

# CUTANEOUS MANIFESTATIONS OF TUBEROUS SCLEROSIS

By

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Tuberous sclerosis refers to nodules of potato-like appearance and consistency that are found distributed over the cortex of the cerebral hemispheres. Its most striking manifestation is adenoma sebaceum which very frequently occurs with mental deficiency and epilepsy comprising the triad. It usually manifests itself early in life and occurs with equal frequency in both sexes<sup>2</sup>. The dermatologic aspects of tuberous sclerosis have been beautifully recorded by Butterworth and Wilson<sup>1</sup> and Butterworth and Streat<sup>2</sup>. The clinical reappraisal of the entity has been nicely done by Lagos and Gomez<sup>4</sup>, while Yakovlev and Guthrie<sup>5</sup> and Critchley and Earl<sup>3</sup> discussed neurocutaneous syndromes in their papers in details. Recently, we had an opportunity of seeing a case who reported with some of the interesting clinical manifestations which are recorded along with the histological observation in the present communication.

## CASE REPORT

S. D., 20 years female was referred to Dermatologic out-patient section of Sir Sunderlal group of hospitals by the junior author (N. S.) with nonitchy papular eruptions over the face. These eruptions were first noticed by the parents of the patient when she was only of five years of age. Since then the lesions have been progressively increasing in size and number to assume the present form. No history of associated epileptic fits could be elicited at any time during the illness. All the other members of the family were normal and no history of such a disease was present in the maternal or fraternal relations of the patient.

Examination of the skin surface revealed multiple, erythematous papular eruptions of the size varying from pin-head to pea. These lesions were distributed on the side of the nose, naso-labial folds, cheeks, forehead and chin, bilaterally disposed (Fig. 1). Many skin tags were also seen on the back and side of the neck extending on to the shoulders (Fig. 2). The skin of the back showed localized areas of thickened skin especially in the lumbo-sacral region the so called 'shagreen' patches. The mucous membranes and the nails were conspicuously normal. On general physical examination no abnormality in any system could be found.

Investigations: The examination of the urine, stools and blood did not show any deviation from normal. The serum-proteins, the fasting blood sugar (99 mgm. %), blood-urea (27 mgm %), serum-cholesterol (136 mgm. %), and cerebro-spinal fluids were also reported to be within the normal range. The skiogram of the chest and abdomen did not show any abnormality, while radiograph of the skull showed a localized area of calcification in the antero-posterior and lateral views (Fig. 3). The electrocardiogram was also normal.

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### HISTOPATHOLOGICAL OBSERVATIONS

The skin sections from a papule on the forehead stained with hematoxylin and eosin showed changes in the dermis characterised by an increase in the number of immature and mature hair follicles while sebaceous glands were normal. The collagen tissue was greatly increased and condensed in the whole of dermis including the papillary region. The epidermis was found to be atrophied at places (Fig. 4). The skin sections from a 'shagreen' patch showed a great increase and condensation of collagen tissue in the dermis, while the epidermis was found to be atrophied at places (Fig. 5). The Van Gieson stained preparation showed an increase in the collagen tissue with condensation of the fibres in the dermis.

### DISCUSSION

The interesting cutaneous manifestations seen in the present case were comprised of adenoma sebaceum, skin tags and the so called 'shagreen' patches. Surprisingly, the patient has not so far developed the cerebral signs despite the evidence of calcification in the skull. It has been pointed out by Lagos and Gomez<sup>1</sup> in their recent publication that the calcification and mental symptoms may be unrelated to each other. Further, Butterworth and Streat<sup>2</sup> suggested that there is no apparent relationship between the number of cutaneous lesions and the extent of the cerebral sclerosis. It seems possible that patient may develop the other ingredients of the triad at a later date. The clinical and histological features, however, conform to Pringle's disease.

### SUMMARY

A case of tuberous sclerosis (Pringle) demonstrating cutaneous manifestation of adenoma sebaceum, skin tags and shagreen patches along with histological and radiological features is reported in a twenty year old female.

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