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Dermatology Case Reports

Case Report

Median Canaliform Dystrophy of Heller Affecting Multiple Nails: A Rare Clinical Entity

Harris Shaafie

ERA University, Lucknow, India

ABSTRACT

Malaria remains a leading cause of ill health in Africa and Nigeria. From the world malaria report, 2018 53 million annual cases in Nigeria (1 in 4 persons), contributing 25% global burden and 53% of cases in West Africa. In Nigeria alone, 81,640 deaths are recorded annually (9 deaths per hour), which accounts for 19% global malaria deaths (1 in 5 global malaria deaths) and 45% malaria deaths in west Africa. The Nigeria Malaria Strategic Plan (NMSP) 2014-2020 has as its goal-to reduce malaria burden to pre-elimination levels and bring malaria-related mortality to zero.

In North-East Nigeria, malaria transmission is perennial with a marked seasonal peak from July to November each year. Since malaria is highly endemic in the north-east, increasing the burden on health resources and elevating the risk of morbidity and mortality among the affected population, particularly children under five who are one of the vulnerable groups, Seasonal Mass Chemo-Prevention (SMC) during the rainy season to reduce morbidity and mortality in emergency settings was deployed in 2018 and the results were enormous 6.5% reduction in fever cases and confirmed malaria, when compared with previous years in children under five who benefited from SMC as obtained from the study of effect of SMC on malaria morbidity conducted in Adamawa State in 2018. To achieve a better impact on malaria control, a combination of preventive measures (robust surveillance, indoor residual spray, using insecticide-treated nets), effective case management and improved capacity of personnel is recommended.

Keywords: Familial; Median canaliform dystrophy of heller; Tacrolimust

INTRODUCTION

Median Canaliform Dystrophy (MCD) is a rare clinical entity first described by Heller in 1928 [1]. It is characterized by a midline or a paramedian split and trench development in the nail plate. Small cracks or fissures that extend laterally from the central canal give the appearance of an inverted fir tree or Christmas tree. It is hypothesized that the condition results from a temporary defect in the matrix which interferes with nail formation [2], however the exact pathophysiology remains unknown. The majority of the cases are idiopathic, but cases implicating trauma in the form of habitual picking of the nails [3], oral isotretinoin use [4,5] and familial history [2,6,7] have been reported in the literature. Spontaneous remission is often seen after months to years, although the disease has a tendency to recur. To date, no therapy has proven to be consistently successful in effectively managing the condition. Commonly utilized treatments have been intralesional triamcinolone acetonide injections into the dystrophic nail [8], topical application of 0.1% tacrolimus ointment [9,10] and topical tazarotene 0.05% ointment [8], but lack considerable supporting evidence. Here, we report a case of a 14-year-old male presenting with MCD affecting multiple fingers and toe nails.

CASE REPORT

A 14-year-old Indian boy presented to us in the Dermatology outpatient department with complaints of asymptomatic lesions over multiple fingers and toe nails since one year. A detailed clinical history ruled out habitual picking of nails, nail-biting, prolonged contact with irritants or allergens, psychological stress, oral retinoid use or any existing co-morbidities. The patient's mother had a history of similar lesions on the toes during her teenage years which resolved spontaneously on their own. There

Correspondence to: Harris Shaafie, ERA University, Lucknow, India, E-mail: harris.shaafie@gmail.com

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was no history of psychiatric illness in the other family members. On examination, both the great toe nails showed well-defined, median longitudinal grooves extending from the proximal nail fold to the distal nail edge with transverse furrows stretching from either side of the groove in a fir-tree configuration with enlarged lunulae (Figure 1). The finger nails of the second and third digits showed a single transverse groove on both sides with normal-appearing lunulae (Figure 2). The lateral and proximal nail folds were normal in all affected nails. A 20% potassium hydroxide mount prepared from the nail plate and subungal scrapings were negative for fungal elements. The biopsy was not performed as it would not aid in the therapeutic management of the patient and the diagnosis of MCD is primarily based on clinical grounds. Keeping in mind the results of previous case reports, the patient was started on 0.1% tacrolimus ointment topically under occlusion at night along with jojoba oil application once daily for nourishment and moisturization. The patient was also advised to avoid forceful pushing back of the nail cuticle and keeping the nail length short to avoid the jagged edges catching on any clothing material or while walking. The patient was followed up every month for the next 3 months.

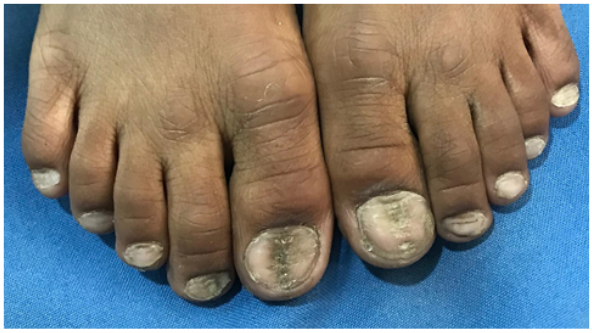

Figure 1: Symmetrical, median longitudinal grooves extending from proximal nail folds to the distal nail edges on the both the great toes with transverse furrows stretching from either side of the groove in a fir-tree configuration. The lunulae were enlarged.

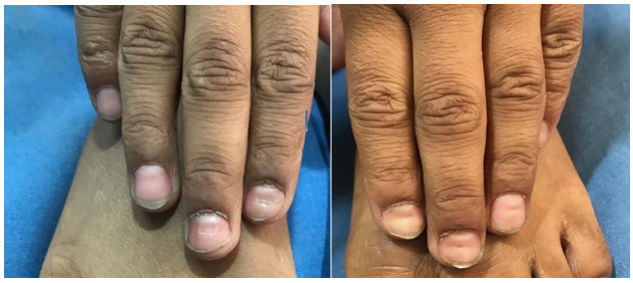

Figure 2: The finger nails of the second and third digits bilaterally showed a single transverse groove with normal-appearing lunulae. The proximal and distal nail folds were normal appearing.

DISCUSSION

Median canaliform dystrophy (MCD) of Heller, also known as Dystrophia Unguis Mediana Canaliformis and Nevus Striatus Unguis is a dystrophic condition of the nail in which a longitudinal groove extends from the proximal nail fold to the end of the nail plate. Lateral extensions of this groove give an inverted fir tree like appearance. Thickening of the proximal nail fold, enlargement, and redness of the lunula have also been reported. The etiology of median canaliform nail dystrophy is unknown [11-13]. It usually is an acquired condition with no racial predilection. The majority of the cases described have onset during the late teen years [3-5,11,14]. The condition is usually symmetrical most commonly affecting the thumbs [4,9,15], albeit other fingers and toes may be affected. To our knowledge, only one such case has been previously reported in the literature with the involvement of both great toe nails [10]. Histopathology shows parakeratosis and accumulation of melanin within the longitudinal groove of the affected nail plate and between the nail bed keratinocytes [1,16]. Systemic syndromes have not shown any association with familial median canaliform nail dystrophy. The familial occurrence of median canaliform nail dystrophy has rarely been described. As per our literature survey, only four families with median canaliform nail dystrophy have been described to date [6,12,17]. In all four families, the mother was a common member affected. The differential diagnosis of median canaliform nail dystrophy includes habit tic deformity, digital mucous cyst, lichen striatus, nail-patella syndrome, nail pterygium, Raynaud disease and trachyonychia all of which show longitudinal ridging of the nail plate [3]. The habit tic deformity is the closest differential which produces transverse ridges along the central nail plate depression (Figure 3) [18] instead of a longitudinal groove with lateral projections [5] as seen in MCD (Figure 4). It is caused by the constant or habitual rubbing of the thumb's proximal nail fold by the tip of the second digit. Subungual skin tumors such as glomus tumors [13] and myxoid tumors have also been described to cause longitudinal grooving, lifting off the nail plate from the nail bed resulting in a tube-like structure (solenos) distal to it. Hence, MCD has also been referred to as solenonychina. Co-existing conditions have been observed in some patients with median canaliform nail dystrophy. Sutton RL Jr. [19] described the case of a 19-year old woman who presented with a fragment of fleshy tissue within the dystrophic nail canal. The extraction of this tissue resulted in the resolution of the nail deformity, however, the condition reappeared. The management of MCD is a therapeutic challenge as no therapy has been shown to be consistently successful. Many patients present quite late for a dermatology opinion due to the relatively asymptomatic nature of the disease and many times irreversible damage to the nail sets in. If the patients have a known psychiatric illness, for example, impulse-control or obsessive-compulsive disorder, a psychiatry consultation must be sought. Tacrolimus, formerly known as FK506, is a macrolide antibiotic with immunosuppressive properties. It exerts its effects principally through impairment of gene expression in target cells. Tacrolimus binds to an immunophilin, FK506 Binding Protein (FKBP) (Figure 5). This complex inhibits calcineurin phosphatase. The drug inhibits calcium-dependent events, such as interleukin-2 gene transcription, nitric oxide synthase activation, cell degranulation, and apoptosis [20].

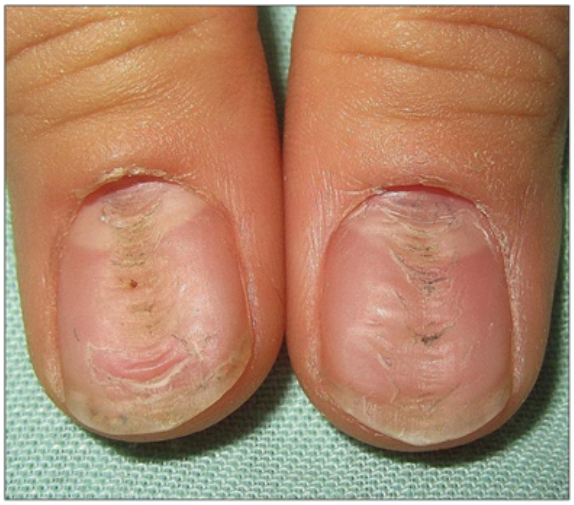

Figure 3: Transverse ridges along the central nail plate depression in habit tic deformity.

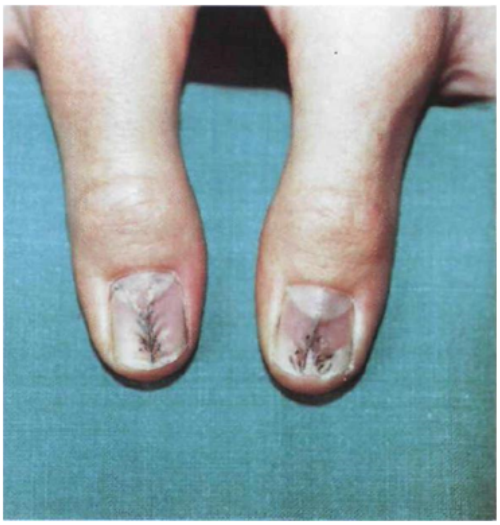

Figure 4: Longitudinal groove with lateral projections in MCD.

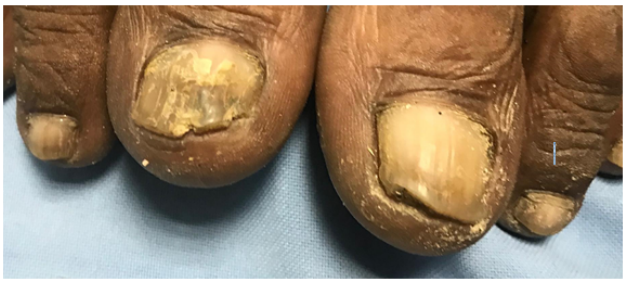

Figure 5: After 3 months of follow up, the lesions showed substantial resolution in the great toes after topical tacrolimus 0.1% ointment.

CONCLUSION

Familial MCD has rarely been reported. MCD involving the great toe nails is an even rarer entity. In our study, where the patient had involvement of both his great toe nails, a positive family history in the mother, no psychiatric co-morbidity and no significant drug history, the exact etiology cannot be effectively determined. Tacrolimus through its immunosuppressive action has proven to be effective in the management of our case thereby raising the possibility of an immune-mediated mechanism underlying the pathophysiology of MCD. Genetic studies need to be performed in order to determine the mode of inheritance for familial MCD.

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