

UNUSUAL CUTANEOUS MANIFESTATION OF TUBEROUS SCLEROSIS (A case report)

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Summary

Cutaneous manifestations are found in 60 to 70 % cases of tuberous sclerosis and consist of adenoma sebaceum, periungual fibromas, Cafe au lait spots, shagreen patches and white macules. Our patient showed unusual skin manifestations like spotty pigmentation on the chest, back and abdomen and hyperkeratosis palmaris et plantaris.

Pringle is credited with the first description of skin lesions of adenoma sebaceum in 1890. Von Recklinghausen described the scleromatous brain lesions in 1863. Bournville grouped together adenoma sebaceum, mental deficiency and epilepsy in 1880. This symptom complex is now known as Bournville's Syndrome. Hallopeau-Leredde in 1895 emphasized the sclerosis and described shagreen patches. Sherlock in 1911 used the term epiloia for the ensemble of convulsions, mental retardation adenoma sebaceum and tumours of the brain and other organs. Van der Hoeve first described the ocular findings of this condition in 1920. Chao¹, Gold², Freeman² and Lagos³ described various skin manifestations of tuberous sclerosis but it appears that the description of the syndrome is still incomplete. It is a neurocutaneous disorder. The syndrome of tuberous sclerosis exhibits some of several of its well known manifestations like epilepsy, mental retardation, ade-

noma sebaceum, periungual and sub-ungual fibromas, shagreen patches, white macules, tumors of the retina (phacomias) and calcification in the basal ganglia. Other rare manifestations of this disease are fibromas of the gums, palate, tongue, larynx, pharynx, lungs, heart, kidney, congenital rhabdomyoma and cyst like lesions of the phalanges. Irregular thickening of cortex of metatarsals, metacarpals and vertebral lesions may also be present. Associated developmental abnormalities are also reported. Skin lesions are found in 60-70% of cases. Our patient showed skin lesions which are hitherto not described in literature.

Case Report

A 50 years old male patient attended skin department of New Civil Hospital, Surat on 14-7-79 complaining of growths from nailfolds of hands and feet and small lesions on the face for eight years. The lesions had shown increase in size for about 2 years prior to the hospital visits. Family history revealed that all three of patient's children were epileptics receiving regular anticonvulsive therapy. Patient had swellings of the gums earlier and was

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Fig. 1 Shows adenoma sebaceum on the left nasolabial fold and left side of the face.

treated for the same by a dental surgeon. On examination skin showed small pinhead sized reddish discrete papules on the face suggestive of adenoma sebaceum (Fig. 1) Multiple, smooth, firm, flesh coloured, fibromatous, nontender periungual and subungual tumours were seen arising from under all nail folds of hands and feet (Figs. 2 & 3). The nails were thick,

brittle and distorted. A shagreen patch was found on the lumbosacral region. Cafe¹ au-lait patches and white macules were not present. Spotty pigmentation was present all over the trunk. Palms and soles showed hyperkeratosis, (Fig. 4).

Respiratory system revealed diminished air entry in both the bases with fine crepitations. Spleen was enlarged 2 finger breadth below left costal margin. C. V. S., C. N. S. and eyes did not show any abnormality.

Investigations

X-rays of the hands showed cysts and irregular periosteum of proximal phalanx of right index and ring fingers and cystic recession of the lateral aspect of left fifth proximal phalanx. Exostoses were found on lateral sides of metacarpals proximally. Chest X-ray showed reticulation and interstitial fibrosis of lower zone with infiltrations. Cardiac size and contour were normal. X-ray of skull did not show any abnormality.

Histopathology of the biopsy material from one of the periungual growths revealed fibrous tissues and fibroblasts suggestive of fibromas. Lesion on the face showed regeneration of sebaceous glands, dilatation of capillaries and



Fig. 2 Shows periungual fibromatas from the nail folds of the fingers of both hands.



Fig. 3 Shows periungual fibromas from the nail folds of the toes of both feet. Bandage indicates site of biopsy.



Fig. 4 Shows hyperkeratosis of the skin of the soles.

lymphatic vessels, as well as fibrosis and inflammatory reaction with atrophy around hair follicle. Haematological investigations were normal.

Discussion

This case is presented due to the interesting hitherto unrecorded findings

of (1) pigmented spots on the trunk. Perhaps this represented a freckling seen in those cases which are associated with neurofibromatosis (2) presence of palmo-plantar keratoderma. Both these findings were absent among the rest of patient's family members.

References

1. Chao D: Congenital neurocutaneous syndromes in childhood J pediat 1959. 55 : 447.
2. Gold AP and Freeman JM : Depigmented nevi, the earliest sign of tuberous sclerosis, J Pediat 1965, 35 : 1003.
3. Lagos JC and Gomez MR : Tuberous sclerosis, reappraisal of a clinical entity, Mayo Clin Proc 1967, 42 : 26.

Announcements...

International Dermatopathology Symposium

The 2nd International Dermatopathology Symposium, entitled "Dilemmas and directions in differential diagnosis" will be held at Grosvenor House Hotel, Park Lane, London, 12-15 July 1981. The symposium is co-organized by Prof. E. Wilson Jones and Dr. Martin Black of the Institute of Dermatology, University of London, and Dr. A. Bernard Ackerman of the Section of Dermatopathology, New York University School of Medicine. The emphasis will be on histologic clues to diagnosis by conventional microscopy, clinical, electron microscopic and immunopathological aspects of inflammations, neoplasms, malformations and deposits of abnormal materials in the skin.

All enquiries should be addressed to : Marcus Summersfield, London Symposium 1981, Conference Co-ordinates, Regent House, 60 King Street, Twickenham, Middlesex TW1 3SH, with the exception of specific enquiries regarding the medical aspects of the programme which should be addressed to : Prof. E. Wilson Jones, Institute of Dermatology, St. John's Hospital for Diseases of the Skin, Lisle Street, Leicester Square, London WC2H 7BJ.

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