THE FAMILY OF VASCULAR TUMOURS WITH EPITHELIOID ENDOTHELIAL CELLS

- Epithelioid haemangioma (angiolympoid hyperplasia with eosinophilia)
- Epithelioid angiomatous nodule
- Epithelioid haemangioendothelioma
- Epithelioid angiosarcoma

KIMURA DISEASE

- Not the same as ALHE (epithelioid haemangioma)
- Young oriental males
- Deep seated lesions
- Lymphadenopathy
- Eosinophilia
- Rare nephrotic syndrome
- ?Infectious aetiology

EPITHELIOID HAEMANGIOMA (ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA)

- Not the same as Kimura disease
- Relatively rare
- Wide age range (mainly between ages of 20 and 50)
- Can occur in many organs (including bone)
- F>M
- Papule/nodule or multiple lesions
- Recurrence common

Hobnail endothelial cells are not the same as epithelioid endothelial cells
EPITHELIOID HAEMANGIOMA ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA
EPITHELIOID HAEMANGIOMA (ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA)

- Lobular architecture
- Proliferation of vascular channels lined by epithelioid endothelial cells (abundant pink cytoplasm)
- Frequent intracytoplasmic lumina
- Inflammation ++ with lymphocytes and numerous eosinophils
- Germinal centres rare
- Fibrosis in late lesions

EPITHELIOID HAEMANGIOMA (ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA)

- Immunohistochemistry:
  - CD3 and ERG ++
  - EMA and keratins usually negative

- Cytogenetics:
  - Although FOS gene rearrangements have been found in epithelioid haemangioma of bone, they are usually absent in cutaneous lesions. However, immunohistochemistry for FOSB (as in pseudomyogenic haemangioendothelioma) is positive and this is therefore useful in differential diagnosis.
Intravascular epithelioid haemangioma

- **16** PATIENTS
  - 12 M, 4 F
  - 11-71 YRS (MEAN 40.2 YRS)
  - SIZE FROM 2-30 MM (MEAN 13 MM)
- **21** LESIONS
  - 13 SOLITARY
  - 3 MULTIPLE (OF THEM 2 IN THE SAME AREA - DISTAL EXTREMITIES)
- **SITE**
  - EXTREMITIES (13/21 - 62%)
  - HEAD AND NECK (8/21 - 38%)
- **FOLLOW-UP (MEAN 27 MONTHS)**
  - NO LOCAL RECURRENCES

- Wide age and anatomical distribution (mainly trunk and limbs)
- No sex predilection
- Mainly solitary but rare multiple or eruptive lesions may be seen
- Small papule, superficial and mostly less than 10 mm
- No tendency for local recurrence
- Benign, part of the spectrum of epithelioid haemangioma

- Non-ulcerated
- Polypoid with epithelial collarette
- Well-circumscribed
- Sheets of epithelioid endothelial cells
- Regular with a single nucleolus and vesicular nuclei
- May be mitotic
- Little tendency to form vascular channels
EPITHELIOID HAEMANGIOENDOTHELIOMA

CLINICAL FEATURES
- Middle-aged adults
- M = F
- Extremities > trunk > head/neck
- Mainly soft tissues
- Often multicentric (bone, viscerae) in up to 50%
- 20% metastatic rate
- 15% mortality
- Small primary cutaneous lesions (good prognosis)

EPITHELIOID HAEMANGIOENDOTHELIOMA

HISTOLOGICAL FEATURES
- Frequent angiocentricity
- Variable cellularity
- Myxoid or hyalinized stroma
- Strands or nests
- Intracytoplasmic vacuoles
- Variable mitotic rate / cytological atypia
EPITHELIOID HAEMANGIOENDOTHELIOMA
IMMUNOHISTOCHEMISTRY AND CYTOGENETICS

- Positive for CD31, ERG, FLI-1 and D2-40
- 20% positive for keratin and EMA
- WWTR1–CAMTA1 fusion in up to 90% (CAMT1 immunohistochemistry useful in differential diagnosis as tumours positive in up to 86%)
- Rarely YAP1–TFE3 fusions

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Primary cutaneous epithelioid angiosarcoma: a clinicopathologic study of 13 cases of a rare neoplasm occurring outside the setting of conventional angiosarcomas and with predilection for the limbs.

Romero MF, Thome A, Stein S, Rotten G, Ges虇 S

Abstract

Epithelioid angiosarcomas are rare aggressive neoplasms that occur most frequently in deep soft tissues. Primary cutaneous lesions are rare, and there are discordant findings in the literature regarding their behaviour. In this study, we report a series of 13 cases of primary cutaneous epithelioid angiosarcoma and analyze their clinicopathological features. The tumours arose in the conventional settings for conventional angiosarcomas (i.e., in the head and neck region of elderly patients, and these occurring presentation of an association with previous radiotherapy). The tumours were composed of epithelioid or spindled cells, arranged in solid, nested, or trabecular patterns, and measured on average 6.8 cm in diameter. Immunohistochemically, the tumours were positive for CD31, ERG, FLI-1 and D2-40, and negative for CD34, CD117, and DOG1. The ratio of Ki-67 was 20%, and tumours were positive for a variety of markers, including EMA, keratin, and vimentin. The tumours were negative for S-100, desmin, and smooth muscle actin. Follow-up information was available for 11 patients. Six patients died of metastatic disease after a median follow-up of 12 months (range, 3.1–66 months), and 1 patient died of unrelated causes. These findings suggest that primary cutaneous epithelioid angiosarcoma occurring outside the conventional settings of angiosarcoma is a highly aggressive malignant tumour with mortality rates in excess of 50% after 3 years.
EPITHELIOID ANGIOSARCOMA

CLINICAL FEATURES

- Rare
- Mainly in deep soft tissues
- Primary lesions can occur in many organs
- Middle to old age
- M > F
- Haemorrhagic papules / nodules
- Often aggressive

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EPITHELIOID ANGIOSARCOMA

HISTOLOGICAL FEATURES

- Infiltrative and diffuse
- Dermal and often subcutaneous
- Sheets of large atypical epithelioid endothelial cells with abundant pink cytoplasm, vesicular or hyperchromatic nuclei and a single prominent nucleolus
- Frequent mitotic figures/variable necrosis
- Intracytoplasmic lumina with or without red blood cells frequent
- Blood vessel formation rare
- May be intravascular (in such cases rule-out origin from a large blood vessel with embolization)

Immunohistochemical profile: CD31, FLI-1 and ERG positive. In up to 40% or more at least focal positivity for pan-keratin and also for EMA, CD34 variable positivity