

SOCIETY PROCEEDINGS

SWEDISH DERMATOLOGICAL SOCIETY

Meeting at Södersjukhuset, Stockholm, October 29, 1971

Felty's Syndrome

Presented by Eric Sandström

Case history. A woman, 80 years old, with a 40-year history of rheumatoid arthritis primarily affecting joints in the hands and feet, leaving lasting deformities. Over the last 10 years, free of symptoms.

Eighteen years ago the patient had her first pretibial ulcer which healed on routine local treatment. When hospitalized 3 years ago, on account of a recurrence of the pretibial ulcer after a circulatory occlusion the year before, splenomegaly to the umbilical plane, pronounced leucopenia down to $1\,100\text{ WBC/mm}^3$, moderate anemia and ESR around 40 mm were found. Further investigation revealed antileucocyte antibodies; ECG suggested transient pericarditis. At discharge only small ulcers remained.

On admission 2 years later the pretibial ulcer had grown to palm size; the size of the spleen, leucopenia and anemia being unchanged.

When hospitalized 1 year later the ulcer had recurred in the same place; spleen size and laboratory findings were unaltered; ESR around 20 mm; antinuclear antibodies, Waaler-Rose and acryl fixation test giving negative results; a normal ECG. Low IgA was noted at repeated electro-

phoresis. No pathogenic organism could be cultured from the ulcer.

On the ventral side of the tibia a shallow ulcer $18 \times 2\text{ cm}$ was found surrounded by a brownish blue halo of about 5 cm width. At the margins the skin was thin and brittle (photos from 1970) (Fig. 1).

Good healing and marked decrease of the discoloured area was observed with surgical tape over the ulcer three times a week.

Pseudoxanthoma Elasticum

Presented by Taavi Kaaman

A female, 36 years old. Since the late teens, "rough skin" on the neck. A few years ago she noticed progress of the skin lesions on the neck and new lesions appeared in the axillae and the groins. One of her sisters has similar skin lesions.

Findings. On the sides of the neck, the axillae and the groins, yellowish papules coalescing into plaques. Mainly on the sides of the neck, lax and wrinkled skin (Fig. 2 a, b). Arterial involvement could not be detected by plethysmography or oscillometry but clinically there was absence of radial artery pulses. Ophthalmoscopic examination showed typical angoid streaks of the retina. X-ray

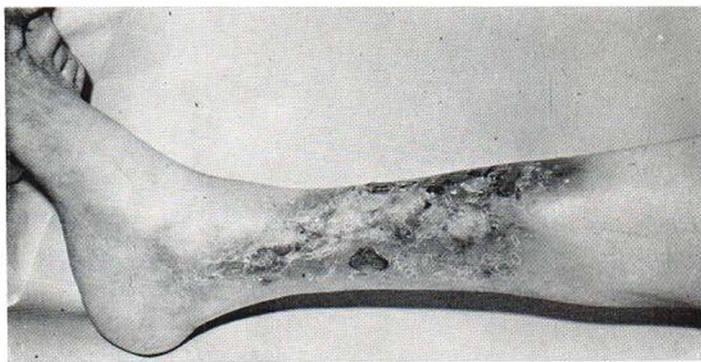


Fig. 1. Leg ulcer in patient with Felty's syndrome.

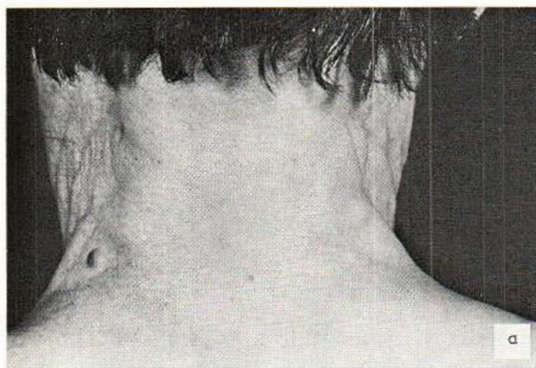


Fig. 2 a, b. Neck of patient with pseudoxanthoma elasticum.

of the skull revealed ossification of the pineal body.

Skin biopsy. The epidermis is somewhat thin.

The mid-dermis and deep parts of dermis showed basophil degeneration of the elastic tissue with diffuse calcification.

Meeting at University Hospital, Uppsala, February 11, 1972

Kaposi's Sarcoma after Immunosuppressant Therapy

Presented by Lennart Juhlin

A 51-year-old housewife with pyelonephritis since 1960 and a progressive decline in renal function until February, 1970, when a new kidney was transplanted into the iliac fossa. The renal homograft functioned well without any sign of rejection and the patient's condition improved remarkably. She is treated daily with 100 mg of azathioprine, 8 mg methylprednisolone and 50 mg dicoumarol. For the last 5 months she has also been receiving 150 mg of hydralazine and 240 mg of propranolol daily for her elevated blood pressure.

In March, 1971, she suddenly discovered a bean-sized, reddish-blue tumour on the breast. This tumour was removed, but since September, 1971, similar tumours have appeared on the chin, buttocks, in the scar from the kidney operation and on the upper part of the left leg (Fig. 3). Biopsies show a typical picture of Kaposi's sarcoma. Some lesions are treated by X-ray with good response. In January, 1972, an X-ray control of the lungs revealed fairly well demarcated, round infiltrate of metastatic type in both lungs. Haematological examinations are normal. Creatinin, 1.2 mg %. Serum electrophoresis shows an



Fig. 3. Kaposi's sarcoma with multiple lesions on the leg.

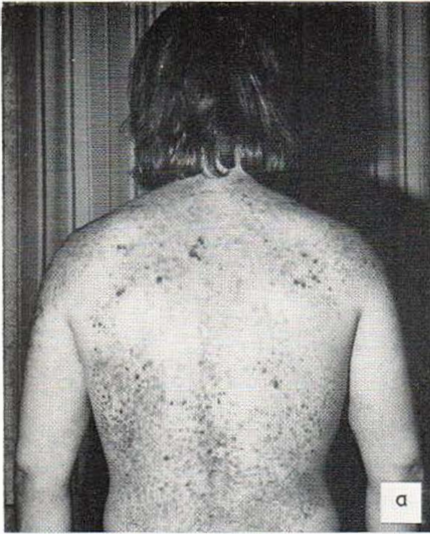


Fig. 4 a. Epidermolysis bullosa albuginosa.

Fig. 4 b. Detail from Fig. 4 a with blister.

Fig. 4 c. Thickened, triangular toenails in epidermolysis bullosa albuginosa Pasini.

increase of α_1 and α_2 globulins and lowered immunoglobulins.

Discussion

L. Juhlin: It is known that immunosuppressive therapy can induce malignancies. It has been described in patients treated after renal transplantation. A case of disseminated Kaposi's sarcoma has been reported by Siegel et al. (1969) during the late rejection of a homograft kidney. Our patient has disseminated lesions in the skin and

possibly also in the lungs. There were no signs of tuberculosis.

N. Thyresson: There have been some cases described with hypernephroma in transplanted kidneys which can develop metastasis. Is there any sign of tumour in the transplanted kidney?

L. Juhlin: No, the kidney functioned well and the scintigram is normal. I think it would seem more probable that her Kaposi's sarcoma is induced by the immunosuppressive therapy than by a tumour from the transplanted kidney.

Addendum: Some skin lesions were treated with X-ray with good response. The patient was later given bleomycin (165 mg). The remaining skin lesions disappeared rapidly and there was a marked regression of the infiltrates in the lungs. She lost her hair on the scalp but is now enjoying the summer in good health and wearing a wig.

References

- Siegel et al.: Disseminated visceral Kaposi's sarcoma. *JAMA* 207: 1493, 1969.
- Shelley, W. B.: Consultation in Dermatology, p. 256. Saunders, Philadelphia, 1972.

Epidermolysis Bullosa Albupapuloides Pasini

Presented by Lennart Juhlin

A 23-year-old man with itching skin lesions since 11 years of age. He was then seen at the Dermatology Clinic in Helsinki where blisters were noticed with a diameter up to 4 cm over various parts of the body, and dark, thick toenails. A biopsy showed changes consistent with epidermolysis bullosa. Later the picture was dominated by milia and lichenoid papules on the trunk and upper arms. A diagnosis of epidermolysis bullosa albupapuloides Pasini was made. Since then his condition has remained unchanged and resistant to various forms of treatment such as locally and orally given corticosteroids, antibiotics, diaphenylsulphone (Dapsone) and penicillamine. Treatment of one-half of the body with UV-light did not affect the lesions. He still complains of severe itching especially when new papules and blisters appear. Alcohol and aspirin aggravate his symptoms. There are no certain seasonal changes. He was fostered in a nursing home and it is only known that his mother, a sister and brother are healthy.

Examination. On repeated examinations the patient has blisters, lichenoid papules and dark-red macules with slight crusts on the trunk (Fig. 4 a, b). There are also atrophic, ivory-white macules and several milia. The toenails have a triangular shape and are thick and yellow in their distal part (Fig. 4 c). Erosions are often seen on the buccal mucosa.

Laboratory studies. Routine analysis of blood and urine were normal. Immunoglobulin E slightly increased (980 ng/ml). Increased excretion of

uroporphyrin in urine (585 $\mu\text{g}/24$ hours) and a slight increase of faecal coproporphyrins (82 $\mu\text{g}/\text{g}$ dry weight). ALA 1.5 mg, porphobilinogen 0.6 mg, and coprophorphyrin 38 μg in urine collected for 24 hours. Protoporphyrin in faeces 34 $\mu\text{g}/\text{g}$ dry weight and in red blood cells 29 $\mu\text{g}\%$.

Biopsy. Subepidermal blister with follicular hyperkeratoses and the presence of granuloma. Milia in some sections.

Discussion

T. Fischer: Is he sun-sensitive?

L. Juhlin: No, he considers himself to be better during the summer despite the fact that he has an increased excretion of porphyrins. It has been shown by Gottron and others that these patients can excrete an increased amount of porphyrin.

K. Wikström: In what group is the porphyrin classed?

L. Juhlin: The determinations were made by Dr Haeger in Malmö and the pattern is not unlike that seen in porphyria cutanea tarda.

Reference

Götz, H. & Meinicke, K.: Zur Klinik und Therapie der Epidermolysis bullosa et albo-papuloidea Pasini. *Derm Wschr* 131: 481, 1955.

Editor's address:

Ö. Hägermark, M.D.
Department of Dermatology
Karolinska sjukhuset
S-104 01 Stockholm 60
Sweden