TUBEROUS SCLEROSIS IN A BANTU FEMALE

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Whereas it is well known that races may vary in their proneness to certain diseases, such differences are probably being increasingly recognized as due to environmental factors. Where apparent differences between races exist in the incidence of some conditions, it may be found that these discrepancies are due to inadequate investigation, and to this group may belong the disease termed epiloia.

The condition known as tuberous sclerosis or epiloia does not thus far seem to have been reported in the Bantu races of Africa. Bourneville, in 1880, first established tuberous sclerosis as a morbid entity. Brain[1], in 1940, categorically stated that it occurred in White people only, but subsequently he amended this view and said that 'it is almost confined to the White races', chiefly the poorer classes.[2] Kinnier Wilson[3] also held the view that tuberous sclerosis seemed to leave the Coloured races alone. Penrose,[4] who suggested that the familial incidence pointed to some genetic mechanism and that it might be due to the coincidence of two or more dominant genes, made no mention of its occurence in different races. Mayer-Gross, Slater and Roth[5] expressed the view that it appeared to be due to a simple dominant gene, and they quoted Gunther and Penrose (1935) as having estimated that a quarter to half of all cases were due to fresh mutation; they did not refer to racial incidence.

The slow recognition of this disease in the South African Coloured races is no doubt due to the fact that there is as yet no special institution for non-European mental defectives in the country, and hence the condition rarely comes under the observation of those at all familiar with it. During 1948 an unmistakeable case of epiloia was recognized on its admission to the Tower Hospital, Fort Beaufort, by Dr. L. van Dam, but not reported by her. This patient is still in hospital, and the case, in view of the alleged rarity of epiloia in non-European races, is thought to be of interest.

The patient was born on a farm near Grahamstown in 1932. She is not known to have had any European ancestors and her appearance is typically negroid. She has the somewhat lighter-shade brown skin commonly seen in South African Natives. Her family history, obtained recently from the owner of the farm

where she grew up, was to the effect that 'her own sister has a sprinkling of warts all over her face; her mother and one of her mother's sisters each had a big bunch of what looked somewhat like warts on the sides of the nose'.

In 1948 at the age of 17 years she was admitted to mental hospital because her father had reported that she suffered from convulsions which were increasing in frequency; according to him the fits had started several years previously and at first occurred about once per month, but had increased to 2 or 3 per day.

When admitted to hospital she was found to be in good physical health but had typical adenoma sebaceum overlying the butterfly area of the face. Her blood Wassermann was positive but the cerebrospinal fluid showed no abnormality. At that time the expression of her face was described as vacant and her replies to questions were slow, but with patience she could be persuaded to reply relevantly to simple questions and could count up to 10 on her fingers. However, she was unable to name many simple objects. In general conduct she was asocial and at times restless and impulsive, but she was not incontinent and could feed and dress herself.

During her 1st year in hospital she had 12 major epileptic fits and during her 2nd year 41. Subsequently the number of fits decreased and during her 5th year she had only 3. During the 6th year no fits were reported.

At present she has characteristic adenoma sebaceum covering the face, but careful examination fails to discover retinal phakomata or tumours elsewhere in the body.

Her intelligence seems to be that of an imbecile (accurate testing on the Binet-Simon Scale is not possible as she is a Native). She now behaves well and does some mechanical tasks in the ward. There is nothing to suggest that she has deteriorated intellectually or in personality.

The radiologist's report on X-ray of the skull was: 'There is fine conglomerate stippling with a more sclerotic focus and other less marked foci of increased density in the area of the parietal lobes; these are consistent with a process such as tuberous sclerosis.

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REFERENCES

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