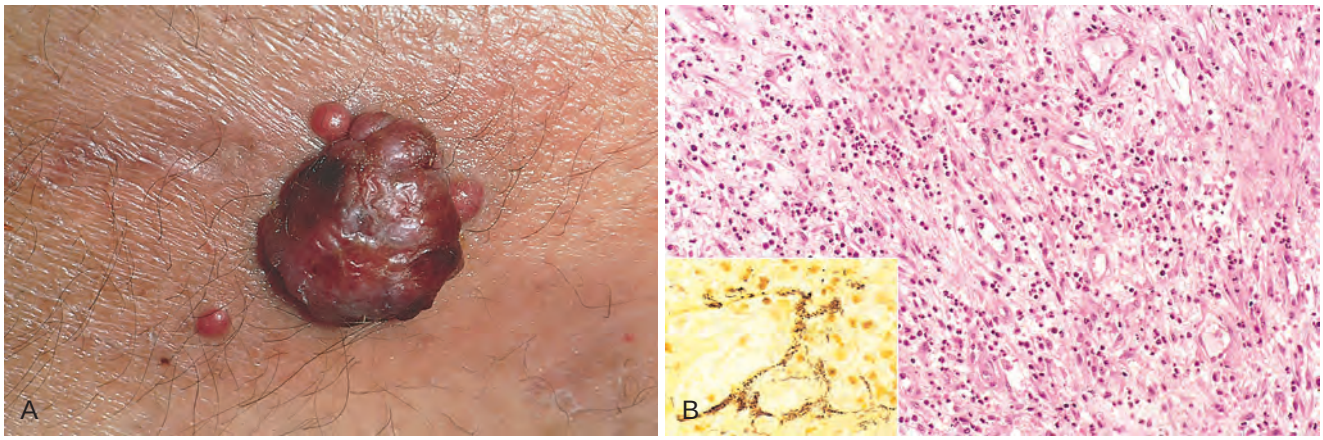

Figure 11-31 Bacillary angiomatosis. **A,** Characteristic cutaneous lesion. **B,** Histologic appearance with acute neutrophilic inflammation and vascular (capillary) proliferation. Inset, modified silver (Warthin-Starry) stain demonstrates clusters of tangled bacilli (black). (**A,** courtesy Richard Johnson, MD, Beth Israel Deaconess Medical Center, Boston; **B** and *inset,* courtesy Scott Granter, MD, Brigham and Women's Hospital, Boston, Mass.)

vascular endothelial growth factor (VEGF) production. The infections (and lesions) are cleared by macrolide antibiotics (including erythromycin).

Intermediate-Grade (Borderline) Tumors

Kaposi Sarcoma. **Kaposi sarcoma (KS) is vascular neoplasm caused by human herpesvirus 8 (HHV8) that is highly associated with acquired immunodeficiency syndrome (AIDS).** It also occurs much less commonly in other settings. Four forms of KS are recognized, based on population demographics and risks:

- *Classic KS* is a disorder of older men of Mediterranean, Middle Eastern, or Eastern European descent (especially Ashkenazic Jews); it is uncommon in the United States. It can be associated with malignancy or altered immunity, but is not associated with HIV infection. Classic KS manifests as multiple red-purple skin plaques or nodules, usually in the distal lower extremities; these progressively increase in size and number and spread proximally. Although persistent, the tumors are typically asymptomatic and remain localized to the skin and subcutaneous tissue.
- *Endemic African KS* typically occurs in HIV-seronegative individuals younger than age 40 and can follow an indolent or aggressive course; it involves lymph nodes much more frequently than the classic variant. In combination with AIDS-associated KS (see later), KS is now the most common tumor in central Africa. A particularly severe form, with prominent lymph node and visceral involvement, occurs in prepubertal children; the prognosis is poor, with an almost 100% mortality within 3 years.
- *Transplant-associated KS* occurs in solid organ transplant recipients in the setting of T-cell immunosuppression. The risk of KS is increased 100-fold in transplant recipients, pursuing an aggressive course that characteristically involves lymph nodes, mucosa, and viscera; cutaneous lesions may be absent. Lesions often regress with attenuation of immunosuppression, but at the risk of organ rejection.
- *AIDS-associated (epidemic) KS* is an AIDS-defining illness, and *worldwide, it represents the most common HIV-related malignancy* (Chapter 6). Although the incidence of KS has fallen more than 80% with the advent of aggressive antiretroviral therapies, it still occurs in HIV-infected individuals at a rate over a thousand-fold greater than in the general population, and affects 2-3% of the HIV-infected population in the US. AIDS-associated KS often involves lymph nodes and disseminates widely to viscera early in its course. Most patients eventually die of opportunistic infections rather KS.

Pathogenesis. Virtually all KS lesions are infected by *human herpesvirus 8 (HHV8),* also known as Kaposi sarcoma herpesvirus. Like Epstein-Barr virus, HHV8 is a γ-herpesvirus. It is transmitted sexually and by poorly understood nonsexual routes potentially including oral secretions and cutaneous exposures (of note, the prevalence of endemic African KS is inversely related to the wearing of shoes). HHV8 and altered T-cell immunity are likely required for KS development; in older adults, diminished T-cell immunity may be related to aging. Because the development and progression of KS are tightly linked to immune function, its molecular pathogenesis is discussed in detail in Chapter 6.

MORPHOLOGY

In **classic KS** (and sometimes in other variants), the cutaneous lesions progress through three stages:

- **Patches** are red-purple macules typically confined to the distal lower extremities (Fig. 11-32*A*). Histology shows only dilated irregular endothelial cell–lined vascular spaces with interspersed lymphocytes, plasma cells, and macrophages (sometimes containing hemosiderin). The lesions can be difficult to distinguish from granulation tissue.
- With time, lesions spread proximally and become larger, violaceous, **raised plaques** (Fig. 11-32*A*) composed of dermal accumulations of dilated, jagged vascular channels lined and surrounded by plump spindle cells. Scattered between the vascular channels are extravasated erythrocytes, hemosiderin-laden macrophages, and other mononuclear inflammatory cells.

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