Shaving and dermabrasion of the facial lesions in tuberous sclerosis

A case report

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Summary

Tuberous sclerosis is a disease characterised by convulsive seizures, mental deficiency and angiofibromas. These angiofibromas are hamartomas consisting of hyperplastic connective and vascular tissue. A case is reported where multiple angiofibromas of the face resulted in significant disfigurement. The lesions were treated by shaving and dermabrasion; the short-term result was very satisfactory.

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Tuberous sclerosis is a disease of dominant inheritance, manifested by the clinical triad of convulsive seizures, mental deficiency and angiofibromas. These angiofibromas may be present in many different areas of the body, the most common cutaneous site being the face.[1] A case is reported in which angiofibromas were treated by shaving and dermabrasion.

Case report

A 14-year-old boy was referred to the plastic surgery clinic of J. G. Strijdom Hospital for assessment of facial lesions. He was severely retarded and institutionalised.

The patient was the first child of an 18-year-old mother, born by normal vaginal delivery after a normal pregnancy. At 7 months of age, the patient was hospitalised for gastroenteritis. During this hospitalisation, the child suffered multiple seizures, which necessitated anti-epileptic drug therapy. At the age of 3 years, computed tomography (CT) of the brain showed vague intracranial calcification and the diagnosis of tuberous sclerosis was made. The child was mentally retarded.

At about this time the patient developed a rash around the nose. From the age of 5 years the child's rash became progressively worse and at 10 years of age, severe adenoma sebaceum was apparent in a butterfly pattern on his face. At the age of 14 years, renal ultrasonography showed typical changes of angiomyolipomatosis, complicating tuberous sclerosis.

On examination after presentation, the patient's face was covered with a great number of papular lesions, which were particularly prominent over the nose, nasolabial areas and cheeks (Fig. 1). The rest of the face was affected to a lesser degree in a butterfly distribution. Papular lesions were also evident on the patient's gums.

Shaving and dermabrasion of a small test area was initially undertaken. Histological examination of a biopsy specimen at this stage demonstrated lesions intermediate between the symmetrical type of smaller angiofibromas and larger symmetrical fibromas.

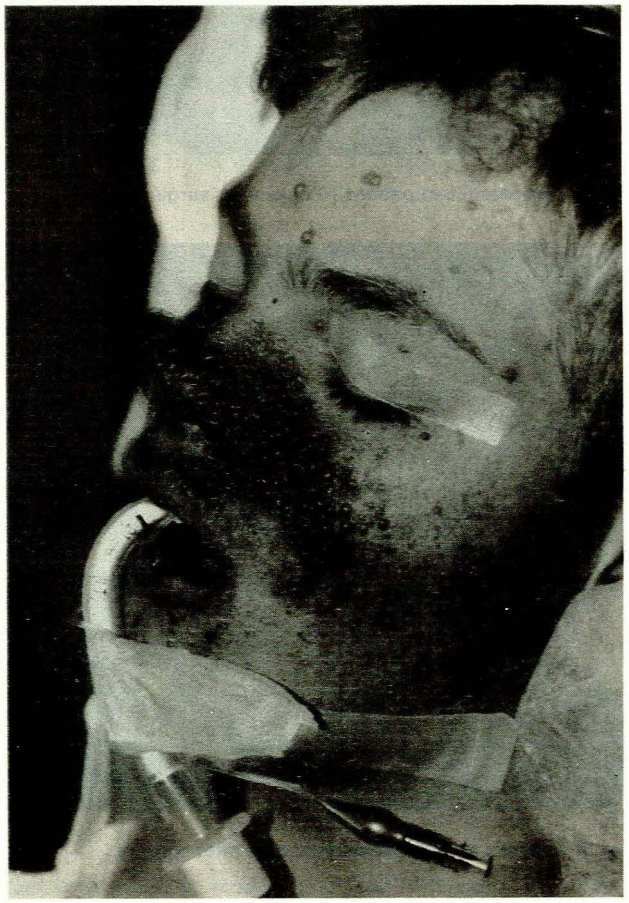

Fig. 1. Pre-operative appearance of patient.

When healing of the test area had occurred, the first formal procedure was performed (3 weeks later). Under general anaesthesia, the large accumulation of papules was shaved flush with the skin. The underlying area was then treated with a Stryker dermabrader with a coarse carborundum cylinder head. Bleeding was moderate. The abraded areas healed uneventfully, despite the dermabrasion having been quite deep into the dermis in some areas (Fig. 2). Follow-up examination after 9 months revealed little regrowth of the fibromas in the affected areas (Fig. 3).

Discussion

Tuberous sclerosis has been estimated to have an incidence of 1 in 20 000.[2] The lesions associated with the disease are fine wart-like lesions predominantly in a butterfly distribution over

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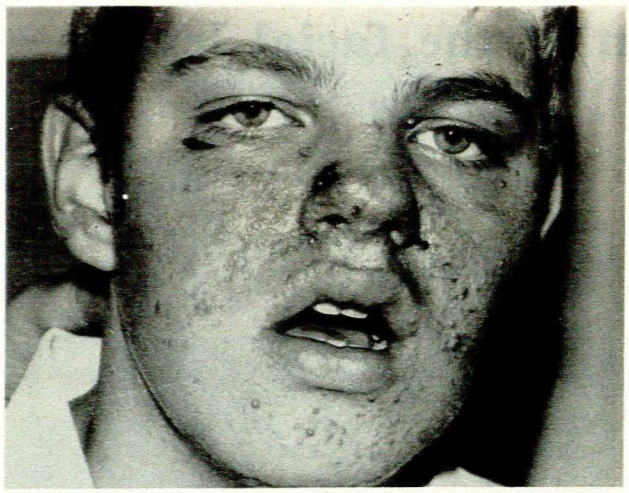

Fig. 2. Appearance of patient 10 days after surgery.

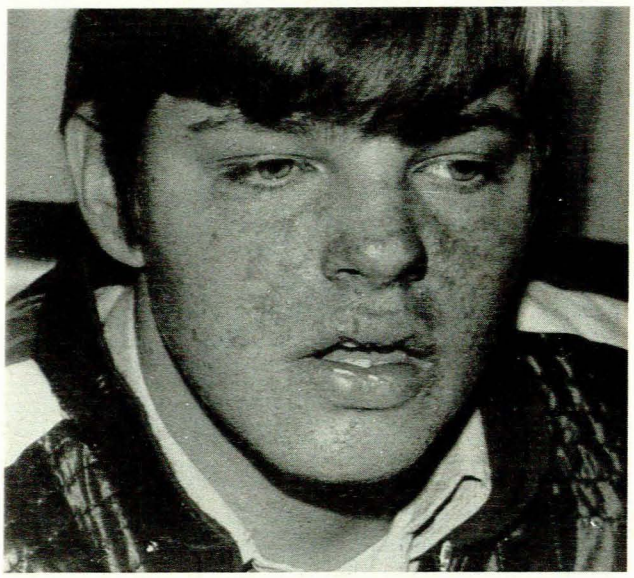

Fig. 3. Appearance of patients 9 months after surgery.

the cheeks and forehead. They are most often found on the face, brain, retina, heart and kidney, but may be found throughout the body. Intelligence varies depending on the extent of the disease. Approximately 50% of cases of tuberous sclerosis occur sporadically;[1] the skin lesions are actually hamartomas consisting of hyperplastic connective and vascular tissue.[3] They were described as adenoma sebaceum by Pringle[4] in 1890; they are, however, neither adenomatous nor sebaceous.

The diagnosis of tuberous sclerosis is aided by skull radiography and brain CT. Calcified nodules occur particularly in the temporal lobes and adjacent to the ventricles. In its fully developed form, particularly in those patients in whom the disease occurs in infancy, prognosis for life beyond the third decade is poor. Death may occur from seizures, associated tumours or other related diseases.[2] However, overall prognosis is dependent on the extent of organ involvement, which may vary from minimal cutaneous effects to diffuse multiple organ disease.

Few reports have been published in respect of the treatment of the skin lesions. Excision of a large facial tumour with excessive blood loss was reported by Alvarez,[5] excision and cautery has been reported,[4] but scant information is available on shaving and dermabrasion of these severe facial lesions.[6,7] Recently articles have appeared reporting satisfactory results using the carbon dioxide laser — this may prove to be a reasonable treatment alternative.[8,9]

The above case demonstrated a dramatic improvement in facial aesthetics from an unacceptable, almost bizarre, appearance to a most satisfactory result. New lesions may recur and further surgery may be necessary, although at this early stage very little recurrence has been noted. Institutionalisation does not protect these unfortunate individuals from occasional cruel social interactions, thus if the potential exists for making these patients more socially acceptable, it should be pursued.

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