Desmoplastic Spitz Naevus

DDx: Desmoplastic melanoma

Reassuring features:
- Lesional circumscription
- Lack of infiltrative growth into subcutis
- Epithelioid melanocytes with Spitzoid features
Deep-penetrating Naevus

- Wide age range; adolescence and early adulthood
- Face, neck, extremities
- Darkly pigmented to grayish symmetrical papule/nodule <1cm
- Closely related to Spitz Naevi
- Mutations in HRAS, but not GNAQ, GNA11
Deep Penetrating Naevus
-Histological Features-

Concerning features:

- Melanocyte atypia
- Lack of maturation
- Scattered mitotic activity
- Deep pigmentation
- Growth along neurovascular bundles
- Extension into subcutis
Deep Penetrating Naevus
-Histological Features-

BUT:

- Circumscription
- Wedge-shaped architecture
- No severe or confluent atypia
- No atypical mitoses
- No expansile growth
- No necrosis
Clonal/inverted type-A Naevus

-Introduction-

Naevus with focal atypical epithelioid component

- Closely related to DPN
- Similar clinical setting
BAPoma

Familial or sporadic

Tumour suppressor gene mutations in BRCA1-associated protein-1 (BAP1) on chromosome 3q21

Autosomal-dominant

Multiple melanocytic tumours with epithelioid cell change

Uveal and cutaneous melanoma

Mesothelioma

Renal cell carcinoma

M, 28 Jahre, Gesicht
Spitz Naevus with Kinase Fusion

Atypical Spitz Tumour/Spitzoid Melanoma

• Poorly defined criteria with poor interobserver agreement

• Constellation of features
Atypical Spitz Tumour/Spitzoid Melanoma

-Concerning Features-

Architecture:
Diameter in mm (>10 mm)
Depth in mm (involvement of subcutaneous fat)
Ulceration
Poor circumscription
Diffuse Pagetoid spread
High cellular density
Lack of zonation and maturation
Asymmetry
Few or no dull pink (Kamino) bodies

Cytology:
High nuclear to cytoplasmic ratios
Loss of delicate or dispersed chromatin patterns
Thickening of nuclear membranes
Hyperchromatism
Large nucleoli

Proliferation:
Significant mitotic rate
Deep/marginal mitoses
Increased mib-1 proliferation index

Modified from: Barnhill RL. Modern Pathology. 2006; 19: S21-S33
2 year old girl; right ankle tumour
10 year old female with an enlarging lesion on the left calf present for 8 months
Spitzoid Melanoma

- No firm established diagnostic criteria
- Distinction from Atypical Spitz Tumour largely arbitrary on histological grounds
- Best avoided in young patients
AST/Spitzoid Melanoma

-Prognose-

In young children (<10 years):

• Favourable behaviour
• Risk for involvement of loco-regional lymph nodes
• Rare disseminated disease and mortality
• Sentinel lymph node biopsy not helpful and should be avoided

AST/Spitzoid Melanoma

-Genetics-

Homozygous 9p21 deletion and P16 loss of expression by immunohistochemistry associated with more aggressive behaviour

Spitz Tumours

-Conclusion-

Challenging aspect of melanocytic tumour pathology

Wide morphological spectrum

Unique underlying genetic abnormalities

Vast majority benign

Behaviour difficult to predict on morphological ground

Atypical Spitz Tumours in children show potential for regional LN involvement but favourable long term behaviour in majority of patients