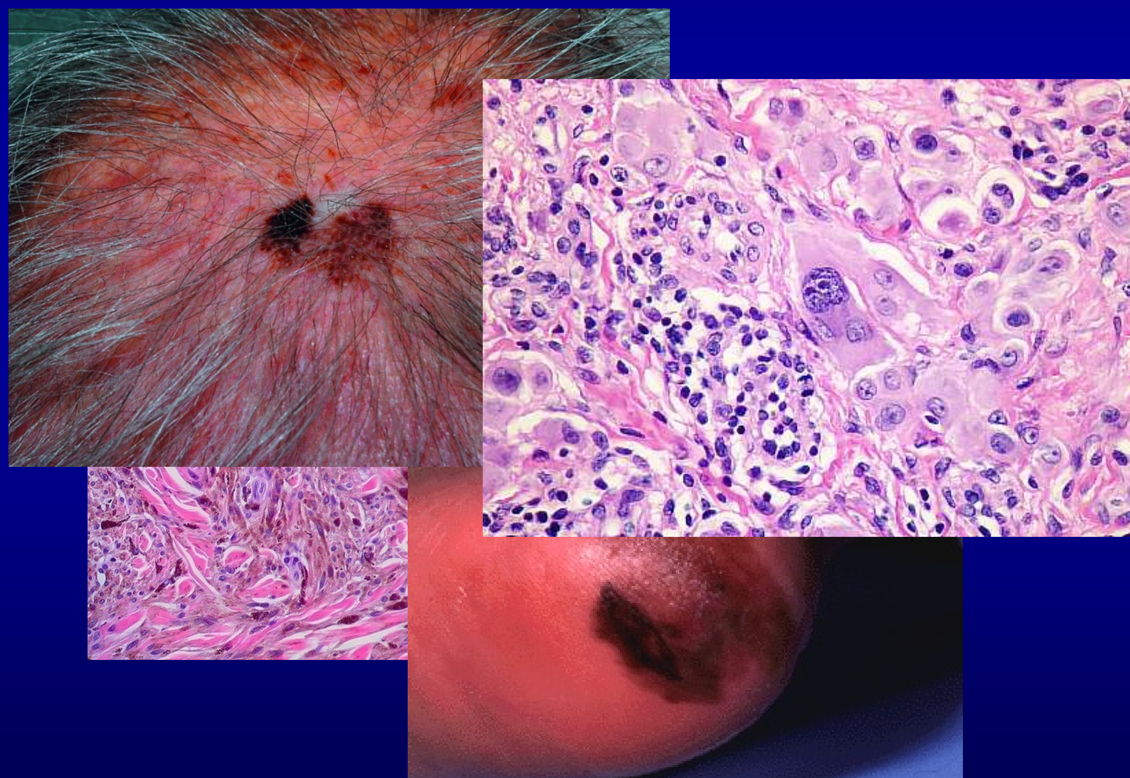
A PRACTICAL APPROACH TO ATYPICAL MELANOCYTIC LESIONS



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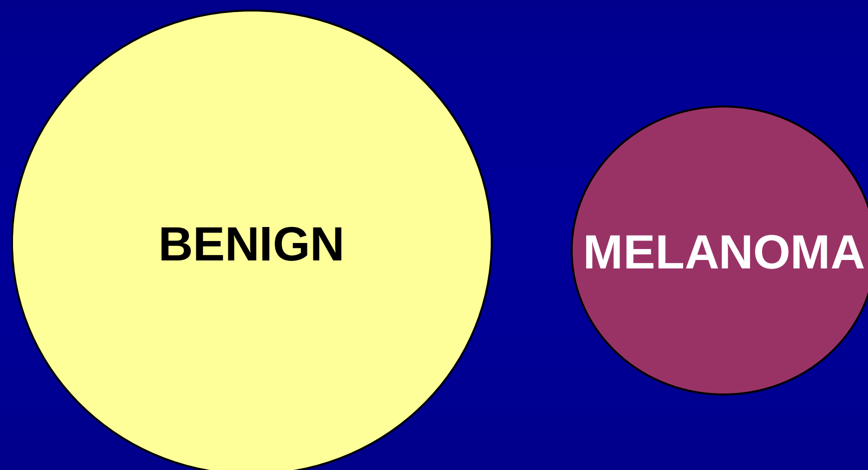
OBJECTIVES

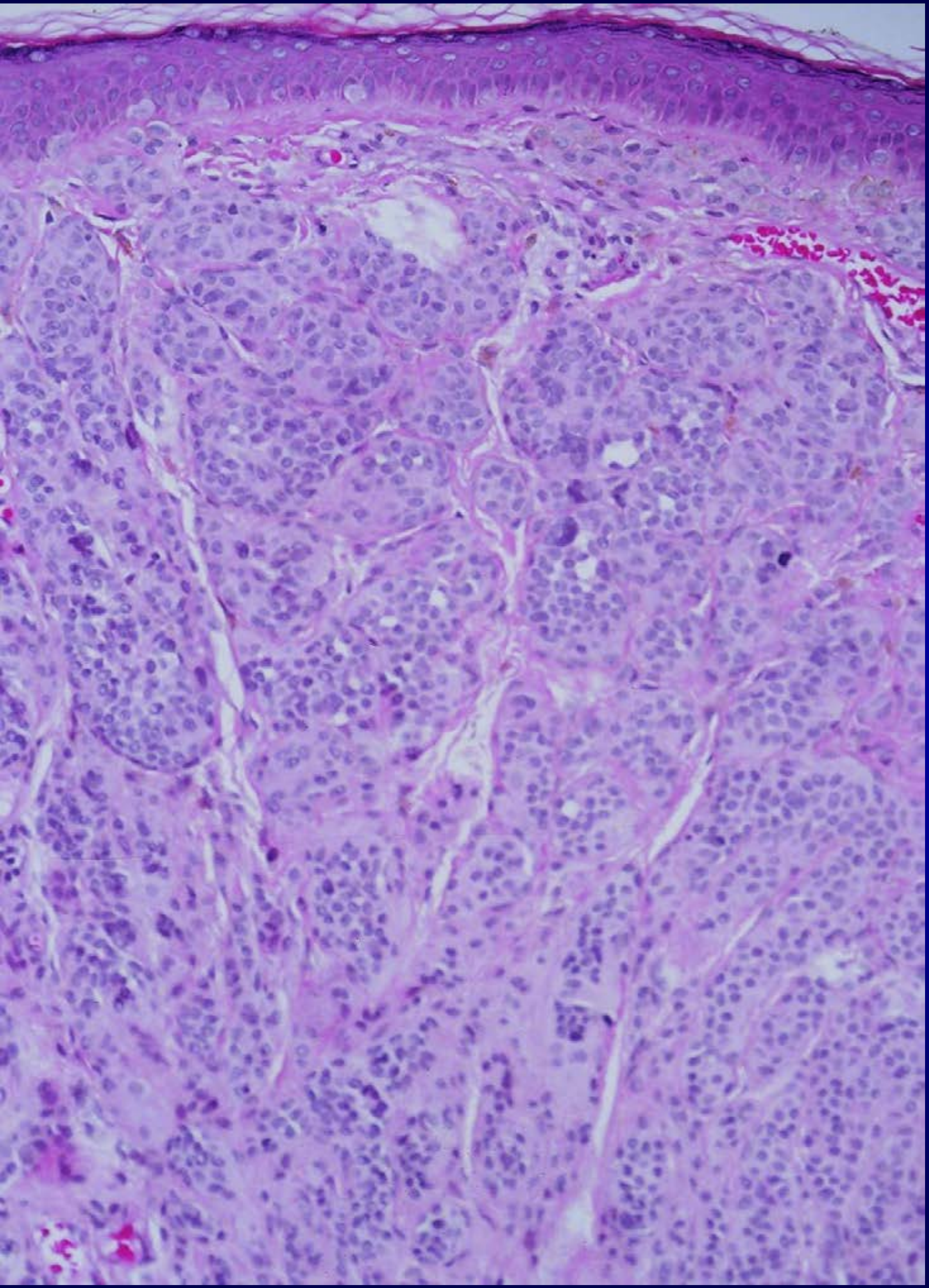
- Discuss current trends and changing concepts in our understanding of atypical melanocytic proliferations.

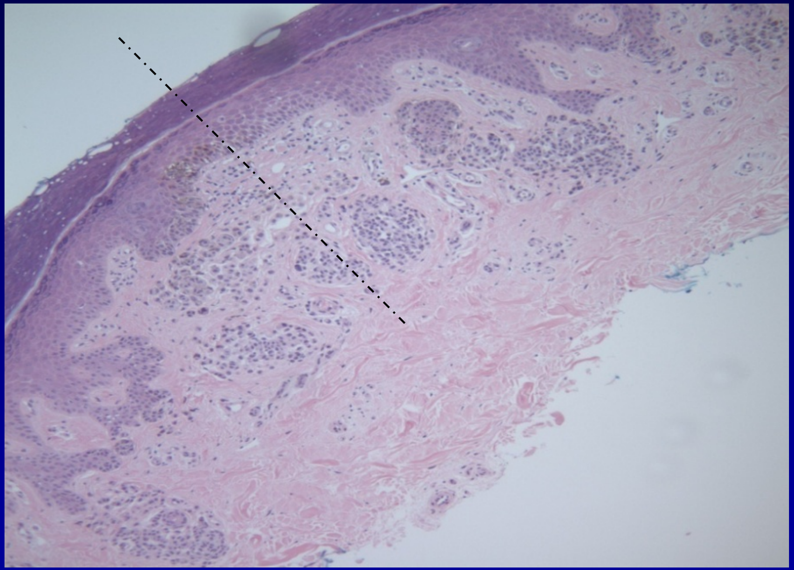
- Suggest a practical approach to the management of atypical melanocytic lesions including Spitzoid melanocytic lesions and dysplastic nevi.

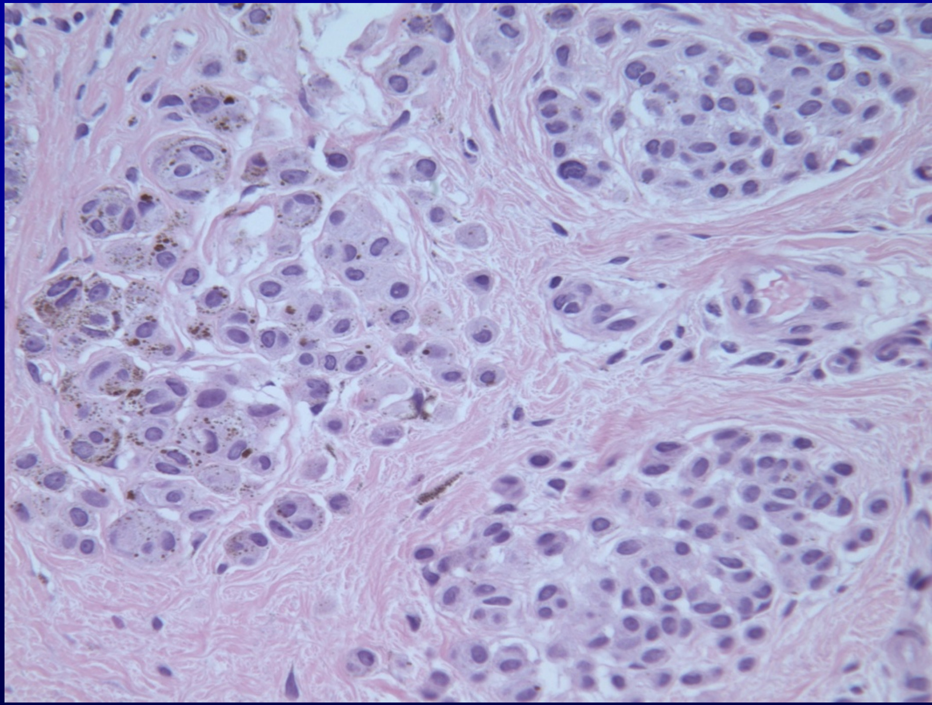
TRADITIONAL / SYMPLISTIC VIEW



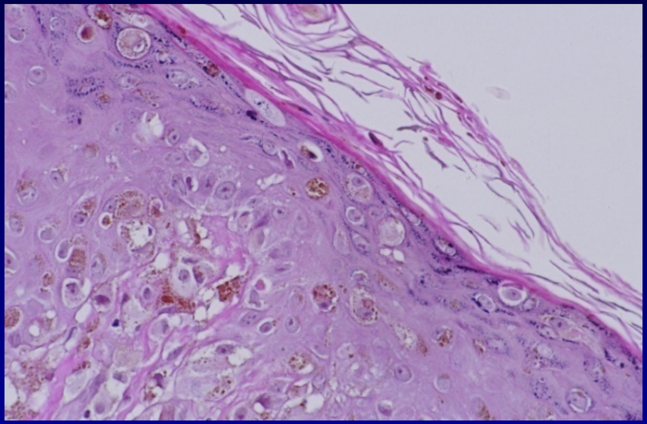
BENIGN NEVUS

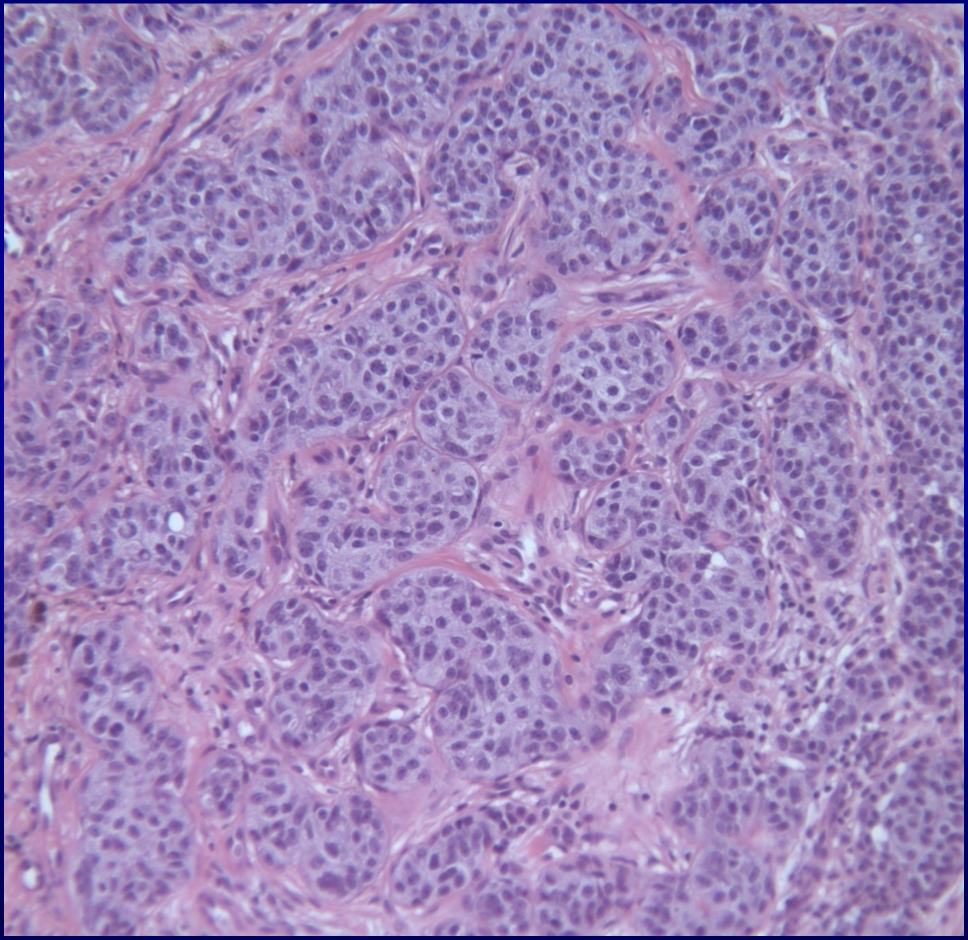


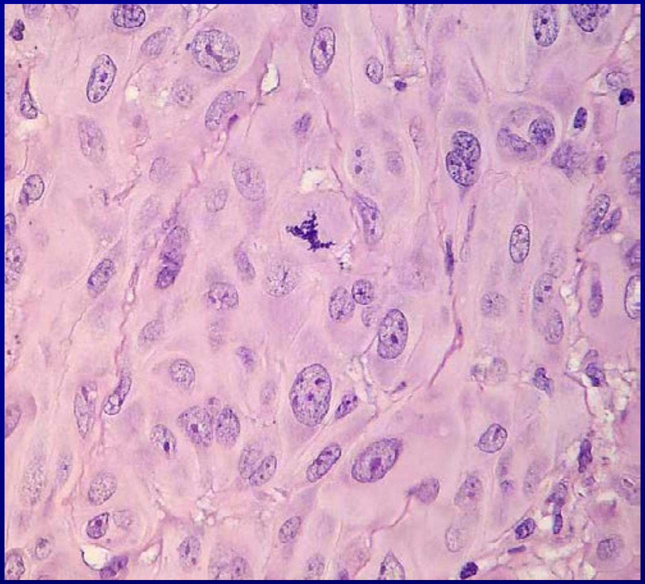




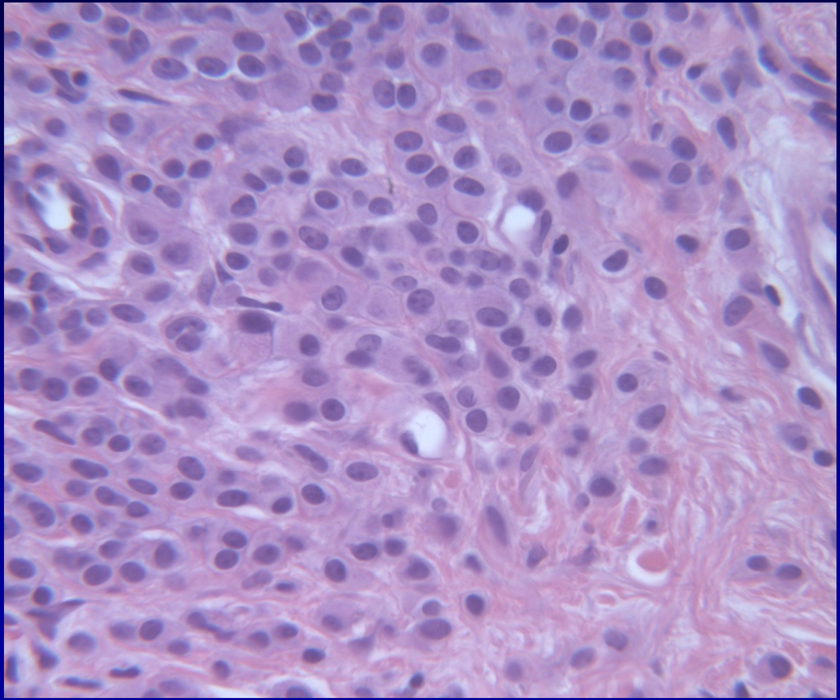
MELANOMA

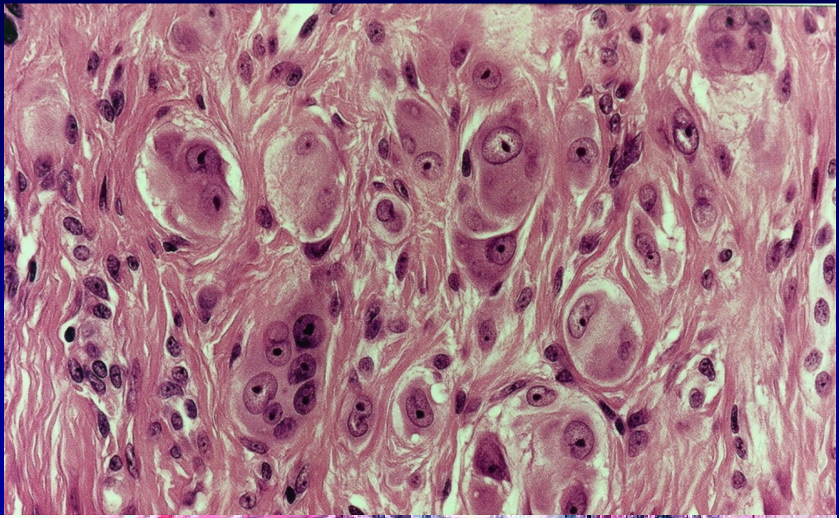


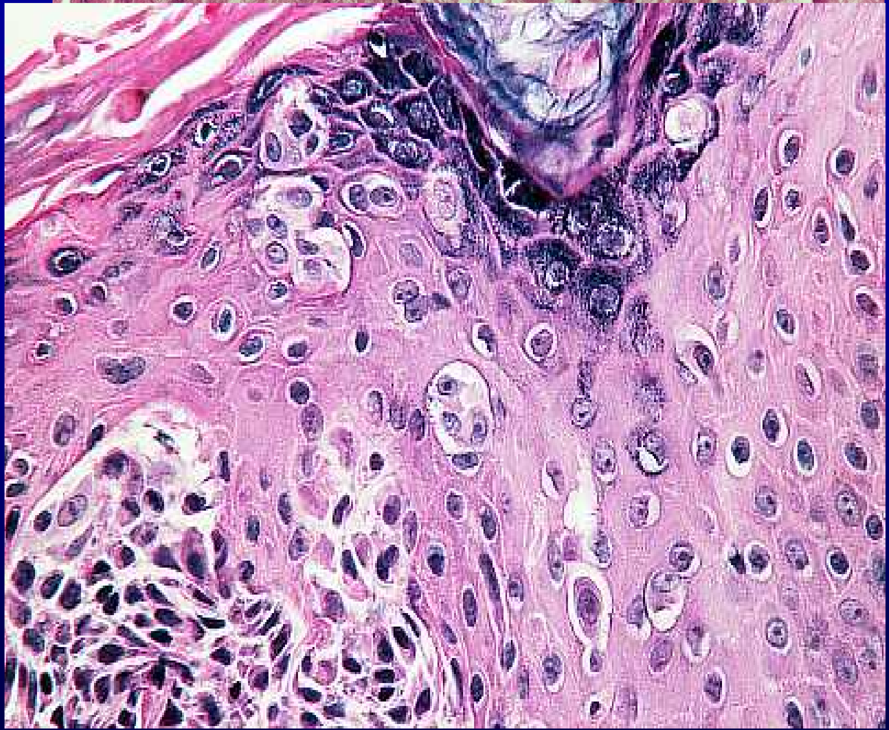




BENIGN OR MALIGNANT?







PROBLEMS WITH TRADITIONAL VIEWPOINT

- Unlike many other malignant neoplasms, the diagnosis of malignant melanoma is based on a constellation of many different features (any of which by themselves may be seen in benign lesions, some of which may not be seen in a given case).

- Even though indeterminate lesions do exist, because of medicolegal pressure many “atypical” cases are placed in the malignant category and are therefore overdiagnosed.

Implications of Overdiagnosis

- Cosmetic disfigurement
- Psychological implications
- Negative impact on insurance

Traditional view

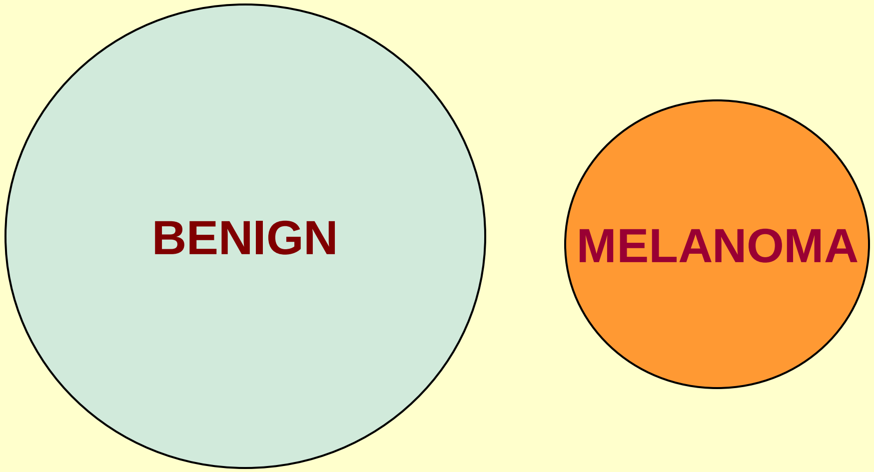
- Regional lymph node metastasis is proof of malignancy!

- Problem: emerging data suggests that some atypical lesions (i.e atypical Spitz tumors in children) may show regional lymph node involvement without further progression.

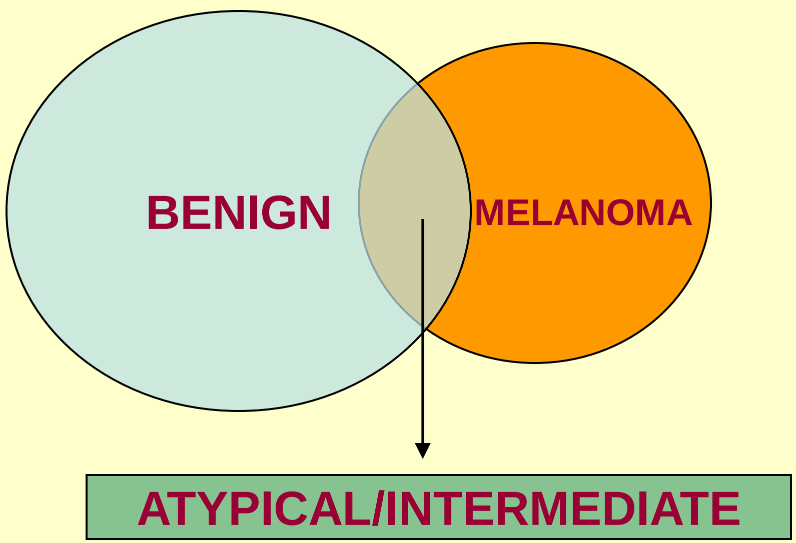
BENIGN

ATYPICAL

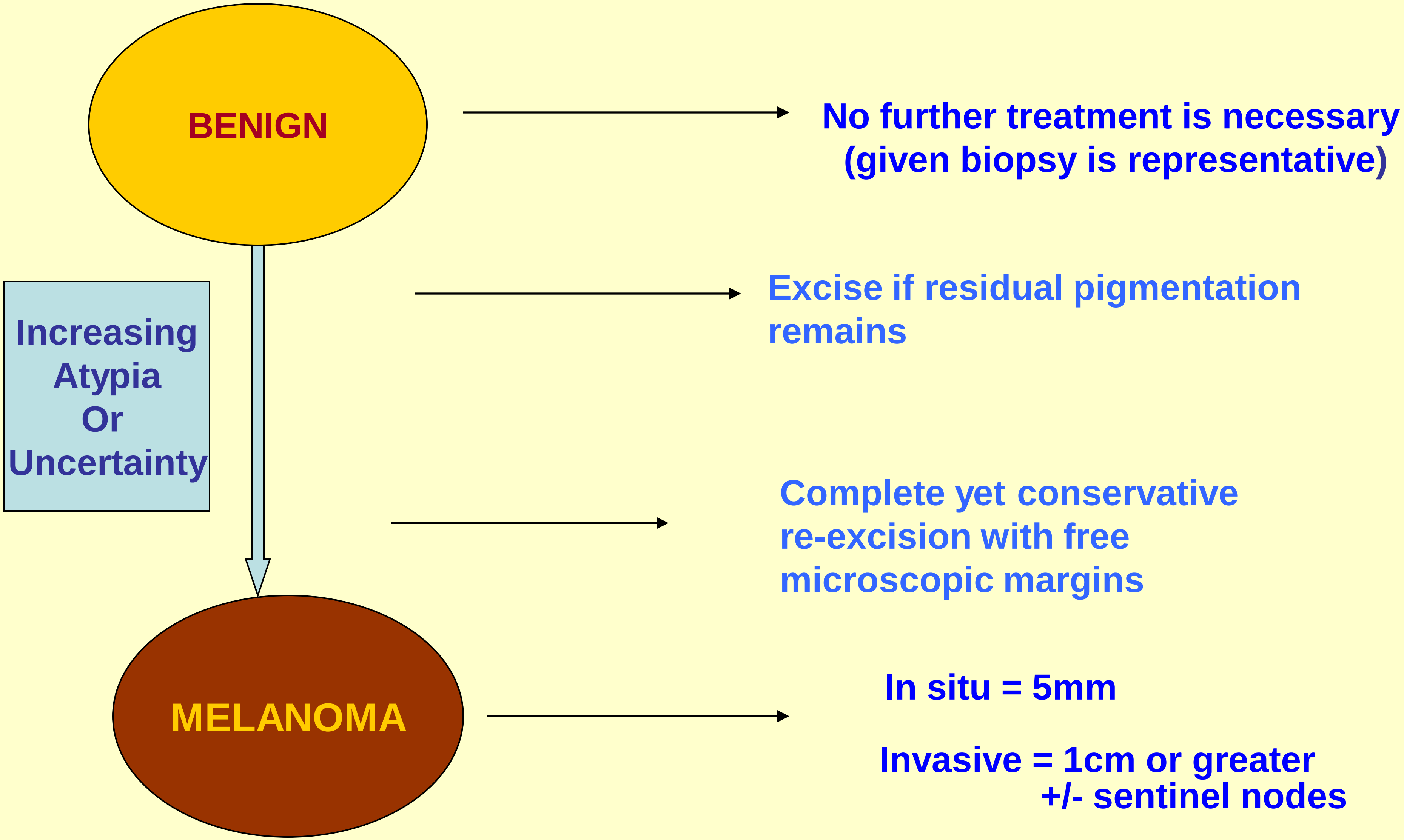
MALIGNANT



TRADITIONAL/ SYMPLISTIC VIEW



CONTEMPORARY/ PRACTICAL VIEW



***Note: Management should take into consideration other clinical risk factors (i.e family or personal history of melanoma, etc.)**

MELANOCYTIC LESION

BENIGN

Clinical/
+/- ancillary studies/
expert consultation

MELANOMA

Clinical/
+/- ancillarystudies/
expert consultation

ATYPICAL

Morphologic simulator (i.e nevi of special site)

No

PROBABLY BENIGN WITH FEW ATYPICAL FEATURES
(i.e low grade dysplastic nevi, ?Spitz nevi

INDETERMINATE
(i.e Atypical Spitz Nevi)

BORDERLINE
(i.e severely dysplastic Nevi)

Approach to Atypical Melanocytic Lesions:
General Principles

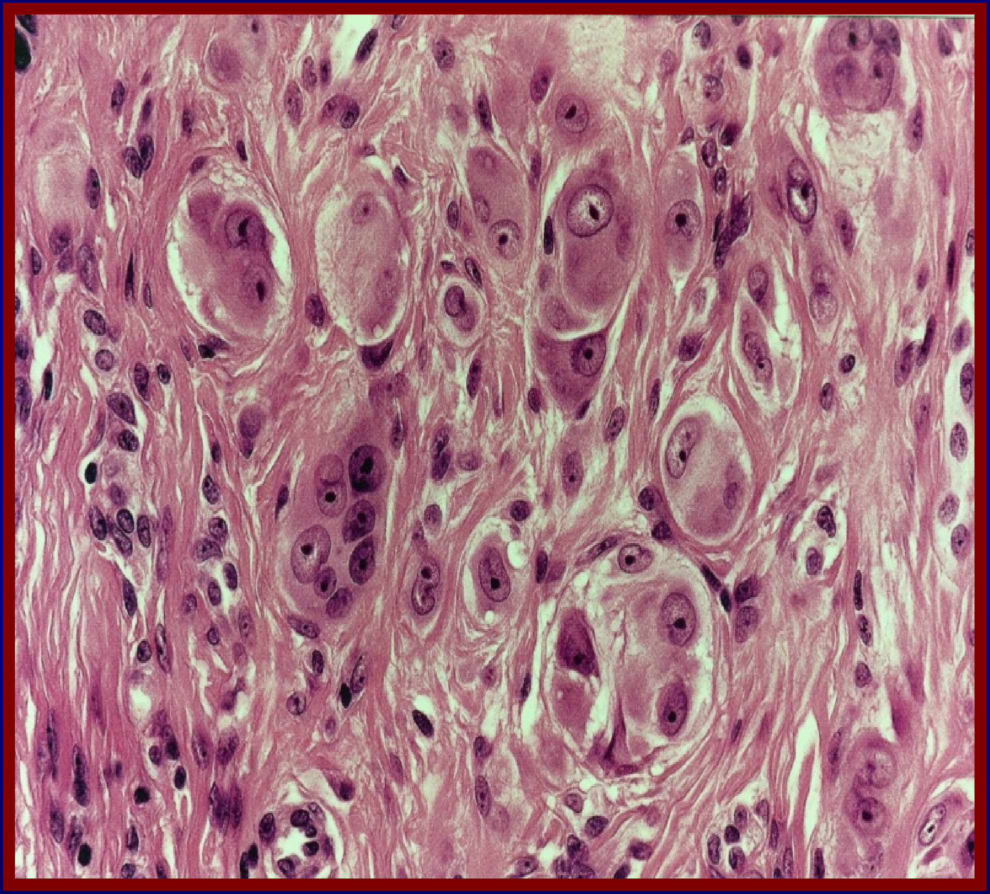
- Diagnostic uncertainty should be expressed in the report and/or directly discussed with treating clinician.
- Treatment may be tailored to the differential diagnosis.
- If melanoma cannot be confidently ruled out (i.e borderline lesion), consider treating as melanoma of similar thickness (consider rendering a descriptive diagnosis, but report melanoma prognostic factors, i.e Breslow depth, mitotic activity, ulceration)

Practical Approach to Spitzoid Tumors

SPITZ NEVUS

- **Melanocytic lesion with characteristic epithelioid morphology, First described by Sophie Spitz in 1948 as "juvenile melanoma" because of its propensity to occur in childhood , morphologically mimic melanoma but exhibit relatively indolent behaviour.**

- **12 of 13 patients diagnosed as melanoma were alive following long term follow-up**



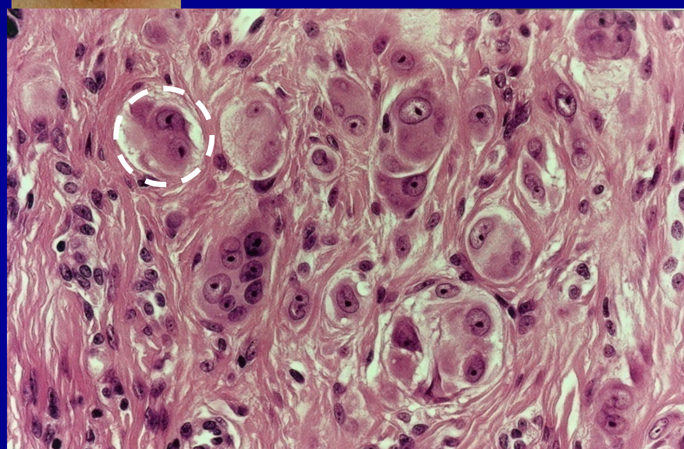
SOPHIE SPITZ

- **“Differentiation histologically between the juvenile and adult melanomas could not be made with certainty in most cases.”**

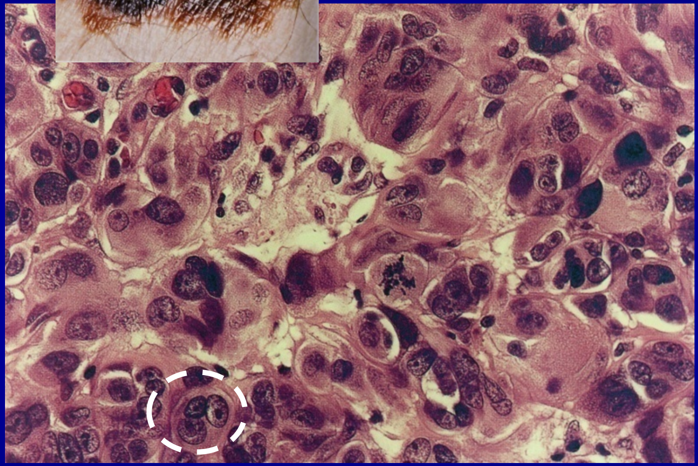
There is no single feature that distinguishes a Spitz nevus from a melanoma

CYTOLOGIC ATYPIA

Spitz Nevus



Melanoma

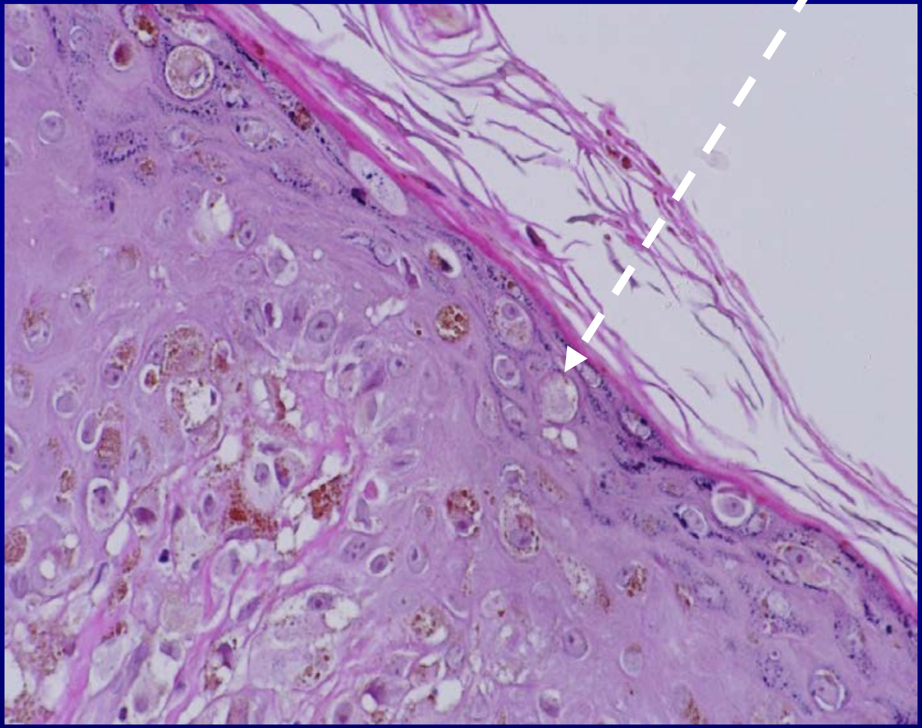


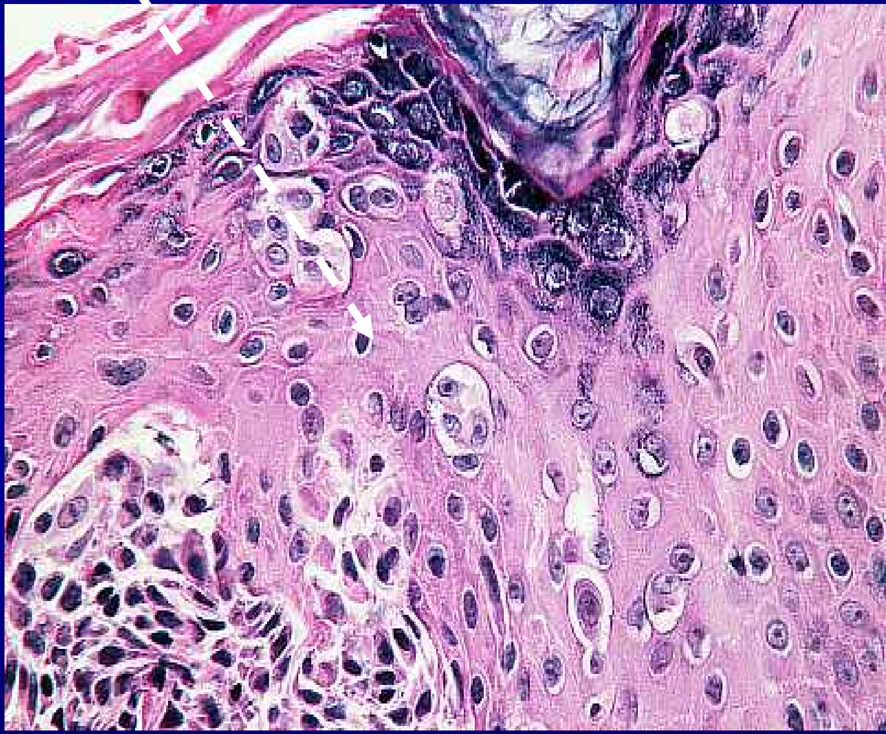
The individual cell that defines a Spitz nevus is by definition atypical

UPWARD PAGETOID SCATTER

MELANOMA

SPITZ NEVUS

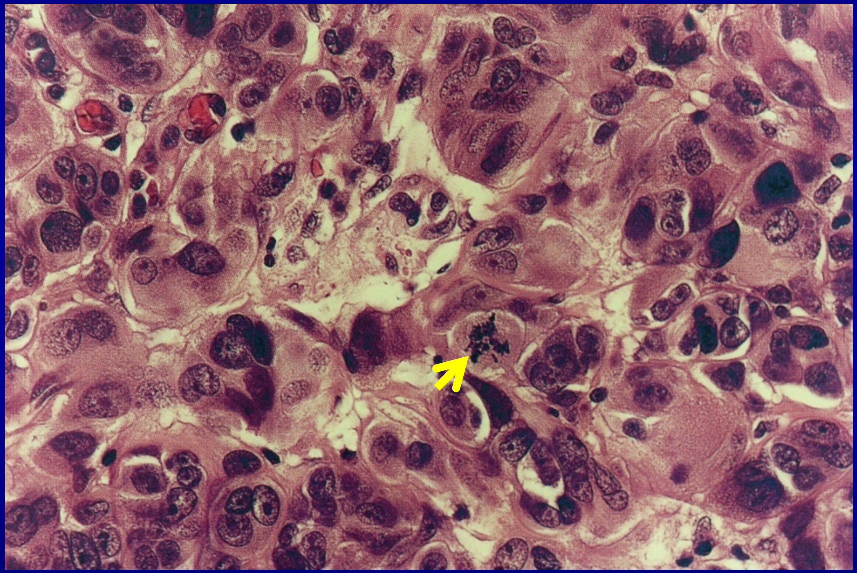




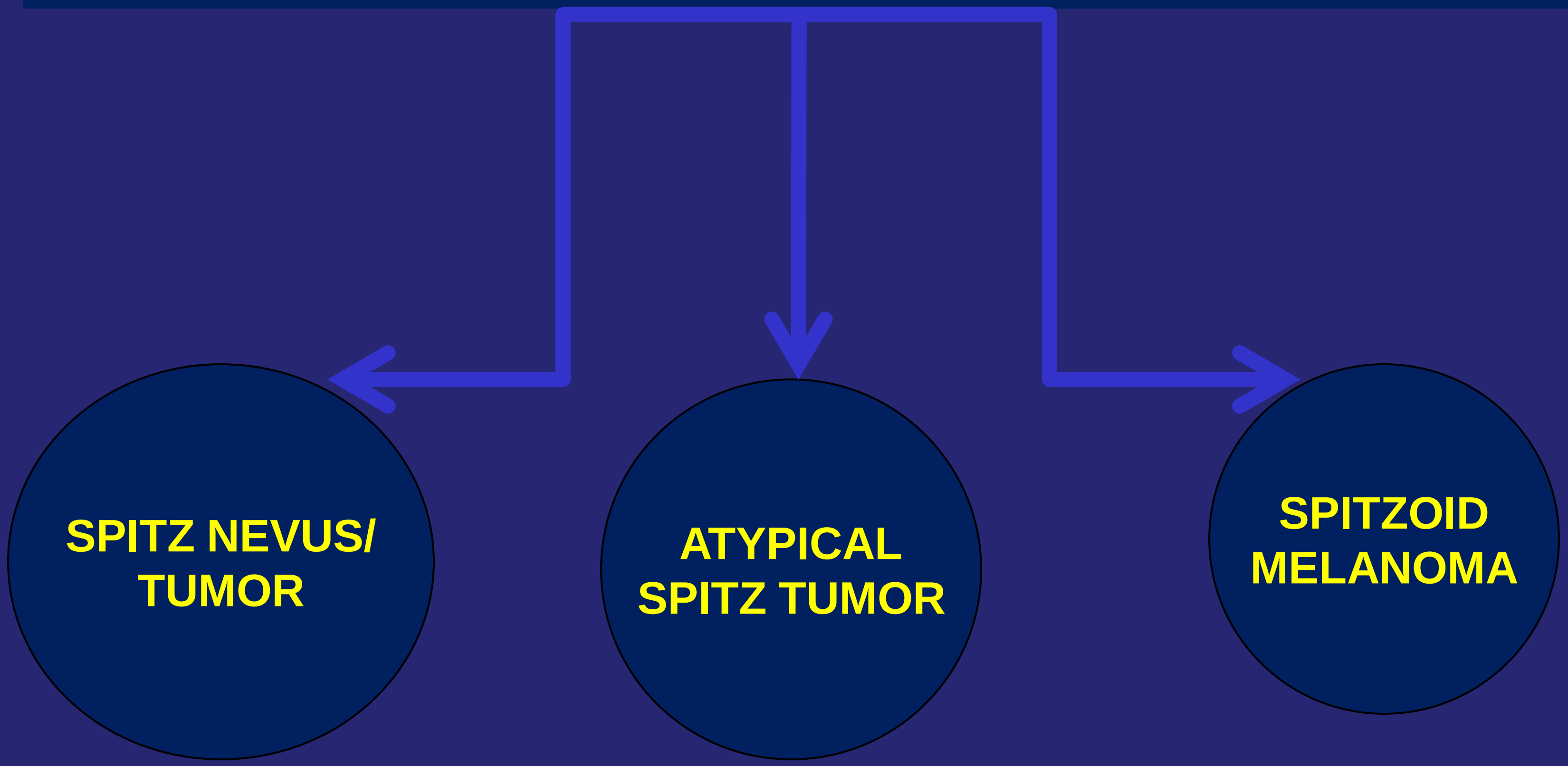
MITOTIC ACTIVITY

SPITZ NEVUS

MELANOMA



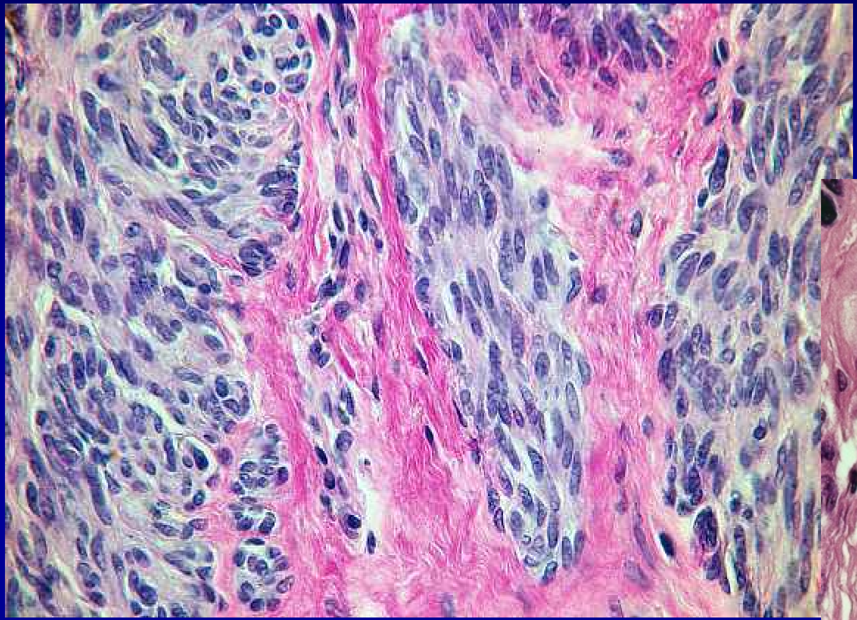
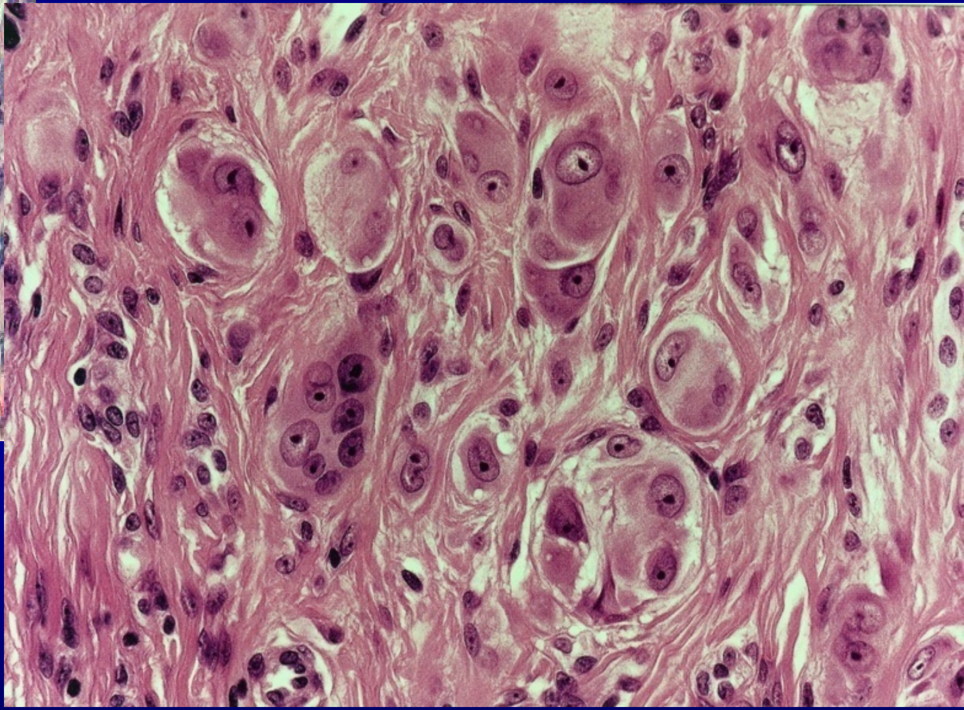
SPITZOID MELANOCYTIC LESIONS

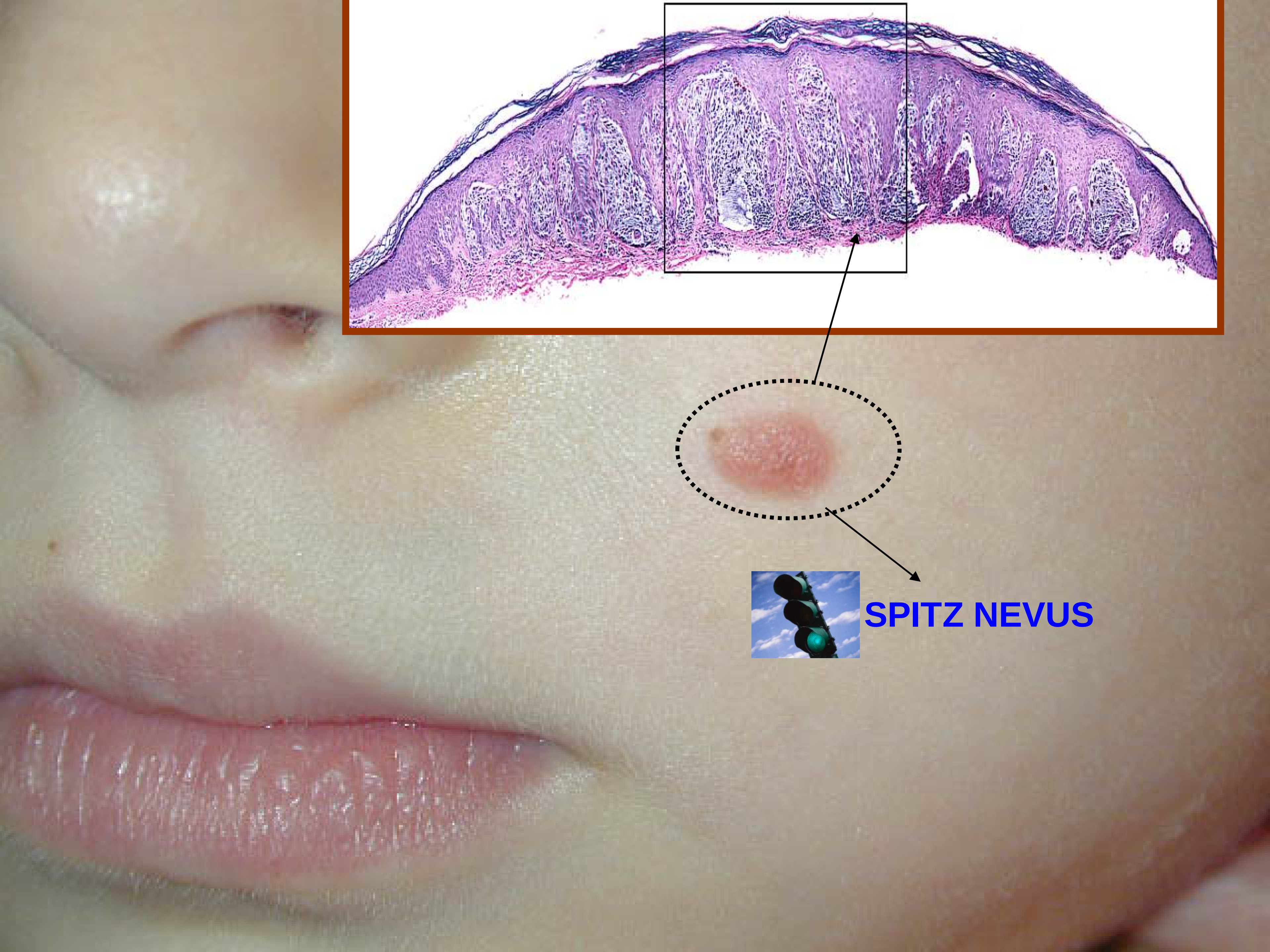


SPITZ NEVUS: Clinical features

- Great majority of lesions occur in childhood or in young adults
- Recent onset and rapid growth
- Usually less than 1 cm
- Pink-tan to reddish nodule
- Clinically symmetric with even borders

SPITZ NEVUS:
Histology


SPITZ NEVUS

Rationale for Complete Excision of All Spitzoid Tumors

- Morphologic overlap with melanoma

- Recurrence of Spitz nevi may demonstrate features which may be very difficult to differentiate from melanoma

- Very rarely Spitz nevi with “classic” morphologic features have been known to metastasize.

SPITZ NEVUS CYTOLOGY AND ARCHITECTURE

+

APPROPRIATE CLINICAL SETTING

SPITZ NEVUS

+

LESION COMPLETELY EXCISED

ATYPICAL SPITZ TUMOR

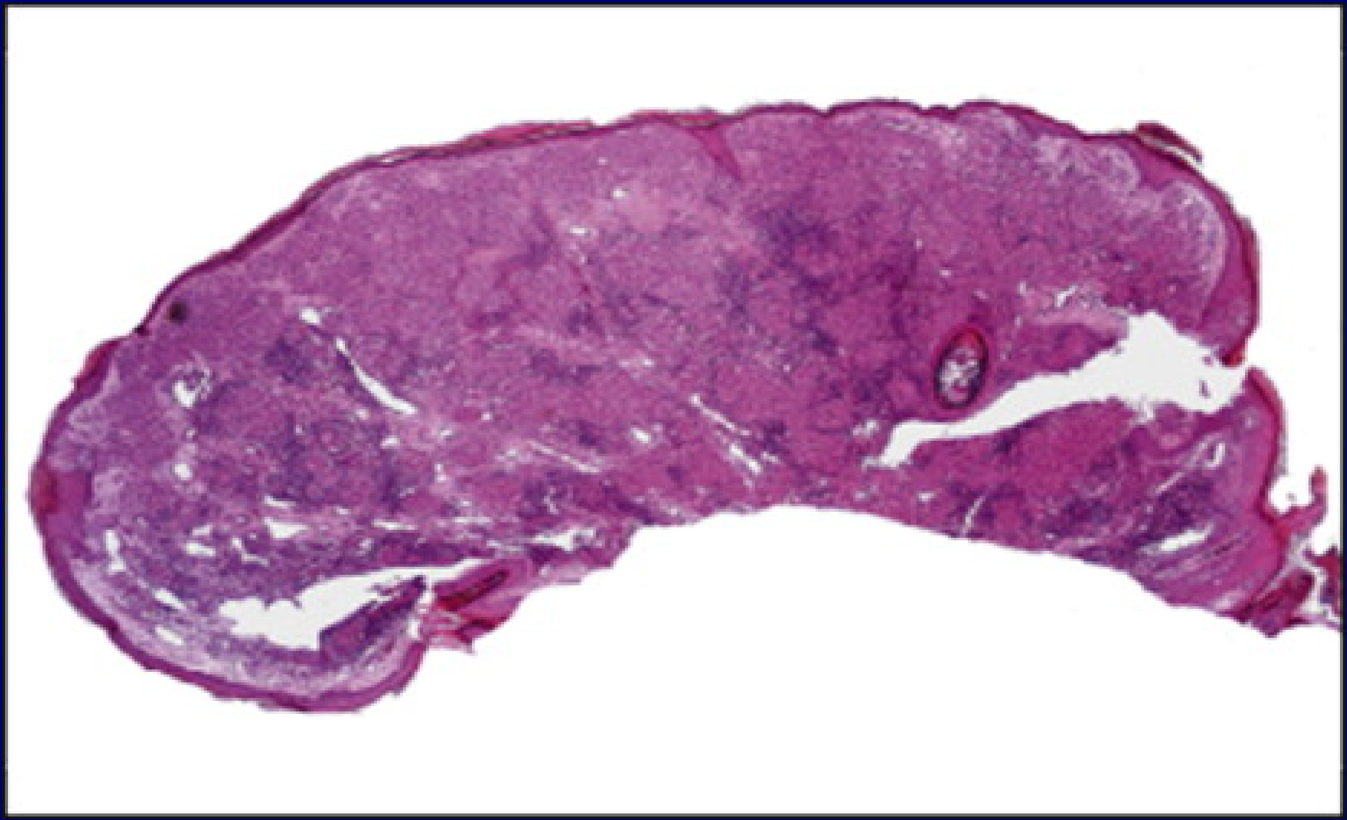
- **Subset of Spitzoid neoplasms with clinically and pathologically disturbing features in which a melanoma cannot be excluded with absolute certainty.**
- **Lesions are more likely to behave more indolently, particularly in childhood, although regional lymph node involvement can occur.**
- **The significance of nodal deposits is unclear and does not necessarily indicate aggressive behavior (especially in children).**

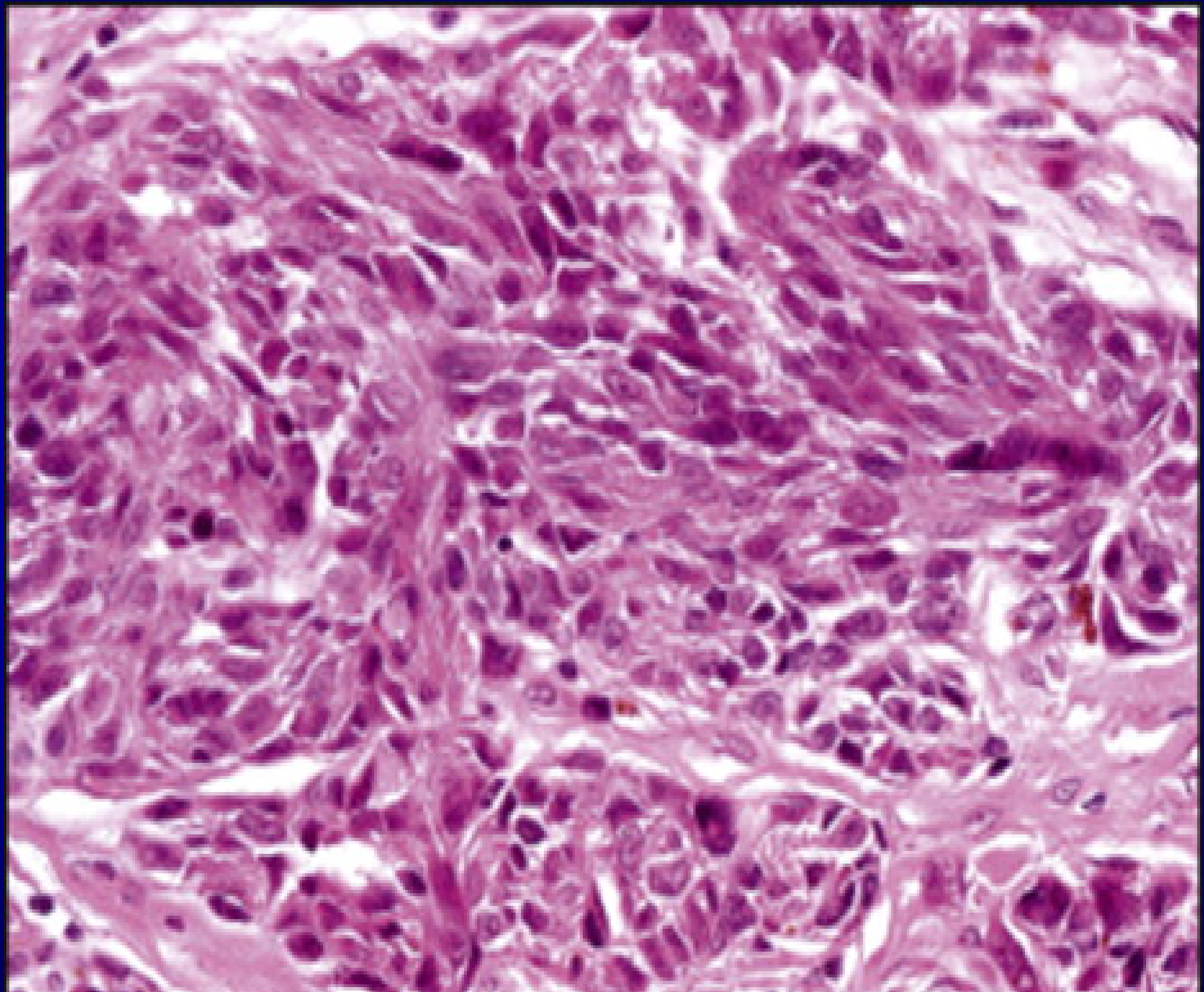
ATYPICAL SPITZOID TUMORS (cont.)

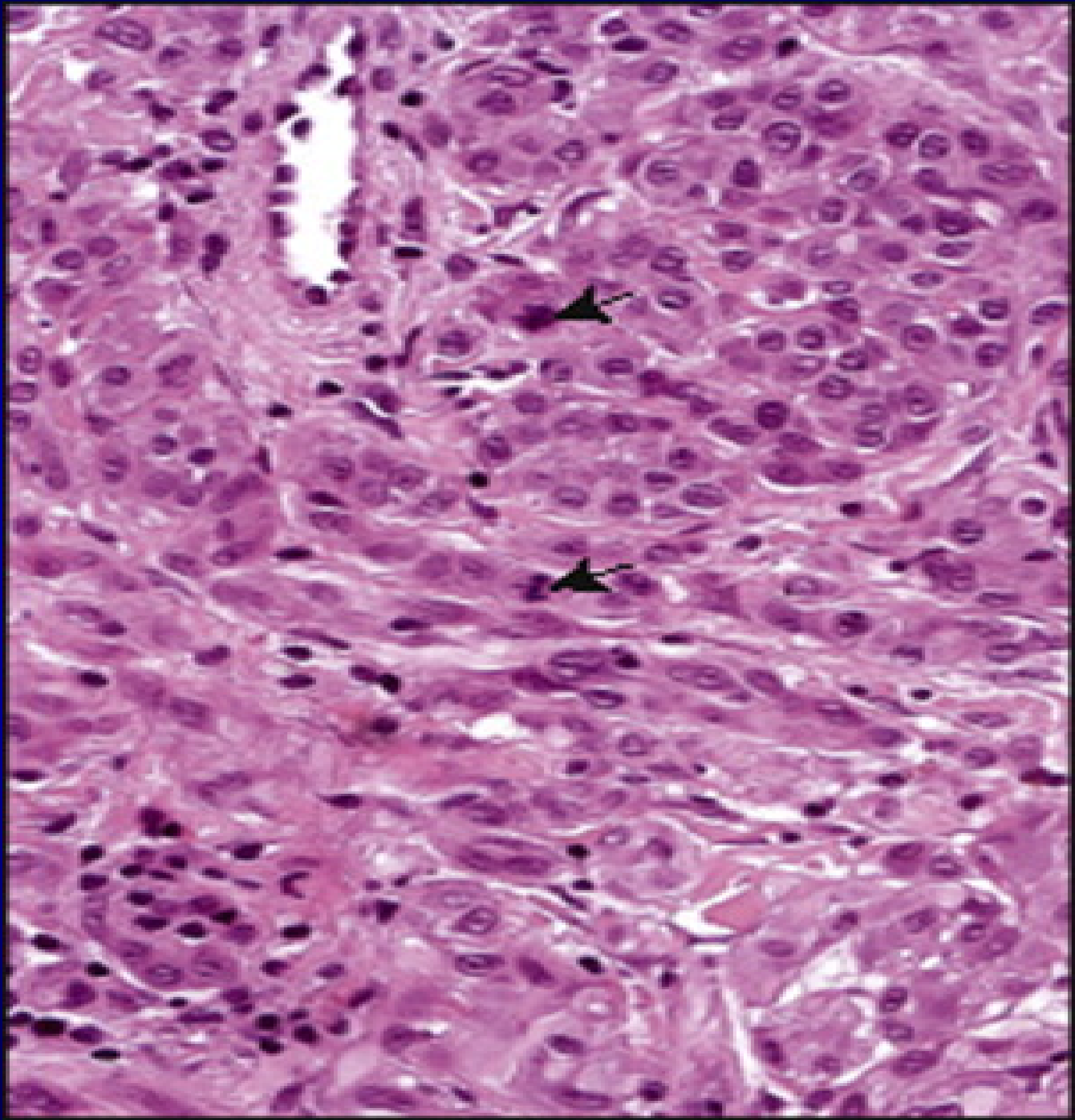
Features with significant risk of nodal metastasis (esp. in children):

- Age greater than 10 years old
- Lesional diameter greater than 1 cm
- Ulceration
- Involvement of subcutaneous fat
- Mitotic activity of at least 6/mm^2

ATYPICAL SPITZ TUMOR:
Histology







SPITZOID NEOPLASMS

BENIGN

SPITZ NEVUS/TUMOR

TX: Excise

INDETERMINATE

ATYPICAL SPITZ TUMOR

TX: Excision +/-
consider treating as melanoma
Generally without systemic therapy

MALIGNANT

SPITZOID MELANOMA

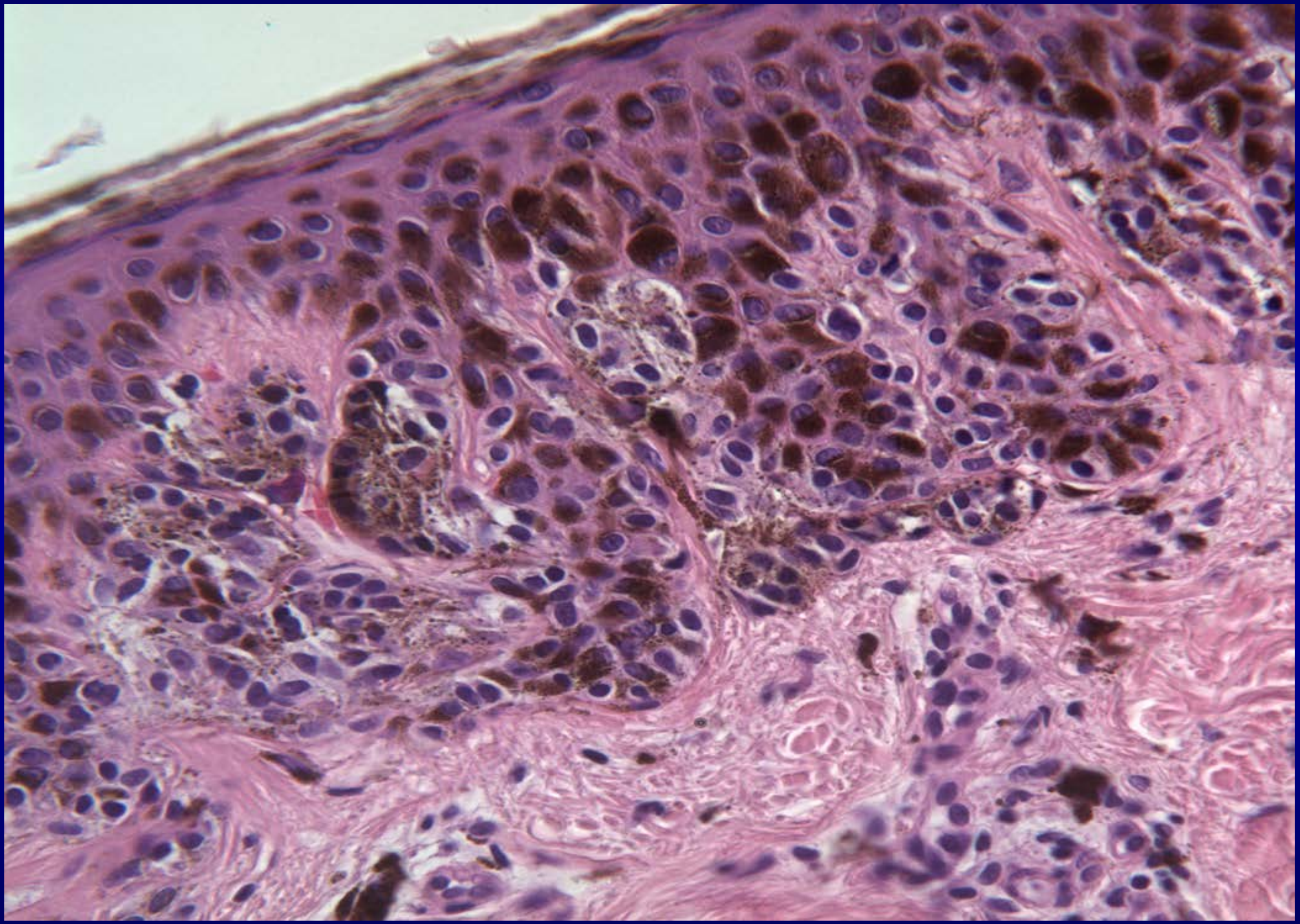
TX: Wide excision +/-
Sentinel node biopsy +/-
Systemic therapy

- Spitzoid melanomas are rare under the age of 20.
- The clinical diagnosis of a benign lesion in a child should be overruled only with very strong histologic evidence to the contrary.
- After age 50, a lesion with features resembling a Spitz nevus is more likely a melanoma than a nevus.
- In an older adult, a junctional Spitzoid tumor is most likely a melanoma.
- Beware of diagnosis of Spitz nevus on severely sun damaged skin (it is probably a melanoma).

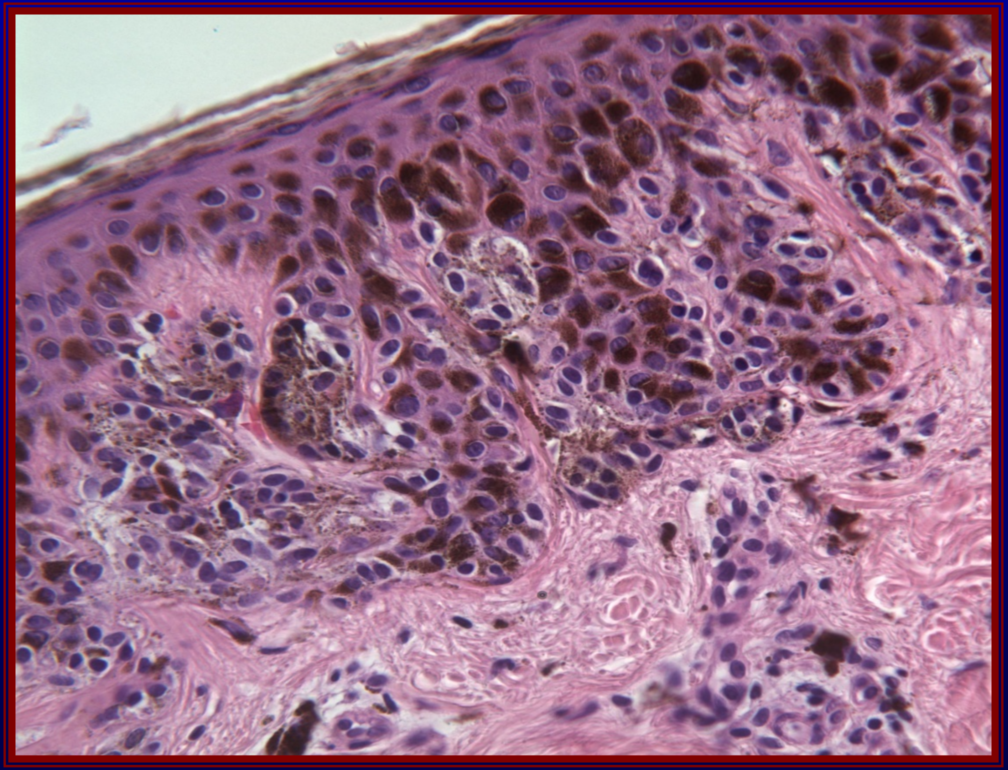
How To Stay Out Of Court

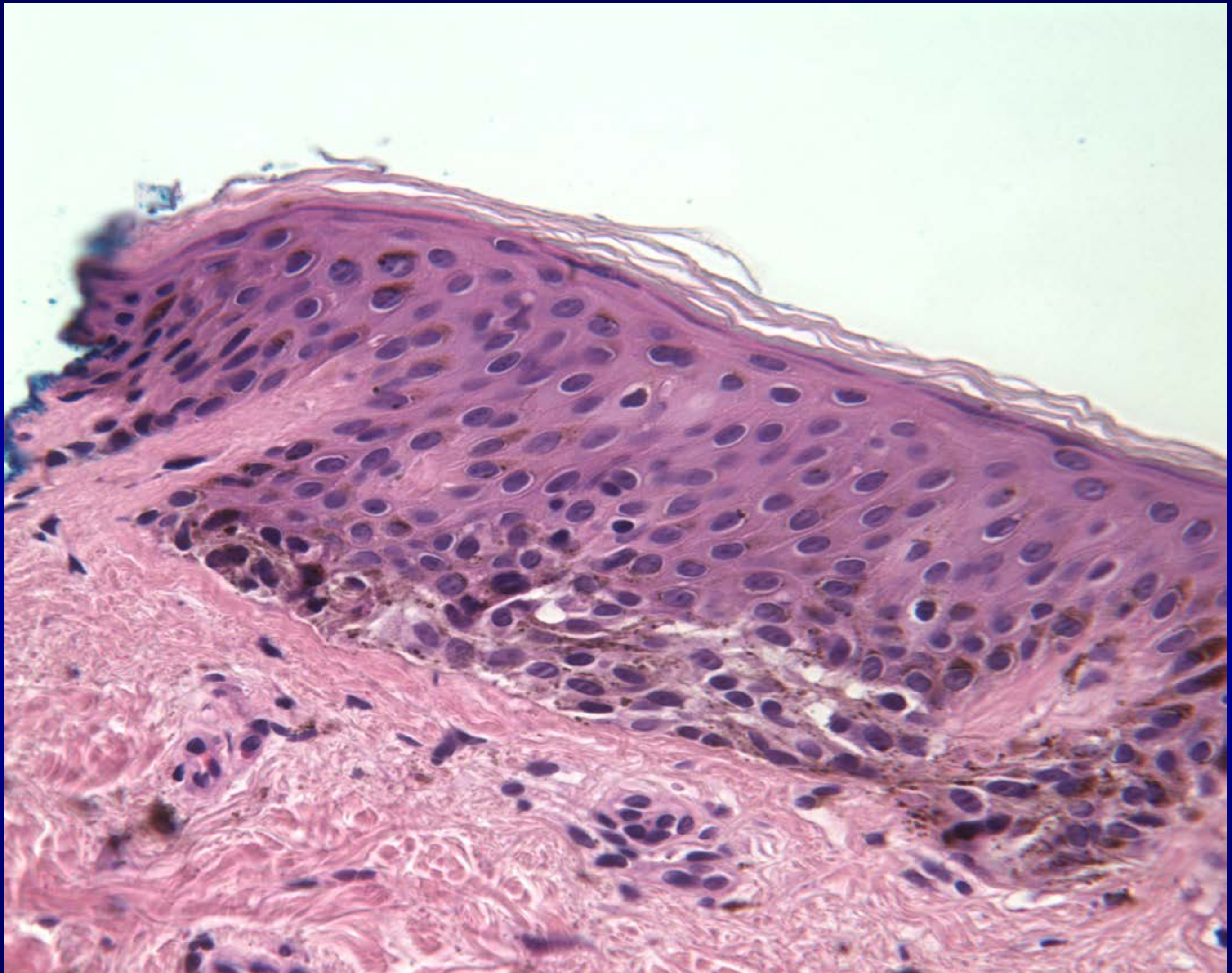
- Spitz nevus is a high risk/low frequency diagnosis (like soft tissue or bone tumors.
- If pathologist does not see Spitz nevi on a regular basis and the patient is more than 20 years old, strongly consider sending to an expert.
- If Spitz nevi are seen on a regular basis but patient is more than 20 years old (unless typical diagnostic criteria are all present) , strongly consider sending case to an expert.
- All Spitz nevi should be completely excised (although exceedingly rare, cases of classical Spitz nevi have been known to metastasize)
- Even experts disagree on Spitz tumors

CASE PRESENTATION



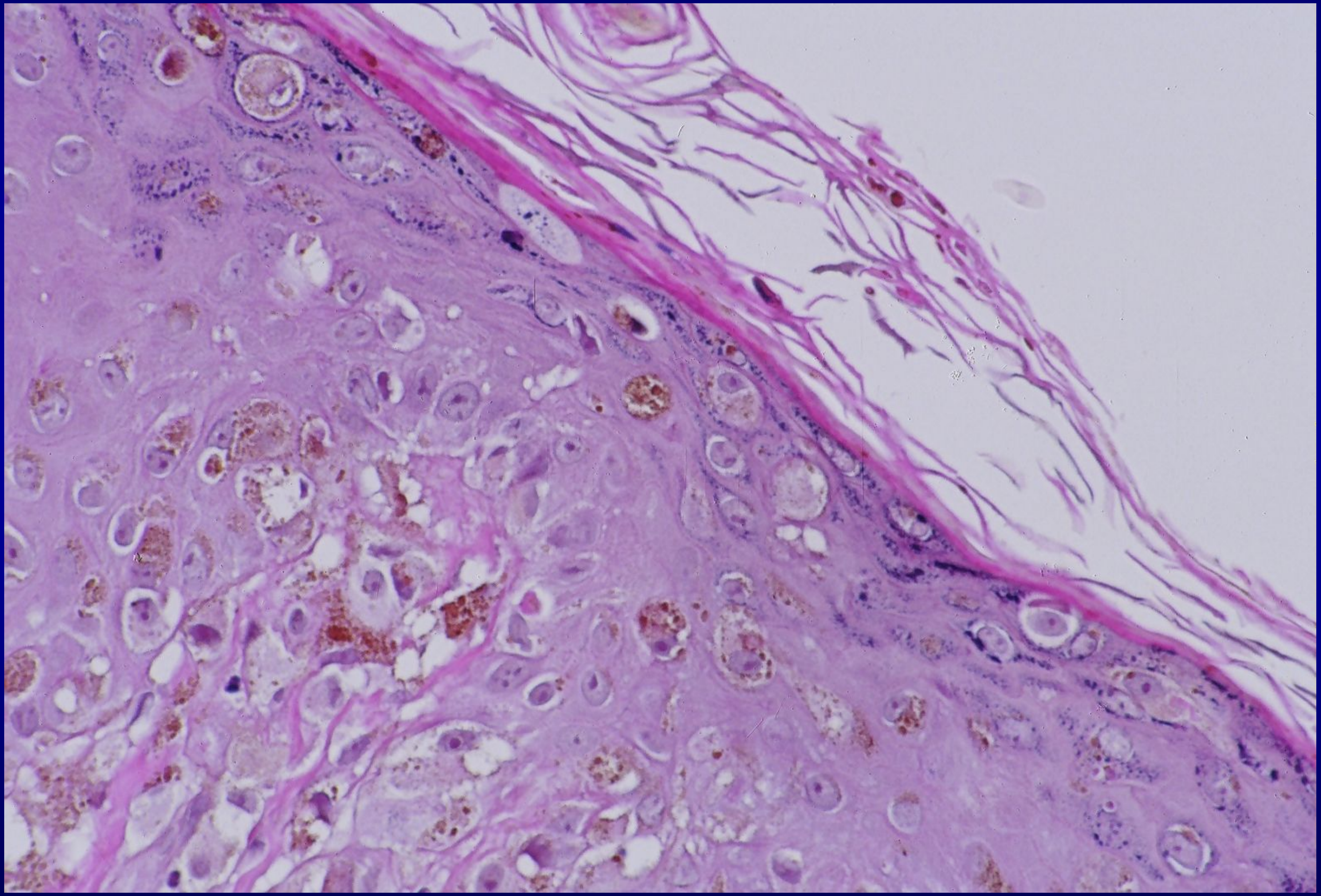
a) JUNCTIONAL MELANOCYTIC NEVUS
b) MILDLY DYSPLASTIC JMN
c) MALIGNANT MELANOMA
d) PIGMENTED SUPERFICIAL BCC





a) Re-excise
b) No further treatment necessary
c) Additional clinical information necessary

- **Additional clinical information reveals that the biopsy was taken from a significantly larger clinical lesion.**
- **Re-excision recommended by pathologist ("re-excision should be considered, as clinically indicated, particularly if residual pigmentation remains").**

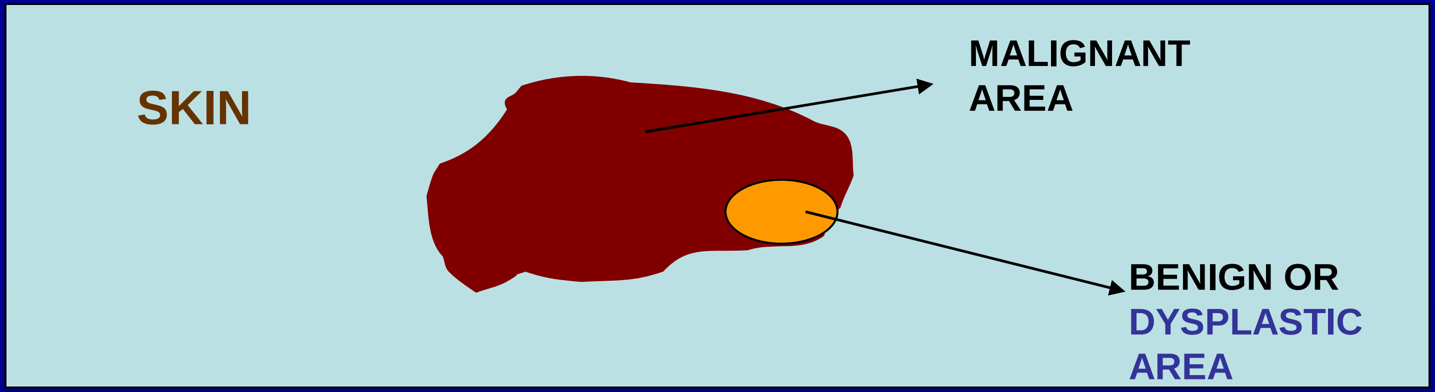


FINAL DIAGNOSIS

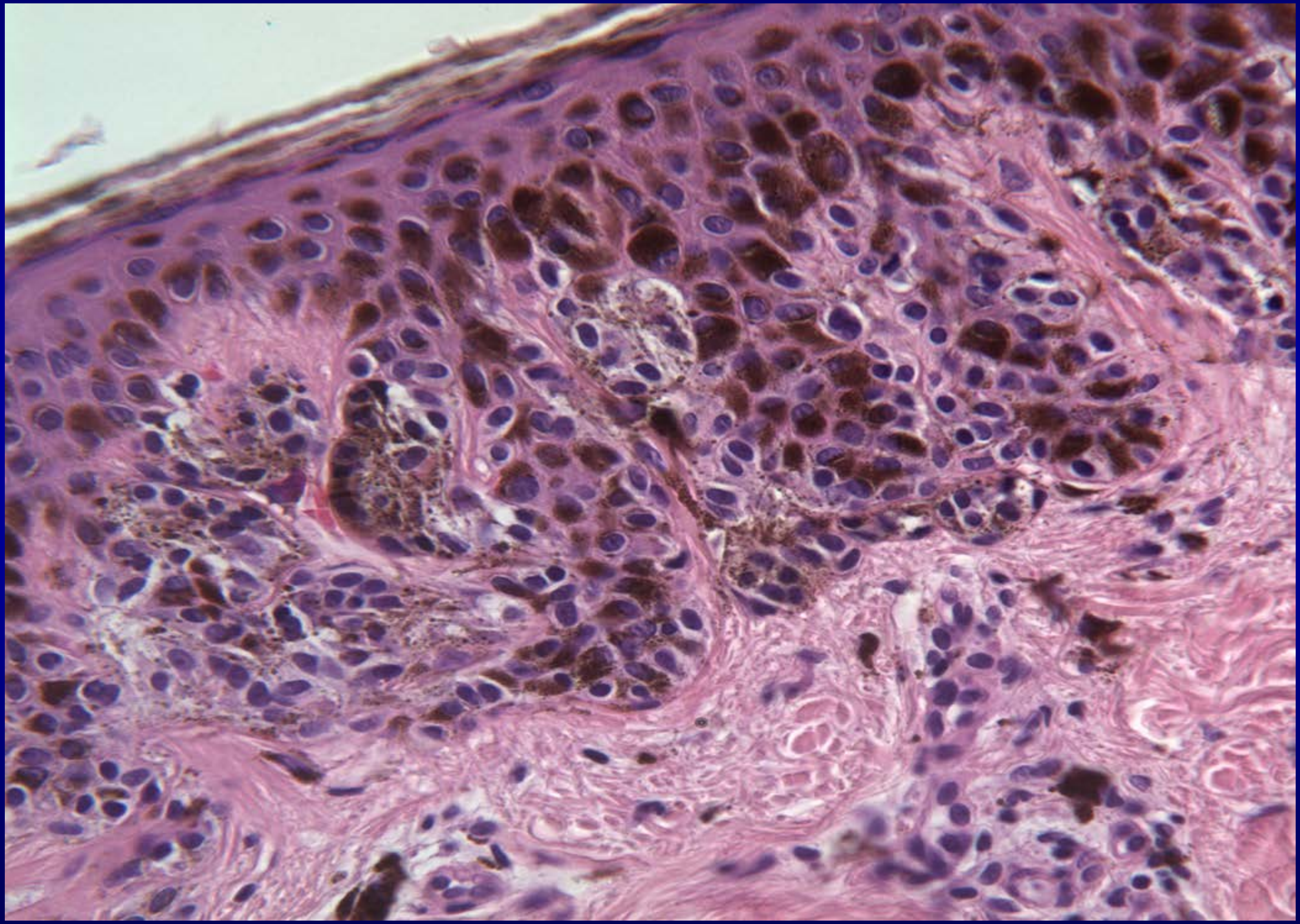
- **Initial biopsy:** Mildly dysplastic junctional melanocytic nevus, involving biopsy margins.

- **Re-excision specimen:** Malignant melanoma in-situ, superficial spreading type, arising in the background of dysplastic nevus..

SAMPLING ERROR



Practical Approach to "Dysplastic" Nevi



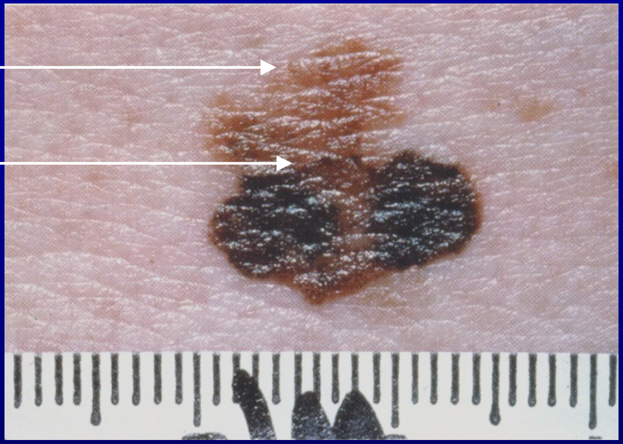
Considerations prior to treatment

✓ Diagnostic pitfall (morphologic overlap with malignant melanoma)

- Association of dysplastic nevi and melanoma in the same lesion

dysplastic nevus

melanoma

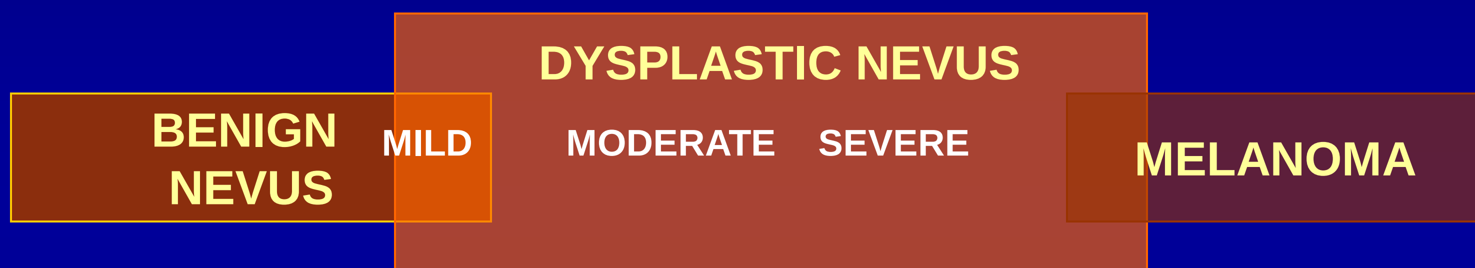


- Interobservor variability in diagnosis

Significance of Dysplastic Nevi

- **Morphologic overlap with melanoma**
- **Marker of individuals at increased risk of developing melanoma**
- **Potential actual precursor of melanoma**

Grading of Dysplastic Nevi



Rationale For Grading of Dysplastic Nevi

Separate slightly atypical but essentially benign nevi from those that:

- 1) might be confused with melanoma
- 2) possibly more likely to progress to melanoma
- 3) may be associated with a higher risk of melanoma

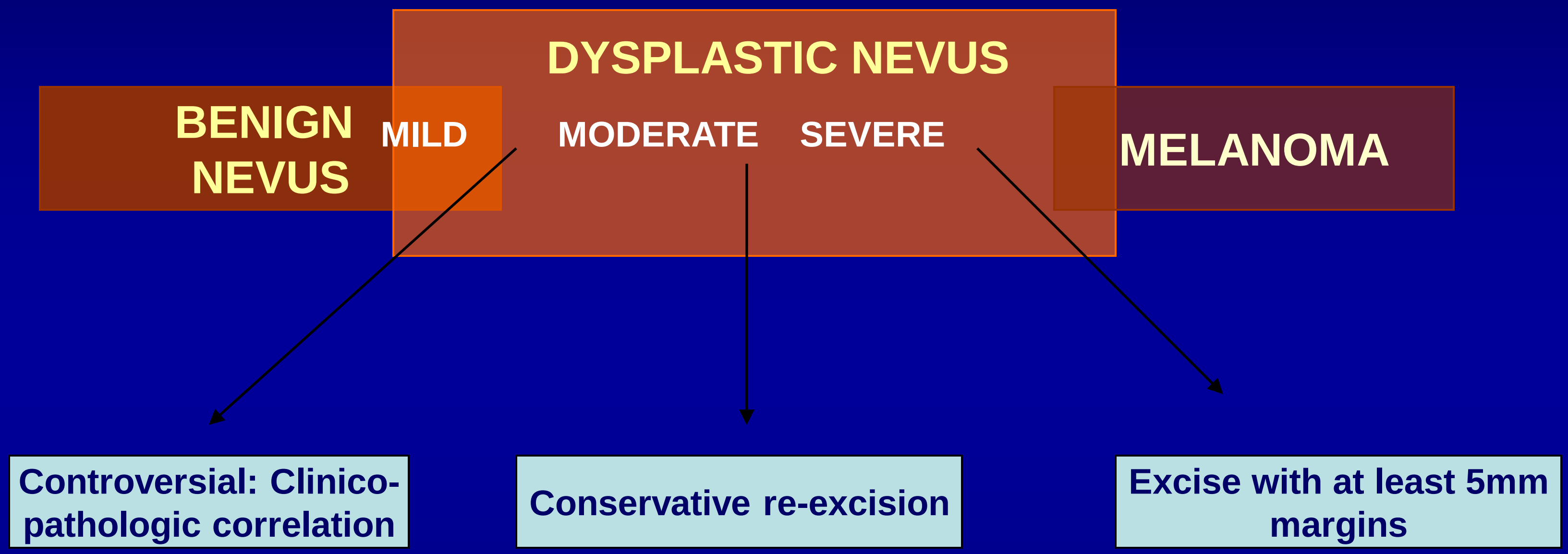
Practical Rationale for Grading of Dysplastic Nevi

Conveys to the dermatologist

- Some information about the pathologist's concern about the lesion.
- The possible need for obtaining a second opinion.
- The decision to perform a complete excision or not (and possible extent of excision).

Problems With Grading of Dysplastic Nevi

- Not highly reproducible from one pathologist to another and therefore highly controversial.
- Some pathologists and dermatologists use the term dysplastic and “atypical” nevi interchangeably.


DYSPLASTIC NEVUS
BENIGN NEVUS
MILD
MODERATE
SEVERE
MELANOMA
Controversial: Clinico-pathologic correlation
Conservative re-excision
Excise with at least 5mm margins

Lessons Learned from Medicolegal Cases.

- The diagnosis pertains only to the tissue submitted and assumes that the biopsy is representative .

- Claims involving dysplastic or atypical nevi appear to result in part from miscommunication between pathologist and dermatologist/clinician and most involve partial biopsies.

- Pathologist may use dysplastic or atypical nevus in a generic sense (i.e cytologically disturbing cells are present in lesion but lack diagnostic criteria for melanoma) and requests re-excision but the dermatologist may be reluctant to do so since to them these terms have a more specific meaning and connote a clinical syndrome or benign clinical entity.

NOTE:

- Optimal width of re-excision for atypical melanocytic lesions/indeterminate lesions are controversial, not based on “hard data” and are based largely on standard of care practices.

TAKE HOME MESSAGE!

- If there is disparity between the clinical and pathologic diagnosis, ask for a second read.
- Know your pathologist and his/her diagnostic tendencies in signing out melanocytic lesions.