Extensive Hemangiomatosis of the Extremities with the Same Histopathological Pattern as the Early Lesion of Kaposi's Sarcoma

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For 13 years a 28-year-old man had on his hands and feet bluish red patches, which had slowly progressed in size. Biopsy specimens exhibited an abnormal vascular pattern similar to that seen in early lesions of Kaposi's sarcoma. Skeletal radiographs of the hands and feet showed lesions indicating hemangiomatosis. *Key words: Hemangioma; Lymphangioma.* (Received March 26, 1986.)

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In 1982 Gottlieb and Ackerman reported on the occurrence of Kaposi's sarcoma in young homosexual men and emphasized the characteristic, diagnostic histological picture of an early lesion of this disease (1). We present here a case of a young man with an unusual kind of hemangiomatosis with a histophatological picture indistinguishable from that considered characteristic of early Kaposi's sarcoma.

CASE REPORT

A male patient, 28 years of age, presented at the Department of Dermatology with skin lesions on the hands and feet. The lesions had first appeared in both areas when the patient was about 15 years old and had slowly progressed. In the last two years he had suffered form edema and pain in the feet when standing for a long time. He was otherwise healthy. He denied homosexuality. To his knowledge there were no hereditary diseases in the family. The patient was of Turkish origin and had immigrated to Sweden a few months earlier.

Clinical findings

There were large bluish red patches with irregular borders on the hands, feet and distal part of the lower legs (Fig. 1 *a*). The dorsal aspects of the feet and toes and the soles were slightly edematous. No plaques or nodules were seen. The configuration of the extremities was normal. The clinical diagnosis was uncertain, but acrodermatitis chronica atrophicans was tentatively suggested.

Histopathological findings

Biopsies were first taken form the dorsal aspect of a foot and from the plantar aspect of a toe. Later on, four more biopsies were taken—two from the soles, one from the dorsal aspect of a hand and one form the distal part of the left lower leg. In the dermis there were numerous thin-walled, irregular vascular spaces. In som areas the vessels formed communicating systems in which protruding buds of connective tissue containing small vessels were seen (Fig. 2 *a* and *b*). The vessels were lined with a thin endothelial cell layer without atypia. Most of them were empty, but some contained red blood corpuscles or a few white blood cells. In one area in the specimen from the lower leg the vascular channels dissected through the dermal collagen, giving rise to a host of small papillary formations (Fig. 2 *c*). In one of the biopsies from the sole an abnormal, thick-walled vessel containing red blood cells was observed in the lower dermis in addition to some thin-walled wide vessels (Fig. 2 *d*). In the biopsy from the lower leg it was obvious that the process also involved the subcutaneous fat. Throughout the dermis there were patchy infiltrates of plasma cells and lymphocytes (Fig. 2 *a*).

Immunological findings

The proportions of total T cells and T cells expressing either helper or suppressor phenotype were within normal ranges. The lymphocyte reactivity to mitogens (PHA, ConA and PWM) was normal. Antibodies against LAV/HTLV-III virus or Borrelia spirochetes were not found.

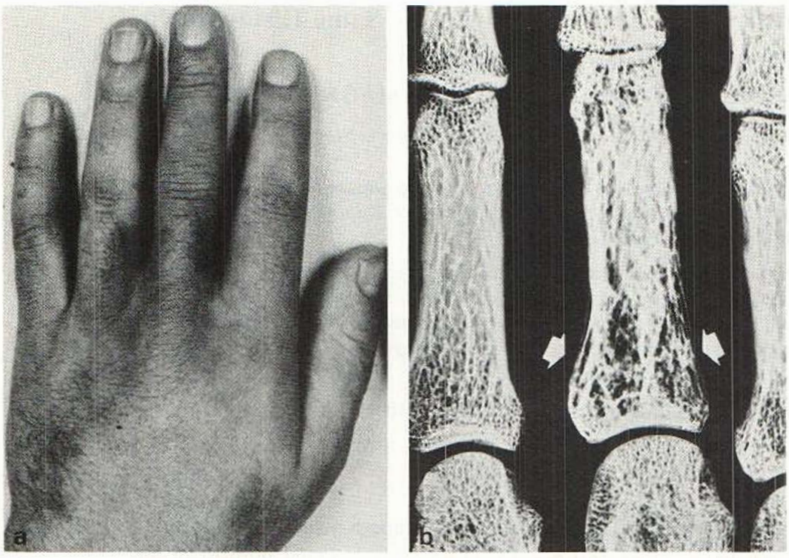

Fig. 1. (*a*) Irregular bluish red patches on the left hand; (*b*) radiograph showing typical hemangiomatous lesion of the proximal phalanx of the third finger, with thinning of the cortex and marked coarsening of the bony trabeculae (arrows).

Radiological findings

Radiographs of the hands and feet produced by an immersion magnification technique revealed angiomatous lesions of the proximal and middle phalanges of the left third finger (Fig. 1 *b*) and the distal phalanx of the left big toe. The cortex was thin, with preserved periosteal margins but an irregular medullary border. There was marked coarsening of the trabeculae of the medullary bone.

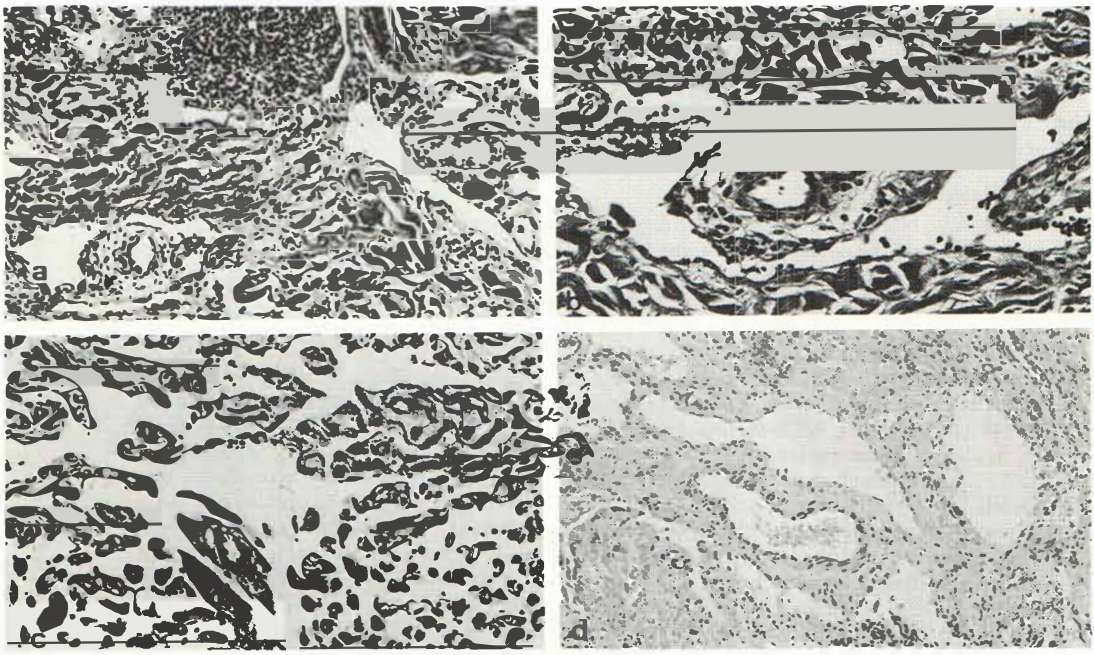

Fig. 2. (*a*) Biopsy from the dorsal aspect of a foot, showing thin-walled vessels, a connective tissue bud protruding into a vessel, and a lymphoid cell infiltrate; (*b*) close-up of an irregular thin-walled vessel with a bud; (*c*) biopsy from the distal part of the lower leg, showing the collagen bundles split up into small papillary fromations by dissecting vascular channels; (*d*) biopsy from the left sole, displaying a thick vessel with red blood cells surrounded by thin-walled vessels.

The zone of demarcation against normal bone was unsharp. The joint spaces were normal. Radiographic examination of the vertebral column disclosed no abnormalities.

DISCUSSION

The first two biopsies examined showed irregular vascular spaces into which protruded buds of connective tissue containing small vessels. There was also a patchy infiltrate of lympocytes and plasma cells in the dermis (Fig. 2 *a* and *b*)—a picture described by Gottlieb & Ackerman as the "promontory sign" of a patch of Kaposi's sarcoma, whether classical or appearing in patients with AIDS. Consequently, the pathologist knowing very little about the patient, proposed early Kaposi's sarcoma in a person with AIDS. The immunological investigations ruled out AIDS. Classical Kaposi's sarcoma was considered unlikely in view of the fact that more than a decade had passed since the onset of the disease without the occurrence of plaques or nodules. The absence of Borrelia antibodies ruled out acrodermatitis chronica atrophicans. Lymphangiomatosis or hemangiomatosis was then discussed. It is often very difficult histologically to decide whether empty thin-walled vascular spaces are blood vessels or lymph vessels (2). The presence of lymphoid cell infiltrates (Fig. 2 *a*) suggests lymphangiomatosis but does not exclude hemangiomatosis. The papillary pattern is compatible with both hemangiomatosis and lymphangiomatosis (Fig. 2 *c*). The presence of a thicker abnormal vessel points towards hemangiomatosis (Fig. 2 *d*). Radiological examination of the skeleton of the hands and feet showed coarsening of the medullary trabeculae and an indistinct border between affected and normal bone (Fig. 1 *b*). This pattern is regarded as characteristic of hemangiomas and differs from the osteolytic lesions of lymphangiomas (3). Altogether, and in consideration of the fact that lymphangiomatosis is an extremely rare disease (2), the most likely diagnosis is hemagiomatosis. Even though the presence of irregular thin-walled vessels and vascular connective tissue buds should first arouse suspicion of early lesions in Kaposi's sarcoma, we would like to stress that they are not unique for this disease. They are merely a sign of intensely proliferating vessels.

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