Acta Dermatovener (Stockholm) 51: 232–240, 1971

SOCIETY PROCEEDINGS

SWEDISH DERMATOLOGICAL SOCIETY

Meeting at Karolinska Sjukhuset, Stockholm, March 18, 1970

Monilethrix

Presented by Göran Wennersten

Spanish female, 3 years. Born with a good growth of hair. According to Spanish custom, her parents shaved off her hair at the age of 4 months. The new growth of hair was brittle and never grew more than a few centimeters long (Fig. 1 *a*). No similar phenomenon in the family. One of her mother's cousins lost her hair completely at the age of 4 years and was said to have been completely bald across the crown of the head at the age of 6, after which time her hair grew back completely.

Findings: Sparse, millimeter-long, brittle dark hair over the capillitium; a few hairs a centimeter or two long near the brow. Intact eyebrows and eyelashes. Follicular hyperkeratosis on the outside of the arms and hair follicles. Microscopic examination of the hairs revealed typically fusiform, bead-like constrictions with a 0.70 mm periodicity (Fig. 1 *b*).

Comment: Two previous cases of monilethrix have been published which periodically had urine argininosuccinic acid (Grosfeld, J. C. M., Mighorst, J. & Moolhuysen, T.: Argininosuccinic aciduria in monilethrix. Lancet 1964, II: 789–91.)

However, amino acid analysis of urine collected from this patient for 8 days disclosed no argininosuccinic acid.

Reiter's Syndrome?

Presented by Gunnar Swanbeck

Concrete worker, 46 years. Ever since 1965, episodes of diarrhoea and severe loss of weight with periods of recovery in between. Since the autumn of 1969 the patient has had diarrhoea about 5 times a day. Skin changes on his lower legs, thighs, genitalia and a few on the body itself started in January 1970. The skin changes began as small, brown, somewhat scaly patches. They then grew in size and had scaly margins with even, central hyperkeratosis. On the penis there was a balanitis of the circinate type. The patient was admitted to the local medical clinic for a study of his malabsorption and was treated with potassium chloride, iron and calcium.

Initial examinations at the medical clinic: faeces culture negative; ESR 82 mm; gamma globulin 3.87 g/100 ml. Other haematological and hepatic values normal. No auto-antibodies found. X-rays: stomach, intestine, kidneys, skull, pelvis—normal.

The consultant dermatologist found that the patient had skin lesions resembling keratodermia blennorrhagica, suggesting Reiter's syndrome. Because of this proposed diagnosis, a urologist was consulted but nothing pathological was found. The patient's history was subsequently penetrated, and it was found that he had previously had ureteral pain, conjunctivitis and diffuse arthralgia. The continued study of his malabsorption showed that the patient had dysfunction throughout the course of the small intestine. Thus, he probably suffered from two independent diseases: Reiter's syndrome and dysfunction of the small intestine.

Multiple Keratoacanthoma (Self-healing Squamous Epithelioma According to Ferguson-Smith)

Presented by Georg Rajka

Female, born 1915. Since 1968, a large number of pruritic tumours, ranging from rice-size to

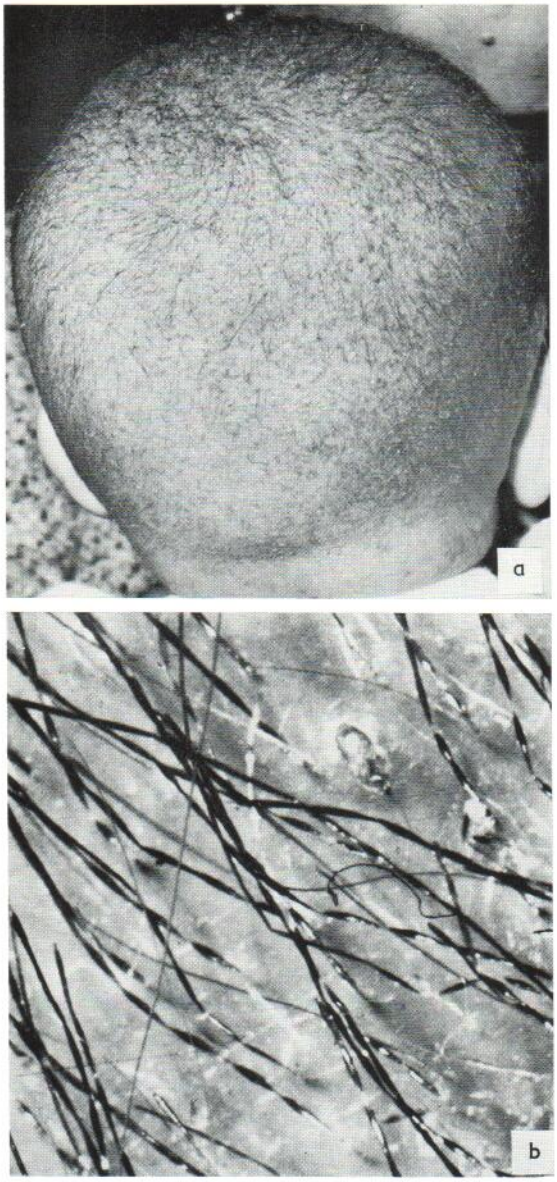

Fig. 1. Monilethrix. (*a*) Survey. (*b*) Close view of hairs.

pea-size, developed on her neck (Fig. 3), occipitally on her face, in hair follicles, on the upper arms and backs of her hands. Several of these tumours displayed a tendency to ulcerate.

Histology: Three earlier and two current biopsies disclosed pictures of rather poorly differentiated squamous epithelioma (Fig. 4).

Course: Some spontaneous regression with scar formation occurred (Fig. 5) but simultaneously some progress, especially occipitally and on the neck.

Treatment: Bucky rays, 300 R × 4 to the neck and face led to some improvement. No cytostatic medication or conventional irradiation were administered.

Diagnosis: Ferguson-Smith's description (Brit. J. Dermatology 46: 267, 1934) of multiple keratoacanthoma is probably the one best suited to this apparently very rare disease without familial occurrence, with a histologically clear picture of squamous epithelioma but with a "benign" course.

Pyoderma Gangrenosum with Systemic L.E.

Presented by Kerstin Olson

Female, born 1949.

1964: Onset of necrotic ulcers on the legs.

1965: Acute illness with pleuropneumonia, myocarditis, hepatitis. Severe breathing difficulties upon admission to hospital. The diagnosis systemic lupus erythematosus was established. Rapid improvement with prednisolone treatment.

1965–67: Repeated attacks of S.L.E. treated with chloroquine and prednisolone.

1967–68: Progress of skin changes with extensive pyoderma gangrenosum ulcers on the buttocks, elbows and knees. Increased ESR as before and elevated gamma globulin in electrophoresis but hepatic values now normal. Treated with prednisolone, chloroquine, salicylazosulfapyridine (Salazopyrin®). Pat. discharged from hospital in good condition.

1969: Recurrence of skin changes in summer without symptoms of S.L.E. Subfebrility, malaise, arthralgia several months later. ESR 80 mm. Hepatic values normal. Anti-nuclear factors (ANF) positive 1/100, immunofluorescence examination (IFL) of glomeruli positive 1/25, and of smooth muscle positive 1/25 suggesting lupoid hepatitis. Therapy with azathioprine 50 mg × 2 was started in Sept. 1969. Prednisolone medication was continued in doses declining from 20 to 5 mg daily. Rapid improvement followed this treatment and the patient was able