Familial Median Canaliform Nail Dystrophy

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GOAL

To understand median canaliform nail dystrophy to better manage patients with the condition

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Recognize the clinical presentation of median canaliform nail dystrophy in patients.
2. Differentiate median canaliform nail dystrophy from similarly presenting conditions.
3. Discuss possible causative factors for median canaliform nail dystrophy.

CME Test on page 156.

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Median canaliform nail dystrophy typically appears as a central nail groove, beginning at or distal to the proximal nail fold, from which small lateral fissures may be found. Although the onset of this nail dystrophy has occasionally been associated with either prior local trauma or the initiation of treatment with isotretinoin, the etiology of this condition remains elusive for most affected individuals. We describe a man with median canaliform nail dystrophy whose brother and mother had the same nail dystrophy, review the other reported cases, and discuss the differential diagnosis of this nail disorder.

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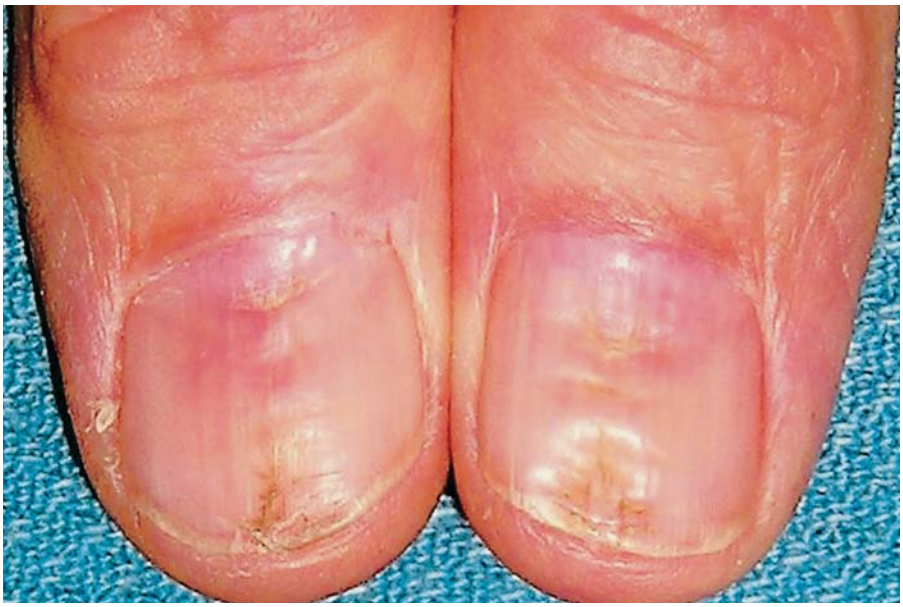
Median canaliform nail dystrophy is a nail abnormality that typically involves one or both thumbnails. The first case of this disorder was recorded by Heller[1] in 1928. Median



Distal longitudinal grooves with small horizontal fissures and red macrolunula on the thumbs of a 68-year-old man with familial median canaliform nail dystrophy.

canaliform nail dystrophy presents as a central longitudinal groove of the nail plate, extending proximally from the end of the nail.[2] This condition is usually not inherited. However, it may be acquired following trauma to the nail. We describe a man with familial median canaliform nail dystrophy and discuss the differential diagnosis.

Case Report

A 68-year-old man presented with bilateral dystrophy of his thumbnails. The nail abnormality initially appeared at 34 years of age with no preceding trauma to the digits. His older brother and his mother also had developed the same nail changes as young adults. Neither the patient nor his brother or mother rubbed their proximal thumbnail fold with the tip of their second finger; the absence of this behavioral activity was repeatedly confirmed by both the patient and his wife during several subsequent office visits.

Examination of both thumbnails showed an asymptomatic distal fissure with a fir tree–like pattern (Figure). Proximally, the nail plates showed transverse grooves. In addition, the lunula was red and enlarged.

Comment

Median canaliform nail dystrophy appears as a long longitudinal groove extending from either the proximal nail fold or a more distal portion of the nail plate to the end of the nail. Lateral extensions of this fissure create a conspicuous inverted fir tree–like pattern. In severe cases, the nail can split along the groove.[3] Thickening of the proximal nail fold, enlargement of the lunula, and redness of the lunula also may occur.[4-9]

The diagnosis of this condition is usually established based on clinical features because pathologic correlation is rarely available. However, specimens for microscopic evaluation have occasionally been provided. Parakeratosis, as well as an accumulation of melanin within and between the nail bed keratinocytes, was demonstrated in the evaluation of an affected nail by Heller[10] in 1927. Subsequently, parakeratosis and intranuclear pigmentation were found within the longitudinal canal of the affected nail plate of a 12-year-old girl with median canaliform nail dystrophy who was described by Robinson and Weidman[11] in 1948.

Median canaliform nail dystrophy may present following trauma to the nail plate or nail matrix.[3-7,12-15] In addition, coexisting conditions such as either soft tissue in the nail defect or dental caries have been observed in some patients with median canaliform nail dystrophy. In one case, a 19-year-old woman presented with a flabby filament of fleshy tissue that was observed within the dystrophic nail canal.[14] The tissue was extracted, and the nail abnormality resolved. Subsequently, the nail dystrophy, including the associated tissue, reappeared.[14] Tooth decay associated with median canaliform nail dystrophy was reported in a 23-year-old woman with a deformity that involved many of the nails on both of her hands. Her nail condition spontaneously cleared after 3 carious teeth were extracted.[16]

Medication was postulated as the causative factor for the development of median canaliform nail

Characteristics of Median Canaliform Nail Dystrophy and Habit Tic Deformity

Characteristic	Median Canaliform Nail Dystrophy	Habit Tic Deformity
Synonyms	Canaliform dystrophy of the nails[14] Canallike median nail dystrophy[24] Dystrophia mediana canaliform(is)[3,21] Dystrophia (unguium) mediana canaliformis (of Heller)[1,2,5,6,8,13,15,16,20,22,25,26] Lateral canallike dystrophy[2] Longitudinal single nail groove[2] Median canaliform dystrophy[12,17] Median canaliform nail dystrophy (current report) Median canallike dystrophy of the nail[18] Median nail dystrophy[4-7,25] Naevus striatus symmetricus of the thumbs[19,27]	Washboard thumbnails[7,28]
Familial	Present[20,22,23]	Absent
Acquired	Present	Present
Onset age	Range: 1–75 y[1,4,11-21,27] Median: 18.5 y[1,4,11-21,27] Mean: 25.72 y[1,4,11-21,27]	Ranging in age from second decade to young and middle adulthood[13,28,29]
Male-female ratio	1:1[1,4,11-17,19-23,27]	No predilection[13,28,29]
Asymptomatic	Present	Present
Appearance	True longitudinal split of the nail beginning near or at the proximal nail fold. Lateral projections, which extend from the central groove, create the appearance of an inverted fir tree–like pattern. Enlargement and redness of the lunula may be present	Transverse ridges along the central nail plate depression that begin at the proximal nail fold. The depth of the central canal depends on the intensity of the inflicted trauma to the proximal nail fold, which also is swollen. Enlargement and redness of the lunula may be present
Possible associations	Macrolunula[4-7] Red lunula[9]	Macrolunula[29] Red lunula[9]
Preceding trauma	20%*	100% (rubbing proximal nail fold of thumb with tip of ipsilateral second finger)
Affected nail	Predominately one or both thumbnails, occasionally nails from other digits	One or both thumbnails
Treatment	1. Keep nail plate short by cutting and smooth by buffing[5] 2. Prevent nail plate catching by covering the nail plate with either tape or a nail wrap[4,7]	1. Apply adhesive bandage to proximal nail fold to prevent direct contact from ipsilateral index finger tip 2. Fluoxetine[29] 3. Serotonin reuptake inhibitors[7]
Reoccurrence	Occasionally resolves spontaneously; however, repeated episodes often develop in these individuals[5,7,15]	Disappears with subsequent normal nail growth after habit tic ceases[28]

*Thirty-five patients were reviewed; preceding trauma was described in 7 of them.[2,4,11-22,27]

dystrophy in 3 patients who were receiving isotretinoin. The first, reported by Bottomley and Cunliffe[17] in 1992, was a 38-year-old woman who developed median canaliform nail dystrophy 6 weeks after beginning treatment with isotretinoin. Her thumbnail returned to normal 4 weeks after she discontinued the drug.[17] The second patient, described by Griego et al[4] in 1995, was an 18-year-old man who developed median canaliform nail dystrophy of both thumbnails after starting therapy with isotretinoin for his acne. The nail disfigurement became distinct after 4 months of treatment; his new thumbnail dystrophy resolved 5 months after he discontinued the medication.[4] A third patient was reported by Dharmagunawardena and Charles-Holmes[12] in 1997. They described a 19-year-old man who developed median canaliform nail dystrophy in both thumbnails within 4 weeks after starting treatment with isotretinoin for his acne. His nails returned to normal 3 months after completing a 5-month course of isotretinoin therapy.[12]

Familial median canaliform nail dystrophy has not been associated with any systemic syndromes. In our patient and his family, the nail dystrophy was not congenital but rather appeared as an acquired abnormality of the nails in adulthood.

The etiology of median canaliform nail dystrophy is unknown.[5,7,13,16-18] It usually is an acquired condition. Nail matrix trauma may precede the onset; however, an associated nail injury has often not occurred.[3-7,12-16,19-21] This nail dystrophy is not considered to be inherited. The familial occurrence of median canaliform nail dystrophy has rarely been described. Indeed, to the best of our knowledge, in addition to our patient, only 3 families with median canaliform nail dystrophy have been described.[20,22,23] In the first such family, a 16-year-old girl with bilateral median canaliform nail dystrophy of her thumbnails since the age of 13 years had a mother with similar-appearing thumbnails.[20] A second such family also included a mother and daughter.[22] Long longitudinal grooves were present in the daughter's left thumbnail since the age of 11 years; her mother had a similar dystrophy involving her right thumbnail that began when she was 12 years old. Her mother, currently 34 years old, still has recurrent episodes of spontaneously resolving median canaliform nail dystrophy. The family had no history of other hereditary diseases.[22] The third family in which median canaliform nail dystrophy occurred was reported by Bossi[23] in the Italian literature in 1965. Our patient and his brother and mother represent the fourth such family.

The differential diagnosis of median canaliform nail dystrophy includes habit tic deformity (Table). It also includes other causes of longitudinal splits in the nail plate such as direct trauma to the nail unit. In addition, digital mucous cyst (synovial cyst), lichen striatus, nail-patella syndrome, pterygium, Raynaud disease, and trachyonychia are other conditions in which a longitudinal nail defect has been described.[5,7,30,31]

Habit tic deformity is usually present in one or both thumbnails and results in alteration of the normal nail growth. It is caused by the constant or habitual rubbing of the thumb's proximal nail fold by the tip of the second digit. The subsequent damage to the nail matrix causes clinical changes in the nail plate that appear different than those of median canaliform nail dystrophy. The habit tic deformity produces transverse ridges along the central nail plate depression instead of a longitudinal groove with lateral projections. The depth of the central nail plate canal depends on the intensity of the inflicted trauma by the index finger to the matrix of the thumbnail. In addition, the lunula may appear red and enlarged.[9,29] Also, the proximal nail fold may be swollen.[5,13]

Median canaliform nail dystrophy has occasionally been described to periodically disappear; often, the nail defect reappears in these individuals.[4-7,13,15,17,24] In some patients, the central nail defect is replaced by a longitudinal ridge[5,6]; however, in most patients, such as ours, the condition does not resolve spontaneously. Keeping the nail length short and buffing the surface of the nail can prevent the edge of the nail plate from catching on clothing and other objects.[5] Covering the nail plate with tape or a nail wrap also can aid in ensuring that jagged edges are not present.[4,7]

Conclusion

Familial median canaliform nail dystrophy has rarely been described. Our patient had adult onset of his condition involving both thumbnails with associated red macrolunula. His brother and his mother also experienced the same nail dystrophy. Including our patient and his family, median canaliform nail dystrophy has only been reported in 4 families. The mode of inheritance for median canaliform nail dystrophy in these families remains to be determined.

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