Merkel cell carcinoma is diagnosed by clinical features and histopathological tests. Adnexal malignant tumors, amelanotic malignant melanoma, and lymphoma are differentiated from it. Metastasis of small-cell lung cancer to the skin presents the same symptoms and pathology as Merkel-cell carcinoma. If this carcinoma is suspected, examination for lung cancer should be performed.

**Treatment, Prognosis**

Merkel cell carcinoma tends to be highly malignant. At the relatively early stages, there is lymph node involvement or hematogenous metastasis. Because of its recurrent tendency, extensive excision and lymph node dissection are conducted. Radiation therapy and chemotherapy are also useful. Cases with spontaneous healing have been reported.

### E. Mesenchymal tumors

#### a. Fibrous tissue tumors

**1. Dermatofibrosarcoma protuberans (DFSP)**

Dermatofibrosarcoma protuberans (DFSP) is a malignant tumor that is thought to derive from fibroblasts, myofibroblasts or histiocytes. The trunk of adult men is most commonly involved. It begins as an intradermal or subcutaneous nodule that slowly forms a dome-shaped or fungiform tumor at the local site (Fig. 22.26). The tumor is firm and dark reddish brown, often accompanied by erosion or crusts. A characteristic swirl arrangement of tumor cells and fibers called storiform pattern is histopathologically observed (Fig. 22.27). The tumor cells are usually factor XIIIa negative and CD34 positive. DFSP rarely metastasizes (fewer than 10% of cases); however, extensive resection is necessary because of its high tendency to recur.

**2. Malignant fibrous histiocytoma (MFH)**

This is the most frequently occurring soft-tissue sarcoma. The proximal limb muscles and retroperitoneum of adults are most commonly involved. Skin is rarely affected primarily. A painless, lobulated, multinodular subcutaneous tumor occurs in most cases. Hematoma-like lesions (angiomatoid type) may form. Malignant fibrous histiocytoma (MFH) is histopathologically composed of highly atypical fibroblast-like cells and histiocytelike cells. It displays various pathological features, including peculiarly shaped giant cells and inflammatory cellular infiltration. MFH is classified by pathological findings into several types: storiform-pleomorphic (the highest incidence), myxoid,
3. Atypical fibroxanthoma

A lesion composed of histiocyte-like cells with low malignant potential forms, most frequently on a sun-exposed area. Some consider it as identical to superficial MFH.

4. Epithelioid sarcoma

This is a rare malignant tumor. It occurs most commonly at the ends of the extremities and progresses relatively slowly. It begins as intradermal or subcutaneous nodules that gradually increase in number and size (Fig. 22.28). Epithelial cells with abundant eosinophilic components histopathologically proliferate in sheet-like formation. The center of the nodule is necrotic in many cases. At the early stages, epithelioid sarcoma resembles granuloma annulare and rheumatoid nodule; differential diagnosis can be made by the immunohistochemical finding of keratin-positive cells in epithelioid sarcoma. Extensive resection is the basic treatment. When epithelioid sarcoma metastasizes, lymph nodes are often involved. The prognosis is poor.

5. Synovial sarcoma

A soft, painful tumor occurs, frequently in the large joint of an extremity, particularly in the knee. In rare cases, it occurs subcutaneously or subfascially. Although synovial sarcoma used to be attributed to abnormal production of synovial tissue, that idea has been disproven in recent years. Chromosomal translocation t (X; 18) (p11.2; q11.2) occurs in the cells of the sarcoma. The tumor grows slowly and metastasizes to the lymph nodes. The prognosis is poor. Extensive resection is essential.

b. Tumors of the fat cells

Liposarcoma

A liposarcoma is a malignant mesenchymal tumor that differentiates into fat cells. It is deep-seated, clearly defined, large and nearly asymptomatic. According to the new WHO classification, the malignant types are the well-differentiated type, myxoid type, round cell type, pleomorphic type, and dedifferentiated type. Well-differentiated liposarcoma has a good prognosis and is also called atypical lipomatous tumor. Extensive resection is the main treatment.