

# Sinonasal Rosettes All Olfactories?

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**Universität Erlangen**



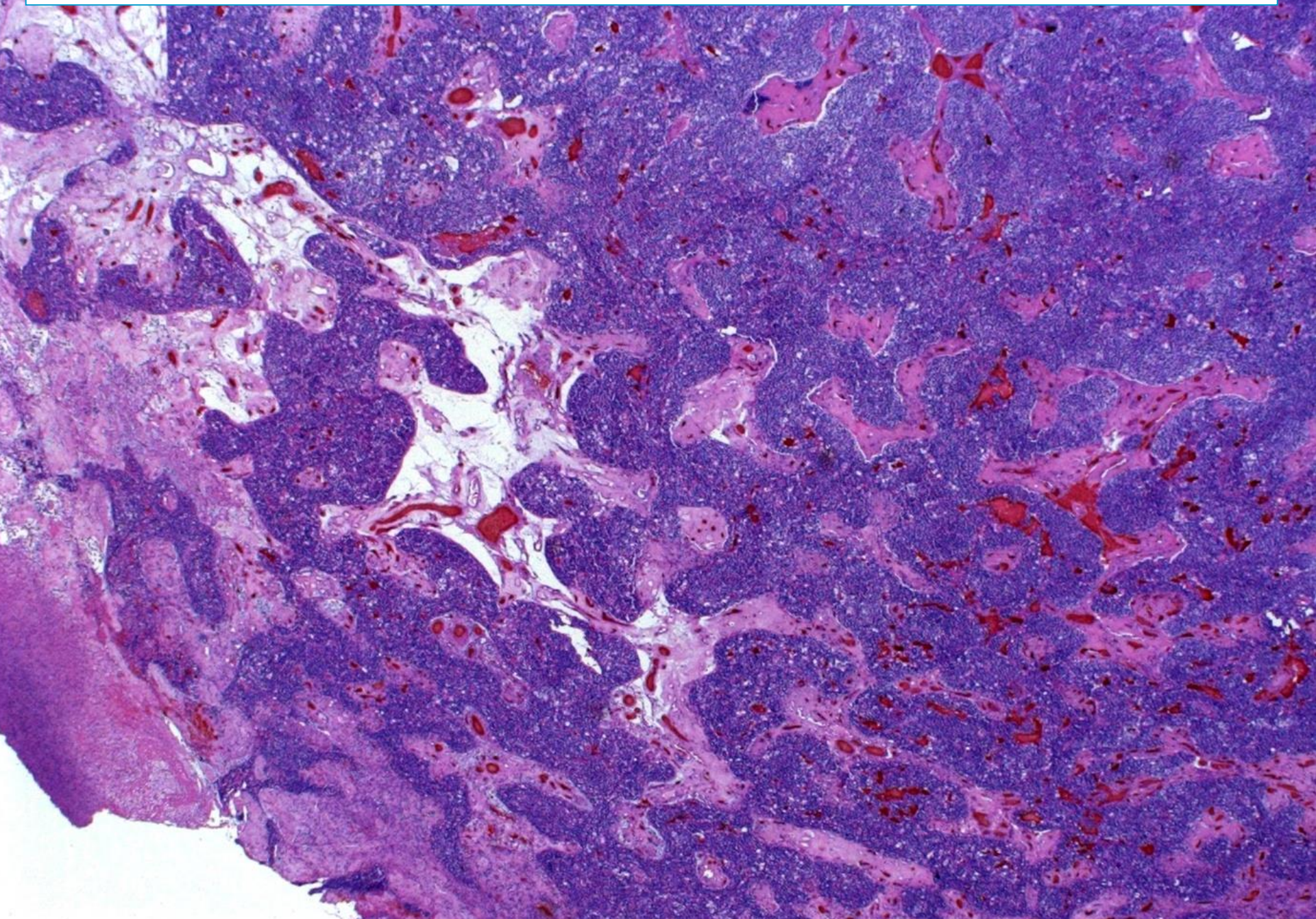
# Olfactory neuroblastoma--basics

- ✓ Very rare: 4/10 millions/yr.
- ✓ 3% of all sinonasal tumors.
- ✓ Upper nasal cavity (cribriform plate).
- ✓ Rarely ectopic in ethmoid & others.
- ✓ Age: 2-90 yrs (males slightly overrepresented)
- ✓ IHC: Syn+, Chromo-A+, CD56+, NSE+ & Calretinin+
- ✓ CK18/cam5.2 + in up to 1/3!!! But different pattern.



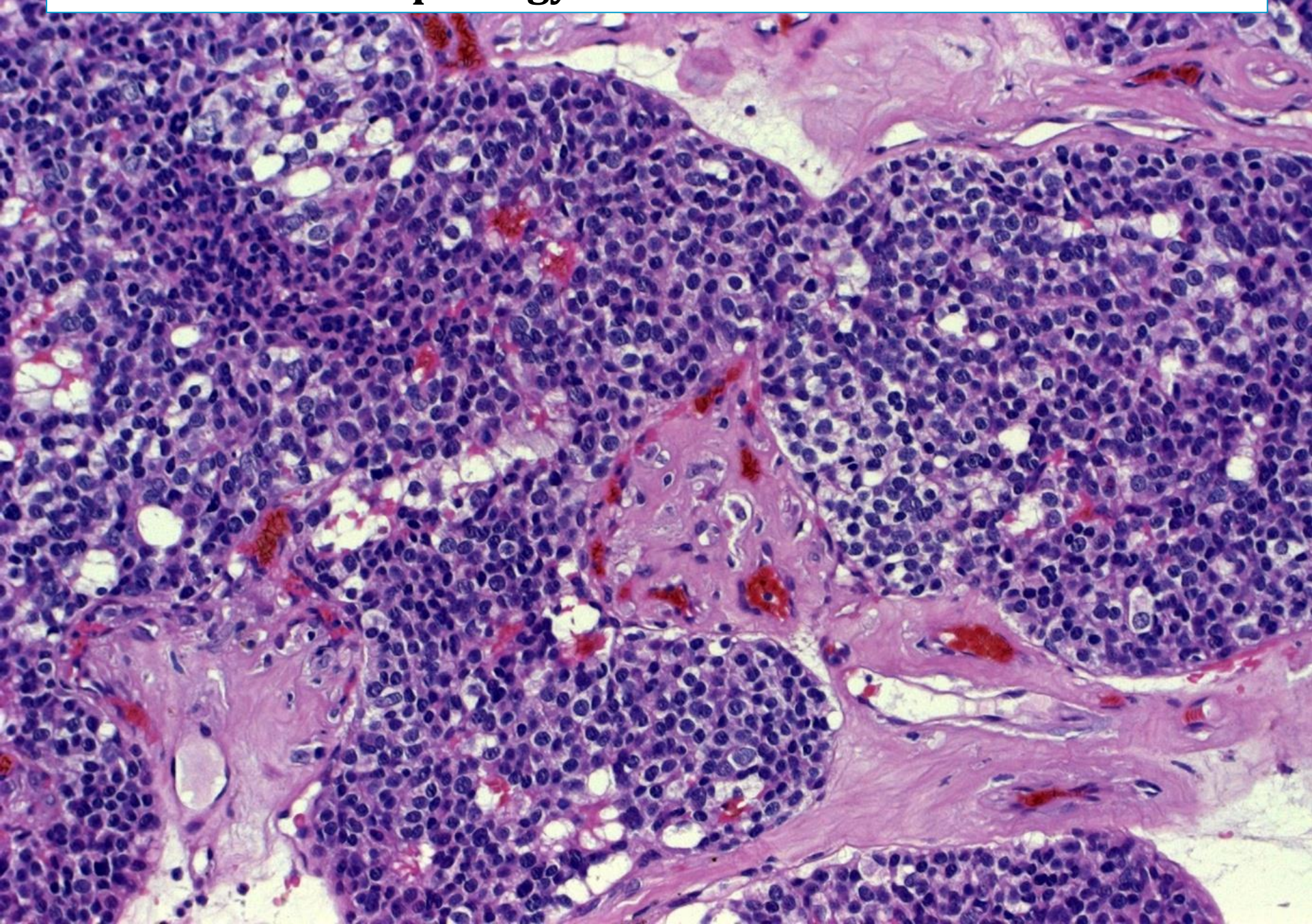


**Prototypical ONB: similar to paraganglioma & well diff NET**

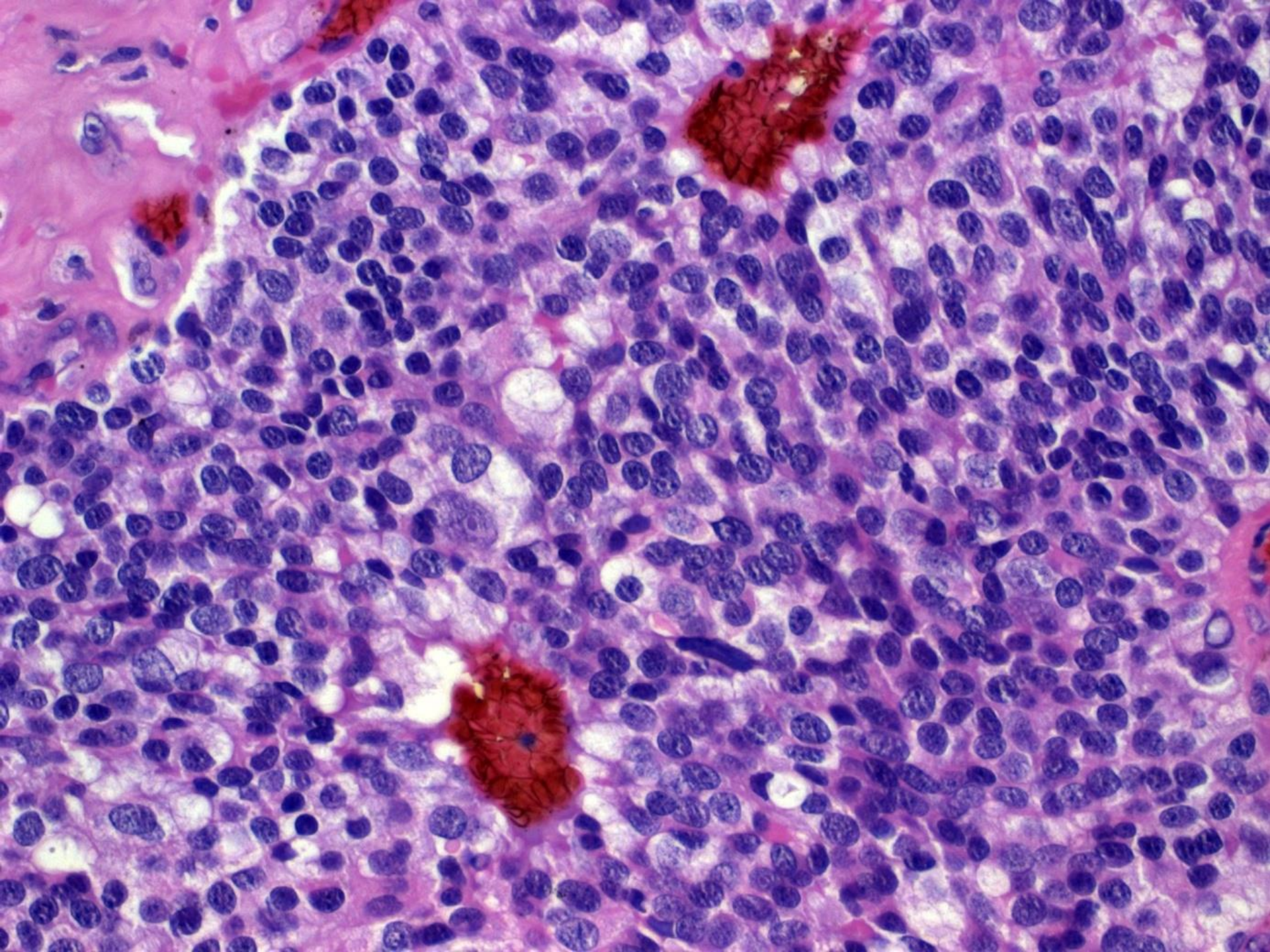




**Both nuclear morphology & architecture = neuroendocrine**

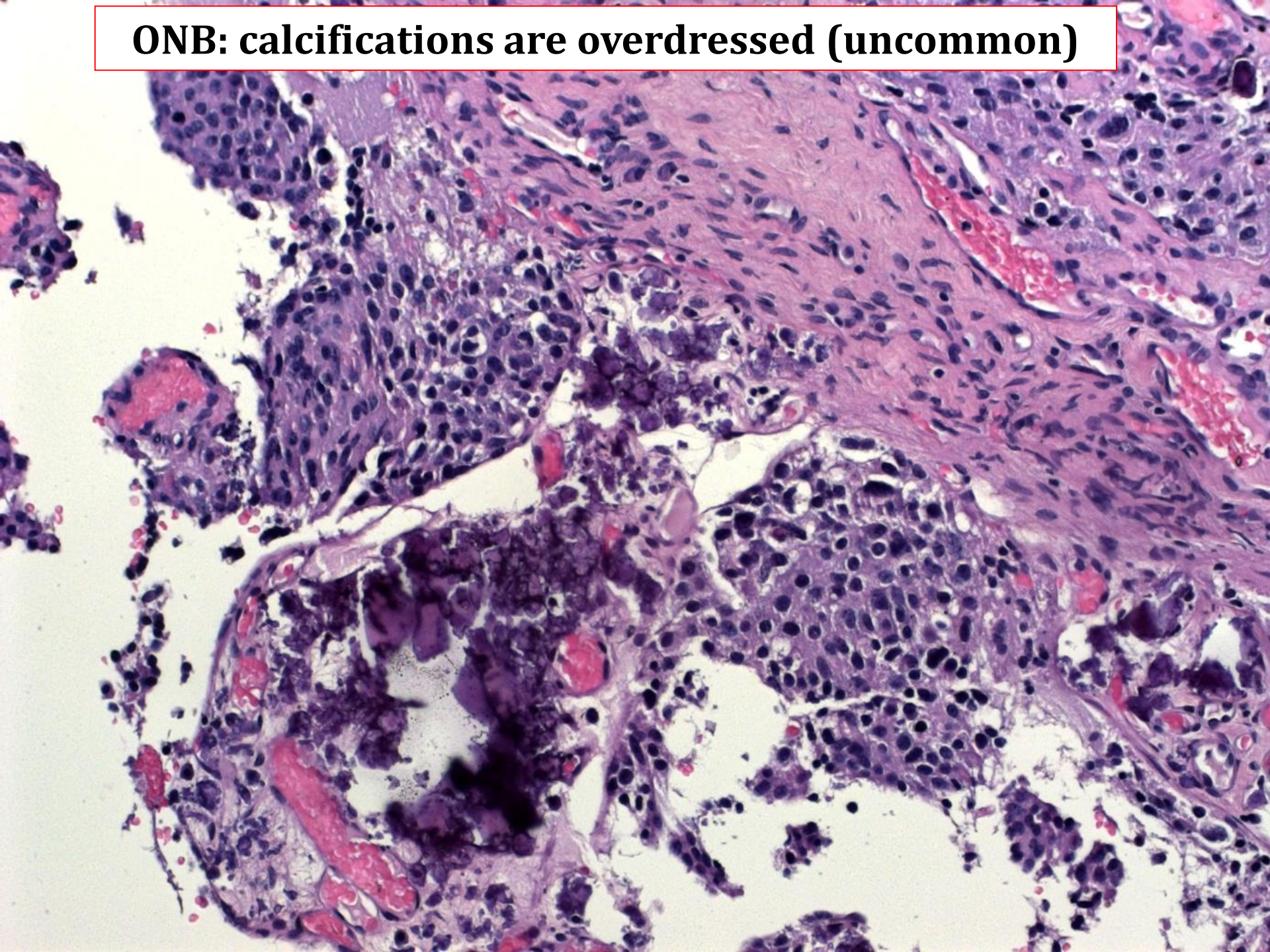






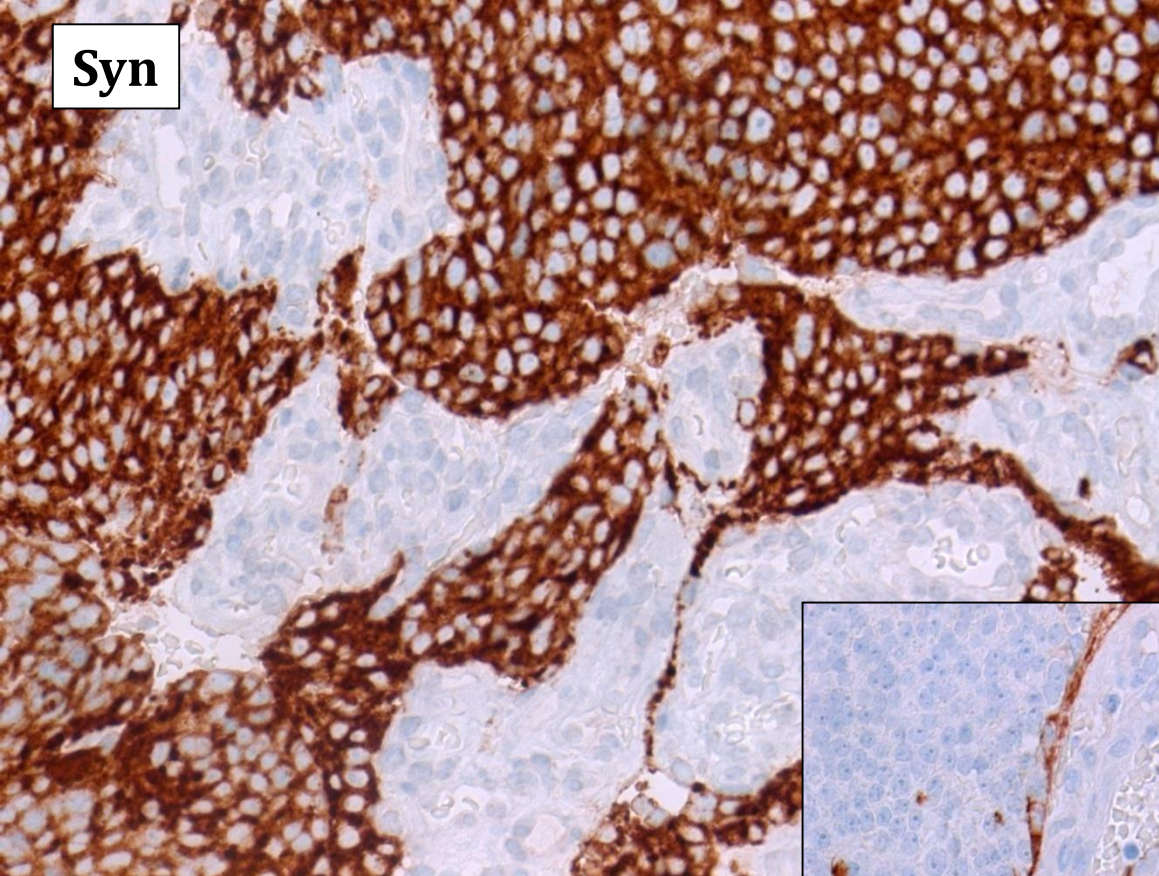


**ONB: calcifications are overdressed (uncommon)**

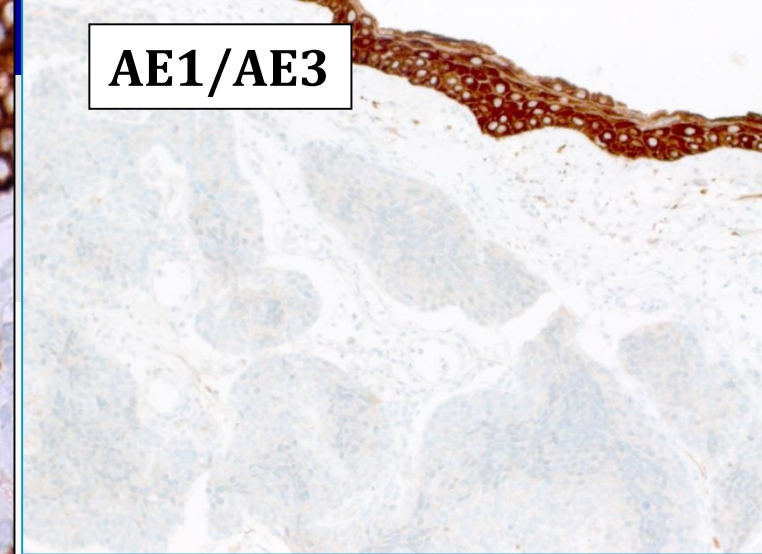




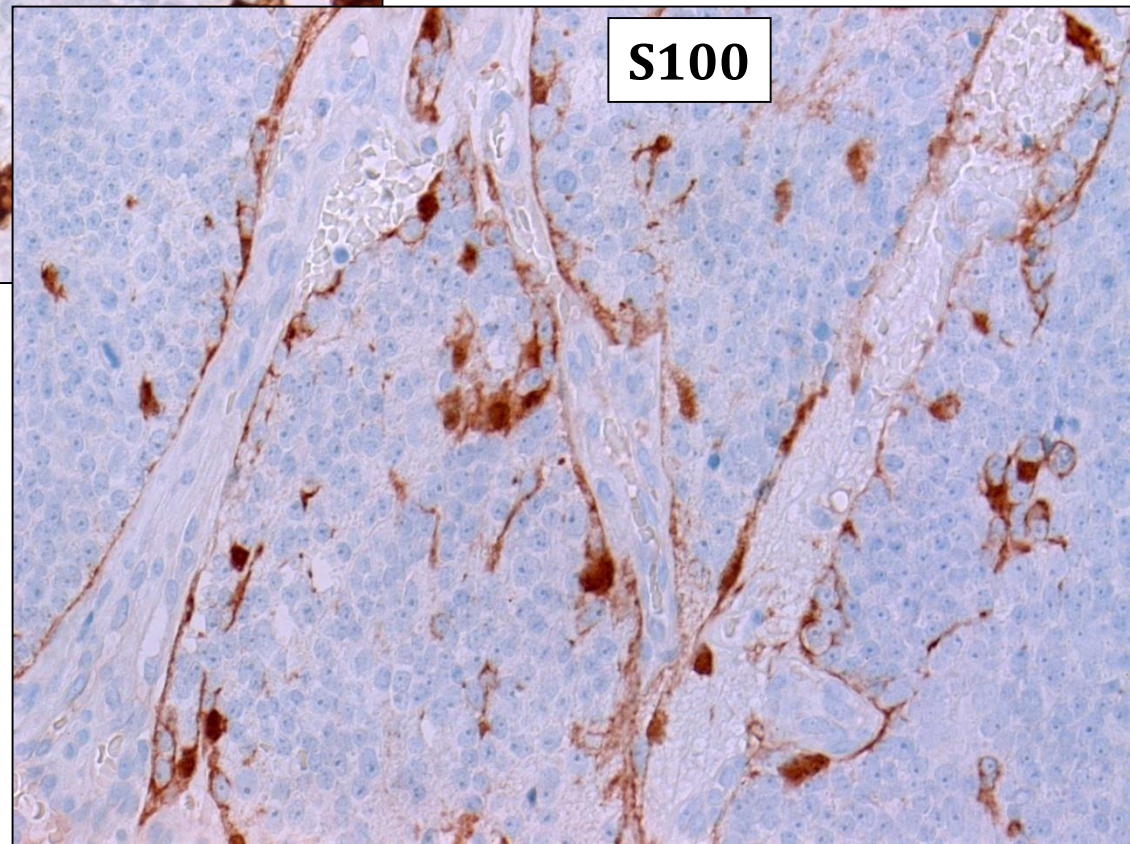
**Syn**



**AE1/AE3**

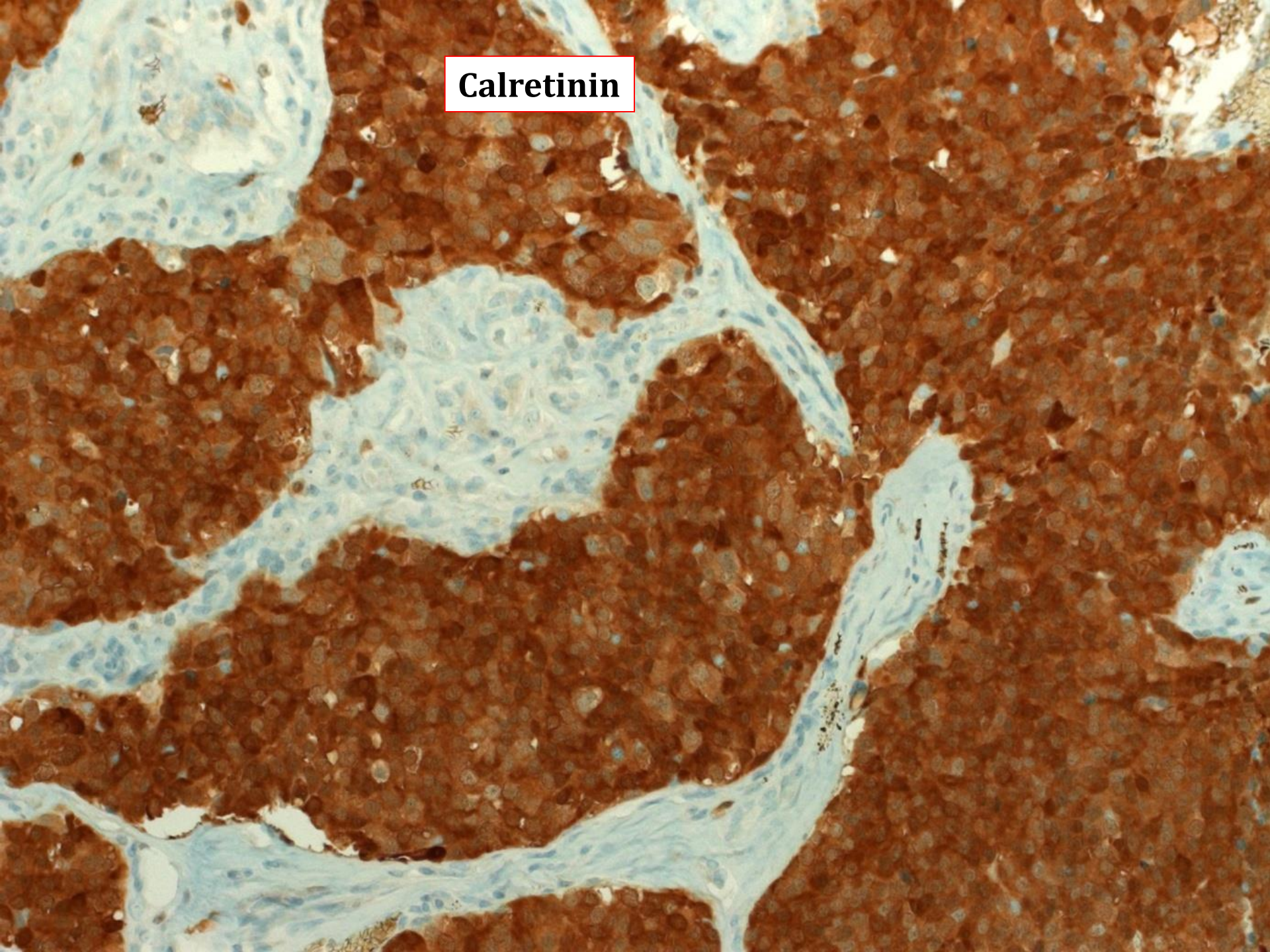


**S100**



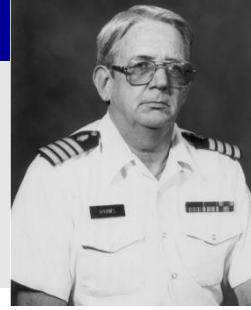


# Calretinin





# HYAMS Grading

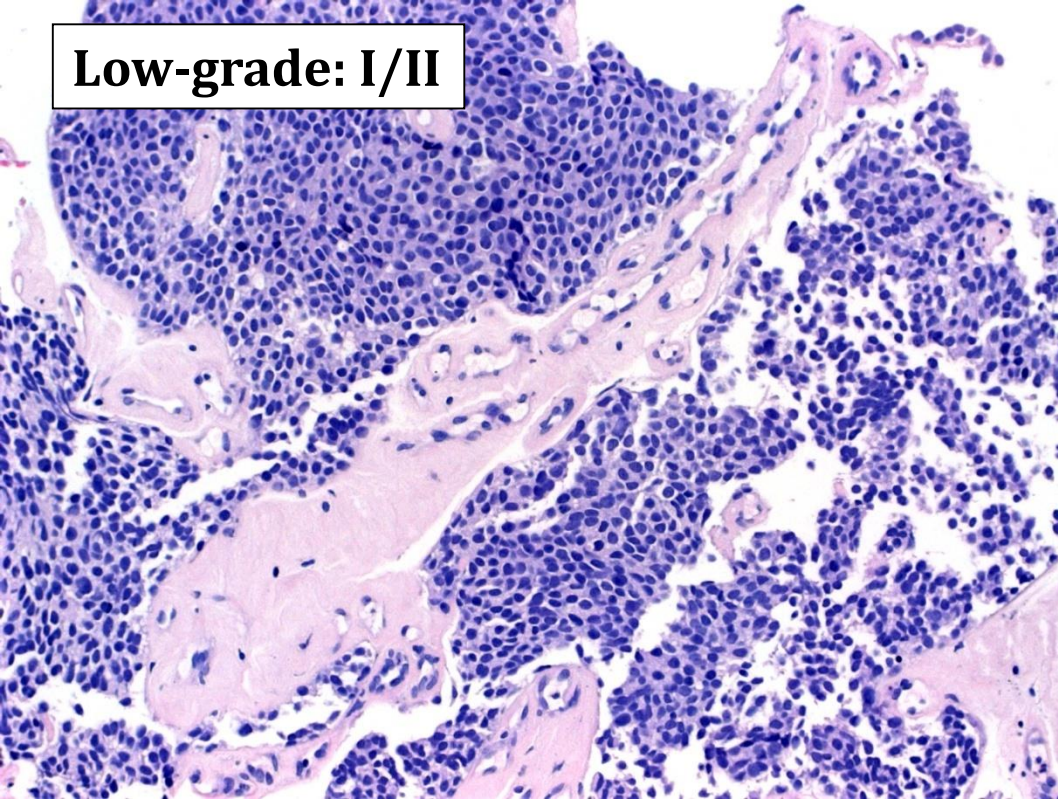


Features	Grade I	Grade II	Grade III	Grade IV
Architecture	Lobular	Lobular	Variable	Variable
Mitoses	Absent	Present	Prominent	High
Nuclear pleomorphism	Absent	Moderat	Prominent	High
Fibrillary matrix	Prominent	Vorhanden	Minimal	Absent
Necrosis	Absent	Absent	May be present	Common
Rosettes	Homer-Wright	Homer-Wright	Flexner-Wintersteiner	Flexner-Wintersteiner

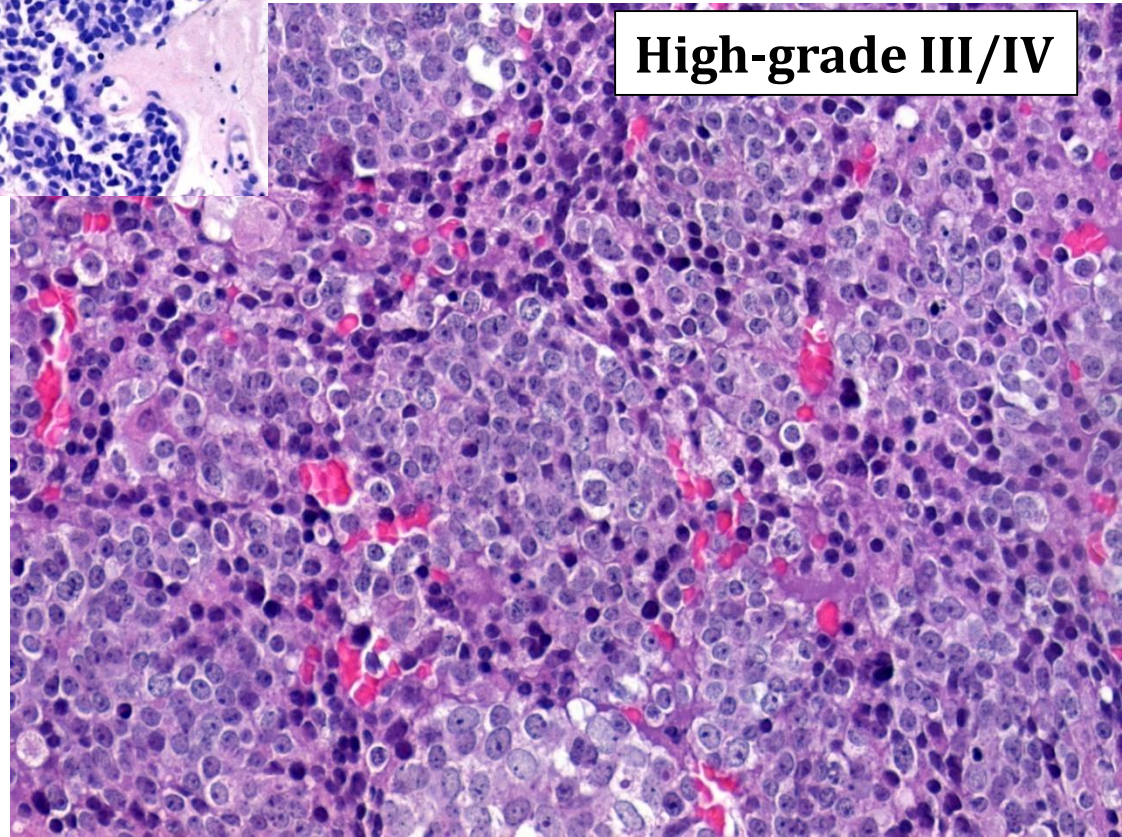




**Low-grade: I/II**

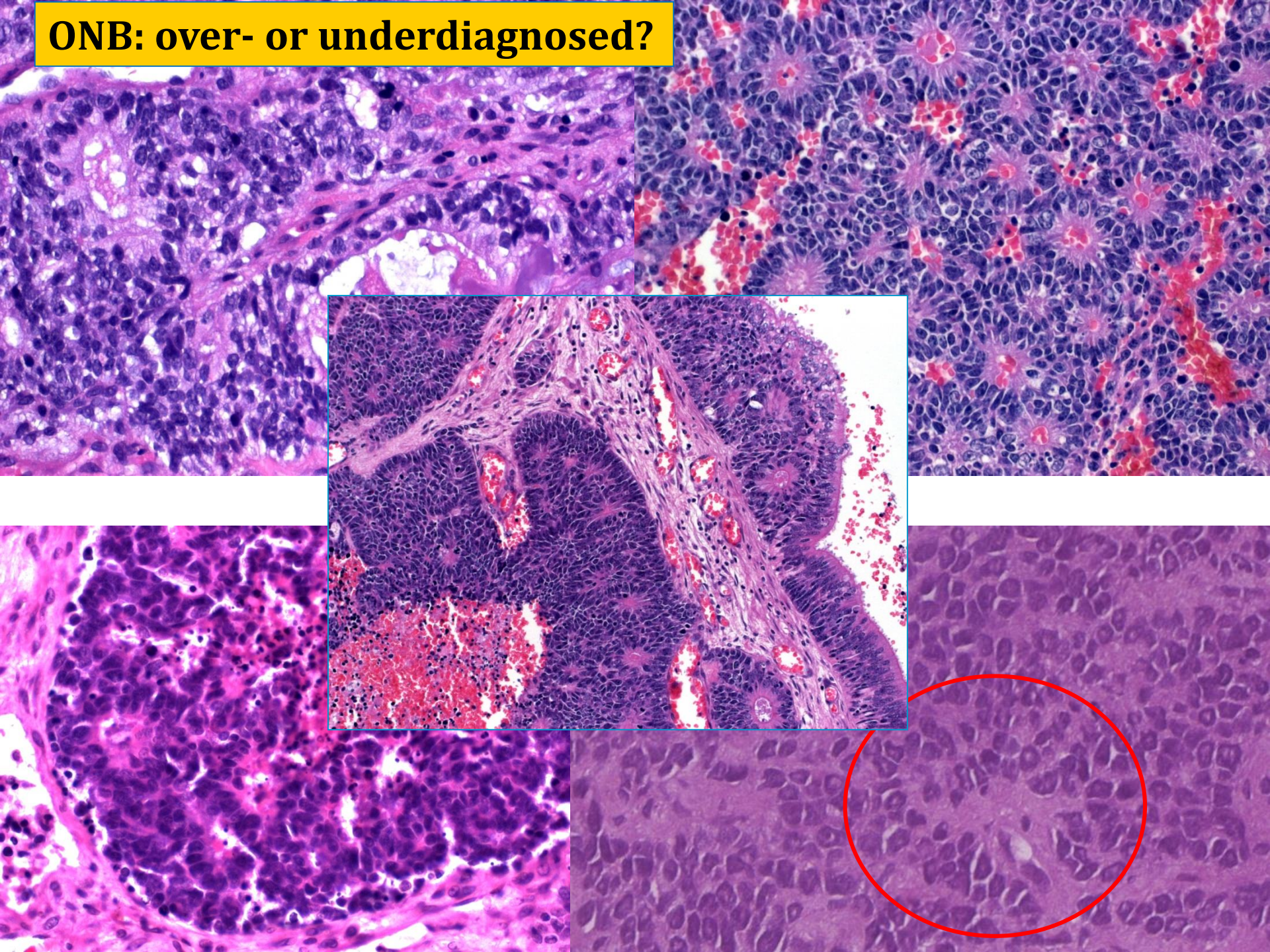


**High-grade III/IV**





# ONB: over- or underdiagnosed?





# Misdiagnoses of ONB (MDACC)

- 12 pts with external diagnosis
- After Review:
- Only 2 ONB confirmed:
  - **ONB: 2 (16%)**
  - SNUC: 2
  - Melanoma: 2
  - NEC: 3
  - Pituitary adenoma: 3

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Showing results for *cohen neuroblastoma marmor*. Your search for *cohen neuroblastoma marmor* retrieved no results.

*Neurosurg Focus*. 2002 May 15;12(5):e3.

**Misdiagnosis of olfactory neuroblastoma.**

Cohen ZR<sup>1</sup>, Marmor E, Fuller GN, DeMonte F.

Author information

**Abstract**

**OBJECT:** Olfactory neuroblastoma (ON) is a rare neoplasm arising from the olfactory epithelium and found in the upper nasal cavity. The authors studied the frequency with which ON is misdiagnosed with other tumors of the paranasal sinuses such as neuroendocrine carcinoma (NEC), pituitary adenoma, melanoma, lymphoma, and sinonasal undifferentiated carcinoma (SNUC). Based on the belief that misdiagnosis commonly occurs, they emphasized the importance of establishing the correct diagnosis, because the treatment regimens and prognosis of these tumor types are often significantly different.

**METHODS:** Twelve consecutive patients in whom ON was diagnosed were referred to the Department of Neurosurgery at the M. D. Anderson Cancer Center between January 1998 and March 2000. Demographic data were collected, physical findings and mode of treatments were documented, and neuroimaging studies were assessed. Pathologists at the authors' institute reviewed the histological specimens. Only in two of 12 patients was the diagnosis of ON confirmed. Lesions in 10 patients were misdiagnosed; there were two cases of melanoma, three cases of NEC, three cases of pituitary adenoma, and two cases of SNUC. Eight of 10 patients in whom lesions were misdiagnosed required significant alteration in the initially proposed treatment plan.

**CONCLUSIONS:** Neurosurgeons should be acutely aware of the variety of neoplasms that occur in the paranasal region. The correct diagnosis should be ensured before initiating treatment to provide the optimum therapy and spare the patients from needless and potentially toxic treatment.





## Diagnose Fall 29 (IAP, Bonn, 2018): Olfaktorius Neuroblastom

### Diagnosevorschläge (HE):

**Esthesioneuroblastom**

**12/38**

Hämangioperizytom

8

Neuroendokrines Karzinom

7

Paragangliom

2

Ewing-Sarkom/PNET

3

Sonstiges

6

**Significantly underdiagnosed by general pathologists!!!  
Significantly overdiagnosed by neuropathologists!!!**



# Misdiagnoses of ONB: why frequent?

- **Low-grade tumors = overlap with**
  - Pituitary adenomas
  - Low-grade NETs/carcinoids.
- ONB with rhabdomyogenic diff = RMS???
- **High-grade tumors = overlap with**
  - SNUC
  - Small & large cell NEC
  - Melanoma with NE differentiation
  - CK+ solid alveolar RMS
  - Teratocarcinosarcoma & other undifferentiated carcinomas.



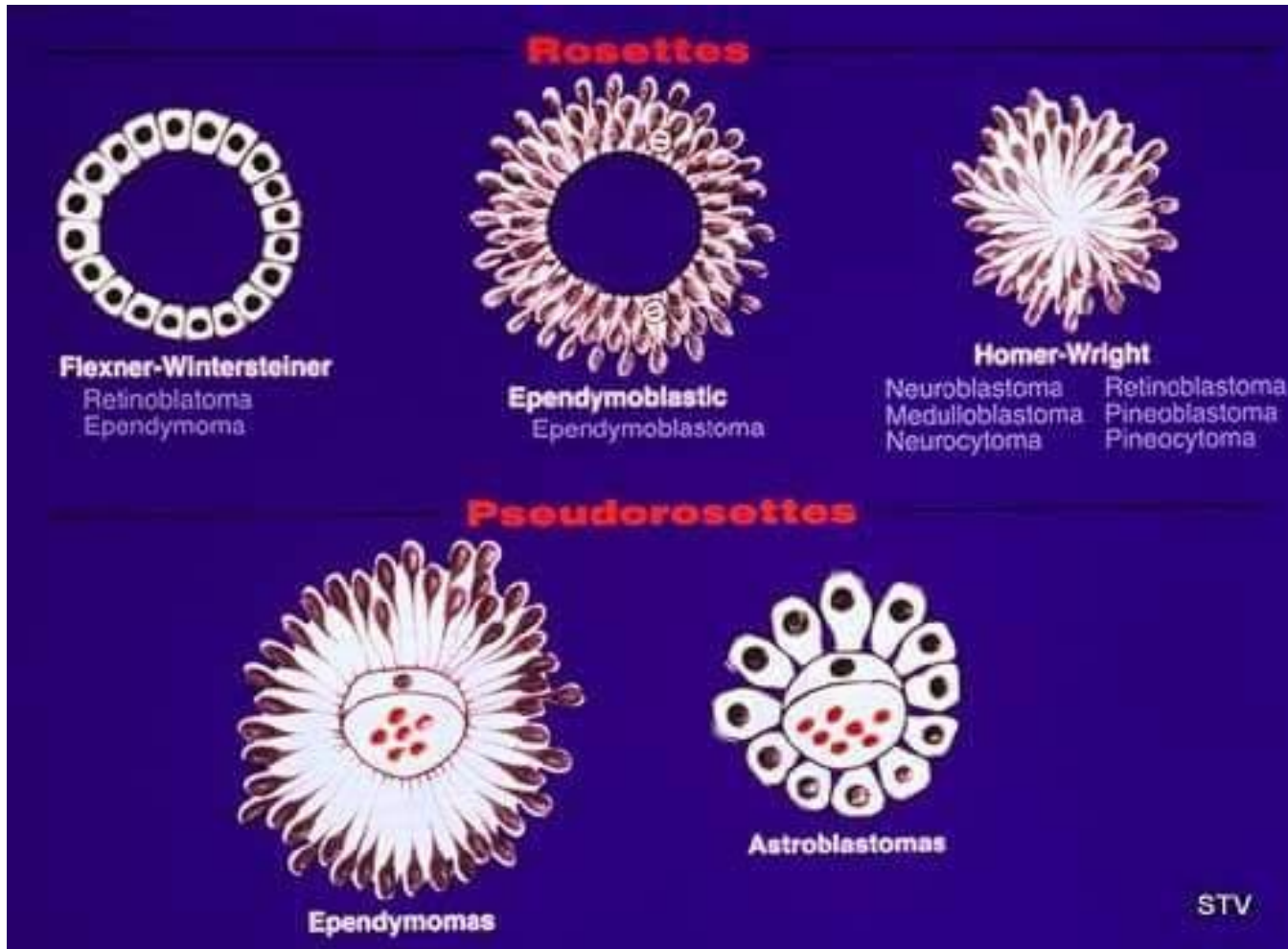
# What is the major problem with ONB



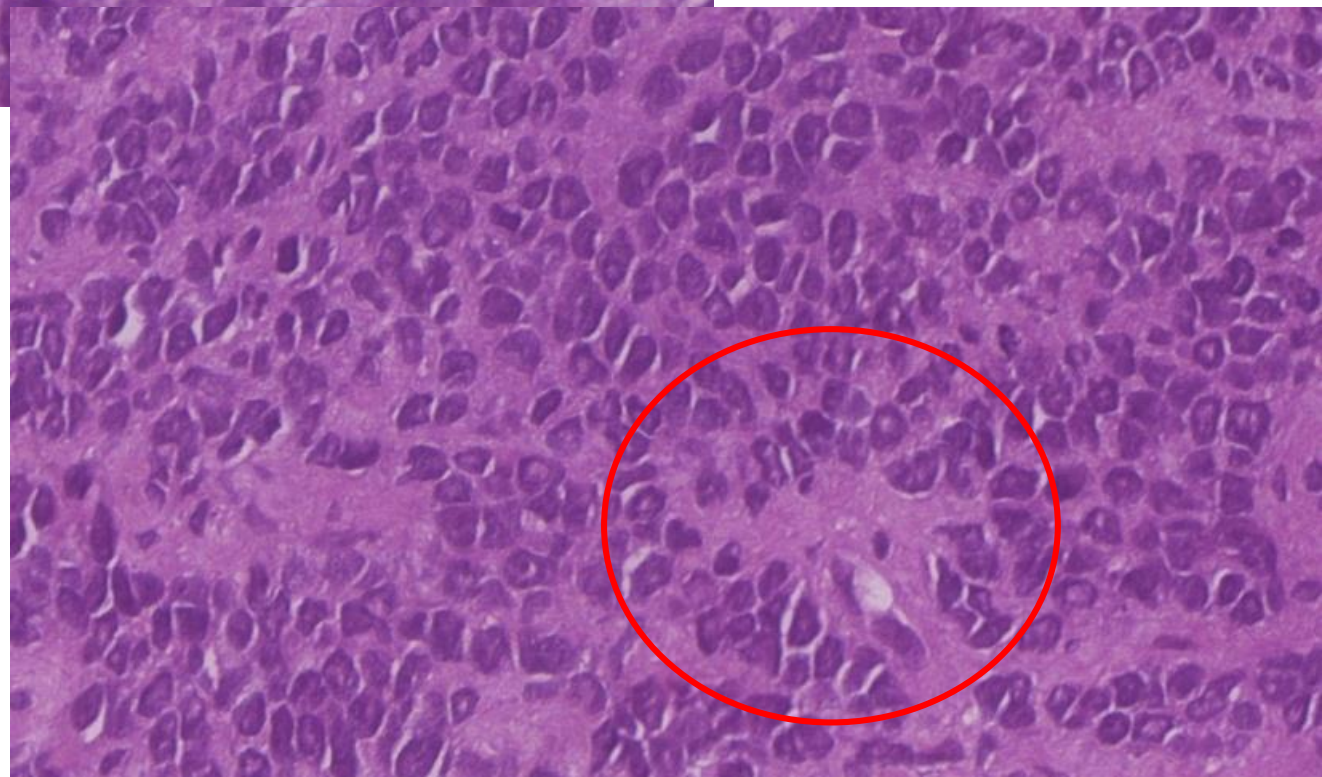
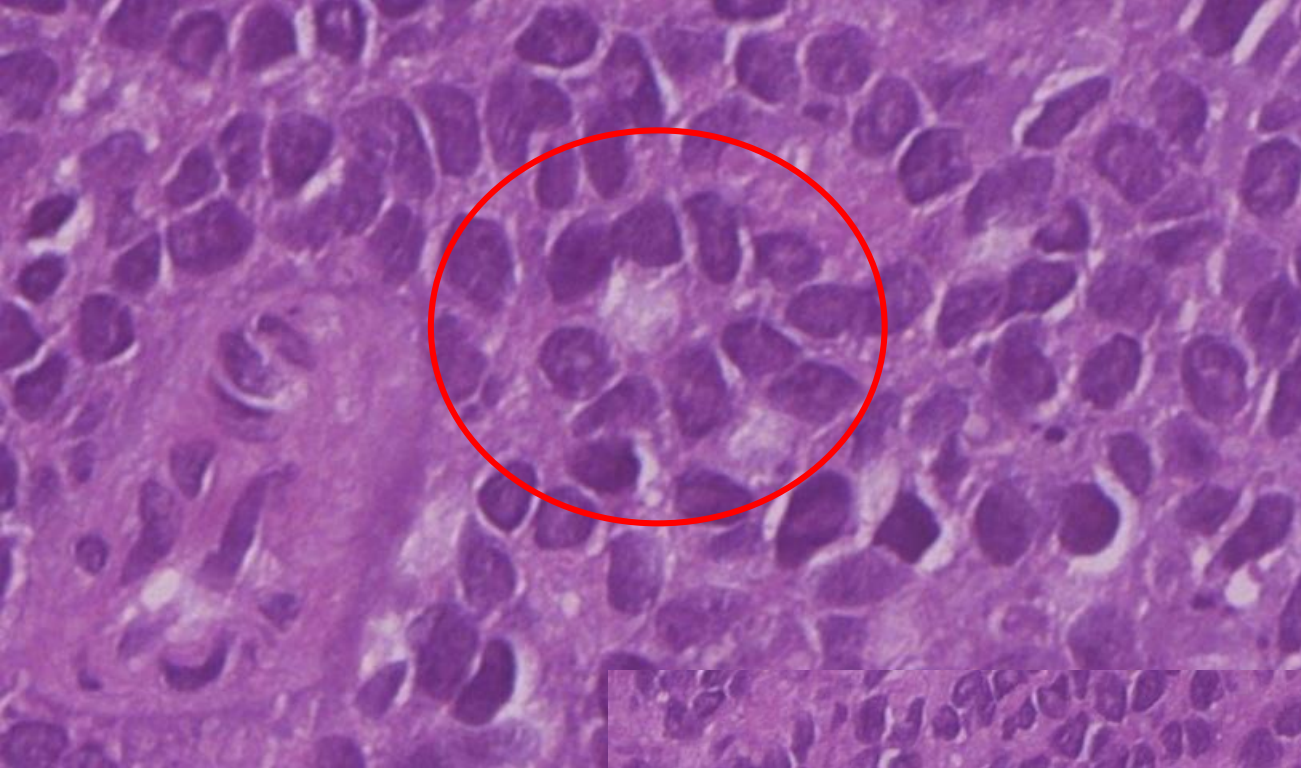


# What is the major problem with ONB

## 1.Problem: the rosettes

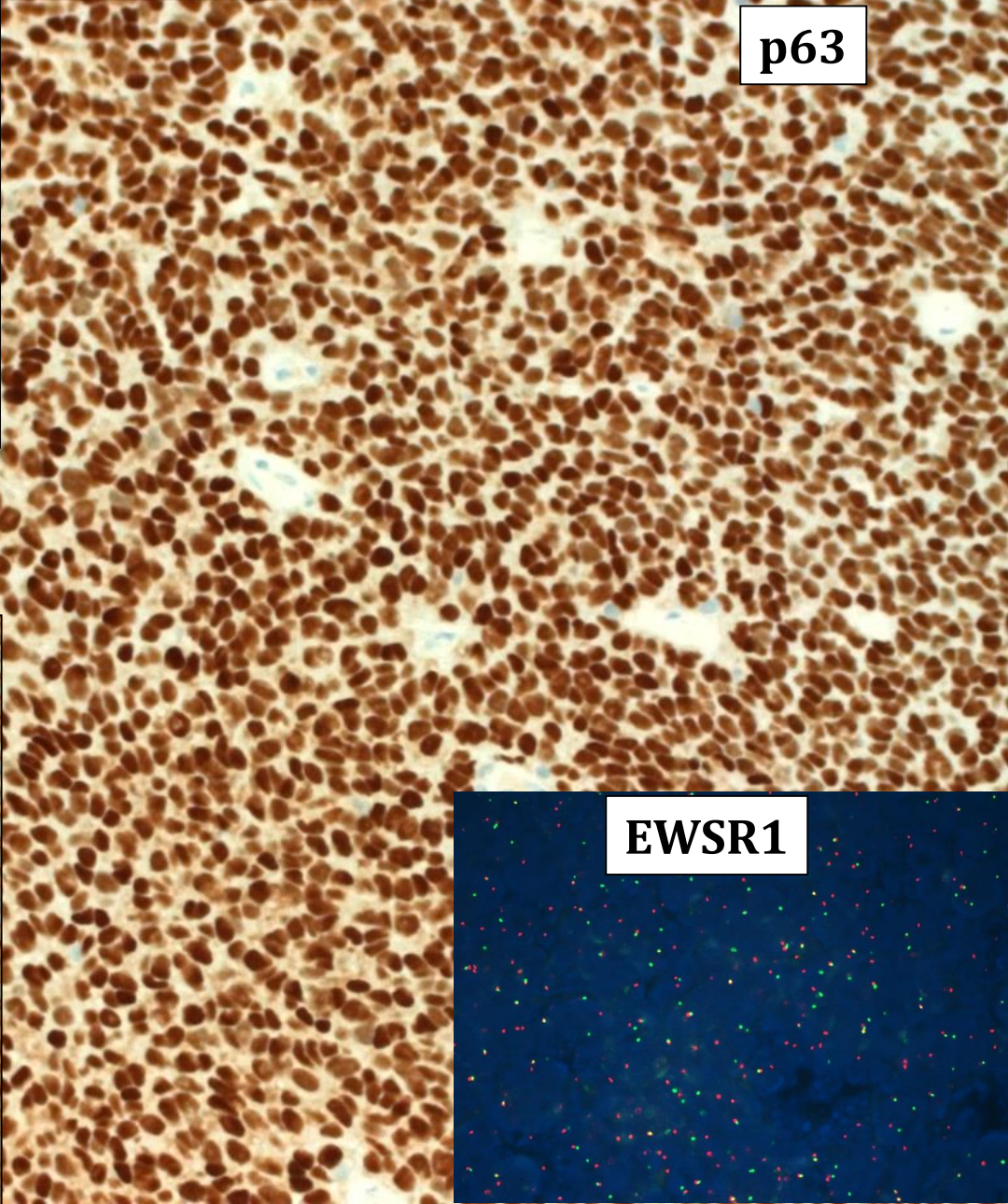
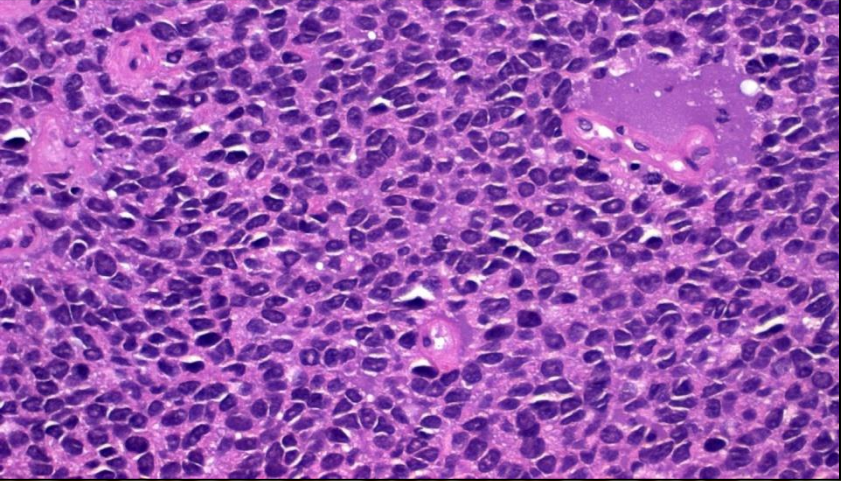




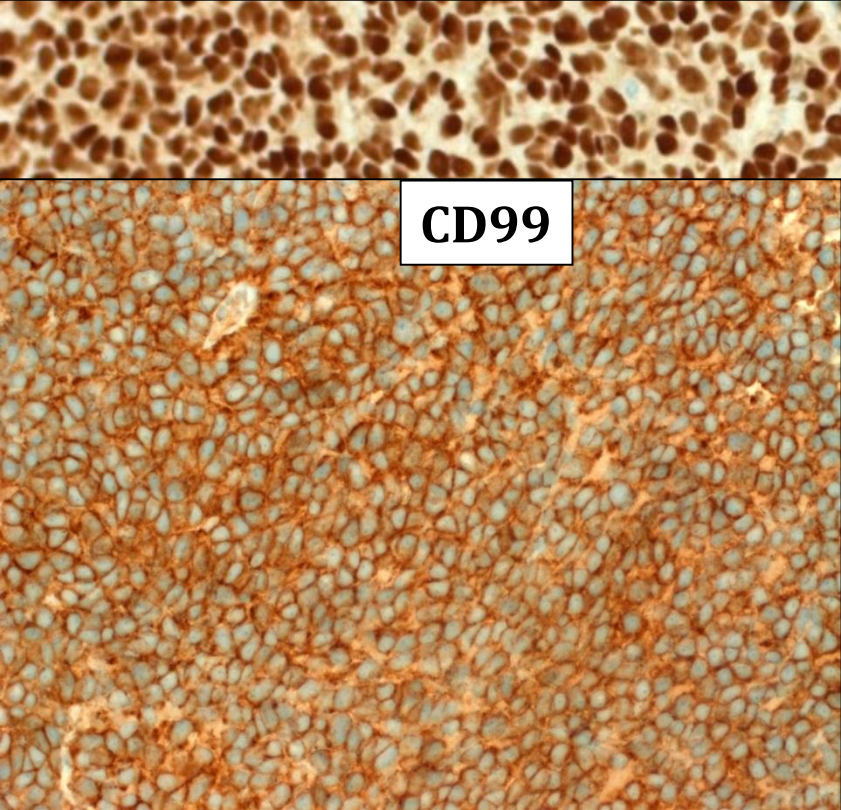




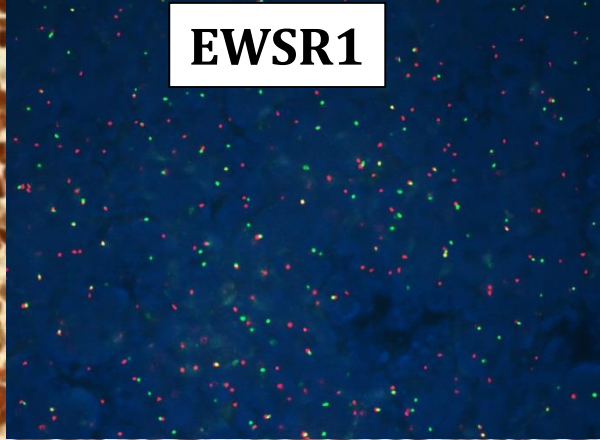
**Ewing sarcoma with epithelial diff  
(adamantinoma-like)**



**p63**



**CD99**



**EWSR1**



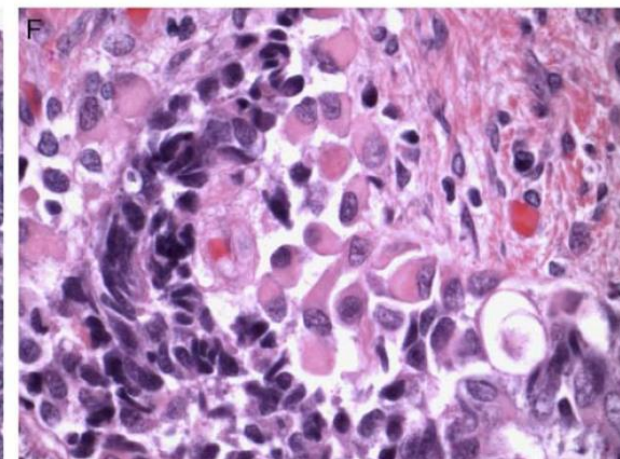
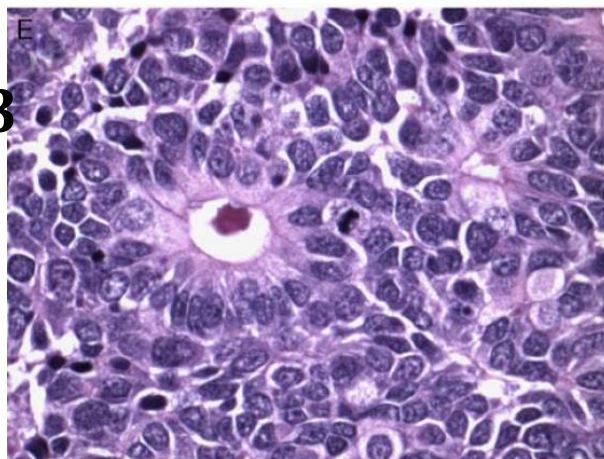
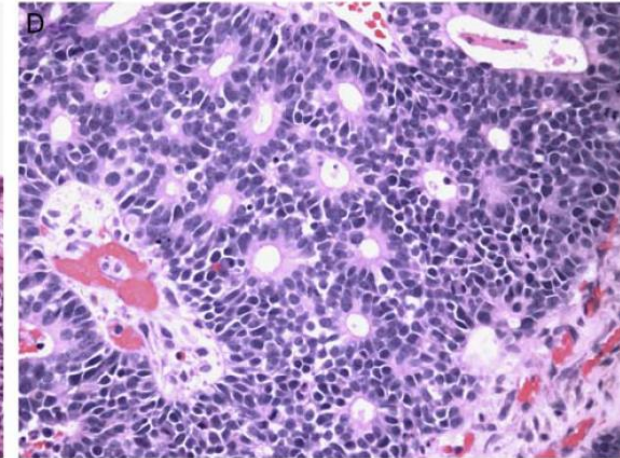
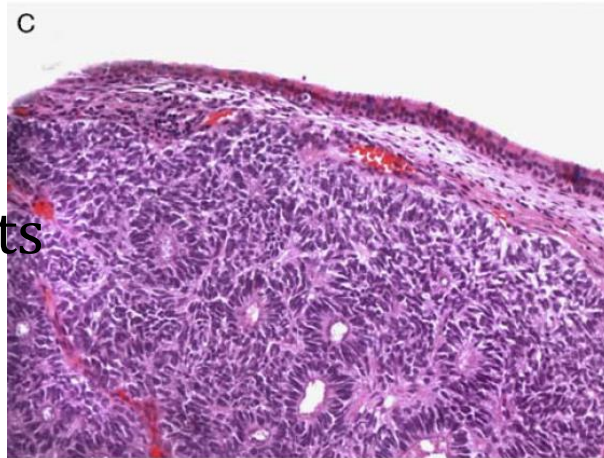
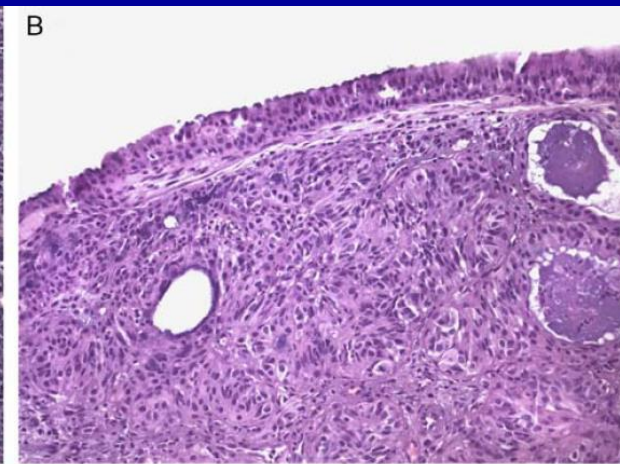
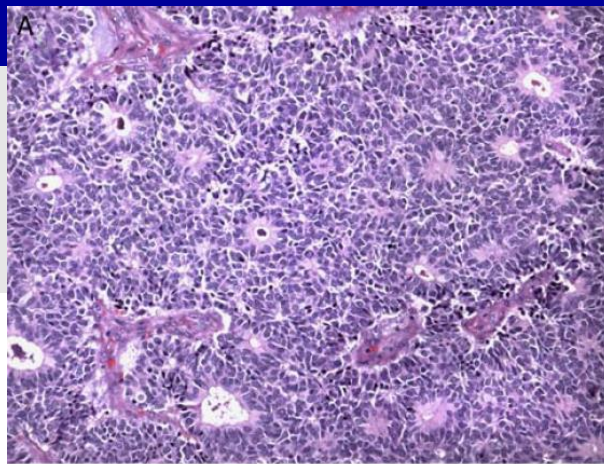
# A Histologic and Immunohistochemical Study Describing the Diversity of Tumors Classified as Sinonasal High-grade Nonintestinal Adenocarcinomas

*Edward B. Stelow, MD,\* Vicky Y. Jo, MD,\* Stacey E. Mills, MD,\* and Diane L. Carlson, MD†*

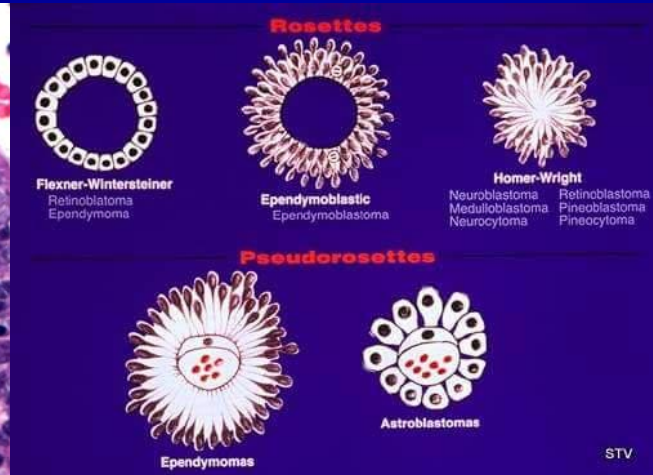
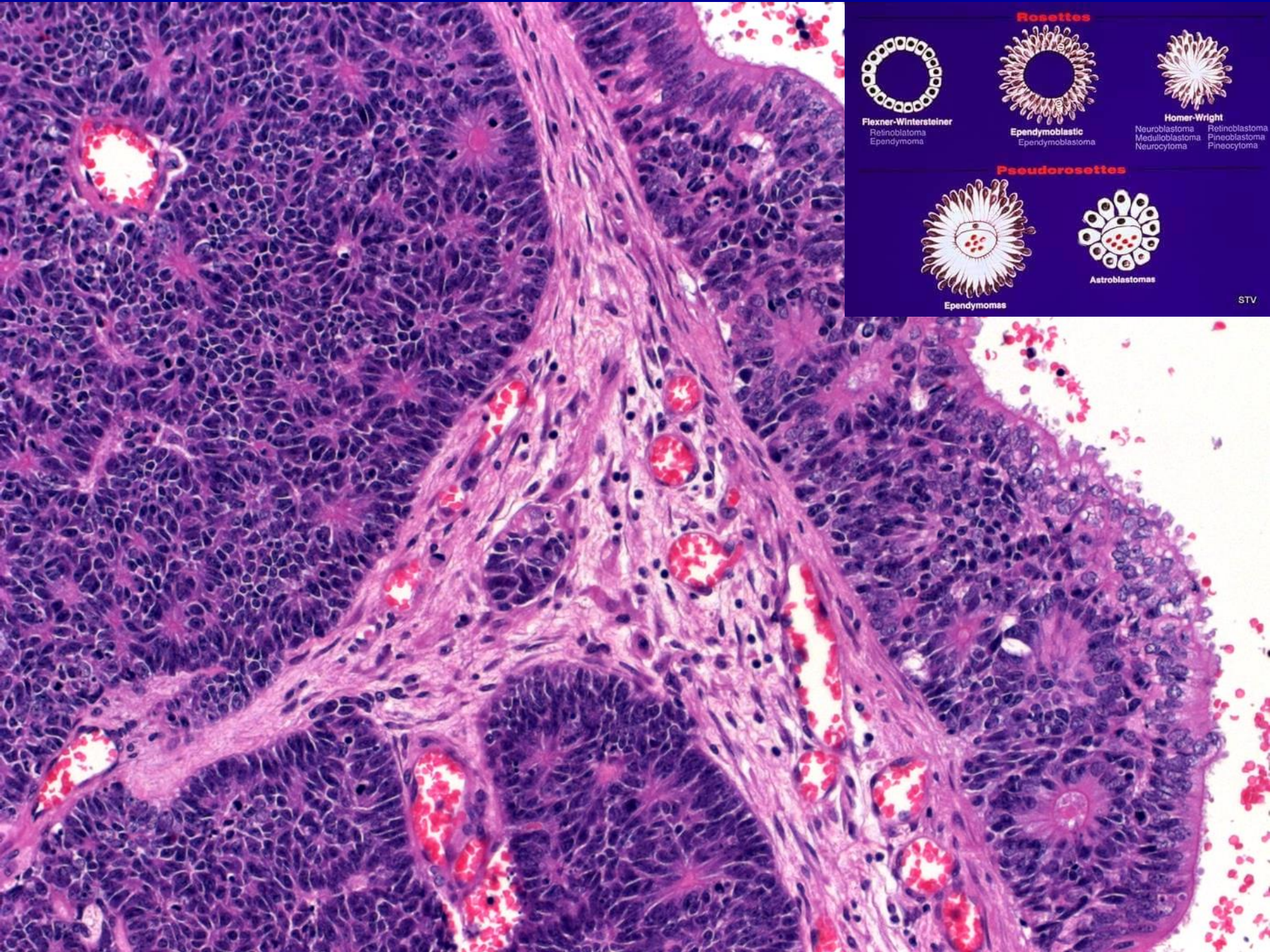


- 9 Pts (7 M/2 F)
- Median age: 56y
- 2 had sparse stroma
- 1 with rhabdomyoblasts
- 1 with neural diff
- Variable IHC +:

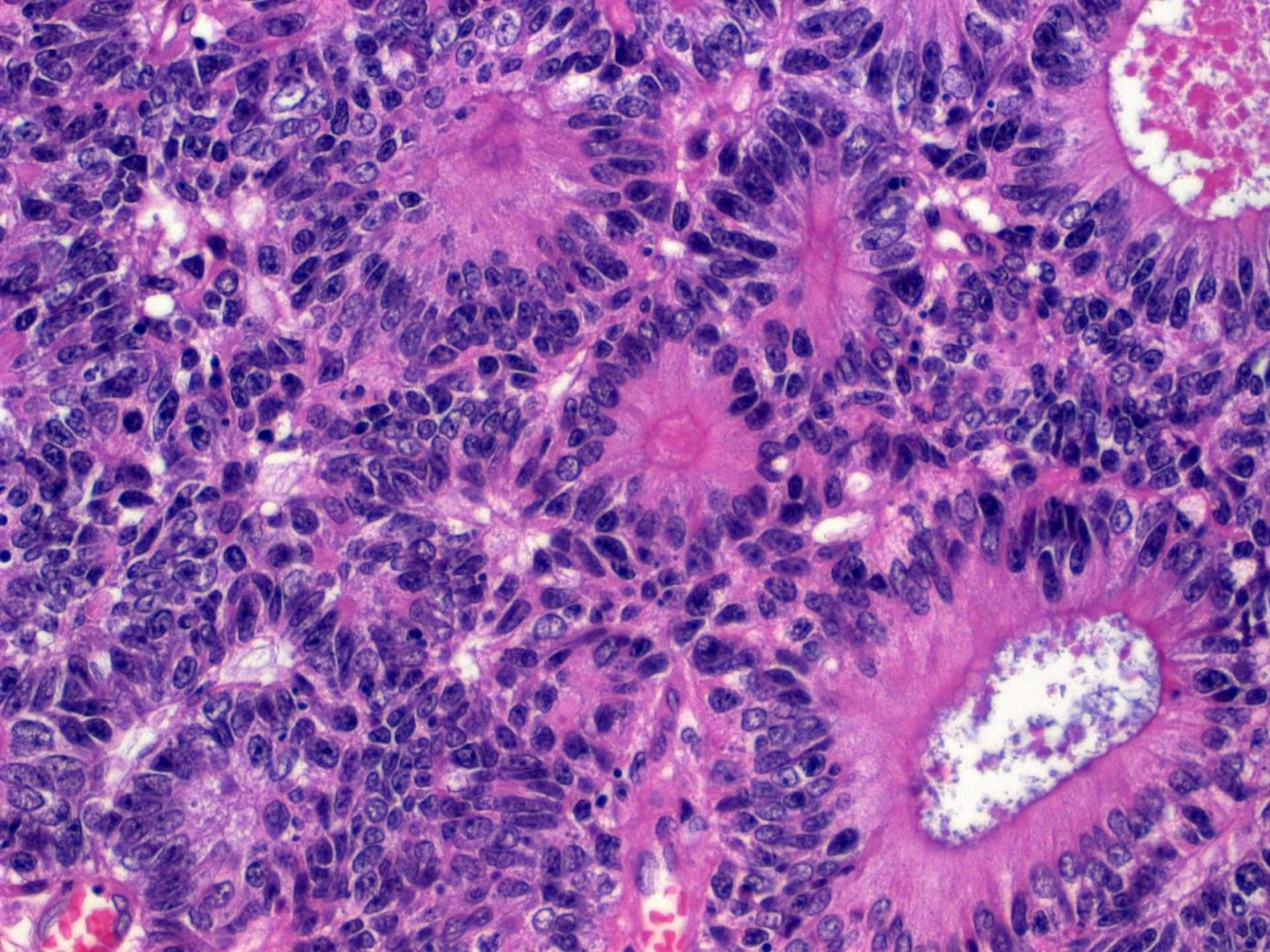
**CK7, S100, *Synapto*, p63**



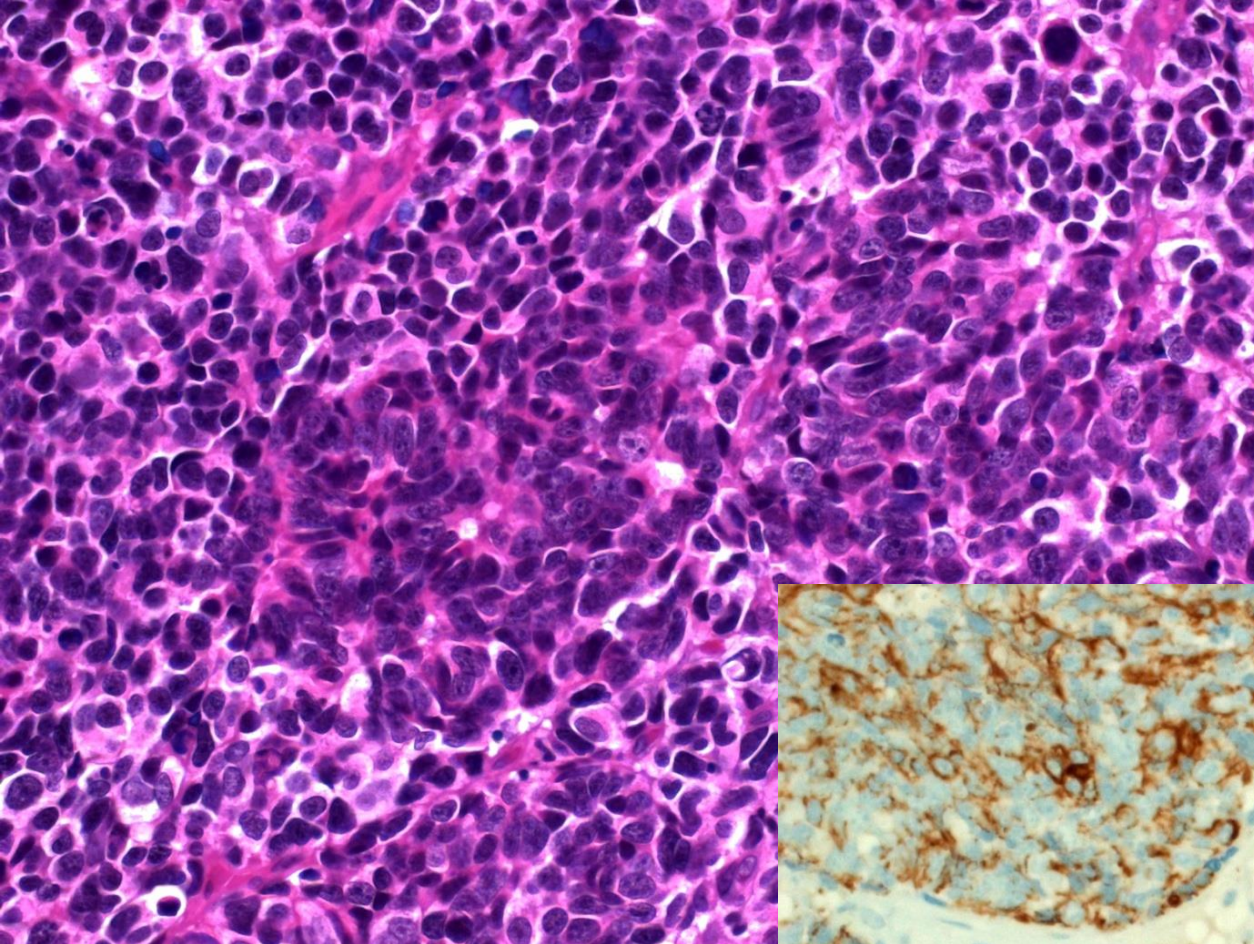




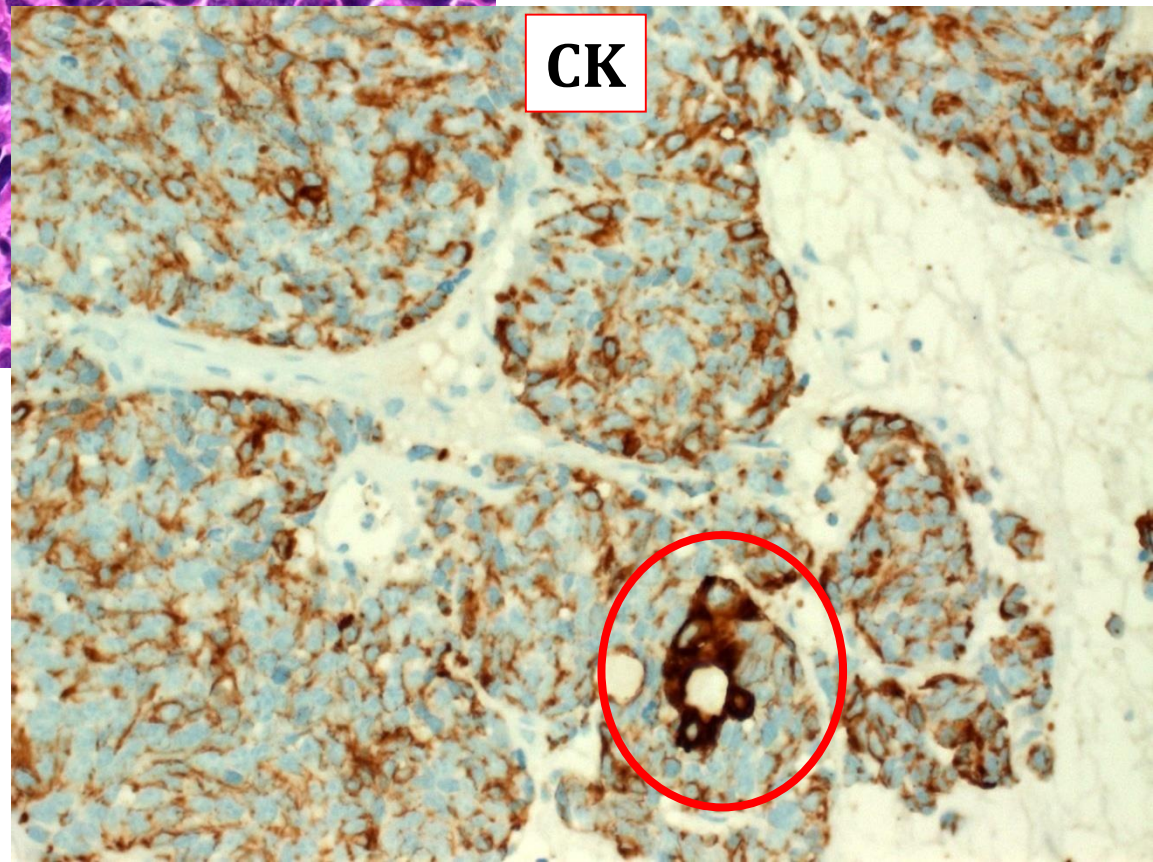








**Contradictorily,  
rosettes are  
Strongly CK+**



**CK**



# *Teratocarcinosarcoma (Malignant Teratoma?) of the Nasal Cavity and Paranasal Sinuses*

## *A Clinicopathologic Study of 20 Cases*

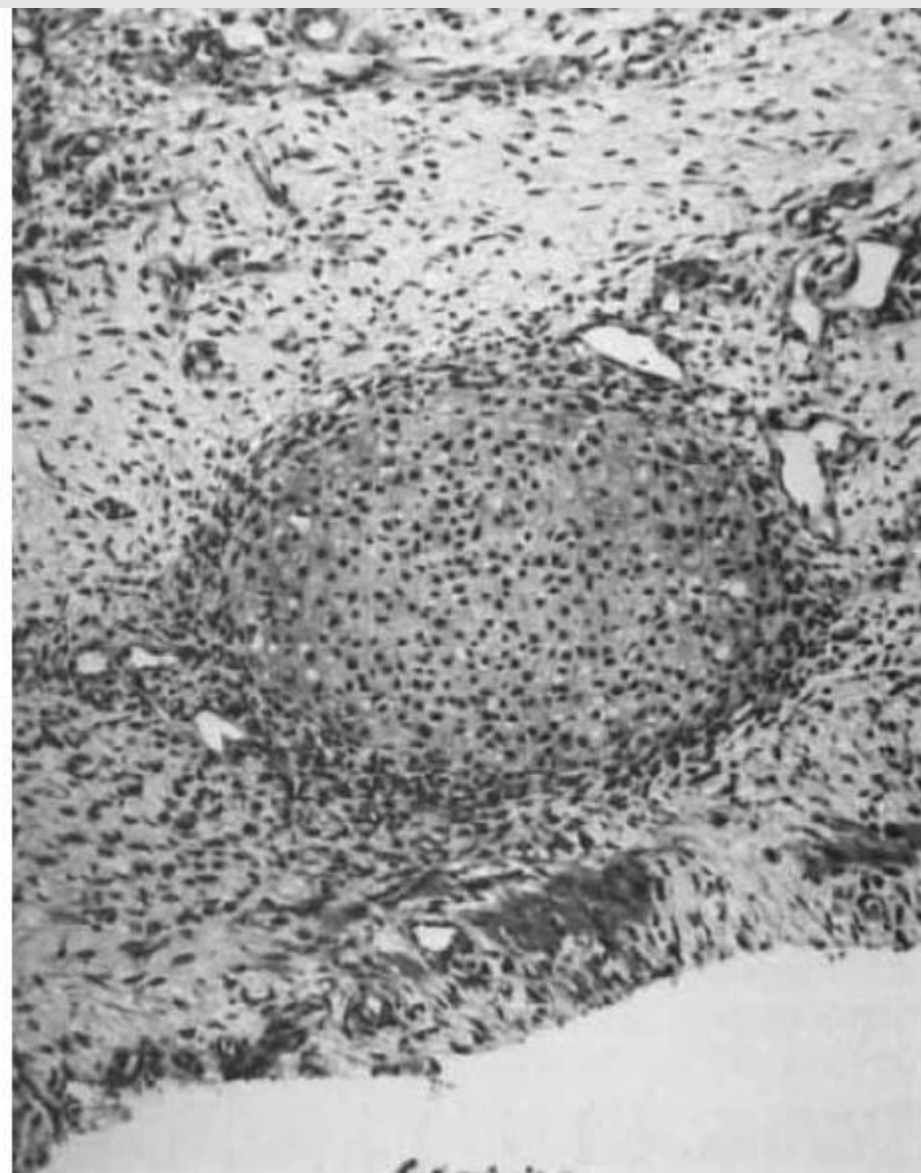
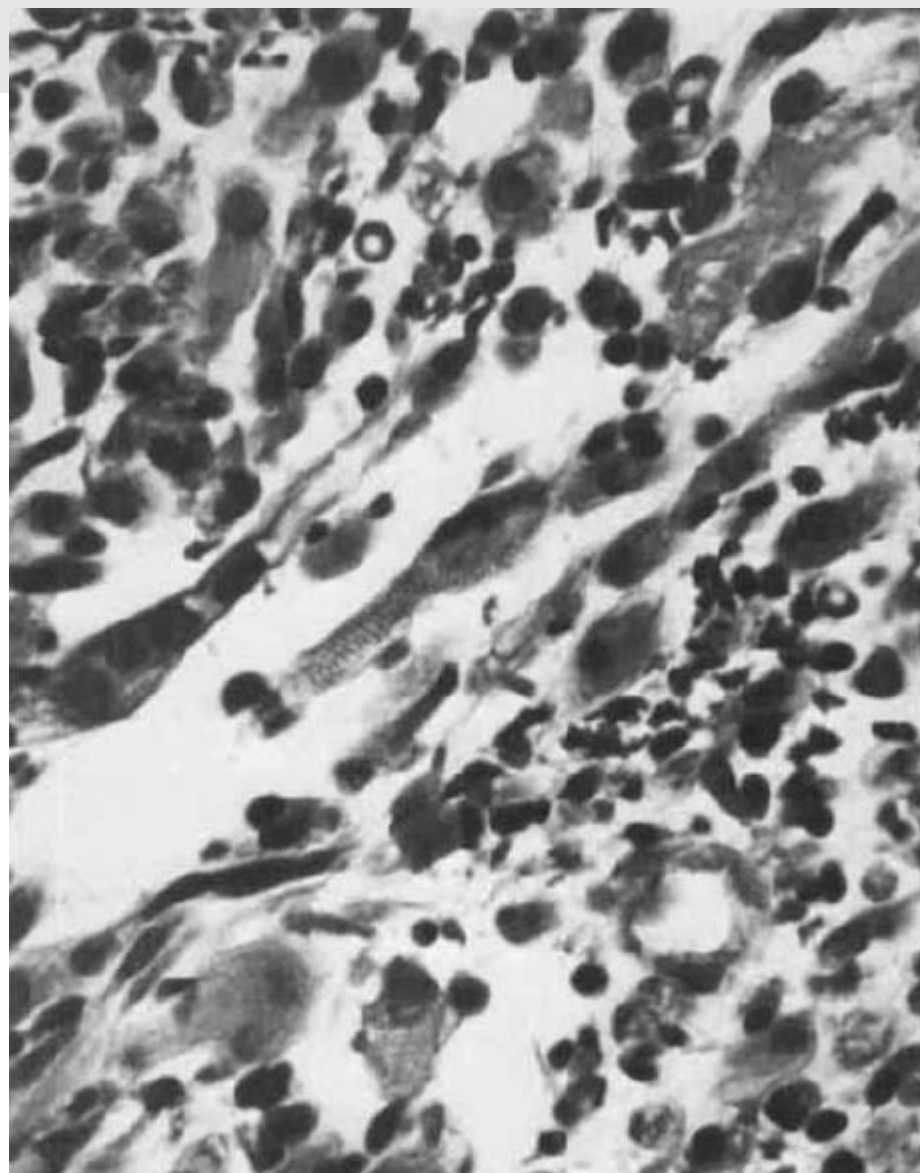
DENNIS K. HEFFNER, MD, CAPT, MC, USN,\* AND VINCENT J. HYAMS, MD, CAPT, MC, USN†

Twenty cases of a unique type of sinonasal tract neoplasm with combined histologic features of carcinosarcoma and teratoma are described and discussed. The term “teratocarcinosarcoma” is proposed and justified. Patients were adults (age range, 18–79 years; median age, 60 years). The variegated histologic components are illustrated and differences from gonadal germ cell neoplasms are delineated. This neoplastic entity is clearly malignant, with 60% of patients not surviving beyond 3 years (average survival, 1.7 years) following diagnosis, regardless of type of therapy. Aggressive therapy (combined surgery and irradiation) seems justified, however, since 40% of patients survived 3 years or longer with no current evidence of neoplasm (average follow-up, 6.1 years).

*Cancer* 53:2140–2154, 1984.

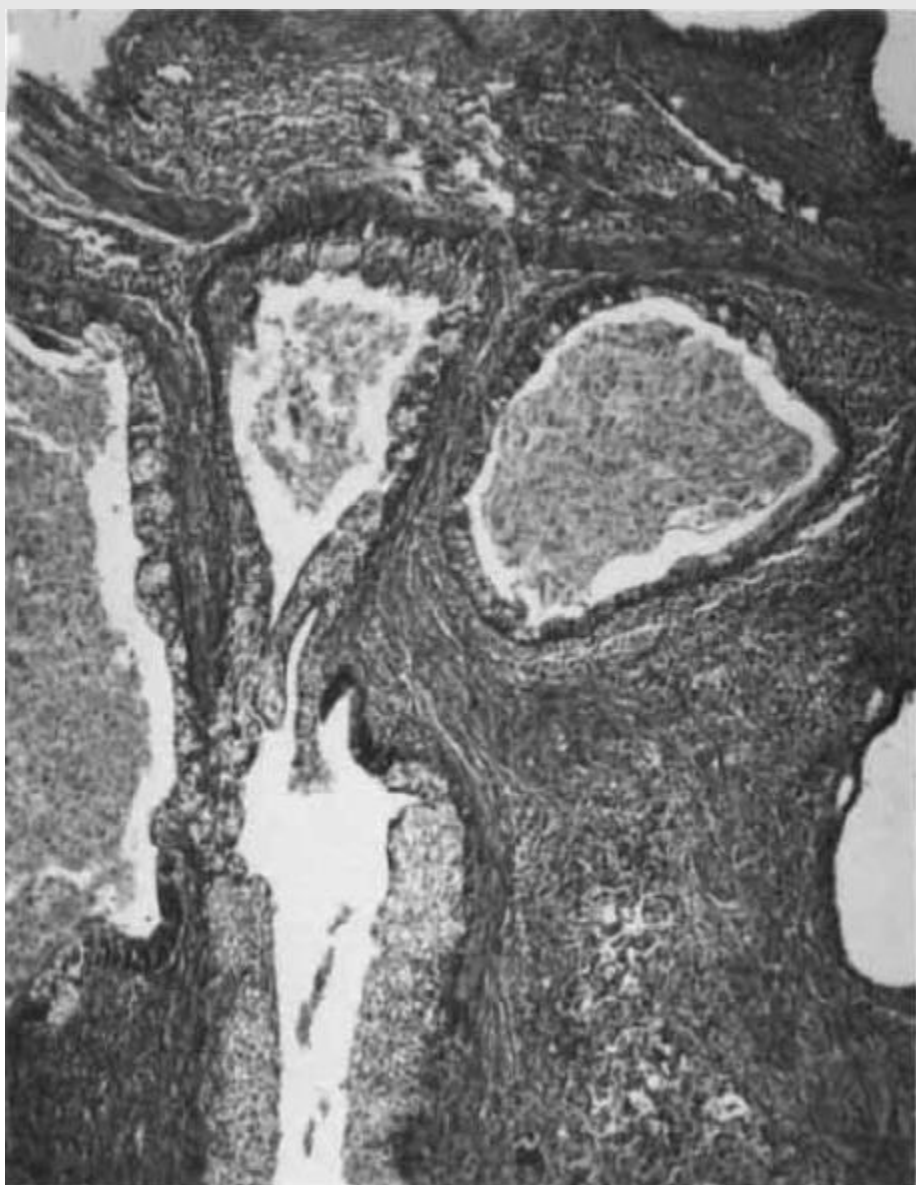


# *Teratocarcinosarcoma (Malignant Teratoma?) of the Nasal Cavity and Paranasal Sinuses*



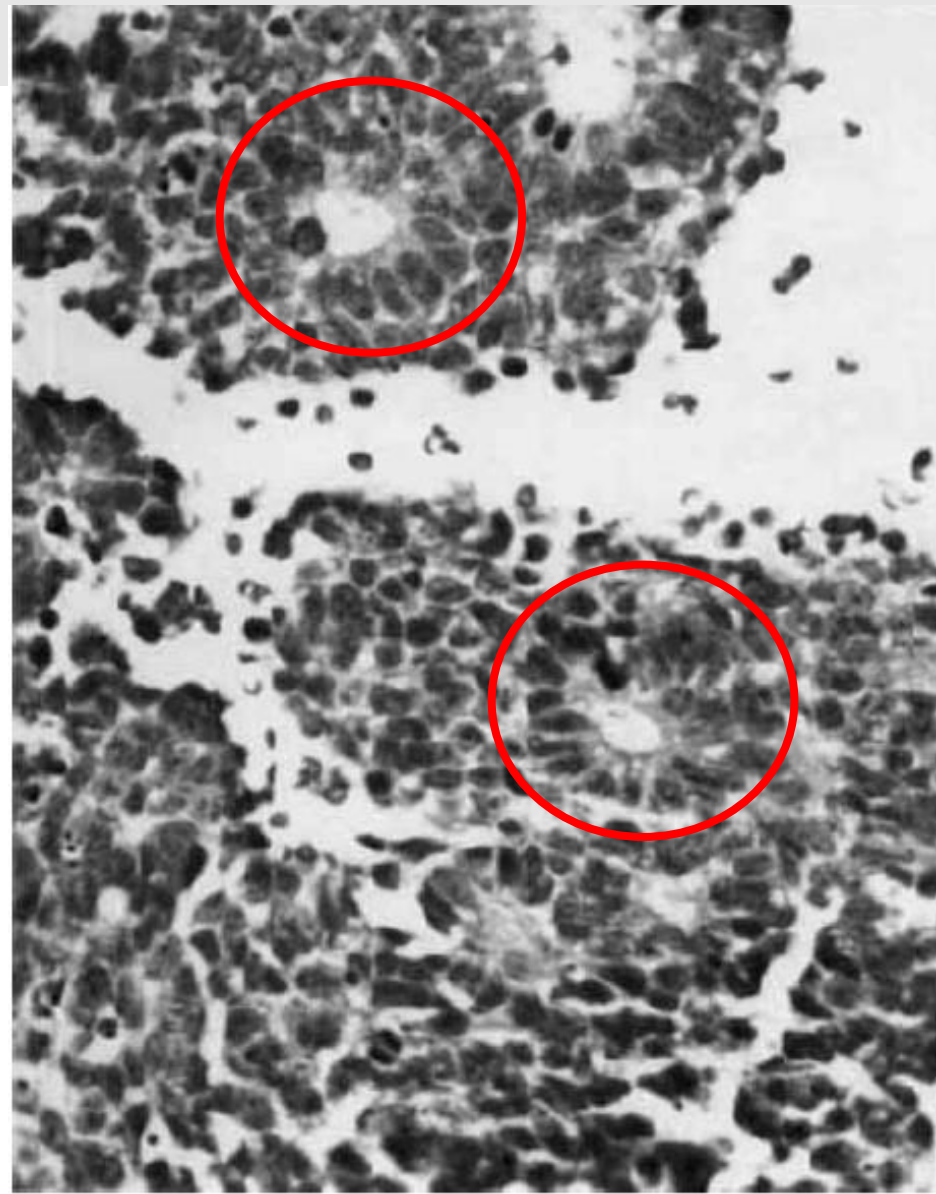
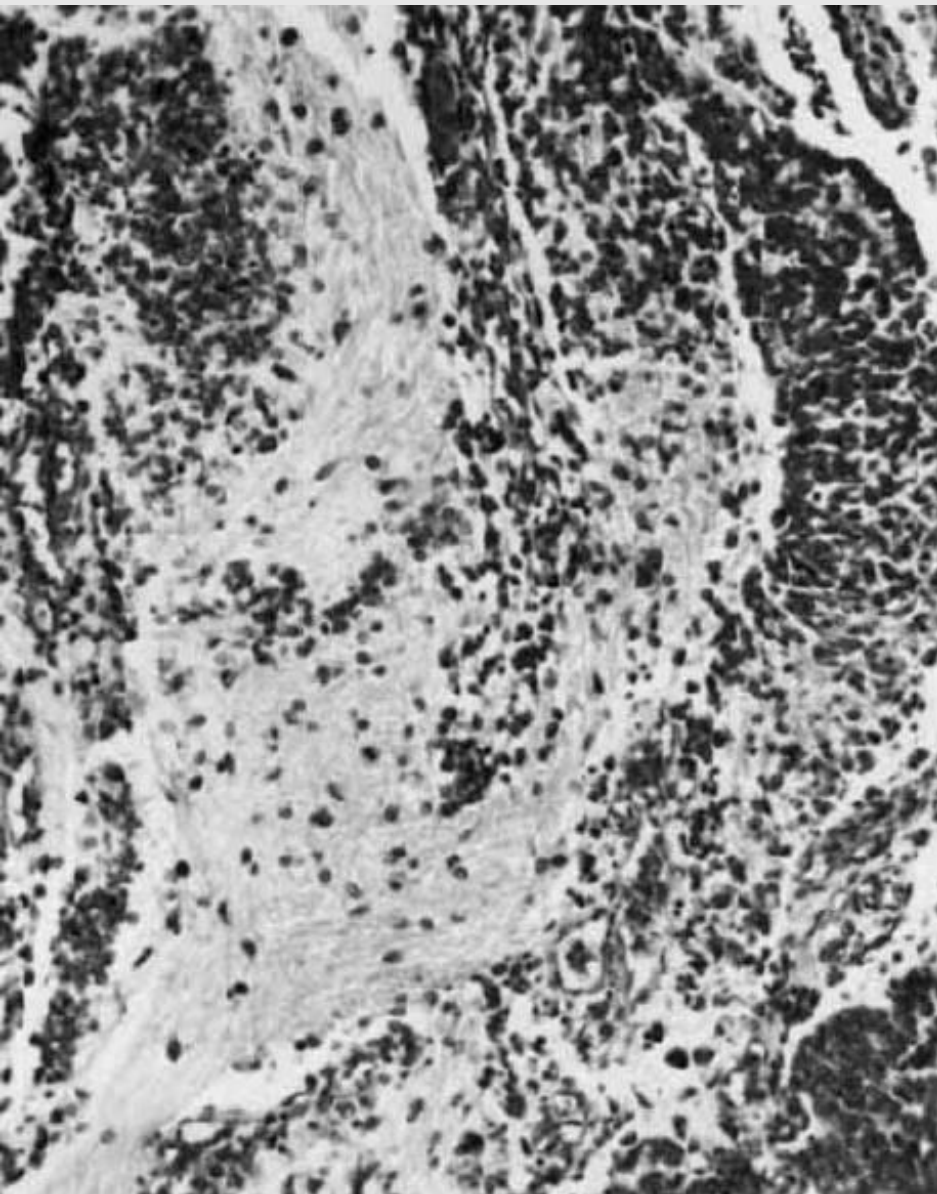


# *Teratocarcinosarcoma (Malignant Teratoma?) of the Nasal Cavity and Paranasal Sinuses*



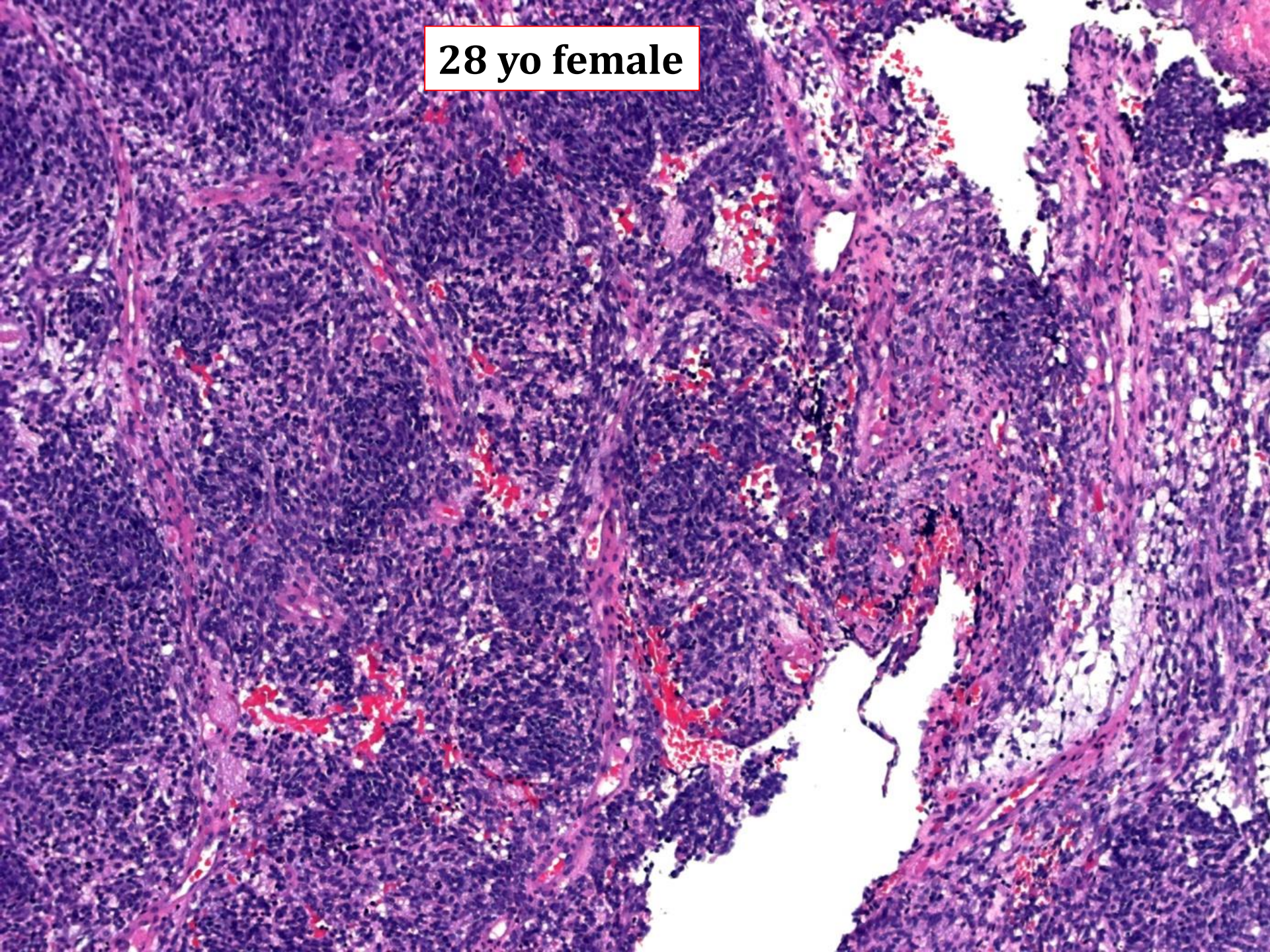


# *Teratocarcinosarcoma (Malignant Teratoma?) of the Nasal Cavity and Paranasal Sinuses*





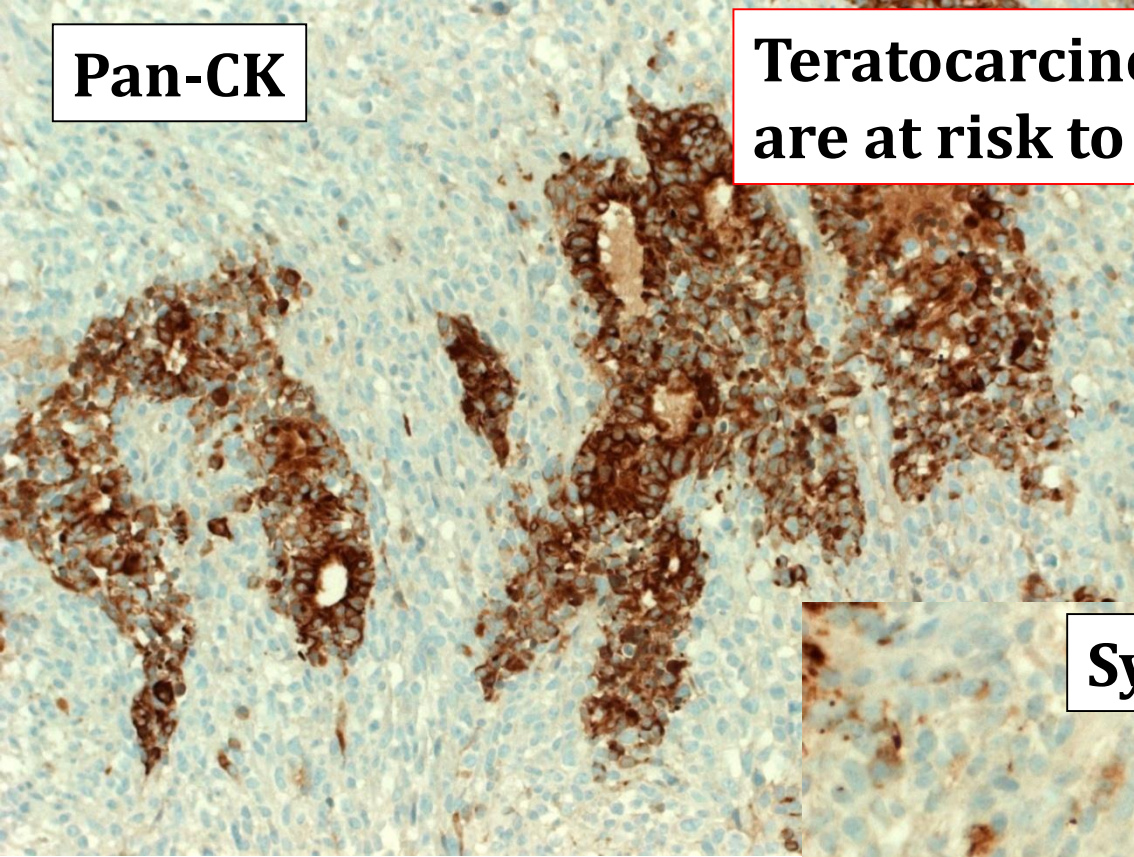
28 yo female



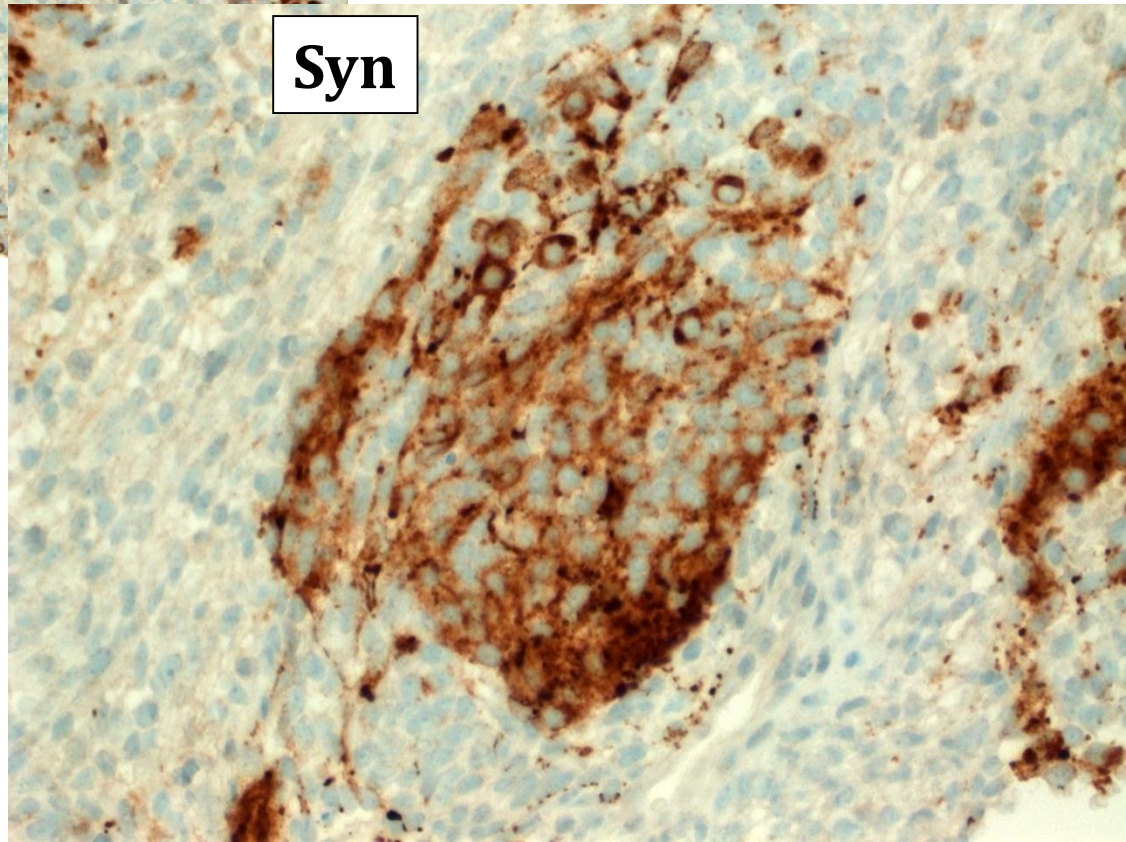


**Pan-CK**

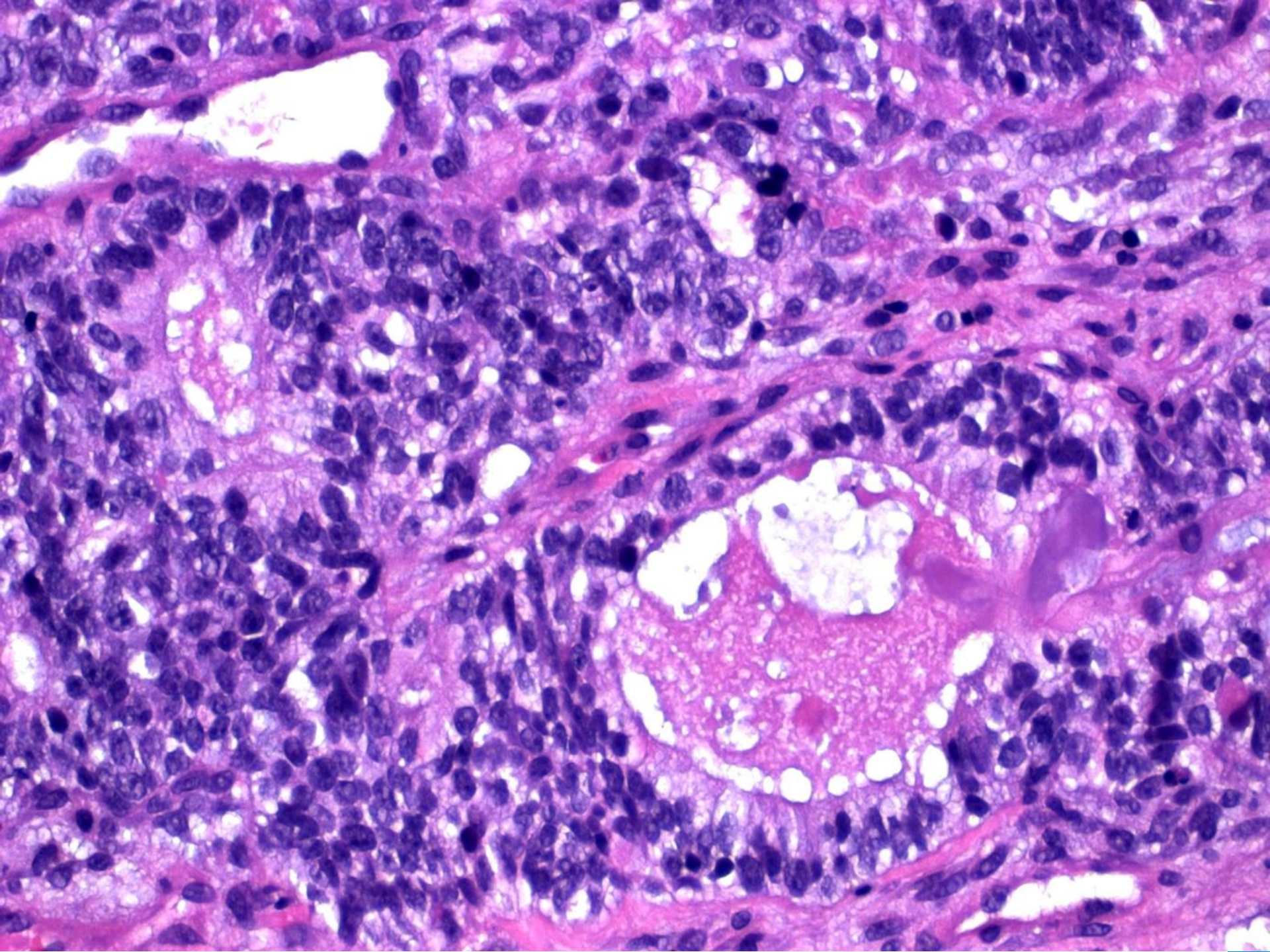
**Teratocarcinosarcomas in biopsies  
are at risk to be misdiagnosed as ONB**



**Syn**

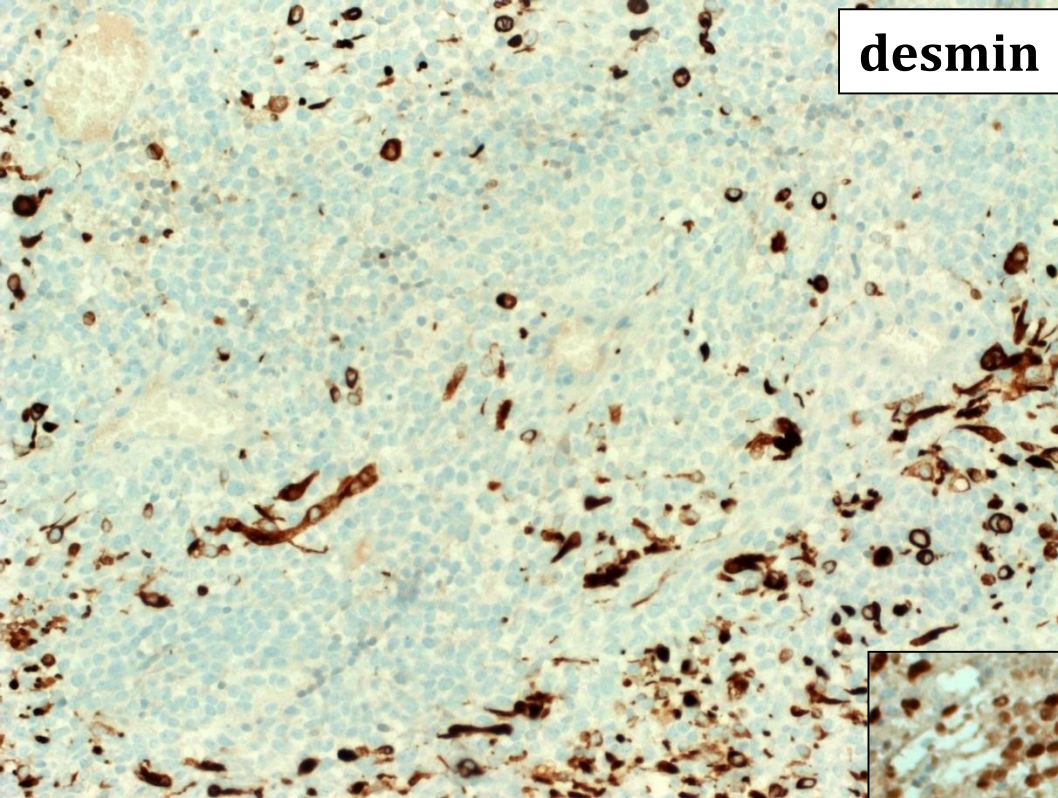




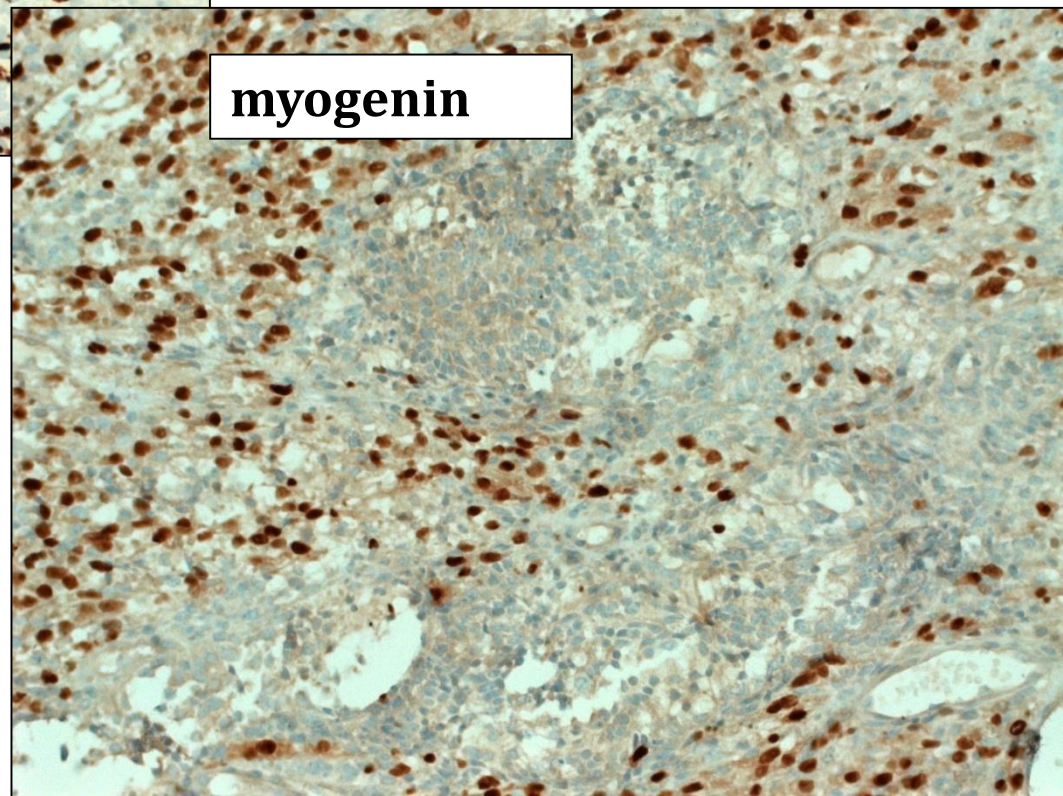




**desmin**



**myogenin**





A high-magnification histological slide of a sinonasal malignancy. The tissue shows a dense population of cells with large, round, hyperchromatic nuclei and prominent nucleoli. The cells are arranged in a microrlobular or nodular pattern, with small, irregular clusters and nests. There are several areas of hemorrhage, indicated by bright red, amorphous regions scattered throughout the tissue. The overall architecture is disorganized, consistent with a malignant process.

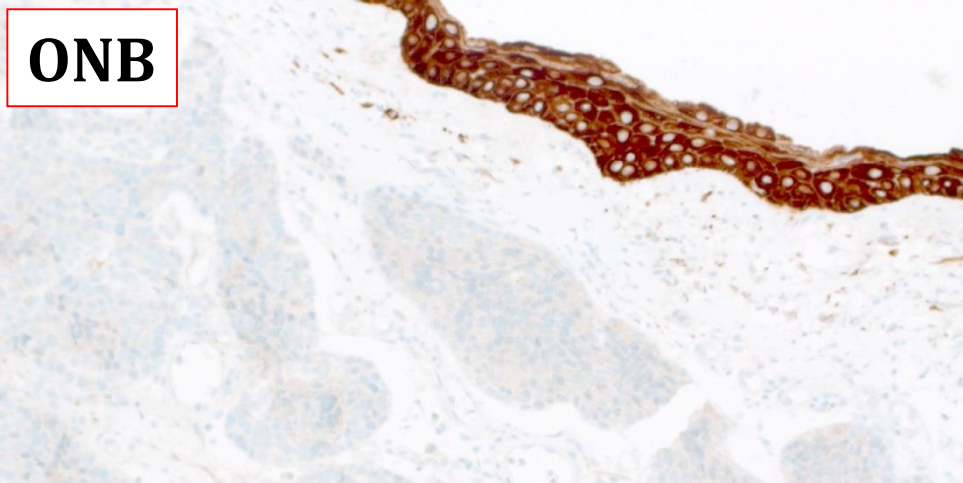
**ONB:**

**2.Problem: the lobules**

**Microrlobular/nodular pattern can be seen in almost any solid-pattern sinonasal malignancy**

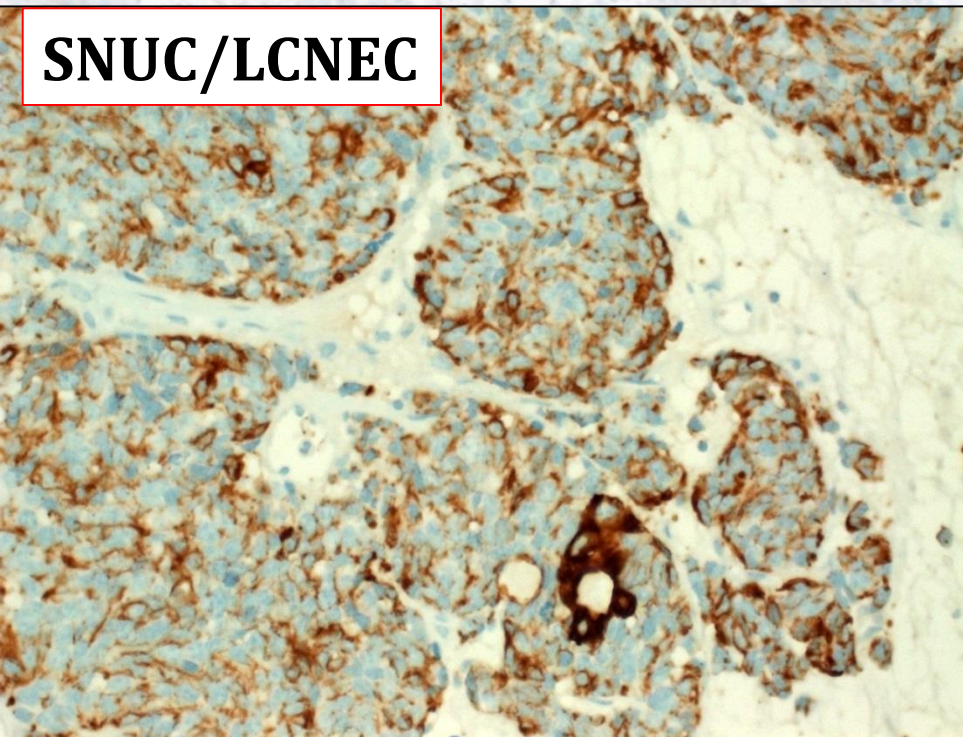


**ONB**



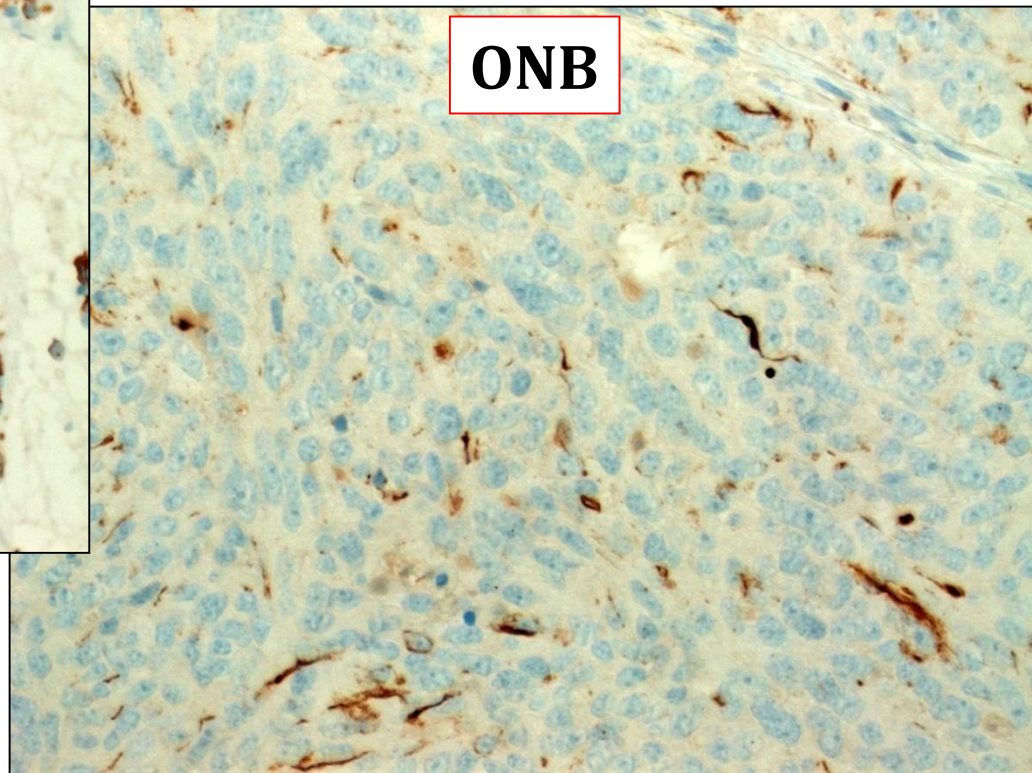
**3.Problem: CK expression**

**SNUC/LCNEC**

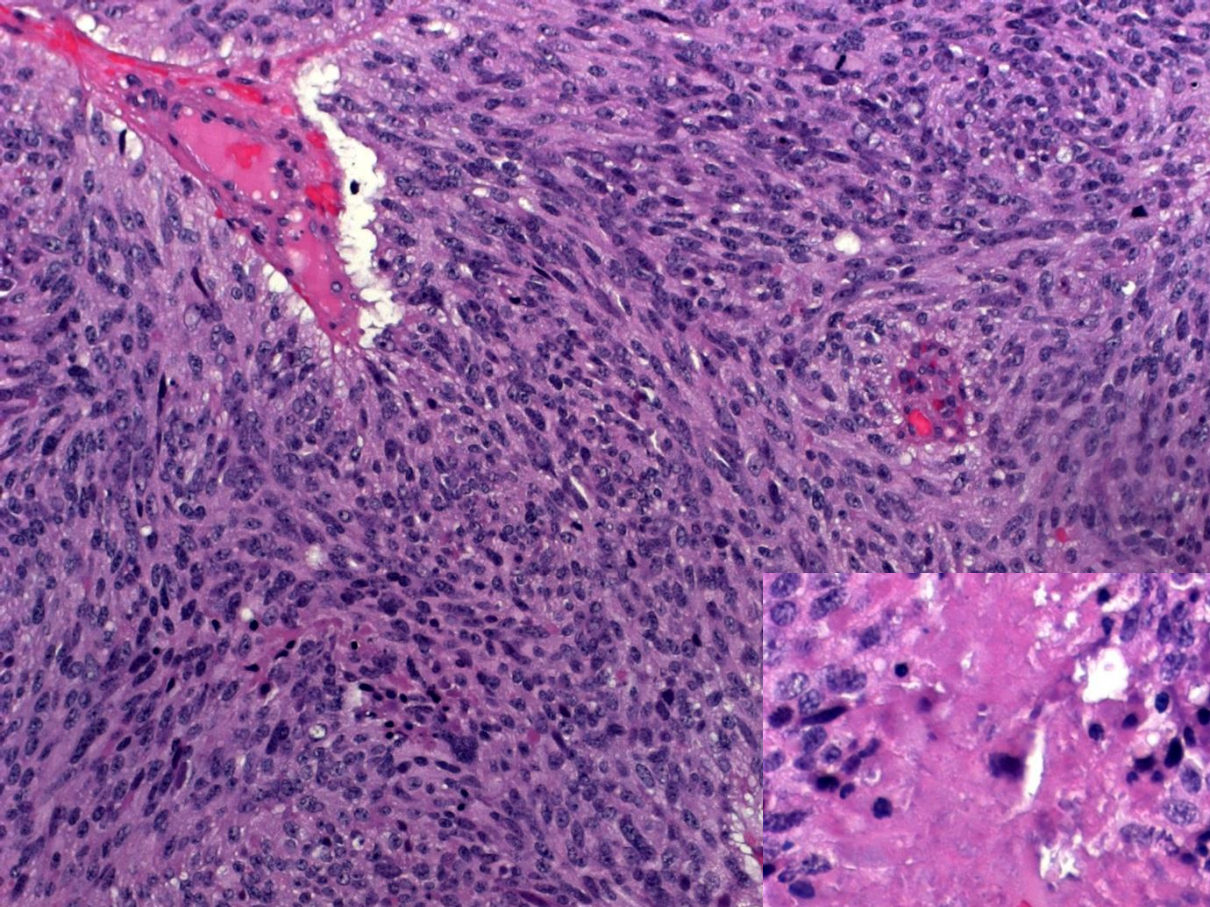


**AE1/AE3**

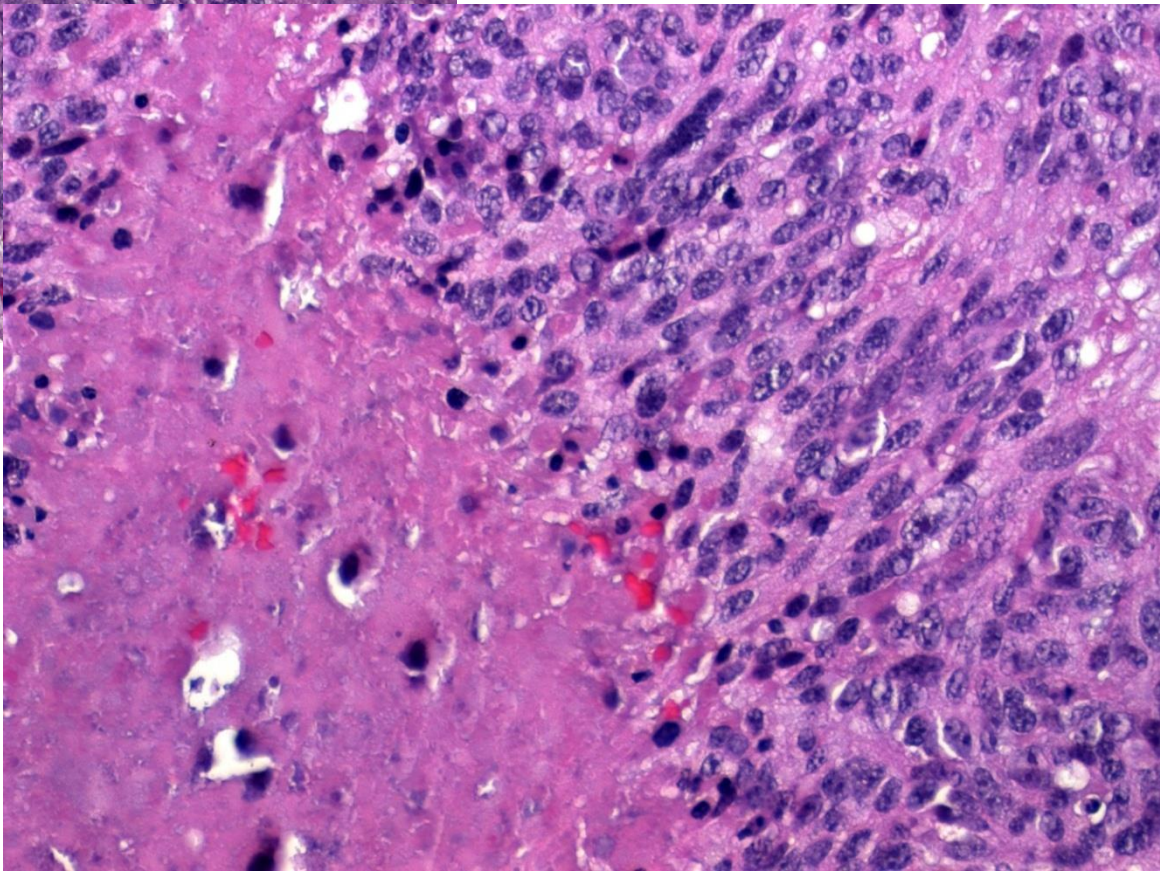
**ONB**







**ONB:**  
**4.Problem: Variants**





# ONB TAHOME

- ✓ No ONB without homogeneous NE marker expression.
- ✓ NO ONB with diffuse strongly cytoplasmic CK expression.
- ✓ All solid sinonasal neoplasms may:
  - Show lobular growth & rosettes.
  - Be variably Synapto+
- ✓ **Grade IV ONB are frequently non-ONB.**
- ✓ In the sinonasal tract, be careful with CK/Syn/Des/Myogen  
& CD99 = **all non-reliable as lineage markers**  
**(=context dependant interpretation)**





# Thank you for your attention

