not detect any abnormality. Potassium hydroxide mount and fungal cultures of the nails were negative. So, the diagnosis of Idiopathic Acquired True Leukonychia Totalis and Partialis was made and the patient was counseled regarding the disease. On 1 year follow up, no other changes were noted in the nails. The leuconychia was persistent.

Discussion

The mechanism of leukonychia is not clear .True leukonychia is thought to result from abnormal keratinization of the distal matrix that causes parakeratosis in the ventral nail plate. Retained keratohyalin granules in the nail plate cells reflect light and cause subsequent loss of nail plate transparency. [2,4]

Leukonychia totalis and partialis can be inherited as an isolated finding or as part of a syndrome or can be acquired. Hereditary leukonychia has been found to be associated with peptic ulcer disease and cholelithiasis, congenital hyperparathyroidism, hypoparathyroidism , cataracts, acanthosis nigricans, pili torti , severe keratosis pilaris , recurrent sebaceous cysts and renal calculi as well as in Bart-Pumphrey syndrome (leukonychia, sensory-neural deafness and knuckle pads), Heimler syndrome, Lowry-Wood syndrome, FLOTCH syndrome, keratoderma-hypotrichosis-leukonychia totalis syndrome, congenital keratosis palmaris et plantaris-deafness-leukonychia totalis syndrome, Bauer syndrome and LEOPARD syndrome.[1-4] Acquired leukonychia can occur due to psoriasis, onychomycosis, local trauma, alopecia areata or systemic illness like cardiac insufficiency, myocardial infarction, renal failure, pleural empyema, liver diseases, protein-losing enteropathies, systemic infections, sickle cell anemia, systemic lupus erythematous, malnutrition, chemotherapeutic expo-

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sure, heavy metal poisoning.[2-4] Removal of cause of acquired leukonychia may lead to resolution of white colour. Sometimes, cause is unknown as in our case.

Idiopathic acquired true leukonychia totalis and leukonychia partialis is a rare condition. Literature review shows that nineteen cases have been reported till date [1-3, 5-18], none reported from Nepal

(Table 1)

Table 1: Reported cases of Idiopathic acquired true leukonychia totalis and leukonychia partialis

Author	Age/sex	Age at onset	Progression	Family History	Comorbidities	Drug or Chemical Exposure	Nail Examination	Histopathology	KO M
Arsiwala [1]	35 M	23 years	started with finger nails, slowly progressed to involve all finger nails and great toe nails	No	No	No	Leuconychia totalis of 10 fingernails and both great toenails		Ne for Ko fur cul
Bakry et al [2]	12 M	5 years	Gradually progressive	No	No	No	Leuconychia totalis of 10 fingernails		Ne for bo Ko an fur cul
Bongiorno and Aricò [3]	34 M	23 years	simultaneous leuconychia partialis of fingers and toenails, gradually progressed to leuconychia totalis	No	No	No	striata and total leukonychia of 20 nails, soft nails		Ne for Ko fur cul

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Author	Age/sex	Age at onset	Progression	Family History	Comorbidities	Drug or Chemical Exposure	Nail Examination	Histopathology	KO Mi
Claudel et al [5]	12 M	11 years	leukonychia on 7 fingernails progressed to involve 8 fingernails and single toenail over 1 year	uncle with alopecia areata	Exercise induced asthma	Short course of prednisolone shortly before nail changes, albuterol inhaler, Leuconychia persisted although asthma resolved and treatment was stopped	total leukonychia 8 fingernails and 1 toenail, occasional pits	globular collections of large immature keratohyalin granules	
D'Souza et al [6]	10 M	4 years	leukonychia partialis to striata and totalis, complete resolution after 7 months of zinc and amino acid supplementation	No	No	No	Leukonychia totalis and striata on fingernails, with leukonychia partialis in both thumbnails		Ne for Ko fur cul
Das et al [7]	14 M	5 years		No	No	No	Leuonychia totalis of 10 fingernails		Ne for bo Ko an fur cul
Dlova and Tosti [8]	20 M	12 years		No	No	No	Leuonychia totalis of 10 fingernails	marked parakeratosis within the nail plate	Ne for Ko fur cul

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Author	Age/sex	Age at onset	Progression	Family History	Comorbidities	Drug or Chemical Exposure	Nail Examination	Histopathology	KO Mi
Eller and Anderson[9]	15 M	14 year		No	No	No	Leuonychia totalis of 10 fingernails		
Kim et al [10]	19 M	19 years	Rapidly progressive (1 month) , central white spots on toenails that progressed to almost total nail whitening.	No	No	No	Leuconychia totalis & partialis of 10 toenails	sparse parakeratosis with globular collection of keratohyalin granules	Ne for Ko fur cul
Park et al [11]	26 M	13 years	progressed slowly from Leuconychia Partialis to totalis	No	No	No	Leuconychia Partialis & totalis of 9 fingernails except left thumb	globular collection of large, immature keratohyalin granules	
Stewart et al [12]	23 M	17 years	slowly progressive from Leuconychia Partialis to totalis	No	No	No	Leuconychia Partialis & totalis of 10 fingernails and second toenails		Ne for Ko fur cul
Chang Nam Lee [13]	26 M	13 years	leukonychia partialis to totalis	No			Leuconychia Partialis & totalis of 9 fingernails except left thumb	globular collection of large immature keratohyaline granules	

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Author	Age/sex	Age at onset	Progression	Family History	Comorbidities	Drug or Chemical Exposure	Nail Examination	Histopathology	KO Mi
Neki NS [14]	29 M	20 years	simultaneous and synchronous whitening of all the finger and toe nails	No	No	No	Leukonychia totalis of finger nails and partialis of toe nails		Ne for bo Ko an fu cu
Freeman SC [15]	17 M	12 years	horizontal bands on several fingernails Gradually, more nails became involved, and some nails became almost entirely white	No	No	No	Horizontal white bands on 9 fingernails	nail plate showed parakeratosis, PAS negative	Ne for Ko
Angoori GR [16]	30 M	Childhood	started in few nails and progressed to involve all fingernails	No	No	No	Total leukonychia of 10 fingernails		Ne for Ko fu cu
	32 M	8 years			Polymorphous light eruption				

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Author	Age/sex	Age at onset	Progression	Family History	Comorbidities	Drug or Chemical Exposure	Nail Examination	Histopathology	KO Mi
Mathachan SR et al [17]	20 M	19 years	progression from partialis to totalis	No	No	No	Total leukonychia of fingernails and partialis in toenails		Ne for bo Ko an fu cul
	18 M	15 years	progression from partialis to totalis				Leuconychia totalis and partialis in all fingernails and toenails		
Verma S [18]	24 M	19 years	progressive from Leuconychia Partialis to totalis, fingernails to toenails	No	No	No	total leuconychia of all finger nails and partial to total leuconychia of all toenails		Ne for Ko fu cul
Our case	17 M	14 years	Gradual progession from partialis to totalis	No	No	No	Leuconychia totalis and partialis in all fingernails and toenails		Ne for bo Ko an fu cul

All cases were young males with age ranging from 10–35 years at the time of diagnosis, with symptoms first manifesting at age 4-23 years. In our case, age at onset was 14 years. Male predilection may suggest a role for androgens in the disease process or a sex chromosome–based genetic predisposition.[15] In most of the cases, progression was gradual, progressing from leuconyhcia partialis to totalis and from fingernails to

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toenails. Similar progression was seen in our case. Rapid progression within 1 month was seen in one case [10]. Fingernails were involved in most cases [2,6-9,11, 13,15,16], and some patients had both fingernails and toenails involved [1,3,5,12,14,17,18], similar to our case . In a single patient, only toe nails were involved [10]. In a case reported by D'Souza et al [6], complete resolution was seen after 7 months of zinc and amino acid supplementation.

In our case, no cause or associations was detected on thorough examination and investigations and thus patient was diagnosed as a case of Idiopathic Acquired True Leukonychia Totalis and Partialis. This is a rare clinical entity with nineteen reported cases. Our case is the twentieth addition to the list. It is the first case to be reported from our country, Nepal. We also want to highlight that timely diagnosis and reassurance may help to avoid unnecessary investigations and prevent psychological stress to the patient and family.

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