TOURAINE'S SYNDROME (ELASTORRHEXIA) IN A BANTU SUFFERING FROM SCHIZOPHRENIA

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Touraine's syndrome or *l'élastorrhexie systématisée* is a rare condition which affects the elastic tissue of many of the organs of the body. It is a degenerative and familial condition and the nature of the pathological changes has given rise to such names as elastosis dysplastica (Parkes Weber) and pseudoxanthoma elasticum (Darier). Because of the widely spread pathological changes a large number of signs and symptoms may occur. These may be conveniently divided into several groups, as follows:

1. Eye changes: angioid streaks in the fundus, retinal haemorrhages, and degenerative changes in the macula, colloid bodies and Bruch's membrane.
2. Skin changes: loss of elasticity and the presence of yellow papules in the skin.
3. Cardiovascular involvement: aneurysms, early arteriosclerosis, or calcification and hypertension.
4. Gastro-intestinal involvement: haematemesis.
5. Neurological symptoms: epilepsy, headaches and vertigo.
6. Psychiatric symptoms: mental instability and depression.
7. Pulmonary involvement: idiopathic pulmonary siderosis with minimal clinical signs but with mottled shadows in the hilar area and diffuse speckling in the lung fields on X-ray.
8. Associated skeletal disease: Paget's disease of bone.

There appears to be no report in the literature of this condition occurring in a South African Bantu nor does there appear to be any record of its occurrence together with schizophrenia. The following case is therefore recorded:

CASE REPORT

E.L. was admitted to Tower Hospital, Fort Beaufort on 10 December 1938. She was single, a Morolong (Bantu) and 30 years of

Fig. 1. Note (1) 'plucked chicken' appearance of the skin of the neck, due to the yellow papules and (2) the large lax folds of the skin of the axillae, abdomen and inguinal regions (mainly the flexor surfaces are involved, but in this case the skin at the back of the neck and elbows is also affected.)

Fig. 2. Note (1) the skin of the thighs, resembling in appearance draped curtaining, (2) the depigmented yellowish rough areas seen best near the inguinal region, and (3) the symmetrical nature of the skin changes.

age. This was her first attack of mental disorder and she had been mentally ill for 4 years before admission.

Family history. She was the fourth of 8 children who survived to adulthood; 3 siblings died in infancy and 1 was stillborn. One sister died of an unknown cause in adulthood, and another died of a 'weak chest'. A third sister is a tuberculosis suspect. The submental skin of yet another sister is 'like that of a plucked chicken and the remainder of her skin is not very smooth'. Her mother suffered from hypertension, had a stroke and subsequently died. After her death the patient's father remarried and had 4 more children. There is no other family history of mental disorder, epilepsy, cardiovascular disease or visual difficulty.

Physical examination and progress. On admission the skin of the patient was noticed to be dry in parts (?pellagra), an apical systolic murmer was heard and a tremor of the hands was observed. Her physical health remained well until June 1953 (14½ years), when she suddenly collapsed and a few hours later vomited roundworms. In 1955 my attention was drawn to a progressive change in her skin. This is well illustrated by the photographs in Figs. 1 and 2.

In May 1956 the following abnormal physical signs were found: Blood pressure 200/140 mm. Hg, an enlarged heart, a loud precordial systolic murmur, and accentuated second pulmonic sound, bilateral arcus senilis, bilateral angioid streaks and haemorrhages in the retinae, and a marked prognathus. X-ray of the skull was normal. In a X-ray the mandible was reported to be 'of more than ample proportions'. X-rays of the long bones and muscles revealed no abnormality; no evidence of Paget's disease was seen. X-ray of the chest revealed a cardiac-thoracic ratio $7\frac{3}{4} : 10\frac{3}{4}$; the report read: 'Cardiomegaly with hilar and perihilar densities and mottlings. Diagnosis — cardiomegaly of hypertensive type with hyperaemia; congestive cardiac failure' (Fig. 3). A blood count showed a reversal of the neutrophyl-lymphocyte ratio, this being 34 : 58% in a total number of 6,000 leucocytes. On lumbar puncture the pressure was normal and analysis of the cerebrospinal fluid showed no abnormality.

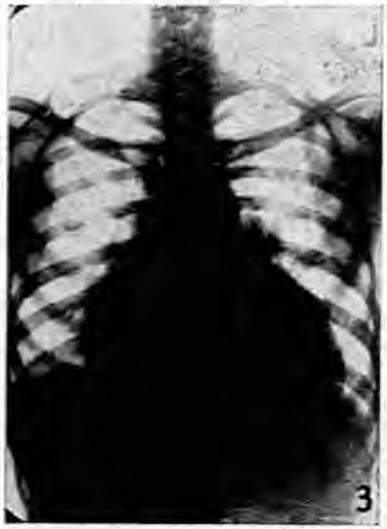

Fig. 3. X-ray of the chest showing cardiomegaly and hilar and perihilar densities.

On 12 December 1956 the patient collapsed, complained of abdominal pain, and vomited bile-stained fluid. She was pale and cold and the blood pressure was 150/100 mm. Hg. A resonant tender mass was found in the left hypochondrium, the palpation of which caused the patient to cough. It extended from the right costal margin to below the left costal margin and to within an inch of the umbilicus. Her pulse waxed and waned and her breathing varied in rate, rhythm and depth. She recovered slowly from this episode and the 'mass' disappeared. It was diagnosed as an acute dilatation of the stomach. On 11 January 1957 she again collapsed and was found to be in congestive cardiac failure. Despite treatment her condition steadily deteriorated and she died on 5 September 1957.

Psychiatric examination and progress. On admission the patient was withdrawn and apathetic. She was visually and aurally hallucinated and expressed delusions of persecution. Her conversation was apt to be rambling and disconnected. She became steadily more withdrawn, apathetic and inaccessible. At times she was cataleptic. Five years after admission she was resistive and abusive when approached. Five years later (1948) she was reported to alternate between phases of catatonic excitement and semi-stupor. At interviews only very brief replies could be obtained from her because she was completely preoccupied with hallucinatory experiences. She began to exhibit echopraxia, and she performed bizarre movements and adopted abnormal stances. Her mental condition steadily deteriorated and in the latter years of her life she muttered to herself all day long, displaying no interest or response either to her illness or her environment.

In January 1957 a skin biopsy was taken from the right axilla but the specimen was unfortunately lost in transit to the laboratory and another could not be obtained. A post-mortem examination was not possible.

DISCUSSION

Etzine and Ovedoff[1] refer to the large variety of signs and symptoms that may occur in Touraine's syndrome. To this may now be added the occurrence of acute dilatation of the stomach.

The problem raised by this patient's mental disorder is an interesting one. Generalized elastorrhexia may affect the cerebral blood vessels, giving rise to psychiatric signs and symptoms included among which must be a schizophreniform picture, since this picture is known to occur in organic disease of the brain, e.g. cerebral tumour, traumatic psychosis and general paralysis of the insane. While it cannot be clearly excluded as the aetiological factor in this patient's mental illness, the long and classic course of the illness in the absence of other signs of organic brain disease make any diagnosis other than that of true schizophrenia or dementia praecox unlikely.

SUMMARY

A case of Touraine's syndrome in a Bantu female suffering from schizophrenia is reported. The following features are illustrated: (1) positive family history, (2) skin changes, (3) cardiac and pulmonary pathology, (4) ocular pathology, (5) gastric symptoms, (6) mental disorder.

My sincere thanks are due to the Rev. C. H. Mariman of Potchefstroom for investigating the family history. I wish to thank Dr. L. R. Brumberg, Tower Hospital, Fort Beaufort for his interest and Dr. B. P. Pienaar, Commissioner for Mental Hygiene for permission to publish.

REFERENCE

1. Etzine, S. and Ovedoff, D. (1956): Med. Proc., 2, 28.