

Giant Koenen tumors involving 20 nails



Figure 1: Multiple, smooth to verrucous, firm, flesh-colored excrescences emerging from proximal nail folds of all nails



Figure 2: Multiple, erythematous to hyperpigmented, polypoidal and digitate growths. Garlic clove morphology with a yellow-brown scale at the tip of the central growth, and longitudinal striations on nail (Dermoscopy using Dermlite DL200 Hybrid, ×10)

A 63-year-old lady presented with multiple periungual growths which progressively increased in size and number to involve all 20 nails for last 30 years. There was no history of seizures or psychosis. Both her daughters had similar but smaller lesions on a few nails. On examination, she had multiple smooth to verrucous, firm, flesh-colored excrescences emerging from proximal nail folds of all nails, largest on the left thumb being 4 cm × 3 cm in size [Figure 1]. Dermoscopy revealed multiple erythematous to hyperpigmented polypoidal and digitate growths, garlic - clove morphology with a yellow-brown scale at the tip of the central one and multiple longitudinal striations on nail [Figure 2]. She also had multiple firm, red-brown papules on nasolabial folds and cheeks. There were multiple soft to firm skin-colored plaques on lumbosacral area and multiple ovoid hypopigmented macules on trunk and extremities. Histopathology from one of the nailfold growths revealed a central core of dense collagen with prominent interspersed fibroblasts and dilated capillaries at the distal end with overlying orthokeratosis suggestive of angiofibroma. Fundoscopy, computer tomography scan of brain, magnetic resonance imaging of brain and ultrasonography of abdomen were normal. Diagnosis of tuberous sclerosis complex was established with four major features that included facial angiofibromas, ungual fibromas (Koenen tumors), hypomelanotic macules and shagreen patch. Patient was offered excision of large angiofibromas to reduce functional impairment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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