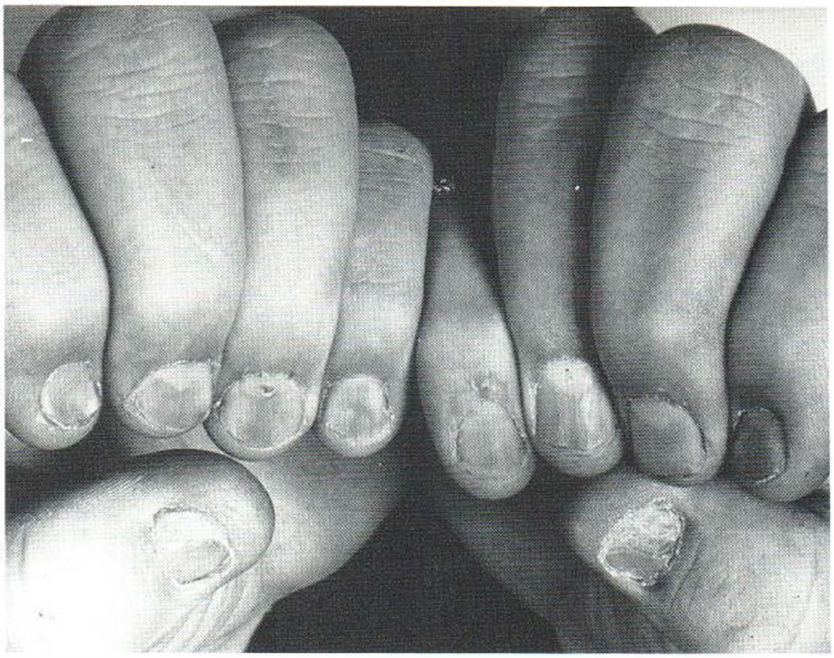

Fig. 2. The patient's nails.

DISCUSSION

Trichoepithelioma multiplex is often familial. Our patient, however, had no family history of trichoepithelioma on his mother's side. His father was unknown. The nail dystrophy seems most likely to have been inherited from his mother. Her mother had had the same kind of dystrophic nails.

The simultaneous appearance of trichoepitheliomas and nail dystrophy may be an example of coincidental genetic linkage or it may be a phenotypic expression of the same genotype of genetic defect.

The treatment with dermabrasion and electrocautery was deemed satisfactory.

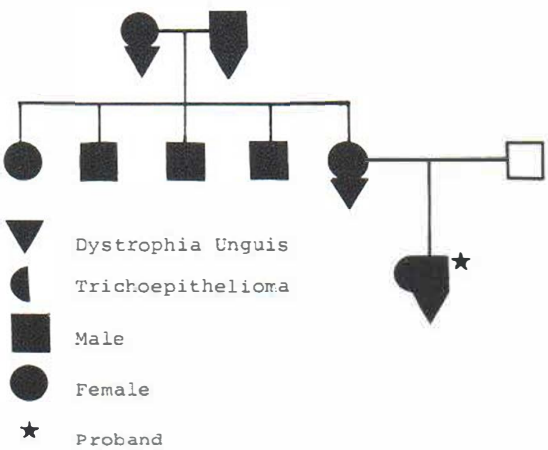

Fig. 3. Pedigree of the patient, showing the hereditary pattern of trichoepitheliomas and nail dystrophy.

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Megalopinna in Naevus Uniuslateris: A Case Report

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Abstract. A female patient, aged 10 years, with naevus uniuslateris limited to the left half of head and neck includ-

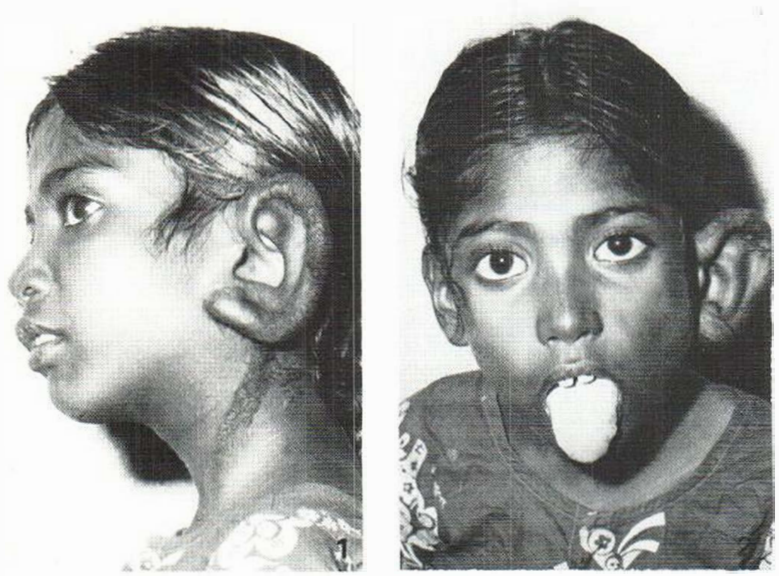

Fig. 1 and *2*.

ing the mucosa of tongue is reported, since she had megalopinna of the same side, which, so far as is known, has not been reported previously in the literature.

CASE REPORT

A 10-year-old girl presented with asymptomatic lesions on the head and neck which had been present since birth. There was no history of delayed developmental milestones or seizure disorders. The family history was negative as regards similar lesions. Examinations revealed linear, brownish-black verrucous lesions over the back and sides of neck extending into the left pinna and posterior scalp (Figs 1 and 2). The lesions were limited to the left side. The left half of the tongue showed hypertrophy, with verrucous papules on its dorsal surface (Fig. 2). The pinna of the left ear was three times the size of the right (Figs. 1 and 2). There were no signs suggestive of an angiomatous component in the naevus. Other physical, otolaryngological and psychiatric examinations were normal.

INVESTIGATIONS

Routine haematological and urinary examinations were normal. Skull X-ray revelaed no bone abnormality. Biopsy of papules on the pinna and tongue showed acanthosis papillomatosis suggesting a verrucous naevus. No angiomatous or neural element was detected.

DISCUSSION

Naevus uniuslateris is a congenital developmental defect of the epidermis usually present from birth or from early childhood as verrucous warty pigmented lesions. The extent of the lesion may vary, from a single localized lesion, to multiple disseminated ones affecting one-half of the body. Affection of mucosa of tongue, lips, and vagina may also occur. Histopathology usually reveals hyperkeratosis, acanthosis and papillomatosis. The inflammatory type and naevus showing vacuolar degeneration of stratum malpighii and granular layer are other histological types of verrucous naevus (1). The naevus may also show angiomatous and/or neural elements. Maceration in flexures and malignant degeneration are a known complication (3). Hypoplasia of the affected parts, skeletal abnormalities, central nervous system abnormality such as epilepsy, hearing loss, and Horner's syndrome have been reported as features associated with naevus uniuslateris (2).

This case is interesting since it showed typical naevus uniuslateris limited to the left half of head and neck including the mucosa of the tongue and was associated with megalopinna of the same side which, as far as I know, has not been reported earlier.

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Clinical and Histopathologic Morphea with Immunological Evidence of Lupus Erythematosus: A Case Report

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Abstract. A 14-year-old girl developed skin changes after BCG vaccination. The clinical and histopathological findings revealed morphea. The immunfluorescence microscopy showed IgG, IgM, IgA, C3 and fibrinogen deposits at the dermal-epidermal junction area in both involved and uninvolved skin. The antinuclear antibody tests were all negative.

CASE REPORT

The patient was a 14-year-old girl who had allergic rhinitis. She had taken her BCG vaccination on her left shoulder in February 1979 and some weeks later she discovered som hypopigmented areas on the left shoulder and the left arm. She entered the hospital in March 1980. On examination we noticed several brown coloured patches and in the centre of these patches the skin was indurated and the colour was yellow to white. The patches were situated on the left side of thorax, her left shoulder and the left arm and hand, and they varied in size from about 1 cm to about 4 cm. The girl did not have other symptoms except her exanthema.

Immunologic investigations

ANA test, Waaler-Latex test, LE cell preparation, anti-RNP, anti-Sm and anti-DNA (double- and single-stranded), were all negative. C3: 0,60 g/l, C4: 0,20 g/l, CH50: 80 u/ml, IgG: 11 g/l, IgA: 1,6 g/l, IgM: 1,5 g/l, IgD: 0,03 g/l, IgE: 300 u/l, i.e. all these values were normal.

Other laboratory results were within normal ranges. Histological examination of two different biopsies of involved skin showed in both some hyperkeratosis in epidermis. The dermis showed an increase of collagen and seemed sclerotic. A perivascular mononuclear cell infiltrate with lymphocytes was also observed. Direct immunofluorescence microscopy of biopsies showed in two species of involved skin a granular deposit in a linear fashion of IgG, IgM, IgA, C3 and fibrinogen at the dermal–epidermal junction. Uninvolved skin showed the same deposits of immunoglobulins at the junction area as did involved skin. These immunofluorescence findings were consistent with systemic lupus erythematosus but the patient's blood tests were not consistent with this disease.

DISCUSSION

This paper describes a young girl having clinical and histopathologic findings of morphea, where immunofluorescence microscopy findings (but not the serologic tests), showed systemic lupus erythematosus.

Winkelmann and associates (6) found by biopsy of involved scleroderma skin that IgM-band immunofluorescence was present in most patients with inflammatory scleroderma ("mixed connective tissue disease"). He also pointed out that patients with progressive systemic sclerosis and two patients with generalized morphea all showed negative immunofluorescence. Christianson et al. (2) reported 191 patients with either plaque type of morphea or linear scleroderma. None of these patients developed multisystem disease. However, Chorzelski & Jablonska (1) mention 3 cases of scleroderma with coexistence of lupus erythematosus – the discoid form in one and the systemic form in 2 patients. Dubois et al. (3) observed 3 patients with morphea who had positive LE cell preparations. All these 3 patients developed systemic lupus erythematosus several years later. Similarly, Tufanelli and co-workers (5) had a patient with linear scleroderma who developed systemic lupus erythematosus 12 years later. Mitchell et al. (4) described a child with circumscribed scleroderma with immunologic evidence of systemic lupus erythematosus. They stressed the importance of continued serologic investigations of patients with local scleroderma.

Possibly, the BCG vaccination may have been the trigger for the skin changes of this patient.

In summarizing, our patient with clinical morphea demonstrated immunological signs which can be interpreted as arousing a suspicion of systemic lupus erythematosus. We must therefore follow her carefully in the future to see if she develops symptoms of systemic disease. Similar cases were