LETTERS TO THE EDITOR

ADENOMA SEBACEUM DEVELOPING WITHIN A HYPOMELA-NOTIC LEAF-SHAPED MACULE OF TUBEROUS SCLEROSIS

A 9-year-old boy developed an asymptomatic well-defined, dull-white, hypopigmented, leaf-shaped 3×2 cm oval patch on the left side of the front of neck, since birth. The patch did not show any sensory impairment and the vascular response to stroking was normal. At the age of 4, he developed multiple, discrete 0.5 to 2 cm sized, pinkish nodules with smooth glistening surfaces on the nose, cheeks and chin in a bilaterally symmetrical fashion. He developed generalised seizures since 2 years and was on daily phenobarbitone and dilantin. A nodule closely resembling those on the face, developed gradually within the hypopigmented leaf-shaped macule since the last 2 years (Fig. 1). A skin-



Fig. 1. Multiple pinkish dome-shaped papules of adenoma sebaceum on the face, a hypomelanotic leaf-shaped macule on the left side of front of neck, with a pinkish nodule of adenoma sebaceum that developed within the hypomelanotic leaf-shaped macule.

coloured elevated flat 3×6 cm plaque with knobby surface was seen on the lumbar area. All other systems were clinically normal. All laboratory tests including skiagrams of the skull and chest were normal. ECG and EEG did not show any abnormality. Biopsy of the nodule revealed hyperplasia of the dermal connective tissue, vascular elements, hair follicles and sebaceous glands.

The presence of cutaneous lesions viz adenoma sebaceum, shagreen patch and congenital hypomelanotic leaf-shaped macule, in association with epilepsy suggested a diagnosis of tuberous sclerosis in our patient. The hypomelanotic leaf-shaped macule may permit a prospective diagnosis of tuberous sclerosis even if other cutaneous findings are absent. Its presence since birth, dull-white incomplete loss of pigmentation rather than ivory-white depigmentation, absence of surrounding hyperpigmentation and persistence without any alteration in shape and size with age, help to differentiate it from vitiligo. Non-obliteration of the margin on diascopy and normal vascular response to stroking easily differentiate it from nevus anaemicus. The interesting feature observed in this case is the development of adenoma sebaceum (angiofibroma) within the hypomelanotic leaf-shaped macule. To the best of our knowledge such a phenomenon has not been recorded previously. Its mechanism remains unknown, may be a simple chance occurrence.

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