Lymphoid cells, plasma cells form a rim around solid histiocyctic areas, Blood.

Vasculature
Sheets of histocytic cells, bordered by a lymphoplasmaclastic rim
Solid, serpiginous, multinodular areas of histolytic cells
Central rounded nuclei
Monotonous
Immunohistochemistry

❖ CD68: positive
❖ CD99: positive, focal
❖ Desmin: negative (positive in half of AFH)
❖ SMA: negative
❖ S100: Negative
❖ EMA: weak blush
❖ ERG: negative
Angiomatoid Fibrous Histiocytoma

- Subcutaneous lesion
- Nodular proliferation of epithelioid and spindled cells with pseudovascular spaces
- Peak incidence first 2 decades
- Occurs where nodes are usually found (antecubital fossa, popliteal fossa, axilla, inguinal area, neck), limbs, trunk, H&N
- Systemic symptoms: anaemia, weight loss, fever
- Size: 2cm, circumscribed mass, simulates a node, haemorrhagic
- **4 key histologic features** 1) nodules of spindled or histiocytoid cells, syncytial growth 2) pseudongiomatous spaces filled with blood and surrounded by tumour cells, 3) thick fibrous psueudocapsule, 4) cuff of lymphoplasmacytic cells, occasional germinal centres
- A subset dominated by small cells with hyperchromatic nuclei
- IHC: Desmin, EMA, CD68, CD99
- t(2;22) EWSR1-CREB1 fusion in 90% (same as clear cell sarcoma)
- 2-10% local recurrence, <1% metastasis to regional nodes. No predictors of outcome
66 year old woman. Painful blotchy rash chest wall. Status post WLE & XRT for breast cancer 12 years ago

Case 8
Angiectatic vessels. Lymphoedema. Low grade cells
❖ Expanded Lymphatics. Lined by 1-2 layer of endothelial cells. Tombstone protrusions/hobnailing into lumen.
Simple vessels and solid areas
Punch biopsy - Clock face. Only 1 involved.

Epidermal hyperplasia, sclerosis, multi nodular areas of a cellular infiltrate.
Low grade cells. Solid & trabecular growth pattern, suggestion of collapsed lumina.
Differential Diagnosis

- Vascular: AVL, Haemangioendothelioma, Angiosarcoma (primary cutaneous, secondary, mammary parenchymal), Kaposi’s
- Epithelioid sarcoma
- Carcinoma (recurrent breast, other)
- Naevomelanocytic
- Fibrohistiocytic (DFSP?)
Additional Investigations

- Immunohistochemistry
- ERG, CD31, CD34: Positive, diffuse
- HHV8: Neg
- AE1/3: Neg
- INI-1: Neg
- S100: patchy, cytoplasmic
- HMB45, Sox 10, Melan A: Neg
- GATA3, ER, PR: Neg
- P53: weak blush
- CMYC IHC and FISH: Neg
Resection

❖ WLE scar. Diffuse area of blotchy induration in LLQ, extending to inferior margin
Diffuse expansion of the dermis by a firm, congested, pale tumour.
Final Diagnosis: Cutaneous Angiosarcoma, Post Irradiation

❖ Infiltrating vessels, atypical cytology
Atypical Vascular Lesion

- Crops of small (<1cm), pink cutaneous papules on the skin
- Context: post XRT (many for mammary carcinoma)
- Histo: circumscribed dermal proliferations of simple vascular channels. 1) Lymphatic type: may resemble lymphangioma circumsictum- ectatic lymphatics within superficial dermis. May show extension and infiltration into the dermis. Minimal atypia or multilayering. 2) vascular type: uncommon, resemble capillary haemangioma. Blood filled capillaries, haemosiderin.
- Local recurrence: 10-20% develop further similar lesions
- Lack C-MYC amplification (DDx AS)
- Risk of progression to AS poorly defined, appears rather low, particularly for the more common lymphatic type AVL
- Mgx: lymphatic type AVL: diagnostic biopsy. VT-AVL: complete excision. If atypia: excision and follow up advised as may be a precursor to AS
Retiform Haemangioendothelioma

- Uncommon, skin and subcutaneous tissues, most <5cm, extremities
- Most *de novo*, rare association with lymphedema, XRT
- Local recurrence rate 60%. No distant metastases reported
- Is a component of composite haemangioendothelioma (together with more solid, low grade angiosarcoma like areas)
Angiosarcoma, Cutaneous, Post Irradiation

- Increasing problem due to breast conservative surgery and adjuvant XRT for breast cancer, 5-14 per 1000 patients. Short latency, most within 5 yrs.

- Vasoformative tumour, multifocal, centred in the dermis, may encroach breast parenchyma. Anastomosing vessels, dissecting the stromal planes, atypia, solid areas. May be epithelioid. 30% CK pos.

- MYC amplification found in 56-100% angiosarcoma secondary to irradiation or lymphedema. Not found in AVLs or primary or deep soft tissue AS. Associated with P53 abnormalities.
  - FLT4 (VEGFR3) co-amplification in 25% secondary AS.
  - VEGF1 (KDR) mutations found in 10%. Particularly in breast and primary.

- Grading may not predict outcome: I: single endothelial layer & minimal atypia, II: multilayering and atypia, III: sheets of cells, high grade nuclei, blood lakes.

- Survival: 5 year overall survival 61%, correlated with size 5cm but not many other factors.