KAPOSI'S SARCOMA WITH OCULAR MANIFESTATIONS*

BY

NECDET SEZER AND CELÂL ERÇIKAN

Department of Ophthalmology, University of Istanbul, Turkey

KAPOSI's sarcoma is rarely mentioned among the skin lesions of the eyelids, most of the literature being devoted to the visceral lesions, the connexion with endotheliomata, the very much debated pathogenesis, and the treatment.

The condition was first described in Kaposi (1872) as "sarcoma idiopathicum pigmentosum" and later as "sarcoma idiopathicum hemorrhagicum". The histology is so varied that the condition was given different names by many of those who studied it.

In Turkey the first case was reported by Siğindım (1947), followed by Kınacıgil (1948), Abaoğlu (1954), and Egeli and Yalçın (1958). Ocular lesions were described by Graham (1942) and Sacks (1956), who each reported one case.

Clinical Appearance

The typical skin lesion is an erythematous, angiomatous plaque presenting in the beginning a macular appearance with well-defined irregular borders. The colour varies from dark red to bluish red. The lesions are hard to the touch but soften increasingly as time goes on. Some seem to be buried under the skin while others are pedunculated. They are usually smaller than a walnut but may conglomerate to form large plaques.

The skin lesions appear first on the hands and feet and spread towards the trunk; similar lesions appear on the lids and conjunctiva at this stage. The lymph nodes are also attacked and a lymphatic block ensues, with resulting elephantiasis of the extremities. Lesions are frequently encountered in the stomach, intestines, liver, spleen, lungs, heart, bones, and even the brain, and are considered to be part of the disease process and not metastases. Paraesthesia and neuralgic pains are among the few subjective symptoms reported by the patient.

Spontaneous involution of the lesions is rare. Radiotherapy may give transient benefit, but usually the patients die as a result of profuse haemorrhages provoked by intestinal lesions, of infection spreading from visceral lesions, or of the complications of malignant lymphoma. The survival period varies from 5 to 10 years from the onset of the disease.

Secondary anaemia, monocytosis, and more rarely eosinophilia and lymphocytosis have been noted in some cases. McCarthy and Pack (1950) observed the association of Hodgkin's disease, Brill-Symmer's syndrome, and lymphosarcoma with Kaposi's sarcoma in their patients. Fungoid mycosis, lymphatic leukaemia, and giant follicular lymphoma have also been found.

Histological Appearance

A typical lesion may show three different factors: angiomatous, sarcomatous, and infectious. Lever (1954) favours the part played by proliferation of the capillary perithelium and endothelium: proliferation of the endothelial cells giving an angiomatous appearance and that of the perithelial cells a fibroblastic appearance.

* Received for publication June 24, 1963.

Lever (1954) classifies these lesions as granulomatous early lesions and neoplastic late lesions. The granulomatous lesions show new formation of capillaries, blood extravasation, perivascular inflammatory cell infiltration, and even fibroblastic proliferation. The neoplastic lesions show an angiomatous appearance and blood extravasation. Cicatrization and fibrosis are dominant in regressing lesions.

Kaposi's sarcoma is regarded as a malignant degeneration of an infectious or granulomatous process of unknown aetiology by Belloni (1949), but Choisser and Ramsey (1939), and Thiers, Potton, and Tramier (1952) have shown experimentally that it is not infectious.

Case Report

A 53-year-old man came to the hospital with a small nodule on his nose (5 or 6 years old) and a bigger one on the left side of the neck (3 years old). Similar lesions had started to appear on the left leg and right foot, and then on the trunk. The patient had never complained of pain or itching. He had entered hospital in a neurological clinic 4 years before with paraesthesia of the arms and had been hospitalized at various times for observation for his skin lesions.

The family history was negative.

Examination.—There were numerous round or oval plaques hard to the touch on his face, trunk, and legs (Figs 1, 2, and 3); they were dark red in colour and varied in size from a pea to a nut.

FIGS 1, 2, 3.—Lesions on face, body, and legs.

A small slightly raised lesion, the size of a lentil, and a large bilobulated lesion were seen on the right eye, and there was a small round flat lesion inferiorly (Fig. 4, opposite). Two other lesions could be seen on the left eye, one on the upper lid nasally and the other on the lower lid temporally. The eyelid lesions were softer than the others. The conjunctivae were hyperaemic but presented no lesions like those on the skin.

Biomicroscopic and ophthalmoscopic examinations were negative as were the laboratory examinations, except that a chest x ray disclosed the presence of a round mass at the hilus of the left lung (Fig. 5, opposite). The bones appeared normal.

The erythrocyte sedimentation was 4 mm. in 30 minutes, 9 mm. in one hour, and 29 mm. in 2 hours.

The blood count showed erythrocytes 4,050,000, leucocytes 8,000, colour index 80 per cent. The differential count gave neutrophils 64 per cent., stab 1 per cent., eosinophils 2 per cent., lymphocytes 32 per cent., and monocytes 1 per cent.

The Wassermann reaction and Kahn test were negative.

The urine was normal.

The blood urea was 31 mg. per cent., cholesterol 200 mg. per cent., sugar 93 mg. per cent.

FIG. 4.—Lesions on right eyelids.

FIG. 5.—Lung x ray, showing round mass at left hilus.

Histology.—The small round lesion on the right lower lid was excised and sent to the Eye Pathology Laboratory, and the following report was received (Figs 6 and 7):

Figs 6 and 7.—Histological appearances in tumour from right eyelid.

The epidermis presents a hyperpigmentation of the cells of the basal layer. Sections of hair follicles are present in the dermis. Surrounding these follicles one can see areas of cellular infiltration presenting mostly an angiomatous and fibroblastic appearance. Mitotic figures are visible but no pleomorphism is observed. In the round and fusiform spaces among the endothelial cells and the fibroblasts, are scattered erythrocytes, a few neutrophil leucocytes, and lymphocytes. The reticulin stains reveal the presence of numerous reticulin fibres forming bands and networks in the areas of cellular infiltration.

Diagnosis.—Further systemic clinical examinations failed to reveal any other pathological findings in the nervous, circulatory, gastro-intestinal, and urogenital systems, and a diagnosis of Kaposi's sarcoma was made.

Treatment.—When the patient came to our clinic the illness was in an advanced stage, and only symptomatic therapy was instituted.

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