

MSS module

Soft Tissue Tumors



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soft tissue tumor

- soft tissue describes any non-epithelial tissue other than bone, cartilage, central nervous system, hematopoietic, and lymphoid tissues.
- They arise from pluripotent mesenchymal stem cells and are not the result of malignant transformation of mature mesenchymal cells.



Risk factor for soft tissue tumor

- Most soft tissue tumors arise without antecedent causes but many causes may be implicated:
- Virus: Kaposi sarcoma is associated with the human herpesvirus 8.
- Genetic syndromes, most notably:
 - ✓ Neurofibromatosis type 1 (neurofibroma, malignant schwannoma).
 - ✓ Gardner syndrome (fibromatosis).
 - ✓ Li-Fraumeni syndrome (soft tissue sarcoma).
- Radiation exposure, burn injury, or toxin exposure

Sites vs ages

- ❖ 40% of sarcomas occur in the lower extremities, especially the thigh.
- ❖ While the overall incidence of sarcomas increases with age, 15% arise in children.
- ❖ Certain sarcomas tend to appear in certain age groups—for example:
 - ✓ Rhabdomyosarcoma in childhood.
 - ✓ synovial sarcoma in young adulthood.
 - ✓ liposarcoma and pleomorphic fibroblastic or undifferentiated sarcomas in later adult life.

Table 20-4 Soft Tissue Tumors

Tumors of Adipose Tissue ←

Lipomas

Liposarcoma

Tumors and Tumor-Like Lesions of Fibrous Tissue ←

Nodular fasciitis

Fibromatoses

Superficial fibromatoses

Deep fibromatoses

Fibrosarcoma

Fibrohistiocytic Tumors ←

Fibrous histiocytoma

Dermatofibrosarcoma protuberans

Pleomorphic fibroblastic sarcoma/pleomorphic undifferentiated sarcoma
(malignant fibrous histiocytoma)

Tumors of Skeletal Muscle ←

Rhabdomyoma

Rhabdomyosarcoma

Tumors of Smooth Muscle ←

Leiomyoma

Smooth muscle tumors of uncertain malignant potential

Leiomyosarcoma

Vascular Tumors

Hemangioma

Lymphangioma

Hemangioendothelioma

Angiosarcoma

Peripheral Nerve Tumors

Neurofibroma

Schwannoma

Granular cell tumor

Malignant peripheral nerve sheath tumors

Tumors of Uncertain Histogenesis

Synovial sarcoma

Alveolar soft part sarcoma

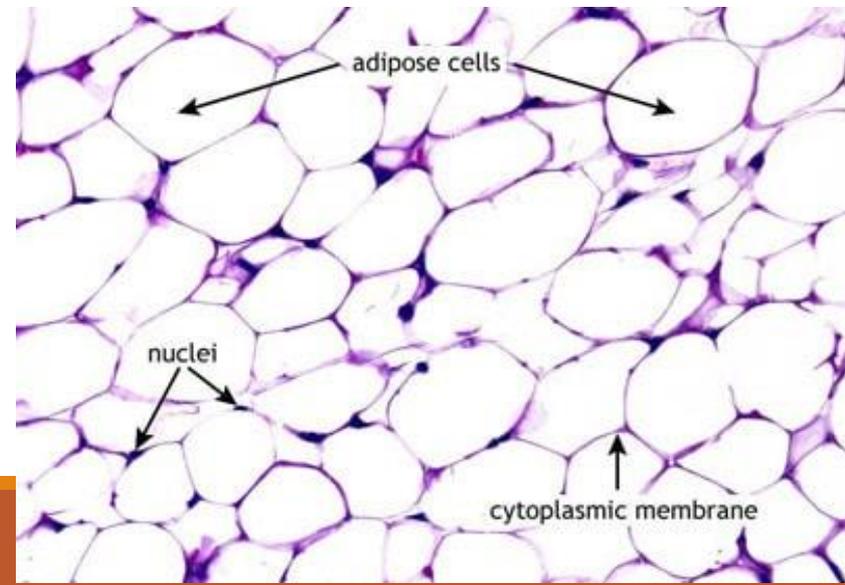
Epithelioid sarcoma

1- TUMORS OF ADIPOSE TISSUE

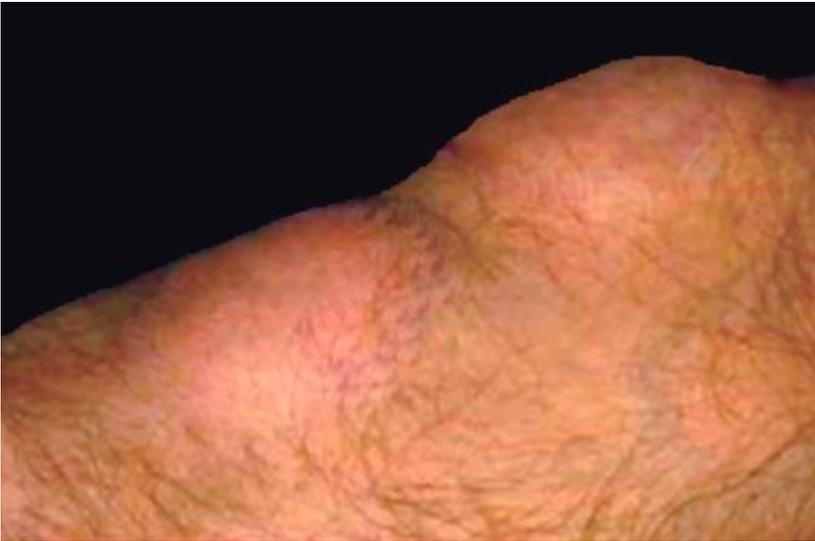
I. LIPOMA:

benign tumors of fat, the most common soft tissue tumors in adults.

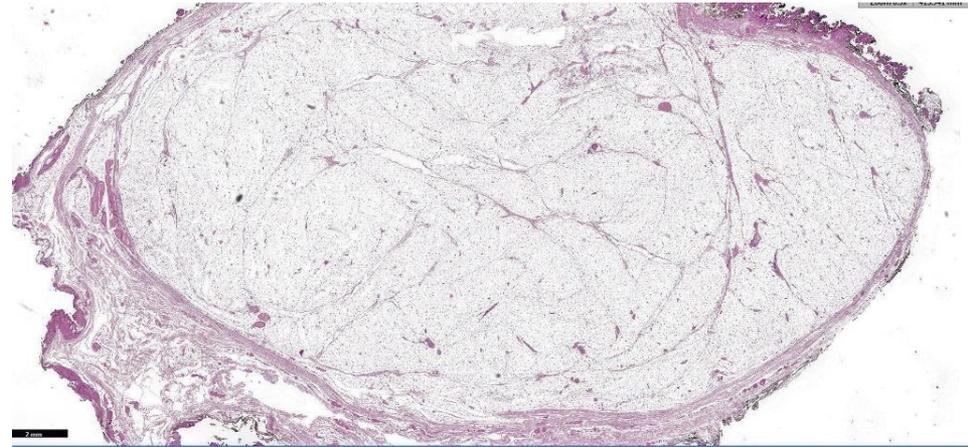
- ✓ Most lipomas are solitary lesions; multiple lipomas usually suggest the presence of rare hereditary syndromes.
- ✓ Most lipomas are mobile, slowly enlarging, painless masses.
- ✓ Complete excision usually is curative.



Gross and histological features



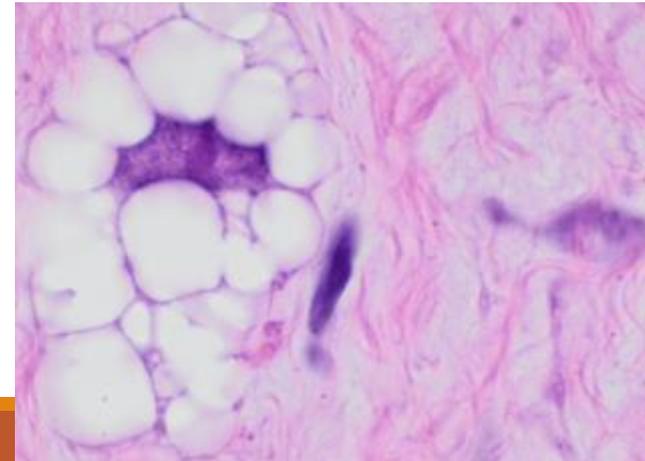
- Well circumscribed, Nodular



- Proliferation of mature adipocytes

II. LIPOMYOSARCOMA:

- Liposarcomas are malignant neoplasms with adipocyte differentiation.
- They occur most commonly in the fifth and sixth decades of life.
- Most liposarcomas arise in the deep soft tissues or in the retroperitoneum.
- In most cases, cells indicative of fatty differentiation known as **lipoblasts** are present; they have cytoplasmic lipid vacuoles.



2- FIBROUS TUMORS AND TUMOR-LIKE LESIONS

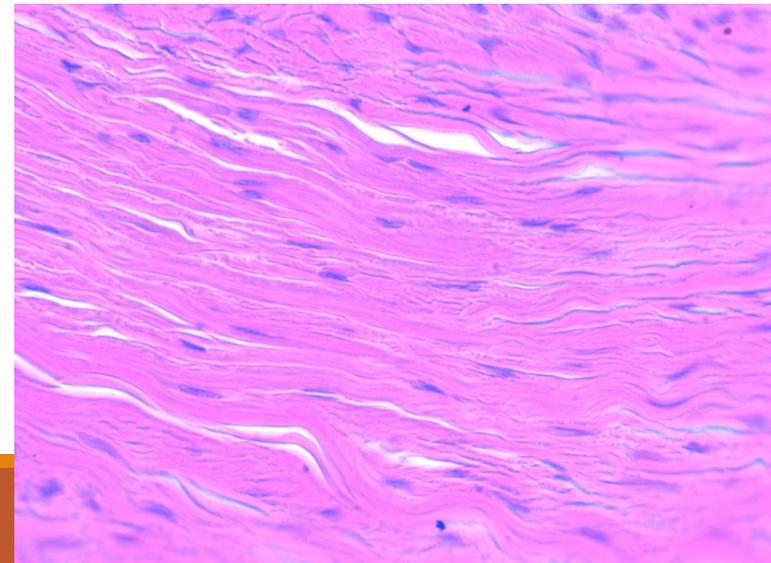
❖ Fibrous tissue proliferations are a heterogeneous group of lesions.

❖ It includes:

1- Reactive Proliferations: Nodular Fasciitis

2- Gray zone lesion: Fibromatoses

3- Neoplasm: Fibrosarcoma

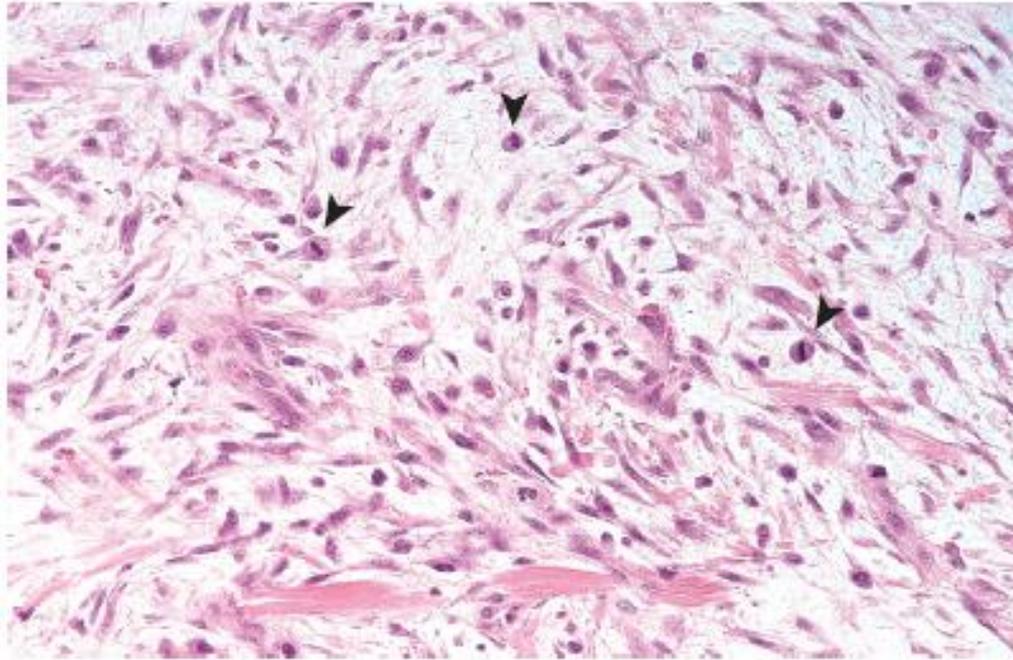


1. Reactive Proliferations

Nodular Fasciitis

- ✓ Nodular fasciitis is a self-limited fibroblastic proliferation that typically occurs in adults on the forearm, the chest, or the back.
- ✓ Patients characteristically present with a several-week history of a solitary, rapidly growing, and occasionally painful mass.
- ✓ Preceding trauma is noted in 10% to 15% of cases.
- ✓ Nodular fasciitis rarely recurs after excision.

Microscopic features



highly cellular lesion composed of plump, randomly oriented spindle cells surrounded by myxoid stroma.
Note the prominent mitotic activity

2. Gray zone lesion : fibromatoses

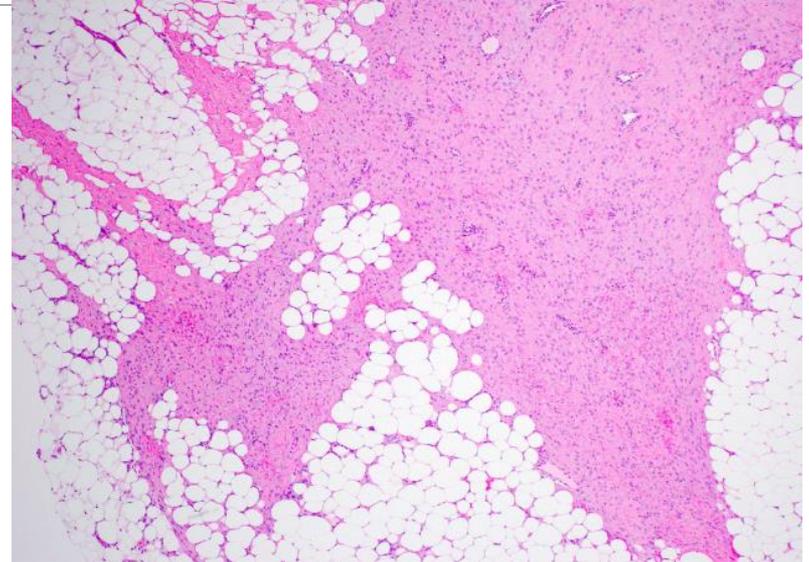
Group of fibroblastic proliferations distinguished by:

- Their tendency to grow in an infiltrative fashion.
- To recur after surgical removal.
- Locally aggressive.
- They do not metastasize.

Gross and histological features



Macroscopically: Fibromatoses are gray-white, firm to rubbery, poorly demarcated, infiltrative masses 1 to 15 cm in greatest dimension.



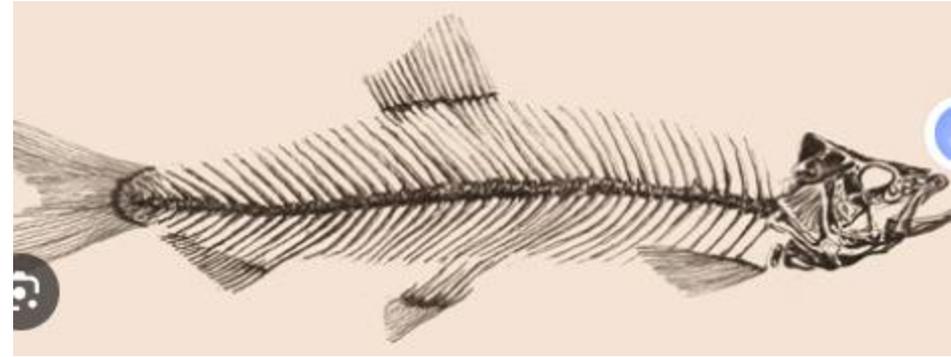
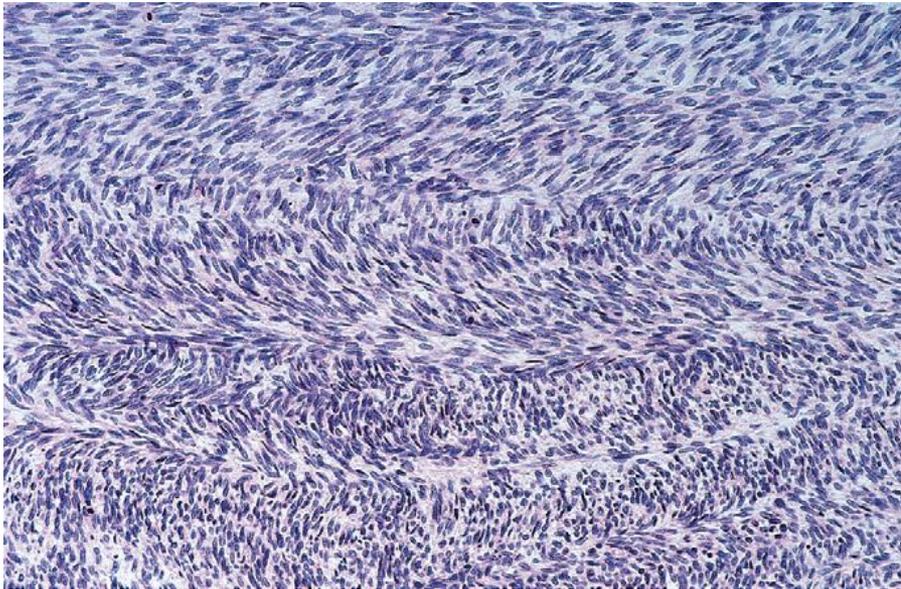
Microscopically:

On histologic examination, they are composed of plump spindle cells arranged in broad sweeping fascicles that penetrate the adjacent tissue; mitoses are few in number.

3. Malignant proliferation: Fibrosarcomas

- ✓ Fibrosarcomas are malignant neoplasms composed of fibroblasts.
- ✓ Most occur in adults, typically in the deep tissues of the thigh, knee, and retroperitoneal area.
- ✓ Fibrosarcomas often recur locally after excision and can metastasize hematogenously , usually to the lungs.

Fibrosarcomas. Malignant spindle cells here are arranged in a herringbone pattern.



3- FIBROHISTIOCYTIC TUMORS

- Fibrohistiocytic tumors are composed of a mixture of fibroblasts and phagocytic, lipid-laden cells macrophages (histiocytes).
- These tumors span a broad range of histologic patterns and biologic behavior, from self-limited benign lesions to aggressive high-grade sarcomas:

1-Benign Fibrous Histiocytoma (Dermatofibroma)

2- Pleomorphic Fibroblastic Sarcoma/Pleomorphic

Undifferentiated Sarcoma

Benign Fibrous Histiocytoma (Dermatofibroma)

- ✓ Dermatofibromas are relatively common benign lesions in adults manifesting as circumscribed, small (less than 1 cm) mobile nodules in the dermis or subcutaneous tissue.
- ✓ On histologic evaluation, these typically consist of bland, interlacing spindle cells admixed with foamy, lipid-rich histiocyte-like cells. The borders of the lesions tend to be infiltrative.
- ✓ They are cured by simple excision.
- ✓ The pathogenesis of these lesions is uncertain.

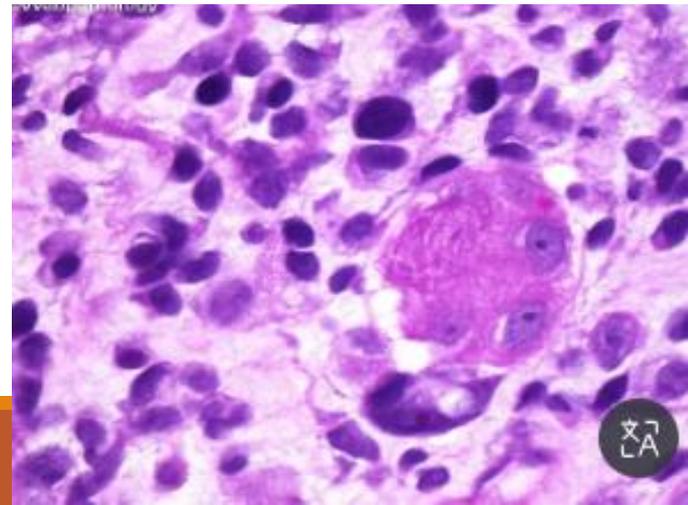
Pleomorphic Fibroblastic sarcoma/Pleomorphic Undifferentiated Sarcoma

- ✓ Undifferentiated sarcoma shows no identifiable line of differentiation when analyzed by presently available technology.
- ✓ They usually are large (5 cm to 20 cm), gray-white unencapsulated masses that often appear deceptively circumscribed.
- ✓ They usually arise in the musculature of the proximal extremities or in the retroperitoneum.
- ✓ Most of these tumors are extremely Aggressive.

Skeletal Muscle Tumors

Rhabdomyosarcoma

- ✓ Rhabdomyosarcoma is the most common soft tissue sarcoma of childhood and adolescence, usually appearing before age 20.
- ✓ Of interest, it occurs most commonly in the head and neck or genitourinary tract.
- ✓ The rhabdomyoblast is the diagnostic cell in all histological sub-types;

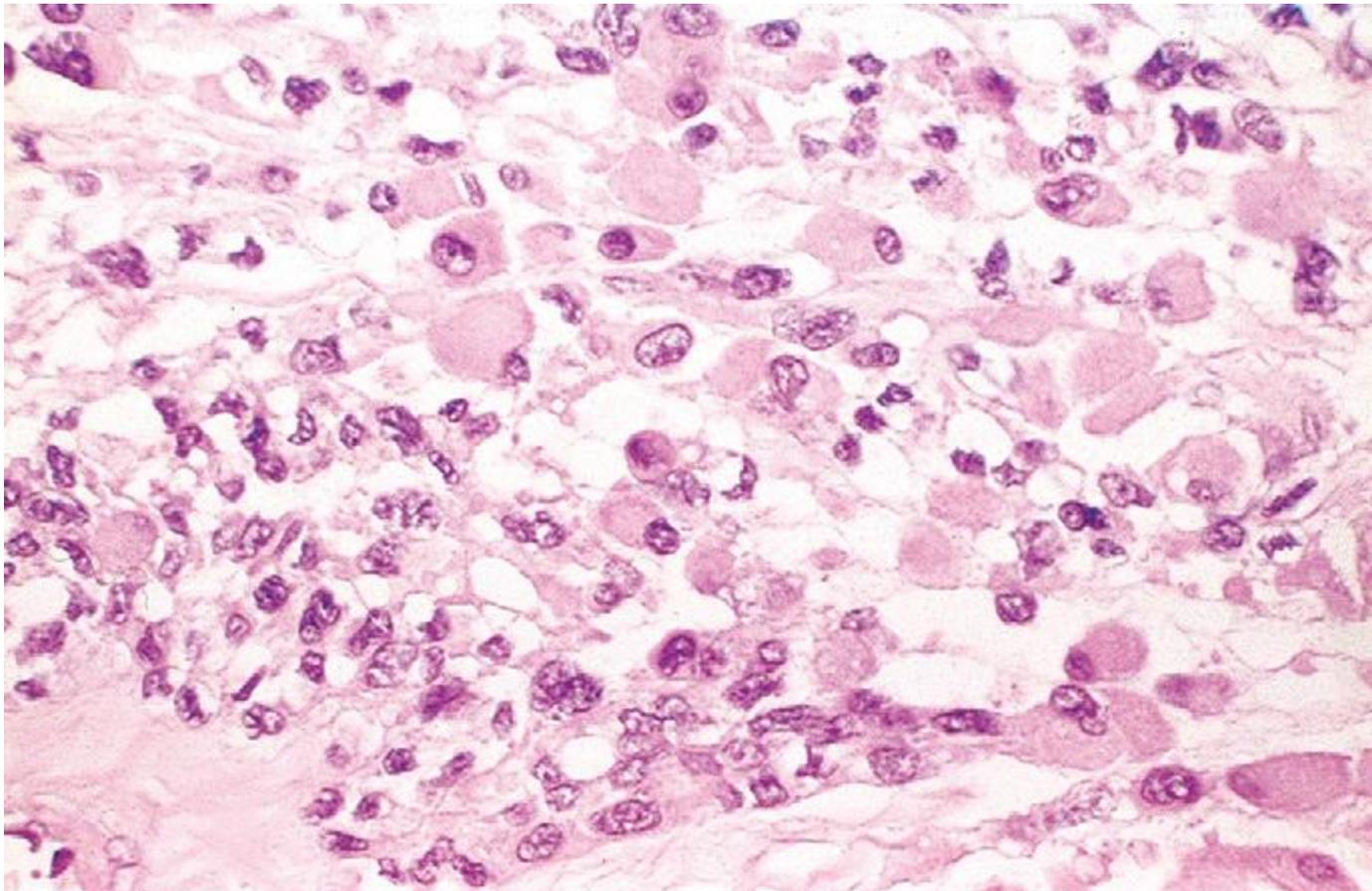


The diagnosis of rhabdomyosarcoma is based on the demonstration of:

- ✓ skeletal muscle differentiation, either in the form of :
 - sarcomeres under the electron microscope.
 - or by immunohistochemical demonstration of skeletal muscle specific transcription factors such as **myogenin** and **MYOD-1**, and the muscle-associated intermediate filament **desmin**.

Rhabdomyosarcoma.

The rhabdomyoblasts are large and round and have abundant eosinophilic cytoplasm.

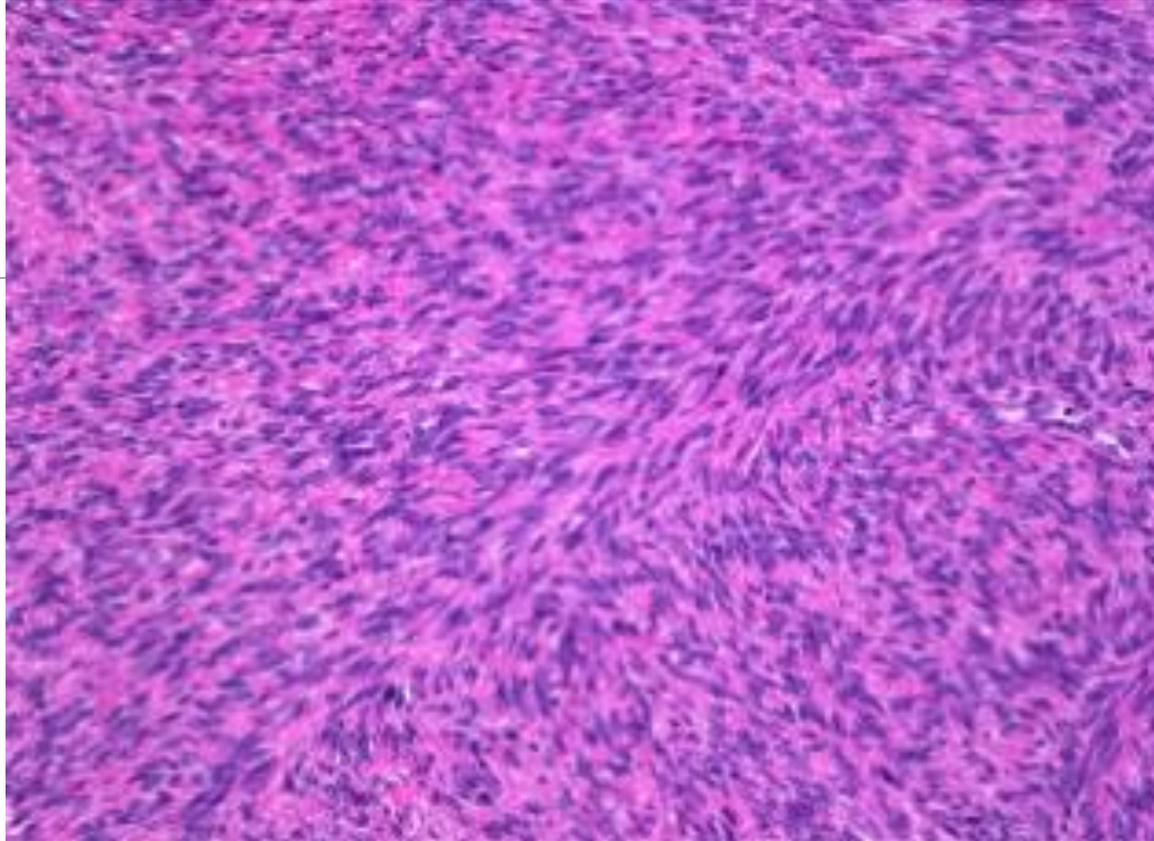


SMOOTH MUSCLE TUMORS

Leiomyoma

- ✓ Benign smooth muscle tumors are common, well-circumscribed neoplasms that can arise from smooth muscle cells anywhere in the body but are encountered most commonly in the uterus and the skin.





Histologic examination shows spindle cells with cigar-shaped nuclei arranged in interwoven fascicles.

Leiomyosarcoma

- ✓ Leiomyosarcomas account for 10% to 20% of soft tissue sarcomas.
- ✓ They occur in adults, more commonly females.

- ✓ Prognosis:
 - ✓ Superficial or cutaneous leiomyosarcomas usually are small and carry a good prognosis.
 - ✓ whereas retroperitoneal tumors are large and difficult to excise and cause death by both local extension and metastatic spread.