

Exuberant Koenen tumors: effective treatment with amputation of the nail apparatus and reconstruction with full thickness skin grafts

Tumores de Koenen exuberantes: tratamento efetivo com amputação do aparelho ungueal e reconstrução com enxerto de pele de espessura total

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ABSTRACT

Introduction: Tuberous sclerosis is a dominant autosomal genodermatosis, classically characterized by facial angiofibromas, fibrotic plaques, periungual fibromas, leaf-shaped hypochromic maculae and fibrous frontal plaques.

Case report: A patient bore this disease, with Koenen tumors in all nail beds of hands and feet, which were resistant to the conventional treatment – simple excision and electrosurgery. Amputation of the nail beds was performed followed by reconstruction with full thickness skin grafts.

Conclusion: The amputation of the nail apparatus and reconstruction with full thickness skin grafts in aggressive, recurrent or high morbidity cases can be an effective therapeutic option.

Keywords: dermatology; dermatologic surgical procedures; tuberous sclerosis; skin transplants

RESUMO

Introdução: A esclerose tuberosa é genodermatose autossômica dominante, classicamente caracterizada por angiofibromas faciais, placas fibróticas, fibromas periungueais, máculas hipocrômicas em formato de folhas e placa fibrosa frontal.

Relato do caso: Paciente portador dessa doença, com tumores de Koenen em todos os leitos ungueais de mãos e pés, recorrentes ao tratamento convencional de exérese simples e eletrocirurgia. Foi realizada a amputação dos leitos ungueais seguida de reconstrução com enxerto cutâneo de espessura total.

Conclusão: A amputação dos aparelhos ungueais e reconstrução por enxerto cutâneo de espessura total para casos agressivos, recorrentes ou que determinem alto grau de morbidade, pode ser uma opção terapêutica efetiva.

Palavras-chave: dermatologia; procedimentos cirúrgicos dermatológicos; esclerose tuberosa; transplante de tecidos

INTRODUCTION

Koenen tumors (KT) are a characteristic and one of the major criteria for the diagnosis of tuberous sclerosis complex (TSC). It usually appears after puberty in up to 50% of patients with TSC.¹

TSC is an autosomal dominant genodermatosis classically characterized by facial angiofibromas, fibrotic plaques (Shagreen patch), periungual fibromas (KT), hypochromic leaf-shaped macules (ash leaves) and fibrous frontal plaque.^{1,2} Seizure, mental retardation, hamartomas of the retina, subependymal nodules

Case report

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and hamartomas of the internal organs are common associations.³ Half the families with TSC are linked to the 9q34 chromosome, with inactivating mutations of the tumor suppressor genes of the protein hamartin (TSC1) and the other half to the 16p13 chromosome with inactivating mutations of the tumor suppressor genes of the protein tuberlin (TSC2). The hamartin/tuberlin complex is an important inhibitor of tumor growth and its absence triggers the loss of inhibition over cell proliferation and migration.¹⁻³

This study has the objective of demonstrating an unusual presentation and multiple florid KT, that were surgically treated.

CASE REPORT

We report the case of a 47-year-old female patient with a history of hypochromic, lenticular, confetti-like and leaf-shaped (ash leaves) lesions on her body since the first decade of life, progressing with papular lesions on the face (angiofibromas) and peduncular keratotic confluent lesions in all nail beds of both feet and hands (KTs), that were bigger in size but morphologically similar to periungual viral warts. She also had recurrent seizures.

The patient complained of pain in the toes and recurrent local infections, difficulty in walking and putting on shoes due to the lesions on the area. Multiple sessions of electrosurgery and simple excision were performed, but the lesions always recurred in less than 2 months of follow up. Because of the clinical picture's exuberance and morbidity, we opted to amputate all nail beds and reconstruct them with full thickness skin grafts (Figure 1). After 24 months of follow up, the patient remains with no lesions (Figure 2), with complete control of local pain and infections.

KT are periungual or subungual fibromas manifested as pink or skin-colored papules and nodules, that can be multiple and affect mainly the toes. On histology, they are characterized by stellate fibroblasts and dense collagen, with numerous ectatic blood vessels.^{1,4}



FIGURE 2:
Before and 24 months
after surgical treat-
ment

They usually vary between 5 to 10 mm in size, but can be much larger. This can be the only clinically obvious abnormality in 50% of TSC cases. The first tumors appear between 12 and 14 years of age, and progressively enlarge in number and in size with age. They sometimes become keratotic, similar to a fibrokeratoma or, more rarely and with few reports in the literature, to periungual viral warts.^{1,2,4} In this case, the patient had a florid clinical picture, with elongated, keratotic and confluent fibromas, similar to viral warts. The tumors were so aggressive that they basically replaced the nail plate in most toes, leading to local pain, recurrent skin infections, difficulty in walking and in putting on shoes.

Various treatment techniques such as dermabrasion, chemical ablation, excision and laser ablation have been described in the literature with variable results in regard to duration, and there is still no consensus on a standard treatment. Recurrence is the norm.⁵ Amputation of the nail apparatus and reconstruction with a full thickness skin graft can have removed the local stimulus for tumor formation, what explains the effectiveness of the therapeutic choice and lack of recurrence in all toes, despite the aggressiveness of the condition.

CONCLUSION

Amputation of the nail apparatus and reconstruction with full thickness skin graft for aggressive, recurrent or high morbidity cases of KT in patients with TSC can be an effective therapeutic option. More cases are needed with reports in the literature to confirm this hypothesis. ●



FIGURE 1: Amputation of the nail beds and reconstruction with full thickness skin graft

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