CONNECTIVE TISSUE TUMORS

MESENCHYMAL TUMORS

- Occur throughout the body where connective tissue is found
 - Form a heterogeneous group that arise from fibrocytes, fibroblasts & specialized mesenchymal cells e.g. fat, muscle
 - May be benign or malignant

WHO CLASSIFICATION

- Fibrous tumor
 - Fibrohistiocytic
- Lipomatous
 - Smooth muscle
 - Skeletal muscle
 - Tumors of blood & lymph vessels
 - Perivascular tumors
 - Synovial
- Mesothelial
- Nasa/tumors
- Paraganglionic tumors
- Ektra-skeletal cartilaginous & osseous
- Plur potent mesenchymal tumors
- Miscellaneous
- Unclassified

CONT.

- Mesenchymal tumors form sheets of cells unlike epithelial cells that form nests or groups of cells
- They form a matrix and have thin walled blood vessels.

MESENCHYMAL VS EPITHELIAL MALIGNANCIES

MESENCHYMAL MALIGNANCIES

EPITHELIAL MALIGNANCIES

- •Sheets of cell
- •Thin walled vascular channels scattered throughout
- •Forms matrix
- •Confusing variety of histologic types stimulating tissue type often corresponding to the grade of the tumor.

- Nests, groups, cords, islands
- •Well-formed vessels supplying the tumor (desmoplastic response)
- •No matrix supported by nonneoplastic stroma
- Mainly squamous glandular, transitional types

CONT.

Malignant mesenchymal tumors are called sarcomas

Benign:

- Lipoma
- Fibroma
- Osteoma
- Chondroma
- Rhabdomyoma
- Hemangioma
- Lymphangioma
- Histiocytoma

Malignant:

- Liposarcoma
- Fibrosarcoma
- Østeosarcoma
- Chondrosarcoma
- Rhabdomyosarcoma
- Hemangiosarcoma
- Lymphangiosarcoma
- Malignant fibrous histiocytoma (MFH)

BENIG MESENCHYMAL TUMORS

- Usually form rounded masses with a near normal appearance
- Sharply defined by thin fibrous capsule formed by condensation of the stroma of surrounding tissue
 - Blood supply is through well formed vessels

LIPOMA

- Benign tumor appearing in the subcutaneous tissue e.g. neck, trunk, face, hands & feet. Sometimes in deeper structures i.e. retroperitoneal, mediastinum, skeletal muscle and GIT.
 - Usually a soft, lobulated mass of fatty tissue 3-5 cm, enclosed by a thin delicate capsule disrupted during surgical excision
- Histology show normal adipose tissue with cells that may be larger and more variable in size.

LEIOMYOMA

- Commonest benign soft tissue tumor seen in the myometrium of up to 20% of women, often multiple
- Spherical sharply demarcated from surrounding myometrium, without fibrous capsule, cut surface is solid with pale grey or pink surface.
- Histology shows smooth muscle lying parallel to one another in interlacing bundles
- Cells are uniform and mitoses are few; well developed blood vessels accompanied by fibrous tissue are distributed through out the tumor
- Tumor cells also respond to sex hormones.
- They develop between puberty and menopause
- Usually symptomless but hen large distort the uterine cavity and cause menorrhagia, abortion, interfere with child birth
- They may also arise in the dermis, GIT and walls of arteries

FIBROMA

- Uncommon, seen in ovaries and GIT
- Rubbery, grey discrete encapsulated masses, cut surface is glistening
 & greyish white
 - Histology shows mature fibrocytes or fibroblasts with no distinct orientation
- Small fibro-fatty nodules are common in the skin and may become peduncalated and are called **skin tags** or fibrous polyps
- Other fibrous proliferation with distinct characteristics are of unknown causes ad present features intermediate between reactive fibrosis and neoplastic proliferations; most are self-limiting i.e. keloids, nodular fasciitis, fibromatoses, desmoid tumors.

CHONDROMA

- Fairly common in small bones of the hands and feet and are rare in soft tissues
- Forms are rounded or ovoid and are encapsulate; they are usually lobulated
 - Resemble hyaline cartilage as they produce chondroid matrix

RHABDOMYOMA

Rare tumor seen in those over 40 years in the tongue, heart & neck

Tumors of striated muscle

SARCOMAS

- They are less common than carcinomas causing only 3% of all cancer deaths
- They show all features of malignancy i.e. rapid growth rate (generally)
 - Frequent sometimes abnormal mitoses
- Various degrees of atypia and anaplasia
- Locally invasive
- Metastases
- They vary in degree of malignancy in that some grow slowly over many years while others enlarge rapidly and metastasise early
- High grade tumors are highly aggressive
- Vascular with many small and medium sized vessels that are poorly formed, have endothelial cells with incomplete basement membrane hence bleeds easily. Cut surface shows hemorrhage & infarction
- Tumor penetrates vessels to form emboli that spread to the lung, liver & bones

CONT.

- Spread through lymphatics occurs less frequently and usually later than carcinomas. Local lymph nodes are free of tumor except in a few rhabdomyosarcoma.
 - They are often coarsely nodular and may be partially capsulated.

LIPOSARCOMA

- Occurs often in the deep tissues of thigh and retro-peritoneum, usually after the age of 30 years
- Spreads extensively by local invasion
 - Gross appearance depends on the degree of differentiation i.e. well differentiated appear fatty, myxoid ones are gelatinous and those with collagen are firm. Varied histologic types.

LEIOMYOSARCOMA

- Occurs in myometrium, GIT, Deep soft tissue
- Rarely may arise in a fibroid (leiomyoma)
- Histology show elongated cells, nuclei have rounded or blunt ends, have numerous mitoses, pink cytoplasm

FIBROHISTIOCYTIC TUMORS

- Fibrous histiocytoma (dermatofibroma)
- Affects 2024 year, skin in the sub-cutaneous tissue especially of the extremities
 - Presents as a nodule that shows fibroblasts and fibrocytes
 - MFH was one of the commonest sarcoma seen in deep tissues of the limbs and retro-peritoneum
 - Shows pleomorphic cells with storiform (cartwheel) appearance
 - Other histologic type include myxoid, inflammatory & angiomatoid.
- TH tumors of borderline malignancy e.g. dermato-fibrosarcoma protuberance. Locally aggressive and rarely metastasizes.

FIBROSARCOMA

- nitially was the commonest sarcoma but separation of nodular fasciitis, fibromatosis and fibrohistiocytic tumors has reduced incidence to 15% of all sarcomas
- Disease of adults, peak in 30-55 years.
- Arises in deep fasciae ad aponeuroses most often in the thigh or around the knee, rarely in the trunk, forearm or leg
- Presents as a deep seated often painless swelling which grows slowly and may reach the surface where necrosis, ulceration & infection are liable.
- It may also arise in peri-osteum or medulla of bones
- May appear as rounded or nodular masses of pale, grey or yellowish white fleshy or firm depending on the amount of collagen.
- Margins may be deceptively well defined, may have out lying masses that may be detached
- Poorly differentiated are more cellular and therefore soft often with hemorrhage and necrosis.

RHABDOMYOSARCOMA

- Rare, highly malignant tumors with four distinct histological and clinical types
- Embryonal rhabdomyosarcoma
- Seen in children, involves genital tract and presets as a bunch of grapes hence called sarcoma botyrides
- Shows undifferentiated small round or spindle cells with hyper-chromatic nuclei and loose myxoid stroma
- Spindle/cell rhabdomyosarcoma a type of alveolar rhabdomyosarcoma
- Alveolar rhabdomyosarcoma
 - Adolescent 10-25 years on limbs and trunk
 - Packet of cells forming alveolar pattern. Occasional giant cells and striated cell may be seen
- Pleomorphic: adult type found on limb. Cells are large and pleomorphic with cross striations, racket cells, spider web cells (giant cells with vacuoles

CHONDROSARCOMAS

- Occur more commonly in bones in a patient of 40-70 ears
- Half occur in the pelvic girdle
 - Male more affected than females
 - Well differentiated kinds ma be confused with chondromas.

OSTEOSARCOMA

- More common in the bones
- Similar histology to those in bone
- Sarcomas that form bone & osteoid (un-mineralized bone) matrix
 - Highly malignant tumors.

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