Malignant Tumors of Connective Tissue-I

Dept Of Oral Pathology & Microbiology

Purpose Statement

At the end of the lecture student should be able to--

- Differentiate between sarcoma and carcinoma
- Describe Classification, incidence, etiology, clinical features & histopathological features of **fibrosarcoma**
- Describe Classification, incidence, etiology, clinical features & histopathological features of **locally aggressive fibrous lesion**
- Describe Classification, incidence, etiology, clinical features & histopathological features of **Kaposi's sarcoma**

Learning Objectives

. N .	Learning Objectives	Do main	Lev el	Crit eria	Con ditio n
	Enumerate clinical features	Cog nitive	Mus t Know	All	
	Write classification	Cog nitive	Must Know	All	
	Write pathogenesis	Cog nitive	Mus t	All	

Contents

- Sarcoma vs Carcinoma
- Fibrosarcoma
- Miscellaneous locally aggressive fibrous lesions
- Fibrous histiocytoma
- Malignant fibrous histiocytoma
- Kaposis sarcoma

Sarcoma Vs Carcinoma

- Age: Relatively younger

- Metastasis: >> Blood
  More wide spread foci
- Rare in oral & maxillofacial region

- Less than 1%

Fibrosarcoma

- Tumor of mesenchymal cell origin
- Cell of Origin: Malignant Fibroblasts
- An increased, abnormal proliferation of malignant fibroblasts in collagenous stroma

Types

Fibrosarcoma

- Soft tissue tumor
- Bone tumor
  - Primary:
    - Central – Medullary canal
    - Peripheral - Periosteum
  - Secondary:
  - Arising from preexisting lesions / after radiotherapy

Fibrosarcoma

- Etiopathogenesis – no definite cause
- Preexisting lesions –
  - Multiple neurofibromas
  - Fibrous dysplasia
  - Chronic osteomyelitis
  - Bone infarcts
  - Paget disease
  - Previously irradiated areas of bone
  - **Current research-genetic mutation**
- More aggressive with poor prognosis

Fibrosarcoma

Retromolar region---showing fibrosarcoma

Clinical Features

Fibrosarcoma

Fibrosarcoma of bone

- Age - any age, but the peak age - 4th decade
- Sex - slightly M>F
- Site – E/o – Lower extremities, femur & tibia I/o – Anywhere in H & N region
- S/S – Pain with swelling
  - Pathologic fracture
  - Longer duration

Fibrosarcoma of soft tissue

- Age range: 35-55 yrs
- Slow growing large painless mass with ill defined margins
- Shorter duration

Histopathologic Features

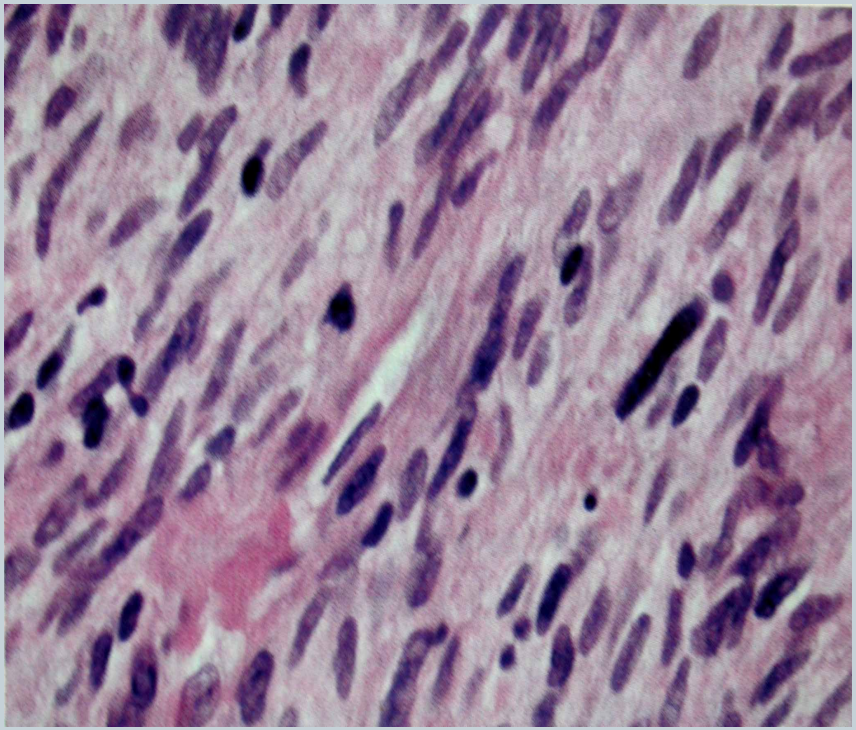
Fibrosarcoma

- Malignant tumor of fibroblasts with no deposition of osteoid or bone.
- 3 grades
  - Well differentiated
  - Intermediate grade
  - High grade

Well-differentiated FS

Fibrosarcoma

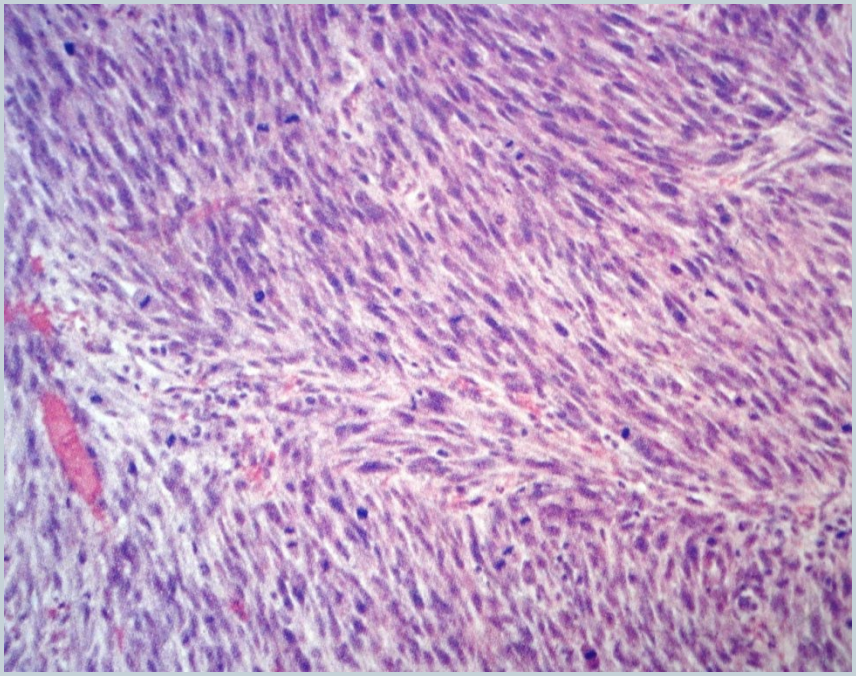
- Multiple plump fibroblasts with pale eosinophilic cytoplasm and deeply staining spindled nuclei with tapered ends.
  - Rich collagen background
  - No pleomorphism



Intermediate-grade FS

Fibrosarcoma

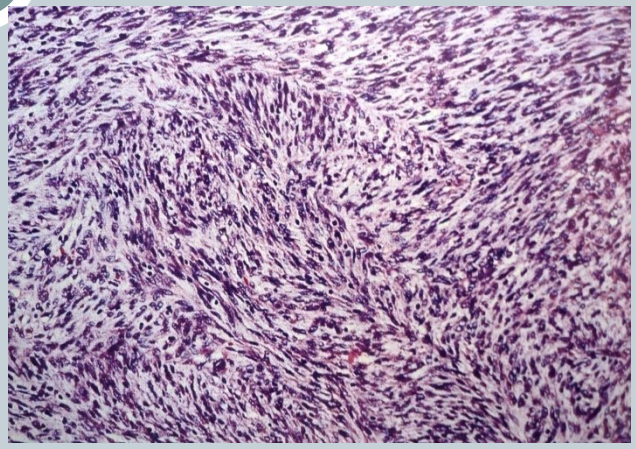
- Heeringbone pattern- showing parallel sheets of cells arranged in intertwining whorls

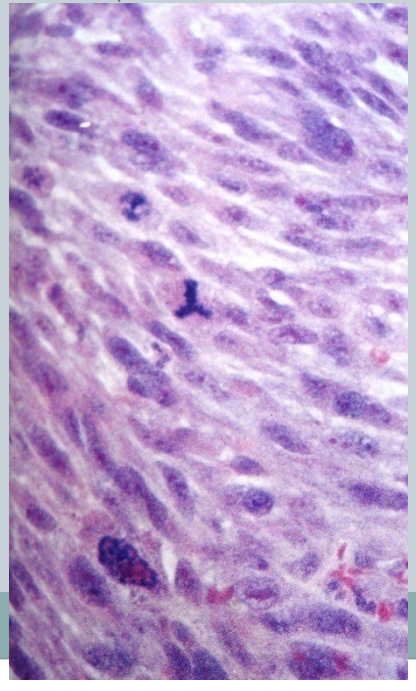


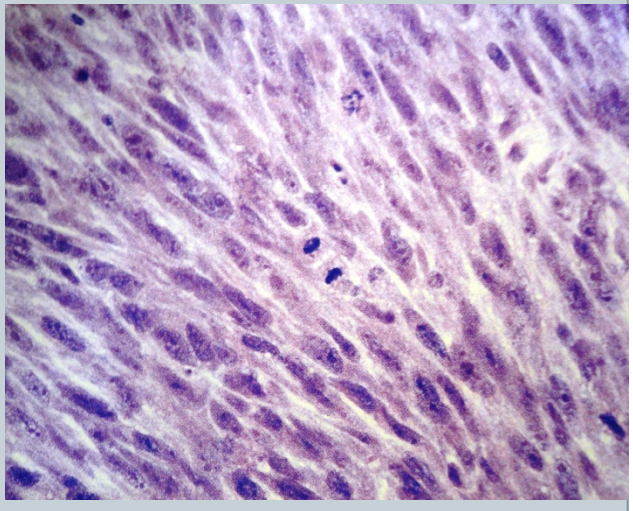
High-grade FS

Fibrosarcoma

- Very cellular, cells are less organized
- Marked cellular atypia and mitotic activity
- Collagen matrix - less







Treatment

Fibrosarcoma

- Treatment: Radical Surgery
- Chemotherapy –bone lesions
- Radiotherapy- soft tissue lesions
- Prognosis -poor

Miscellaneous Locally Aggressive Fibrous Lesions

- Fibrous histiocytoma
- Nodular fasciitis
- Aggressive fibromatosis
- Proliferative myositis
- Atypical fibroxanthoma
- Desmoplastic fibroma of bone

Fibrous Histiocytoma

Fibrous histiocytoma

- Benign but diverse group of neoplasms characterized by fibroblastic and histiocytic differentiation
- Cell of Origin: histiocytes

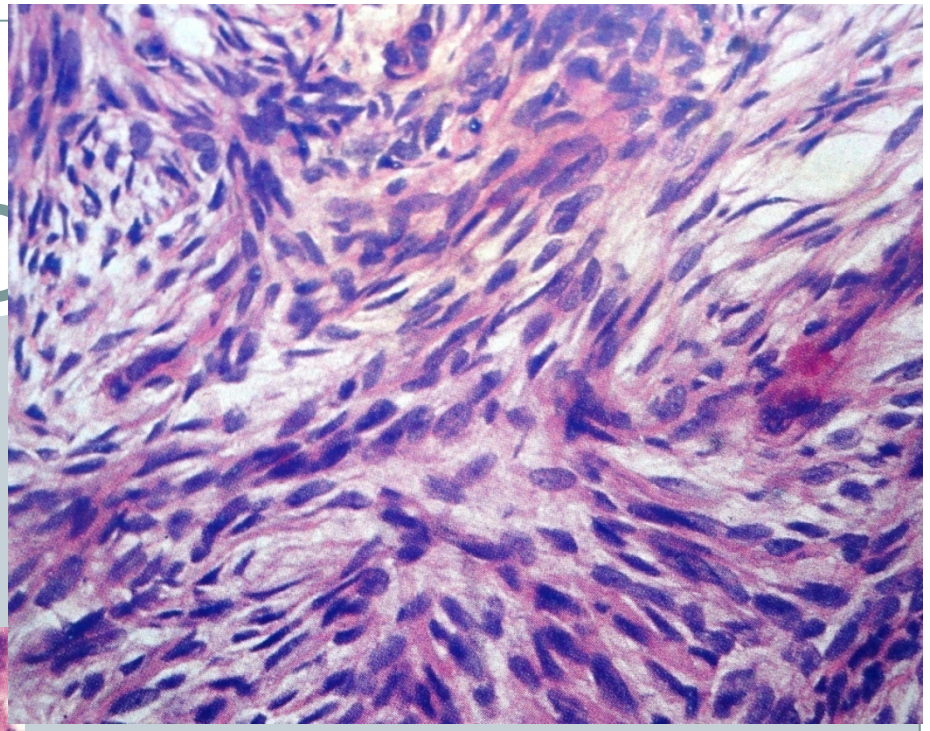
Malignant Fibrous Histiocytoma

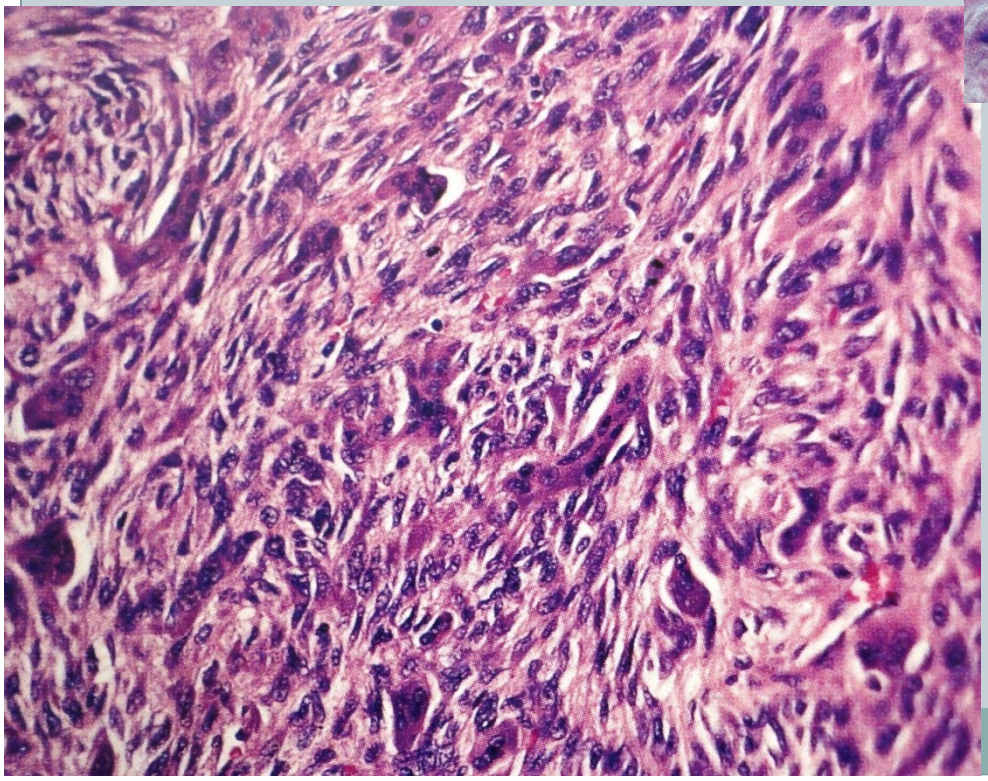
Clinical Features

- Age –Usually older age
- Sex - M>F
- Site – E/o – Skin of extremities,
  Anywhere in H & N region.
  I/o – very rare, buccal mucosa & vestibule
- S/S – Submucosal expanding mass with or without pain & surface ulceration

Histopathologic Features

- Spindle-shaped fibroblasts like cells
- Scattered rounded histiocytic cells
- Touton-type multi-nucleated giant cells
- Spindle-shaped fibroblasts may arranged randomly or streaming in interlacing fascicles,imparting *storiform* pattern





- Liposarcoma - are extremely rare malignant tumors of the adipose tissue

- Hemangiopericytoma and hemangioendothelioma are malignant tumors arising from the components of the blood vessels

Kaposi's Sarcoma

- Also c/as multiple idiopathic hemorrhagic sarcoma of *Kaposi*
- Malignant neoplasm arising from the endothelial cells of the blood vessel
- First described by Moritz Kaposi in 1872

Kaposi's Sarcoma

- *Etiology:* Unknown
  - Immunosuppression – believed to create an environment that allows an opportunistic factor to cause Kaposi's SA
  - HHV 8 or KSHV
  - Reactive lesions?

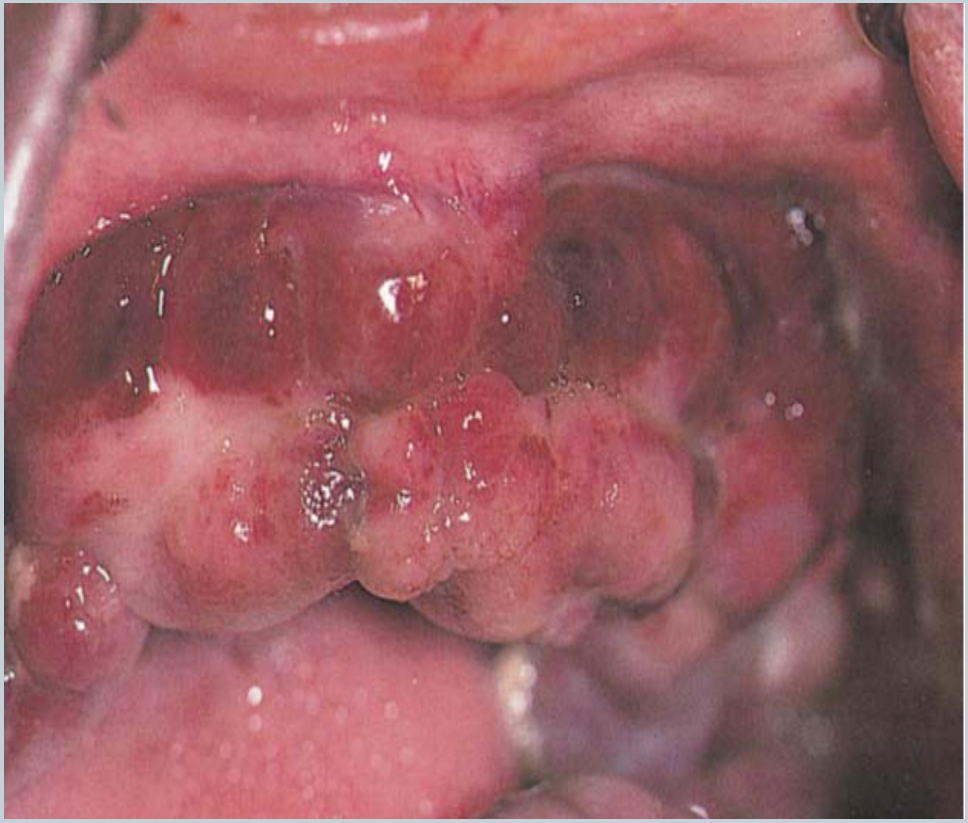
Clinical Features

1. Classic – Cutaneous multifocal blue-red nodule, slowly increase in size and numbers
   oral lesions-unusual, palate & gingiva
1. Lymphadenopathy – endemic to African Children, localized or generalized enlargement of Lymph nodes

C/F

3. Immunosuppression-associated (transplant) - renal transplant patients
4. AIDS-related Kaposi's sarcoma
   - early sign of disease -
   Early oral mucosal SAs are flat and slightly blue, red or purple plaques

Cervical LN and S.G. – enlarged



Clinical stages:

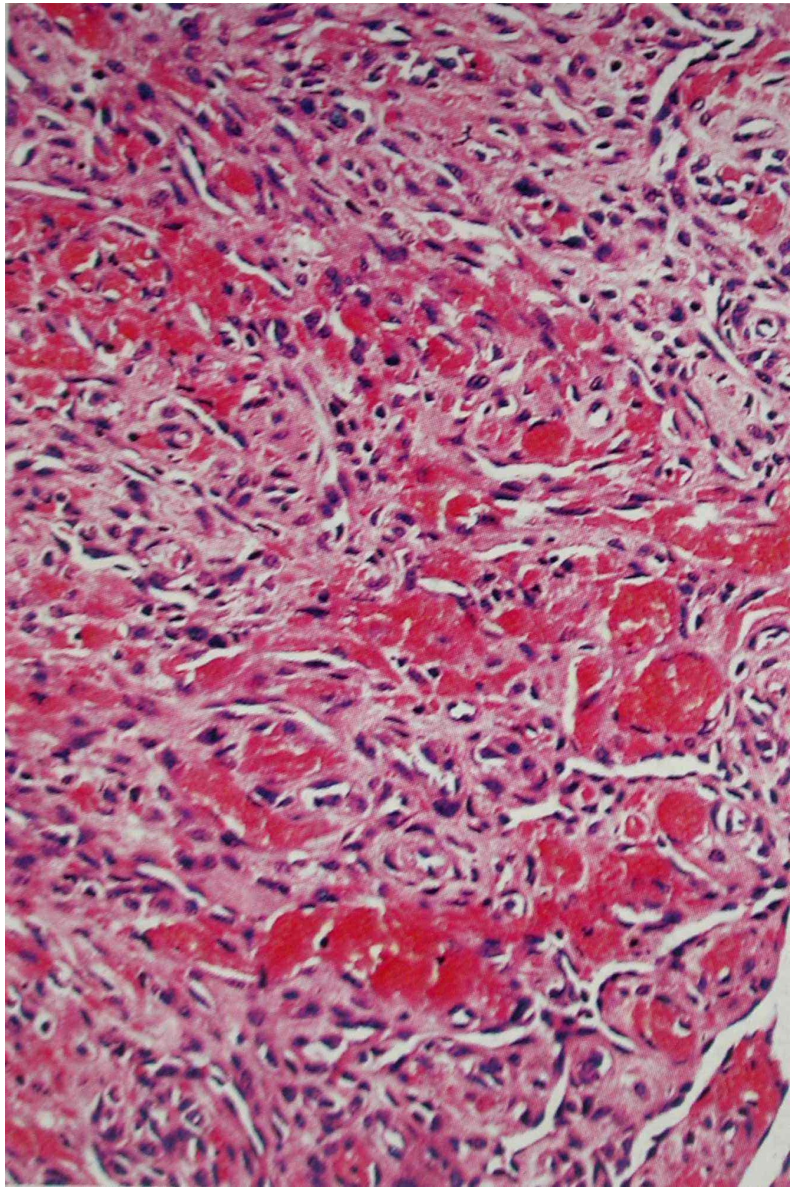
(i) Patch stage

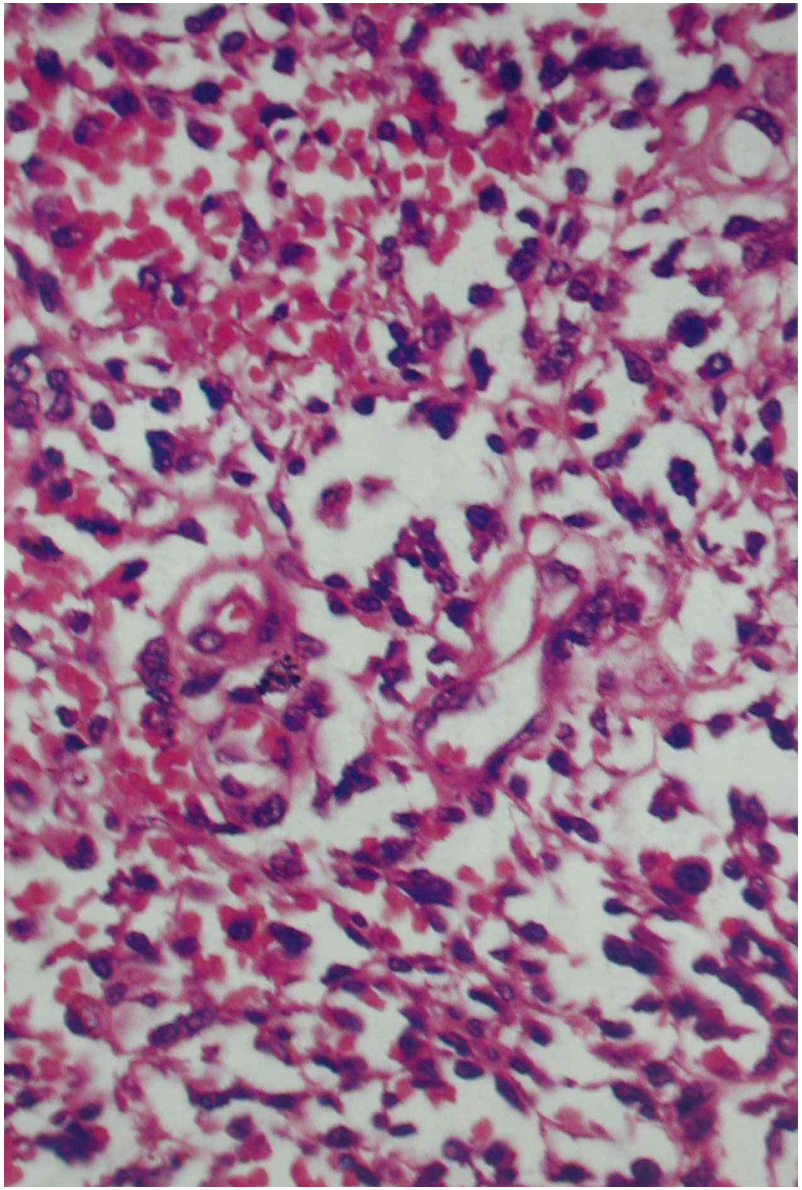
(ii) Plaque stage

(iii) Nodular stage

H/P

- *Patch stage* - initial stage
  Microscopically - dilated, irregular blood vessels, lined by normal appearing endothelial cells.
- *Plaque stage* - ♿ in numbers of small capillaries or dilated vascular channels interspersed with proliferating sheets of sarcomatous or atypical spindle cells, RBCs
- Nodular Stage: all features are more prominent





Summary

1. Difference between sarcoma and carcinoma
2. Classification, incidence, etiology, clinical features & histopathological features of **fibrosarcoma**
3. Classification, incidence, etiology, clinical features & histopathological features of **locally aggressive fibrous lesion**
4. Classification, incidence, etiology, clinical features & histopathological features of **Kaposi's sarcoma**

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Thank You