

QUIZ (1) and (2)

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Quiz (1)

A 65-year-old patient presented with asymptomatic multiple discrete flesh colored shiny small papules affecting face and dorsum of both hands, Fig. (1,2,3,). The rest of the body including – hairs, nails, mucous membranes and genitalia were normal looking.

The pathology showed a circumscribed lymphohistiocytic infiltrate in the papillary dermis surrounded bilaterally by elongated rete ridges.

Answer of Quiz 1:

The diagnosis is lichen nitidus actinicus. The clinical variants of lichen nitidus include confluent, linear, hemorrhagic and vesicular types ⁽¹⁾. Nails may be involved ⁽²⁾. Generalized lichen nitidus is rarely described^(3,4,5).

The generalized lichen nitidus commonly involve arms, forearms, chest, penis and abdomen and face was spared ⁽⁶⁾. Generalized lichen nitidus actinicus was reported to affect children ^(7,8,9).

The present case showed lichen nitidus on face and both hands excited by exposure to sun. He had one lesion of lower lip similar to lichen planus. No proven effective treatment is known for lichen nitidus. Some therapeutic trials of lichen nitidus included the use of steroids, astemizole ⁽⁵⁾ and dinitrochlorbenzene topical application ^(10,11). Some clear spontaneously and some persist and PUVA was tried ⁽¹²⁾.

What is your diagnosis?



Fig.1- Lichen Nitidus actinicus (LNA) of face

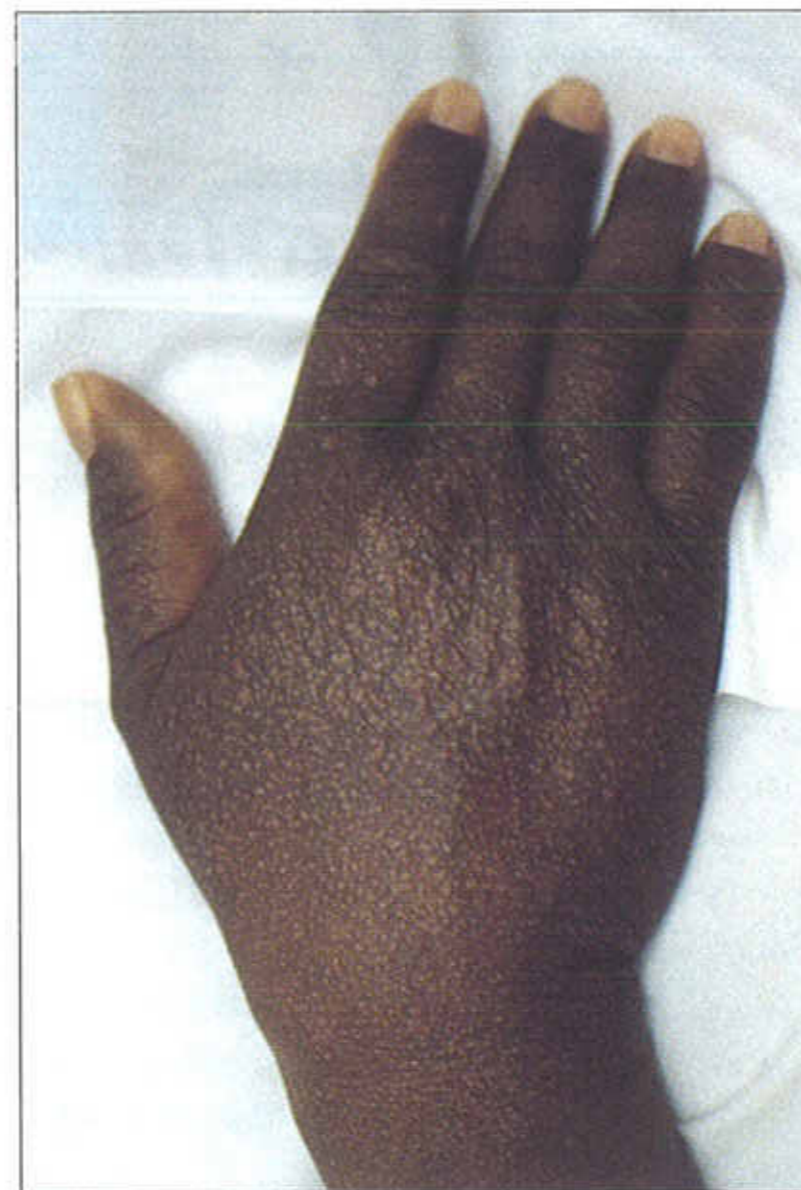


Fig.2- Right hand affected with LNA



Fig.3- Left hand affected with LNA

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Quiz (2)

A female patient 12-years old presented with two small periungual overgrowth protruding from underneath of the proximal nail fold of the right index and middle fingers (Fig.1). A third bigger mass was seen in the subungual region on terminal medial side of right big toe (Fig.2). All masses were asymptomatic.

What is your diagnosis?

Answer of Quiz 2 :

Patient has Koenen tumors, which are periungual fibromas that were recorded photographically by Joannes Henricu Maria Koenen in 1932⁽¹⁾. The periungual fibromas in tuberous sclerosis were first described by Richard Kothe in Munich in 1903⁽¹⁾. Histologically the distal part of the fibroma shows capillaries surrounded by thin collagen bundles and the proximal part is made of dense collagen, which fades into the normal structure of the dermis of the proximal nail fold. The overlying epidermis appears to be connected with the nail fold⁽²⁾.

Such tumors are treated either by complete surgical excision^(3,4) or by Argon laser surgery, which was reported to be without recurrence during a follow-up period of 5-years⁽⁵⁾. Carbon dioxide laser is an alternative treatment and has the advantage of reduced treatment time and reduced bleeding^(6,7,8).

Tuberous sclerosis is an autosomal dominant disorder, which may involve almost any organ system, and periungual fibromas are common skin manifestation and is a source of cosmetic or functional concern⁽⁶⁾.

There is strong evidence for the heterogeneity of tuberous sclerosis with approximately half are linked to a locus on chromosome 9 and half linked to chromosome 16p13.3⁽⁹⁾.

The disease is characterized by wide spread cutaneous and visceral hamartomas. The most frequent finding was ash leaf white macules seen in 97.2% and may be seen at birth or in the first months or first 2-years of age⁽¹⁰⁾. Facial angiofibromas are seen in 74.5%, shagreen patches usually in lumbosacral region in 48.1%, café au lait macules in 28.3%. Molluscum fibrosum pendulum in 22.6%, forehead fibrous plaque in 18.9%, periungual fibroma in 15.1% and wide spread confette

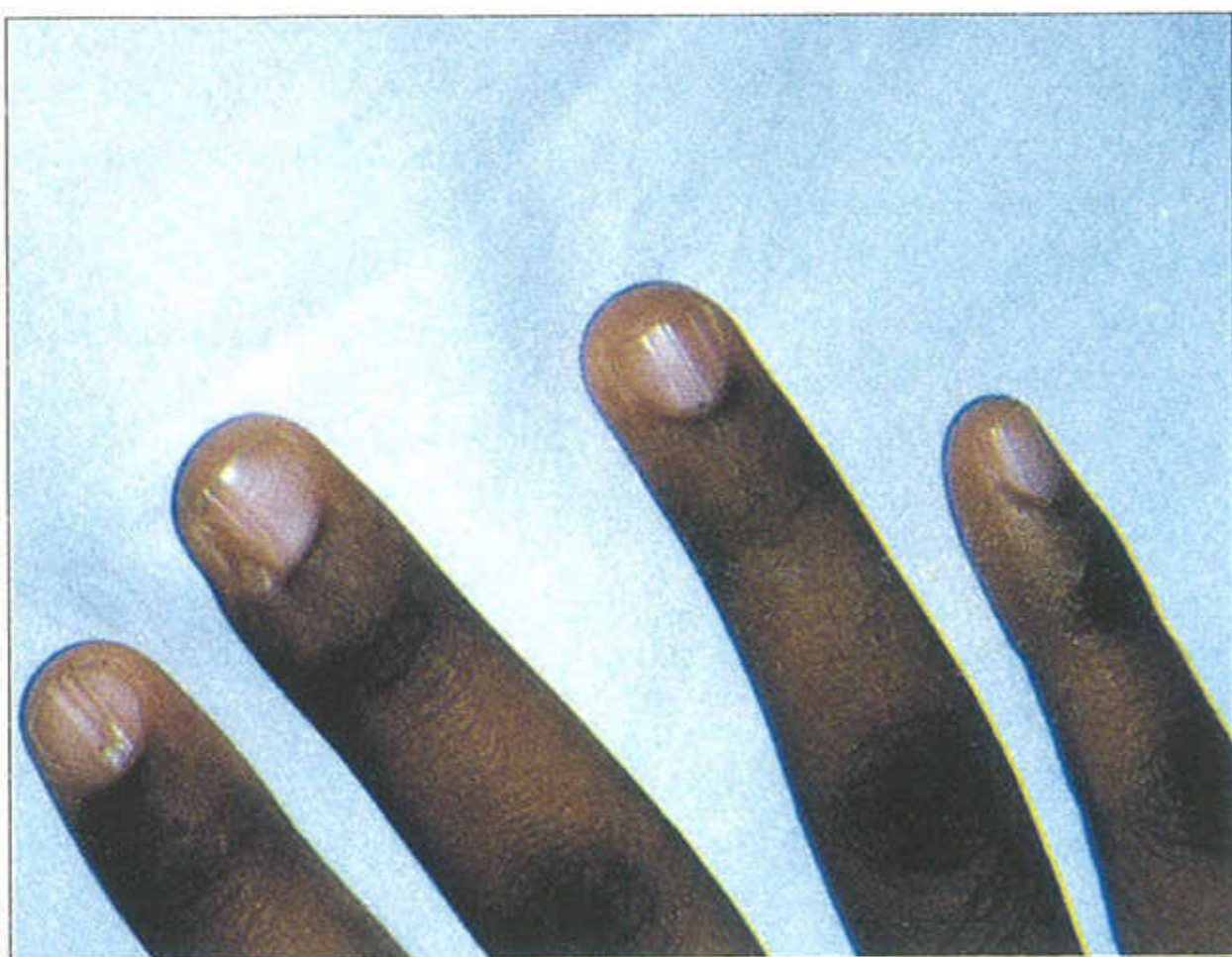


Fig.1 Periungual swelling



Fig.2 Subungual swelling right big toe.

like macules or spots in 2.8%⁽¹⁰⁾. Hyperostosis of the cranium⁽¹¹⁾ and hamartomas of brain, eye, kidney, heart, mental deficiency, epilepsy, paraventricular calcification, skeletal disorders and rarely ophthalmic tumors were described^(7,12). Oral lesions include nodules of mucosa, oral fibromas, enamel hypoplasia, mandibular cysts and subcuta-

neous fibromatosis of subcutaneous tissue overlying the angle of the mandible was described⁽¹²⁾.

Typical cutaneous findings in tuberous sclerosis are present in over 90% of cases and represent one of the earliest markers of the syndrome⁽¹³⁾. The Koenen peri-ungual lesions should take its place along side the major cutaneous signs of this disease⁽¹⁴⁾.

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