

Figure 26-50 Liposarcoma. **A**, the well-differentiated subtype consists of mature adipocytes and scattered spindle cells with hyperchromatic nuclei. **B**, Myxoid liposarcoma with abundant ground substance and a rich capillary network in which are scattered immature adipocytes and more primitive round to stellate cells.

All types of liposarcoma recur locally and often repeatedly unless adequately excised. The well-differentiated variant is relatively indolent, the myxoid/round cell type is intermediate in its malignant behavior, while the pleomorphic variant usually is aggressive and frequently metastasizes.

Fibrous Tumors

Nodular Fasciitis

Nodular fasciitis is a self-limited fibroblastic and myofibroblastic proliferation that typically occurs in young adults in the upper extremity. A history of trauma is present in approximately 25% of cases and the tumors grow rapidly over a period of several weeks or months, typically no larger than 5 cm. Whereas nodular fasciitis was historically considered a reactive inflammatory lesion, identification of t(17;22) that produces a *MYH9-USP6* fusion gene indicates that it is a clonal, but self-limited, proliferation. It appears that the proliferating cells lack some key hallmark of cancer, perhaps the ability to avoid senescence. Intriguingly, ABC (discussed earlier), another tumor that sits in a gray zone between reactive and neoplastic proliferations, also contains *USP6* fusion genes. Nodular fasciitis typically spontaneously regresses and if excised, it rarely recurs.

MORPHOLOGY

Nodular fasciitis arises in the deep dermis, subcutis, or muscle. Grossly the lesion is less than 5 cm, circumscribed, or slightly infiltrative. The lesion is richly cellular and contains plump, immature-appearing fibroblasts and myofibroblasts arranged randomly or in short fascicles reminiscent of tissue culture fibroblasts (Fig. 26-51). A gradient of maturation (zonation) from cellular, loose, and myxoid to organized and fibrous is typical. The cells vary in size and shape (spindle to stellate) and have conspicuous nucleoli; mitotic figures are abundant. Lymphocytes and extravasated red blood cells are common but neutrophils are unusual.

Fibromatoses

Superficial Fibromatosis

Superficial fibromatosis is an infiltrative fibroblastic proliferation that can cause local deformity but has an innocuous clinical course. All forms of superficial fibromatosis affect males more frequently than females. They are characterized by nodular or poorly defined broad fascicles of fibroblasts in long, sweeping fascicles, surrounded by abundant dense collagen. Several clinical subtypes have been identified:

- **Palmar (*Dupuytren contracture*):** Irregular or nodular thickening of the palmar fascia either unilaterally or bilaterally (50%). Over a span of years, attachment to the overlying skin causes puckering and dimpling. At the same time a slowly progressive flexion contracture develops that mainly affects the fourth and fifth fingers of the hand.

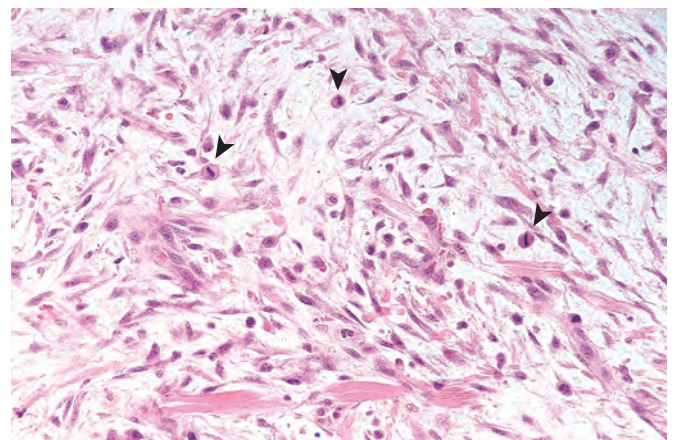


Figure 26-51 Nodular fasciitis with plump, randomly oriented spindle cells surrounded by myxoid stroma. Note the mitotic activity (arrowheads) and extravasated red blood cells.