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Case Report

Familial median canaliform dystrophy of heller affecting multiple nails: a rare clinical entity

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ABSTRACT

Median canaliform dystrophy (MCD); first described by Heller in 1928 is characterized by a midline or paramedian split in the nail plate. Mostly idiopathic, some identifiable causes include habitual picking of the nails, oral isotretinoin use and positive family history. The familial occurrence of MCD has rarely been described. Till 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, topical 0.1% tacrolimus ointment and topical tazarotene 0.05% ointment. Despite available treatment options, the condition has a tendency to recur. Here; we report a case of a 14-year-old male presenting with familial MCD affecting multiple finger and toe nails.

Keywords: Familial, Heller, Median canaliform dystrophy, Tacrolimus, Nail

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. It is hypothesized that the condition results from a temporary defect in the matrix which interferes with nail formation, however the exact pathophysiology remains unknown.[2] Majority of the cases are idiopathic, but cases implicating trauma in the form of habitual picking of the nails, oral isotretinoin use and familial history have been reported in the literature.[2-9] Spontaneous remission is often seen after months to years, although the disease has a tendency to recur. Till 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, topical 0.1% tacrolimus ointment and topical tazarotene 0.05% ointment, but lack considerable supportive evidence.[2,10] Here, we report a case of a 14-year-old male presenting with MCD affecting multiple finger 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 toe nails for 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 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 as shown in Figure 1. The proximal and distal nail folds were normal

in all affected nails. A 20% potassium hydroxide mount prepared from the nail plate and subungual scrapings were negative for fungal elements. 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. 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. After 3 months of follow up, there was substantial resolution of the lesions in all toes following topical daily application of Tacrolimus 0.1% ointment as shown in Figure 2.

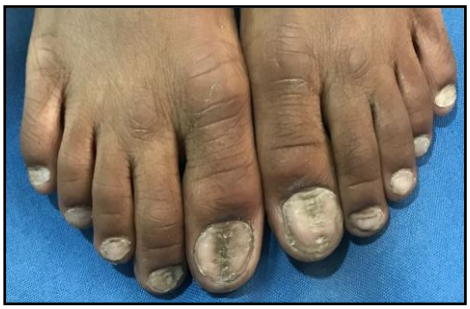

Figure 1: Median longitudinal grooves with transverse furrows stretching from either side of the grooves in an inverted fir-tree configuration.

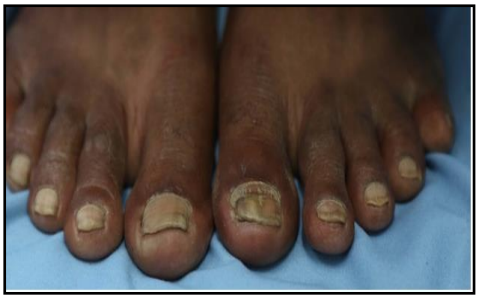

Figure 2: After 3 months of follow up, substantial resolution following topical daily application of Tacrolimus 0.1% ointment.

DISCUSSION

Median canaliform dystrophy (MCD) of Heller, also known as dystrophia unguis mediana canaliformis 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.[6] It usually is an acquired condition with no racial predilection. Majority of the cases described have an onset during the late teen years.[3,5,7,8] The condition is usually symmetrical most commonly affecting the thumbs, albeit other fingers and toes may be affected.[2-8] To our knowledge, only one such case has been previously reported in the literature with the involvement of both great toe nails. Histopathology shows parakeratosis and accumulation of melanin within the longitudinal groove of the affected nail plate and between the nail bed keratinocytes.[1] 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 till date.[4-7] In all four families, the mother was a common member affected. The differential diagnosis of MCD include 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] Habit tic deformity is the closest differential which produces transverse ridges along the central nail plate depression instead of a longitudinal groove with lateral projections as seen in MCD.[5] 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 and myxoid tumors also cause longitudinal grooving, lifting off the nail plate from the nail bed resulting in a tube-like structure (solenos) distal to it.[1] Hence, MCD has also been referred to as solenonychina. 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 a 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 exerts its effects principally through impairment of gene expression in target cells. Tacrolimus binds to an immunophilin, FK506 binding protein (FKBP) and 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.[8] To conclude, Familial MCD has rarely been reported. MCD involving the great toe nails is an even rarer entity. In our study, where the patient had an 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.

CONCLUSION

To summarize, median canaliform dystrophy belongs to a heterogeneous group of a rare nail conditions with far from satisfactory line of management. We report this case to

highlight the fact that often in such cases, the history of 'habit tic' may not be acknowledged by the patient.

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