Myxo-inflammatory Fibroblastic sarcoma

Å AKA

- Acral Myxoinflammatory fibroblastic sarcoma Am.J.Surg.Path 1998; 22; 911-924
- Inflammatory myxoid tumour of soft parts with bizarre giant cells [Pathol.Res.Pract. 1998; 194; 529-533]
- Inflammatory myxohyaline tumour of the distal extremities with virocyte or Reed-Sternberg-like cells [Mod. Path.1998; 11; 384-391]
Myxo-inflammatory Fibroblastic sarcoma

Å M=F, peak middle aged adults
Å 2/3rds hands & wrists, then feet and ankles, but can be more proximal
Å S/c with extension into dermis and underlying tendons
Myxo-inflammatory Fibroblastic sarcoma [MIFS]

- Lobulated tumour composed of varying myxoid and hyaline zones
- Dense inflammation which obscures tumour. Lymphocytes and aggregates of macrophages.
- Cells vary from Multivacuolated lipoblast like, Atypical spindle cells, Large ganglion/Reed Sternberg like cells with large nucleoli.
- CD34 often positive. Nil else
Differential Diagnosis

- PVNS, but lacks atypia
- Hodgkin’s disease
Behaviour

Å Frequent local recurrence, but mets very rare [2%]
Hybrid myxoinflammatory fibroblastic sarcoma/hemosiderotic fibrolipomatous tumor: report of a case providing further evidence for a pathogenetic link.


Consistent t(1;10) with rearrangements of TGFBR3 and MGEA5 in both myxoinflammatory fibroblastic sarcoma and hemosiderotic fibrolipomatous tumor.

Further instructive case
Main Features

Å Infiltrative tumour.
Å Prominent vessels, ectatic with rim thick amorphous material [fibrin] +/- fibrin thrombi. Vessels in clusters.
Å Plump spindle cell proliferation, in sheets and fascicles. Nuclei are hyperchromatic and pleomorphic. Can have intra nuclear inclusions.
Å Mitoses rare.
Å Iron deposition
Diagnosis

Ä Pleomorphic Hyalinising Angiectatic tumour [PHAT].
Clinical Features

• Adults, median age 51, M=F
• Foot/ ankle and lower leg most common site
• Usually subcutaneous.
Conclusion

Á HFLL/HFLT and Myxoinflammatory tumour are related

Á Pleomorphic Hyalinising Angiectatic [PHAT]Tumour; Position not as clear but examples of hybrid HFLT and PHAT described.
HFL - LUMPS

MIFS

HFL

PHAT
40M with cyst on finger
Histological findings

- Multinodular deep dermal tumour on distal extremities.
- Nodules composed of back-back glands with central cystic space.
- Broad papillae composed of heaped up cells are present.
- Mitoses are frequent. Often only mild nuclear atypia.
- Necrosis often present.
- Border can be infiltrative or circumscribed.
- No distinct immunoprofile
Digital Papillary Carcinoma

- Occurs on extremities, particularly fingers and toes.
- Mid-age adults with cyst / lump +/- painful.
- Frequent local recurrence unless radical excision eg amputation.
- Metastasizes in 15%
Problem areas

Benign versus malignant

- At lower end of spectrum, originally called adenoma and carcinoma based on histological features. Longer follow up shown all can behave in malignant fashion, therefore all called malignant in WHO classification.
Differential diagnosis

- Mimic a metastasis from visceral site.
- Papillary eccrine adenoma
  - Circumscribed.
  - Cysts contain delicate papillae with 2 cell layer.
  - Lacks back to back glands/ mitoses and heaped up cells forming papillae.
Cutaneous Digital Papillary Adenocarcinoma

A Clinicopathologic Study of 31 Cases of a Rare Neoplasm With New Observations

Ravi Suchak, MBChB, MSc, MRCP, MRCGP, Dip RCPath,* Wei-Lien Wang, MD,† Victor G. Prieto, MD, PhD,‡ Doina Ivan, MD,† Alexander J. Lazar, MD, PhD,† Thomas Brenn, MD, PhD, FRCPath,‡§ and Eduardo Calonje, MD, Dip RCPath*

Distinctive feature is the myoepithelial cells surrounding glands. Not seen in mets or Hidradenoma
Smooth muscle Myosin IHC.
60 Male with several warty growths, ?SCC
EPIDERMODYPLASIA VERRUCIFORMIS

- Autosomal recessive genodermatosis
- Increased susceptibility to specific types of HPV infection, but not other infections.
- Genes involved have been identified as EVER1/TMC6 and EVER2/TMC8
EPIDERMODYPLASIA VERRUCIFORMIS

- 2 main forms
- HPV3 or 10 - multiple plane warts. No malignant progression
- HPV5 or 8 - Plane warts plus seborrhoeic keratosis like lesions. Risk of progression to Bowen’s and SCC.
EPIDERMODYSPLASIA VERRUCIFORMIS

- Characteristic changes in epidermis.
- Large cells in spinous and granular layer with blue-grey cytoplasm.
- +/- other features of wart virus, perinuclear halo, keratohyaline granules.
37 year old female
6 year history of painful dystrophic right middle finger nail
Hyperkeratosis

Abnormal lunula

Nodule arising from nail bed

Hyperkeratosis

Abnormal lunula