

## KAPOSI'S SARCOMA IN POLAR ESKIMOS

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**Abstract.** Two cases of Kaposi's sarcoma (KS) are described in patients of pure Eskimo trait from the northernmost part of the world. The clinical and histological findings corresponded to KS in other parts of the world. This report on KS in Eskimos from the sparsely populated Arctic areas may support the opinion that KS is more common outside Africa than the literature indicates.

**Key words:** Kaposi's sarcoma; Eskimos

More than one hundred years have passed since Kaposi described the multiple idiopathic haemorrhagic sarcoma of the skin (Kaposi's sarcoma, KS) (7), but still its etiology remains unknown and the histogenesis continues to be a subject for discussion (3, 8, 9, 14, 17). Histogenetically, the basic cell type seems to be an immature, pluripotent vascular cell, developing into either a proliferating endothelial cell or an immature phagocytic fibroblast (5, 13).

From an epidemiological point of view the incidence of KS shows a remarkable variation according to geography and ethnic status of the patients (8, 9, 14, 16), thus giving support to theories of a geographic and/or genetic predisposition.

Although KS has been described in one patient of Eskimo descent previously (11), we consider it appropriate to report the following cases from Arctic Greenland, since KS may be more frequent in northern and even Arctic latitudes than was formerly believed.

### CASE REPORTS

#### *Case I*

Male. Born in Thule, which is a small Eskimo settlement of approximately 400 persons in the northernmost district or Arctic Greenland. He had lived in the district all his life as a hunter and fisherman. At the age of 58 he consulted

the district medical officer because of oedema of the legs and feet. No obvious cause was found, and the patient was treated with diuretics.

1½ years later he was seen again by the medical officer. At that time he had developed numerous dark-bluish skin tumours of varying size on the feet and legs (Fig. 1) and two similar tumours in the right palm and on the right earlobe. There were no signs of internal diseases.

Biopsy from the skin showed a tumourous tissue extending throughout all layers of the dermis. The main part of the tumour was composed of numerous vascular lumina, varying greatly in size and shape, each lined by a single layer of large, proliferating endothelial cells (Fig. 2). The stroma between the vessels contained fibroblasts, extravasated erythrocytes and deposits of hemosiderin. Other parts of the tumour showed interlacing bundles of spindle-shaped proliferating fibroblasts, varying in size and staining qualities and some mitotic figures were present. Deposits of hemosiderin were found within primitive fibroblasts as the result of phagocytosis, and a reticulum stain revealed a rather dense network of reticulum fibres.

The diagnosis KS was established, and the patient was admitted to the Radium Center in Copenhagen for radiotherapy. The immediate result of the treatment was successful. The tumours diminished in size, and the patient returned to Thule.

After a further year there was a relapse, and it became necessary to amputate the left fifth toe because of an ulcerating tumour, but no histological examination was performed. For the next 9 months the patient was not seen by the health service personnel, but he was then admitted to the local hospital with painful ulcerating tumours on both soles and severe oedema of the lower extremities. Chemotherapy was tried with chlornaphazinum (Erysan<sup>®</sup>, Dumex) and later with cyclophosphamidum (Endoxan<sup>®</sup>, Asta), but without any obvious success. As the treatment depressed the leukocytes the treatment was discontinued.

The patient died shortly after. The cause of death was stated as uraemia with universal oedema. No necropsy was performed.

#### *Case II*

Male. Born in a small Eskimo settlement in the southern part of Greenland, where he had lived all his life. At the age of 61 he was admitted to the local hospital, complain-

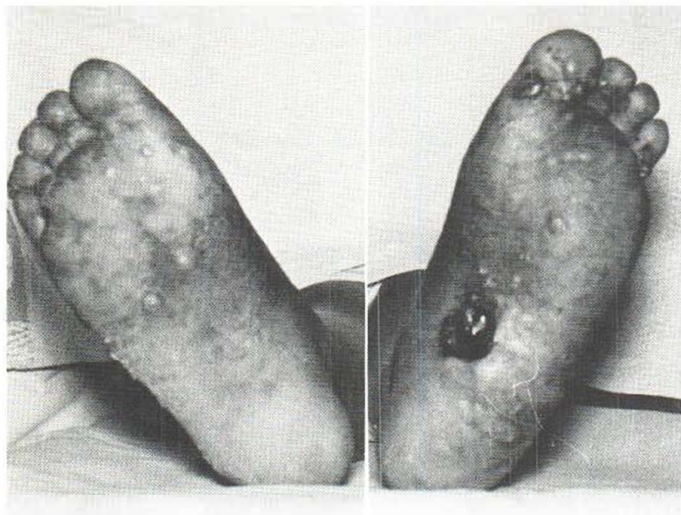


Fig. 1. Case 1. Tumours on the soles.

ing of severe oedema of both legs and hands. Scattered on the extremities were numerous brown-bluish, slightly elevated and compressible tumours. One of the tumours was ulcerated. The patient stated that he had been suffering from painful swellings on the legs and feet for 20 years and that the tumours had developed during the last year. A biopsy was taken, but was accidentally destroyed during the mailing to the laboratory in Denmark.

● On two subsequent occasions, 2 and 4 years later, he was readmitted to the local hospital suffering from cardiac incompetence and was treated with diuretics and digitalis. The skin tumours persisted without treatment.

For the following 10 years he had no contact with the local health service personnel, but at the age of 75 he was readmitted to the hospital suffering from cardiac incompetence and painful ulcerating tumours and oedema of the extremities. No signs of internal malignancy were found. Biopsy from a tumour on the leg showed small groups of capillaries in the dermis, surrounded by proliferating fibroblasts and fibrocytes. Deposits of hemosiderin were found in between these structures as well as extravasated erythrocytes, some of which were located in narrow splits in the otherwise dense fibroblastic tissue. The endothelial cells of the capillaries showed large and dark nuclei. A slight variation in the size and shape of the fibrocytic and fibroblastic nuclei was also present. No mitotic figures were found, however. A few lymphocytes, histiocytes and granulocytes infiltrated the fibrous tissue, but nowhere did the changes resemble granulating tissue. The histological picture was compatible with the diagnosis Kaposi's sarcoma.

The patient did not want to go to Denmark for treatment, and he returned to his settlement, where he died 2 years later. No necropsy was performed.

## DISCUSSION

KS is a very common affection of the skin in some parts of Africa, especially in the Equatorial parts (8,

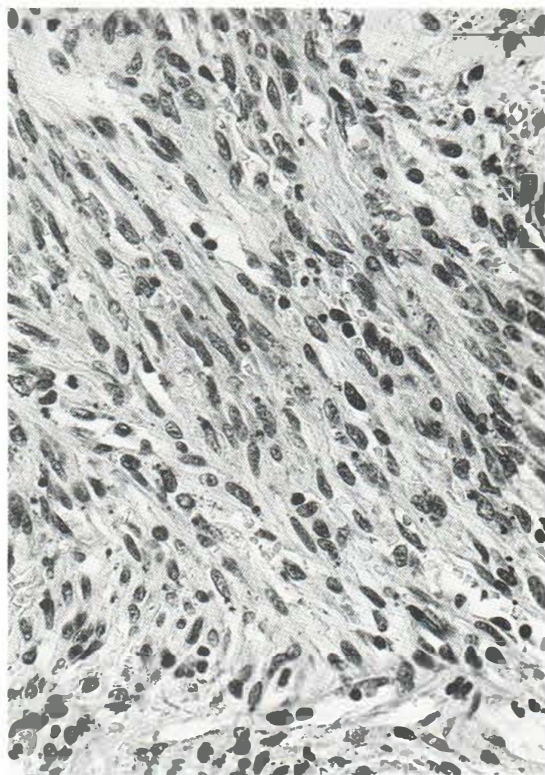


Fig. 2. Case 1. Biopsy from a tumour. Bundles of spindle-shaped fibroblasts are interspersed with slit-like vascular spaces. The nuclei of the fibroblasts show moderate variation in size and staining qualities. H and E,  $\times 305$ .

10, 15). In the USA the disease is more frequent in people of Jewish or Italian descent than in other groups: the incidence in people of negroid race seems to be relatively low (2). In Europe the disease is more frequent in the eastern and southern parts (8, 16); in the northern part, including the Scandinavian countries, KS is a rare disease (1, 6).

During studies on malignant diseases in the Eskimo population of Greenland (4) we found these two cases of KS among 37 histologically proven cases of malignant skin tumours. In 1974 Masse & Glazebrook published a case of KS in a Canadian Eskimo (11). The present report is the second on KS from the sparsely populated Arctic areas.

Our cases throw no light on the complex problems of either the etiology or the histogenesis of KS, but the findings of three cases in the small Eskimo population of the world might support the opinion of McCarthy & Pack (12), and of Brownstein et al. (2) that KS is more common outside Africa than the literature indicates.

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