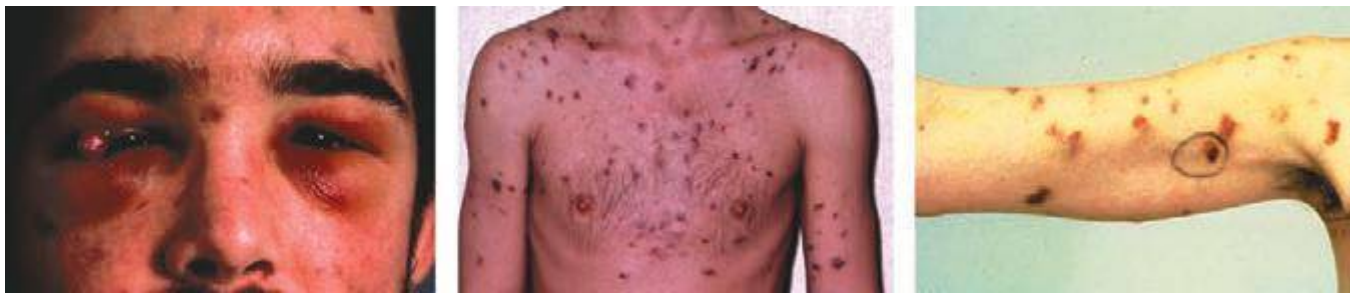

FIGURE 226-41 Kaposi's sarcoma in three patients with AIDS demonstrating (***A***) periorbital edema and bruising; (***B***) classic truncal distribution of lesions; and (***C***) upper extremity lesions.

count. The initial lesion may be a small, raised reddish-purple nodule on the skin (Fig. 226-41), a discoloration on the oral mucosa (Fig. 226-34*D*), or a swollen lymph node. Lesions often appear in sun-exposed areas, particularly the tip of the nose, and have a propensity to occur in areas of trauma (Koebner phenomenon). Because of the vascular nature of the tumors and the presence of extravasated red blood cells in the lesions, their colors range from reddish to purple to brown and often take the appearance of a bruise, with yellowish discoloration and tattooing. Lesions range in size from a few millimeters to several centimeters in diameter and may be either discrete or confluent. KS lesions most commonly appear as raised macules; however, they can also be papular, particularly in patients with higher CD4+ T cell counts. Confluent lesions may give rise to surrounding lymphedema and may be disfiguring when they involve the face and disabling when they involve the lower extremities or the surfaces of joints. Apart from skin, the lymph nodes, GI tract, and lung are the organ systems most commonly affected by KS. Lesions have been reported in virtually every organ, including the heart and the CNS. In contrast to most malignancies, in which lymph node involvement implies metastatic spread and a poor prognosis, lymph node involvement may be seen very early in KS and is of no special clinical significance. In fact, some patients may present with disease limited to the lymph nodes. These are generally patients with relatively intact immune function and thus the patients with the best prognosis. Pulmonary involvement with KS generally presents with shortness of breath. Some 80% of patients with pulmonary KS also have cutaneous lesions. The chest x-ray characteristically shows bilateral lower lobe infiltrates that obscure the margins of the mediastinum and diaphragm (Fig. 226-42). Pleural effusions are seen in 70% of cases of pulmonary KS, a fact that is often helpful in the differential diagnosis. GI involvement is seen in 50% of patients with KS and usually takes one of two forms: (1) mucosal involvement, which may lead to bleeding that can be severe; these patients sometimes also develop symptoms of GI obstruction if lesions become large; and (2) biliary tract involvement. KS lesions may infiltrate the gallbladder and biliary tree, leading to a clinical picture of obstructive jaundice similar to that seen with sclerosing cholangitis. Several staging systems have been proposed for KS. One in common use was developed by the National Institute of Allergy and Infectious Diseases AIDS Clinical Trials Group; it distinguishes patients on the basis of tumor extent, immunologic function, and presence or absence of systemic disease (Table 226-17).

A diagnosis of KS is based on biopsy of a suspicious lesion. Histologically one sees a proliferation of spindle cells and endothelial cells, extravasation of red blood cells, hemosiderin-laden macrophages, and, in early cases, an inflammatory cell infiltrate. Included in the differential diagnosis are lymphoma (particularly for oral lesions), bacillary angiomatosis, and cutaneous mycobacterial infections.

Management of KS (Table 226-18) should be carried out in consultation with an expert since definitive treatment guidelines do not exist. In the majority of cases, effective cART will go a long way in achieving control. Antiretroviral therapy has been associated with the spontaneous regression of KS lesions. Paradoxically, it has also been associated with the initial appearance of KS as a form of IRIS. For patients in whom tumor persists or is compromising vital functions or in whom control of HIV replication is not possible, a variety of options exist. In some cases, lesions remain quite indolent, and many of these patients can be managed with no specific treatment. Fewer than 10% of AIDS patients with KS die as a consequence of their malignancy,

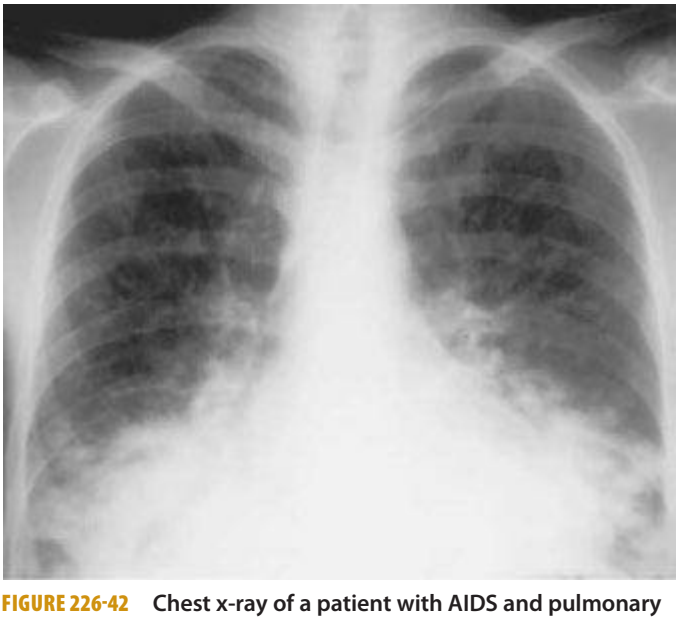

FIGURE 226-42 Chest x-ray of a patient with AIDS and pulmonary Kaposi's sarcoma. The characteristic findings include dense bilateral lower lobe infiltrates obscuring the heart borders and pleural effusions.

TABLE 226-17 NATIONAL INSTITUTE OF ALLERGY AND INFECTIOUS DISEASES AIDS CLINICAL TRIALS GROUP TIS STAGING SYSTEM FOR KAPOSI'S SARCOMA

Parameter	Good Risk (Stage 0): All of the Following	Poor Risk (Stage 1): Any of the Following
Tumor (T)	Confined to skin and/or lymph nodes and/or minimal oral disease	Tumor-associated edema or ulceration
		Extensive oral lesions
		GI lesions
		Nonnodal visceral lesions
Immune system (I)	CD4+ T cell count ≥200/μL	CD4+ T cell count <200/μL
Systemic illness (S)	No B symptoms[a]	B symptoms[a] present
	Karnofsky performance status ≥70	Karnofsky performance status <70
	No history of opportunistic infection, neurologic disease, lymphoma, or thrush	History of opportunistic infection, neurologic disease, lymphoma, or thrush

[a]Defined as unexplained fever, night sweats, >10% involuntary weight loss, or diarrhea persisting for more than 2 weeks.