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Kaposi's Sarcoma in Japan: A Case Report and a Review of the Literature

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This report deals with a patient who had Kaposi's sarcoma which is quite infrequent in Japan, together with a review of the Japanese literature on this subject.

An 87-year-old male Korean long living in Japan had multifocal reddish blue dermal plaques and nodules starting on the right ankle and left forearm. On admission these lesions had progressed up both upper and lower extremities which coincidentally manifested considerable edema. Microscopically, most of the multifocal dermal lesions were ill-defined but distinctly nodular, and were composed of monomorphic spindle cells in vague bundles within which erythrocytes were enmeshed establishing narrow slits. The tumor cells showed mitotic figures of the nuclei but little pleomorphism. There also were areas resembling granulation tissue, suggesting a dermal lesion in the early stage, with numerous proliferating capillaries lined by plump endothelial cells and with scattered immature spindled or rounded cells around these capillaries, accompanying inflammatory cells and hemosiderin granules. Factor VIII-related antigen, a marker for endothelial cells, was found at least in some tumor cells with the immunoperoxidase technique. Electron micrograph revealed, in addition to an unequivocal lumen containing erythrocytes, fine slits adjoining

hyperplastic endothelial cells with perithelial cells around.

Although temporary control was effected by a combined chemotherapy with vincristine, cyclophosphamide and prednisolone, the patient followed a rapidly deteriorating course expiring six months after admission. Autopsy was not done. The patient had suffered from arthrosis deformans and bronchial asthma during the past 20 and 10 years, respectively, and was treated for corneal herpes six months prior to admission. His sera were positive for IgG antibodies to late antigens of cytomegalovirus (CMV) and negative for IgM antibodies to late antigens of CMV and for IgG antibodies to early antigens of CMV. CMV-related antigens were detected in the nucleus or cytoplasm of cells in tissue sections from the biopsies by the anti-complement immunofluorescent test.

Since 1917, about 90 patients have hitherto been reported in the Japanese literature as cases of Kaposi's sarcoma. In many cases, however, it is exceedingly difficult for us to amply justify the diagnosis of Kaposi's sarcoma, because of lack or paucity of illustrations. Less than 10 examples of older male patients with multiple lesions on the extremities could be acceptable as they would certainly be. Judging from the review of the literature, the most common diagnostic error seemed to be misinterpretation of angiosarcoma of the scalp and face of the elderly as Kaposi's sarcoma, both resembling each other only histologically. The next problem in differential diagnosis appeared to be the distinction from lymphangiosarcoma associated with lymphedema usually affecting either the arm or leg. Furthermore, some cases reported as Kaposi's sarcoma were under suspicion of Klippel-Trenaunay syndrome associated with varicosities and hypertrophy of an extremity.