MYC gene

- Myelocytomatosis viral oncogene homolog
- Regulatory gene involved in cell proliferation and differentiation.
- MYC involved in many cancers including breast cancer.
MYC

- Very good correlation with FISH and IHC for MYC
- Remember has to be in correct setting to be diagnostically useful.
75 M Tumour on scalp
AFX VS PDS

Atypical FibroXanthoma [AFX] - considered benign

Pleomorphic Dermal Sarcoma [PDS] - malignant sarcoma
Pleomorphic Dermal Sarcoma

Adverse Histologic Features Predict Aggressive Behavior and Allow Distinction From Atypical Fibroxanthoma

Keith Miller, FRCPath,* John R. Goodlad, MD, FRCPath,† and Thomas Brenn, MD, PhD, FRCPath†

AFX & PDS

- Same clinical picture;
- Elderly, sun damaged sites, esp. head & neck
- Raised cutaneous nodule
AFX & PDS

Å Same cytology-
Å Tumour cell; high grade atypia with pleomorphic cell, atypical xanthomatous cells and spindle cells in varying proportion
AFX & PDS

Å Diagnosis of exclusion; rule out melanoma, carcinoma.
Å Immuno. Panel; Broad spectrum cytokeratins [MNF116 + AE1/AE3], S100, Desmin, CD34.
Å CD10? Useful- No
AFX VS PDS

- Architecture is key - AFX if
  - Circumscribed with pushing margin Not infiltrative
  - Minimal subcutaneous fat extension
  - No vascular invasion
  - No Tumour necrosis
  - No Perineural invasion

- If positive for any of the above - PDS
AFX Rare variants

- Osteoclastic giant cell rich
- Granular cell
- Clear cell
- Spindle cell
- Osteoid/chondroid
PDS

- 32 cases, M > F, sun damaged site.
- Immuno:- Negative for broad spectrum cytokeratins, s100, HMB45, Desmin, CD34
- Odd immuno findings- SMA 70%
- CD31 -48% positive
- EMA can be positive
- Melan A- 6%
- Similar findings in AFX
CYTOKERATINS

Å AE1/AE3 – broadest reactivity
  ï AE3- CK 1-8
  ï AE1 – CK 10,13,14,15,16,19
Å MNF116 – CK 5,6,8,17,19
Å 34 Beta E12 – CK 1,5,10,14 + other unknown.
PDS

• Follow up of 29 patients.
• 28% local recurrence [often incompletely excised].
• 10% metastasize [to skin].
• No disease related mortality
• High grade on cytology, but low grade behaviour.
AFX vs PDS

- One disease process, separating early stage tumour [AFX] from late stage ones [PDS].
- Baby tiger VS adult tiger
- Cell of origin; unknown, but hotly debated.
UNCERTAIN HISTOGENESIS
30 F lump on back
Clear Cell Tumour

Å Epithelial origin
  • SCC/ BCC
  • Adnexal –Porocarcinoma, Sebaceous, Trichilemmal
  • Metastasis - RCC met.

Å Melanocytic
  • Balloon Cell Melanoma

Å Soft tissue
  • Clear cell sarcoma- usually deeper
  • Clear cell dermatofibroma
  • PEComa
  • Distinctive dermal clear cell mesenchymal neoplasm
  • Clear cell AFX
PEComa
Perivascular epithelioid neoplasm

Å Angiomyolipoma
Å Clear cell tumour of the lung ‘sugar’ tumour
Å Lymphangioleiomyomatosis
Å Clear cell Myomelanocytic tumour
PEComa

- Nested tumour
- Prominent vaculature of thin walled capillaries
- Epithelioid +/− spindle cells
- Clear to granular eosinophilic cytoplasm
- Occassional giant cells.
- No pleomorphism
- Immuno; HMB45 positive, MitF Positive, MelanA often positive, Desmin often positive, SMA occasionally, s100 rare focal positivity.
Cutaneous PEComa

- Rare
- F>M, wide age range, often on limbs but anywhere.
- Small tumours < 2cm.
- Dermal based +/- s/c involvement

Cutaneous PEComa

- Majority benign. Very rare malignant cases in the skin.
- No association with Tuberous sclerosis.
- Probably different genetic pathway to visceral cases.
PEComa

- No normal counterpart
- Cells intimately associated with blood vessels
- Dual differentiation to smooth muscle and melanocytes.
DIFFERENTIATION IN PECOMA

- Smooth Muscle
- Fat
- Clear Cell

Blood Vessel