



# Congenital Melanocytic Nevus

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# Key Points

- Small vs. large congenital nevi
- Neurocutaneous melanosis
- Cellular proliferative nodules
- Risk of melanoma

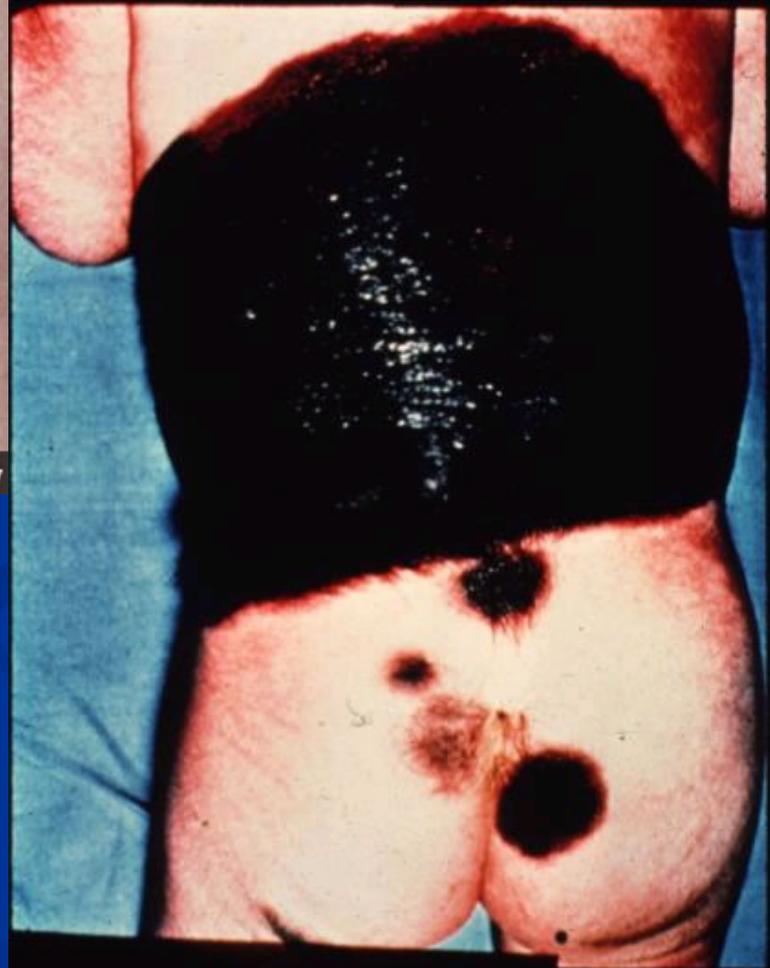


# What is it?

- 1% newborn infants
- Nevi present at birth
- Small  $\leq 1.5$  cm
- Intermediate 1.5 -19.9 cm



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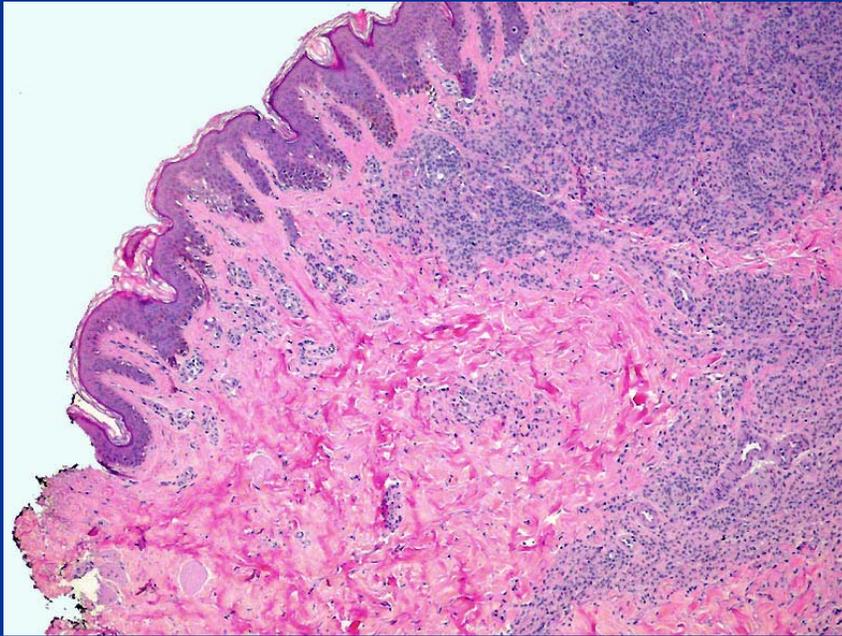
# Large Congenital Melanocytic Nevi

Nevus present at birth that has or is predicted to have a largest diameter of at least 20 cm in adulthood

J Am Acad Dermatol 1979;1:123-130.

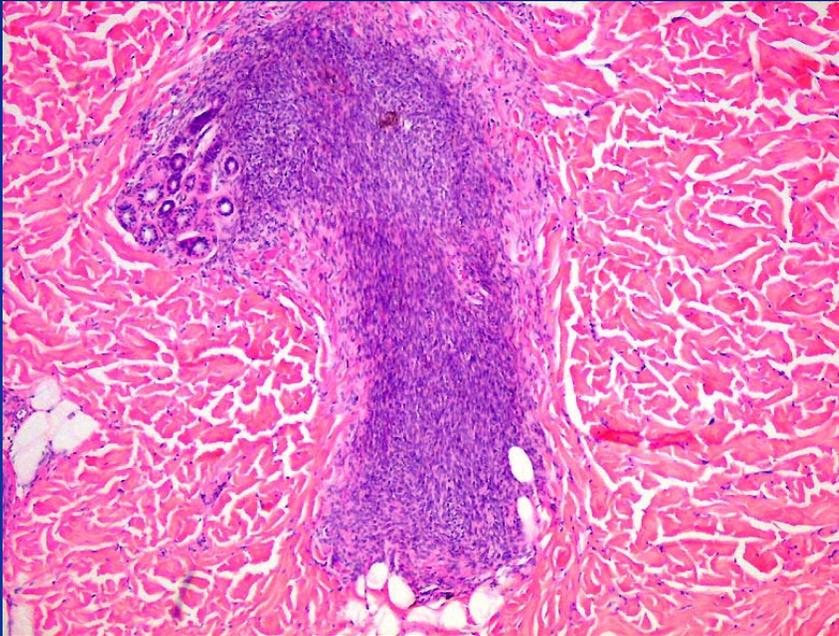
<b>Giant CN</b>	<b>Small CN</b>
>20 cm	1-5 cm
Sometimes garment distrib. Dorsal most common	Often trunk/extremity
Well defined but not sharply demarcated borders	Same
Hypertrichosis	Hypertrichosis
Occipital nevi associated with meningeal and deep penetrating cranial vessel	Absent
Nevus satellitosis	Absent
Soft tissue hypertrophy	Absent

# Histopathology-Epidermal



- Lentiginous junctional proliferation
- Slight atypia
- Rete elongation
- Gentle undulating mammillation
- Junctional component may be absent

# Histopathology-Dermal



- Single cell pattern
- Perivascular
- Periadnexal
- Pilosebaceous units
- SubQ fat with septal/lobular

# How Specific?

- Reliable if  $>3$  cm
- 10% acquired nevi share these characteristics

# Neurocutaneous Melanocytosis (NCM)

# How Common?

- 10% patients with LCMN
- Multiple (3 or more) small to medium-sized CMN, accompanied by benign and/or malignant growth of melanocytes in the CNS
- 5 yr cumulative risk of developing in LCMH patients is 2.5%

# Who Gets It?

- Neurologic signs and symptoms by age 5
- Rarely late adulthood
- Signs of increased intracranial pressure
- LCMN pts. with lesions on head, posterior neck, or paravertebral area at higher risk
- VLLCMN with satellite lesions
- No reports of NCM with LCMN occurring on extremity or without satellite nevi

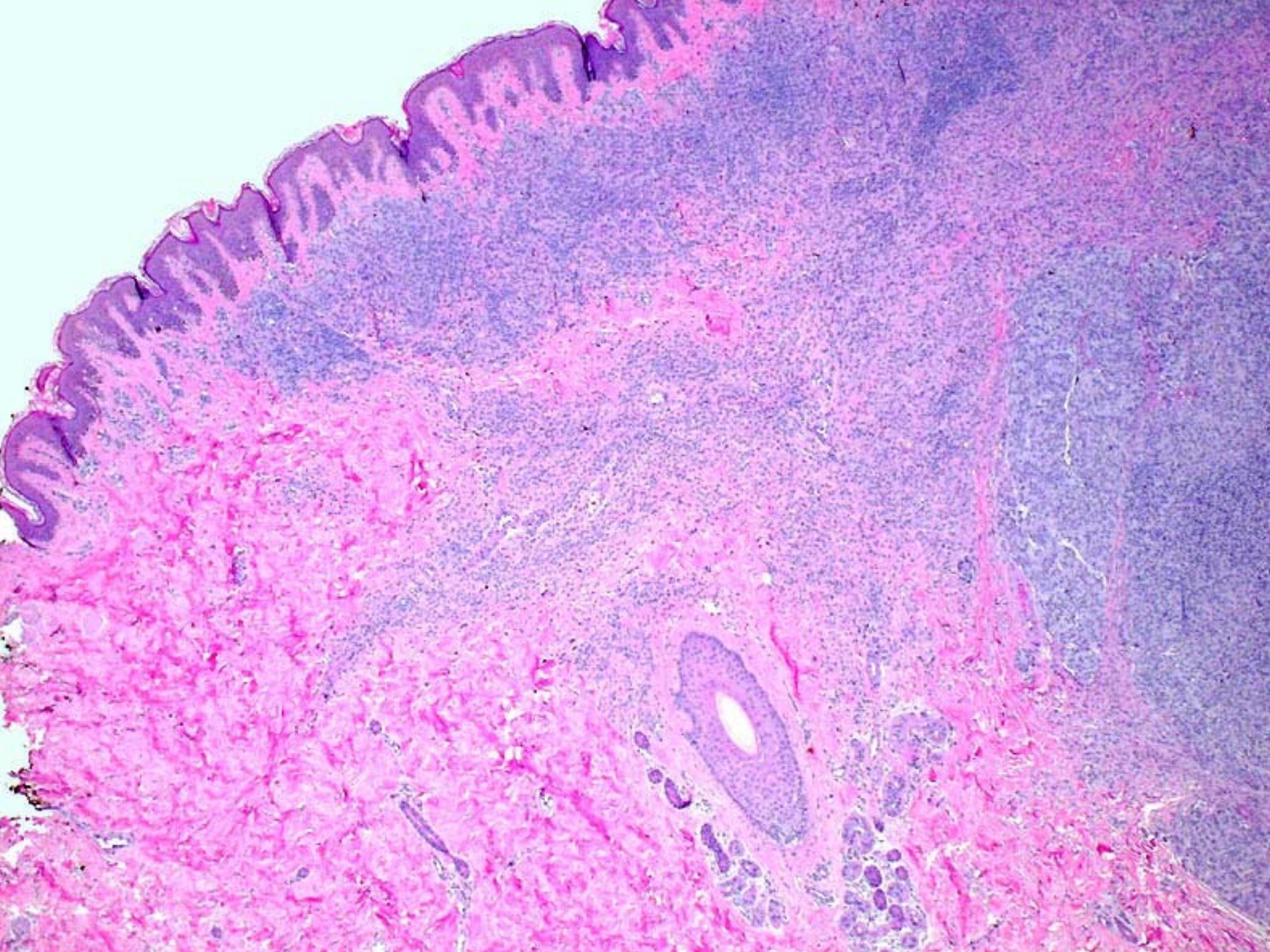
# Risk of NCM

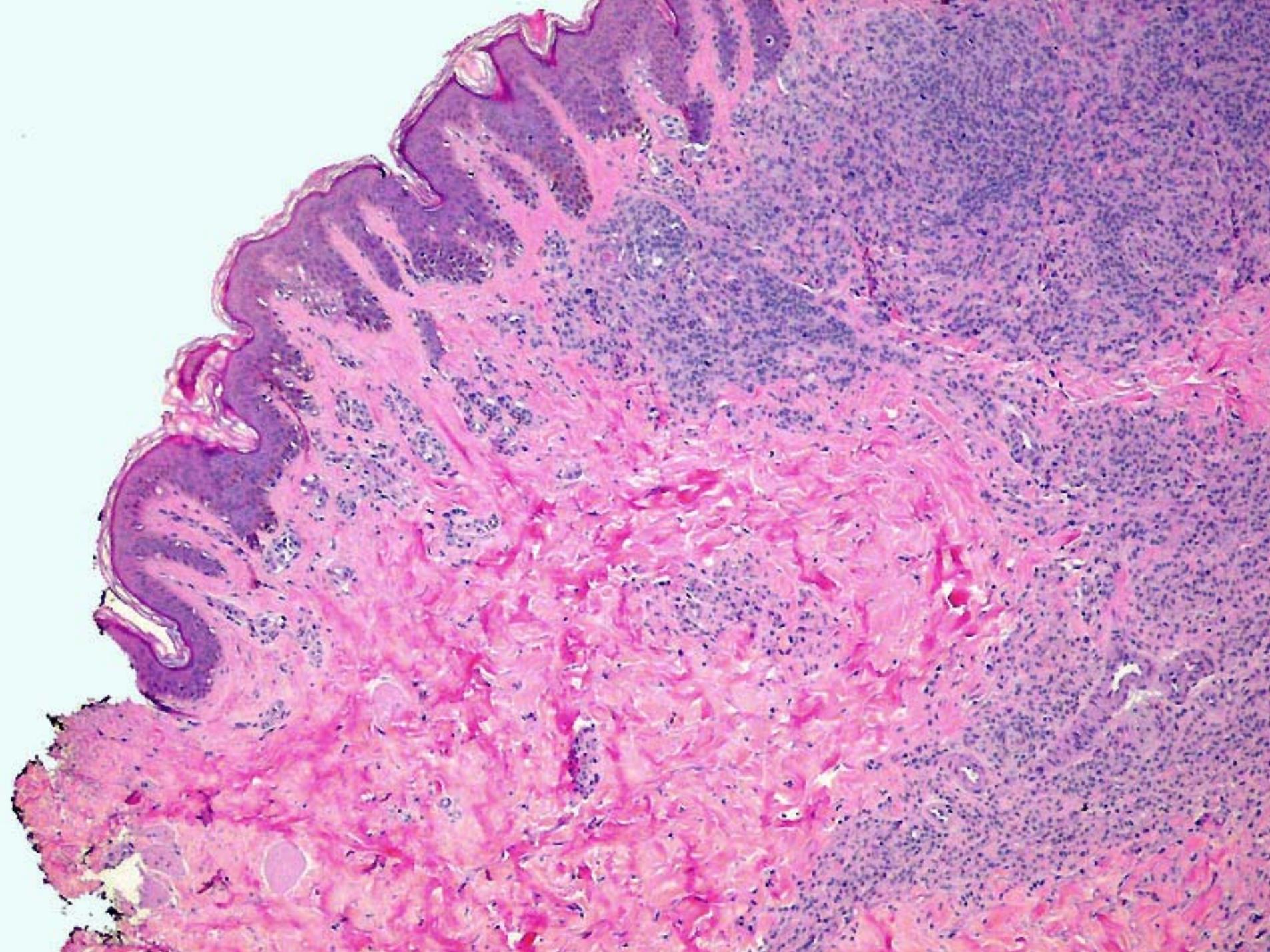
- 40-60% develop CNS melanomas (meninges)
- Neurologic symptoms poor prognosis
- Dead within 3 years of onset neurological symptoms
- 70% die <10 yrs

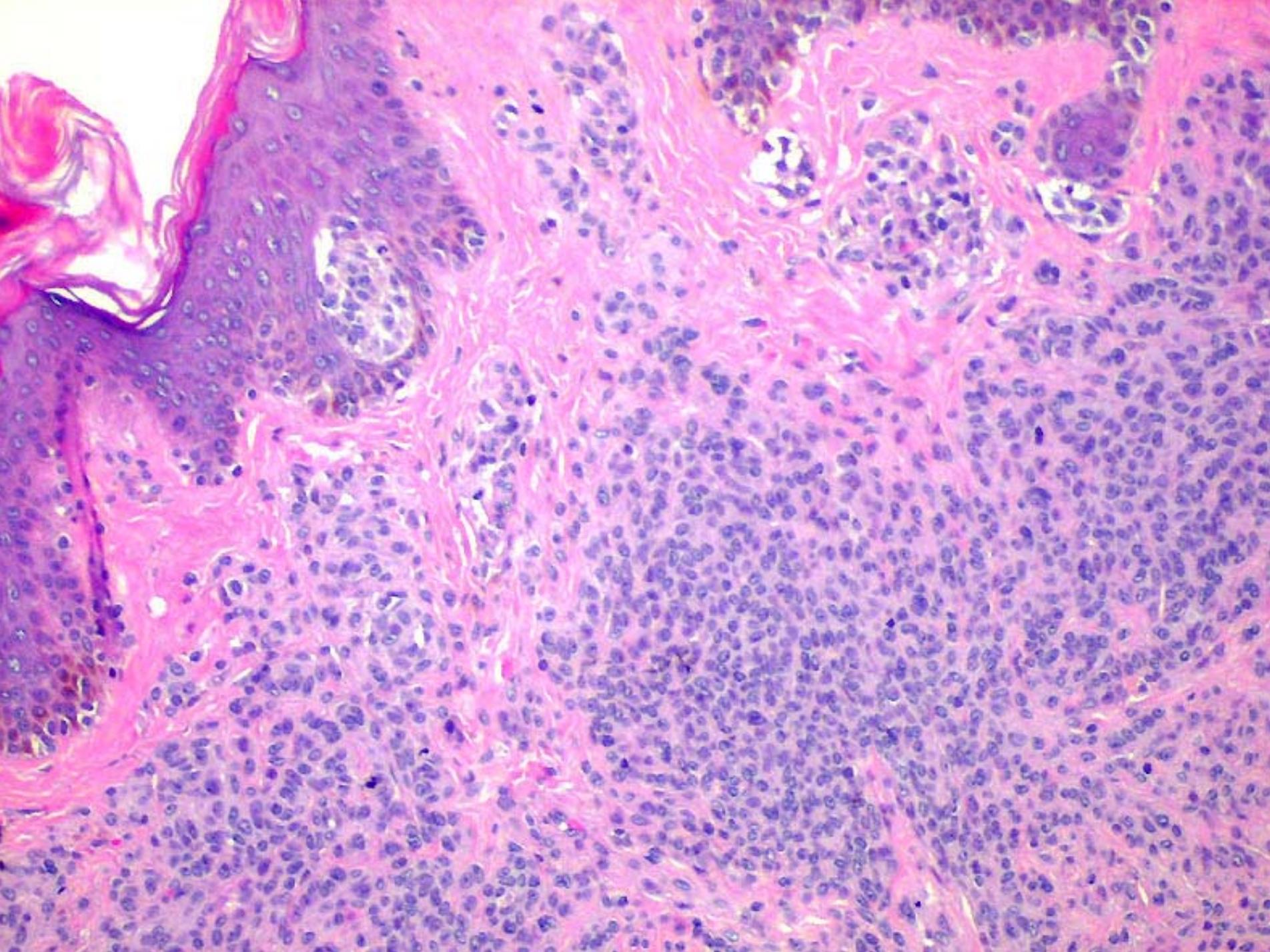
# MRI with Gadolinium

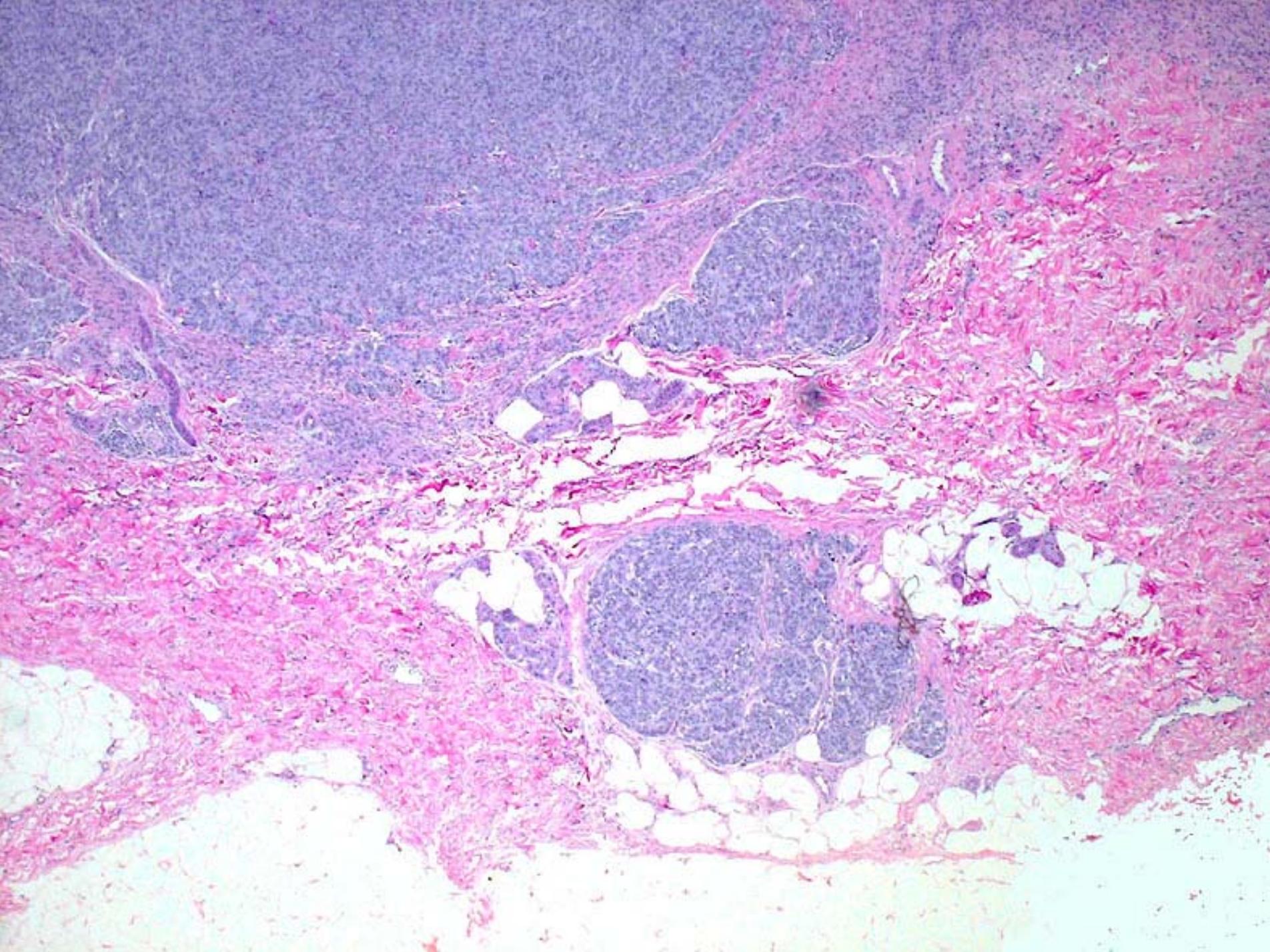
- Identify subset of pts with NCM but with no neurological symptoms
- Prognosis unknown
- Monitor with serial MRI

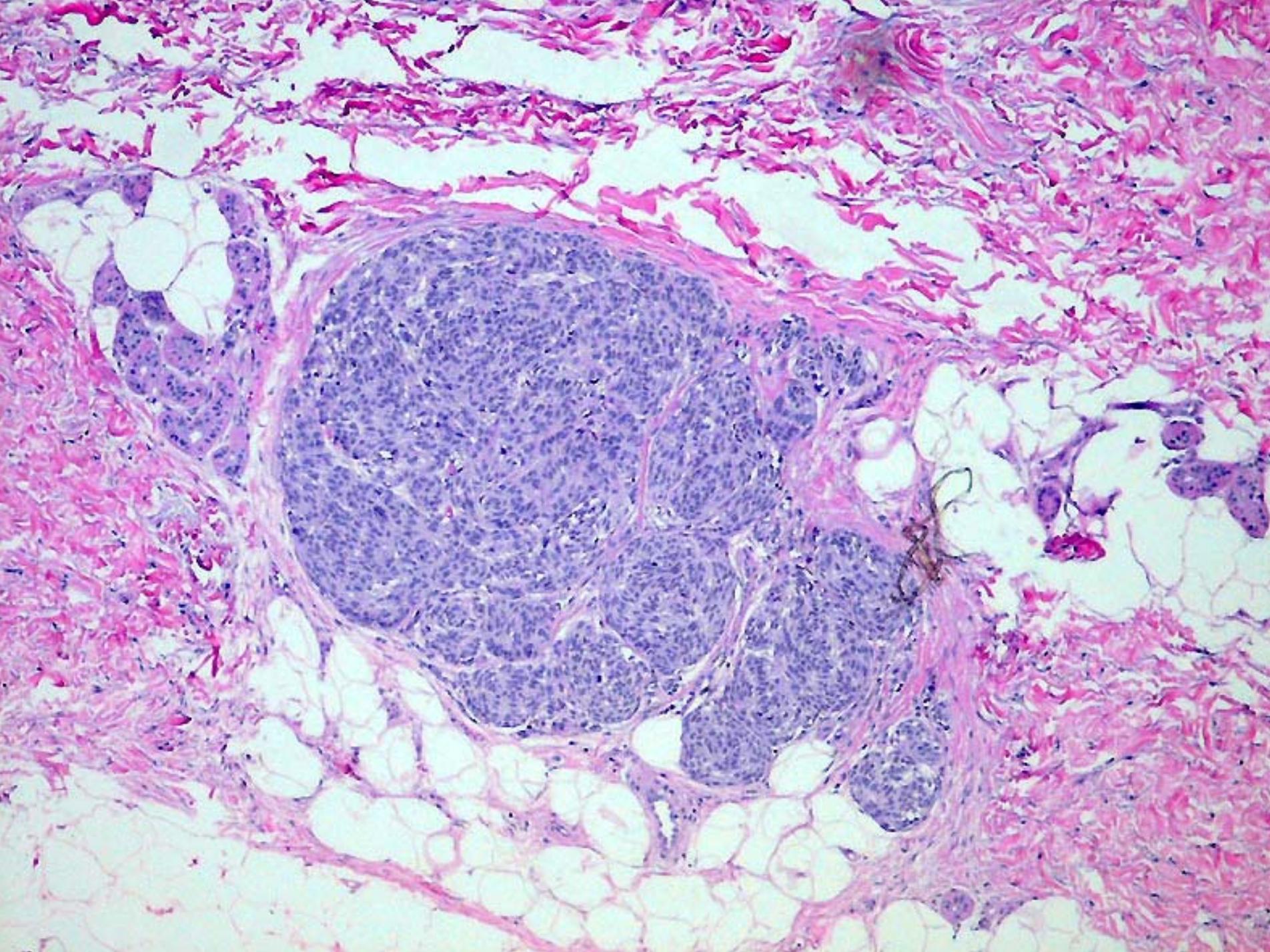


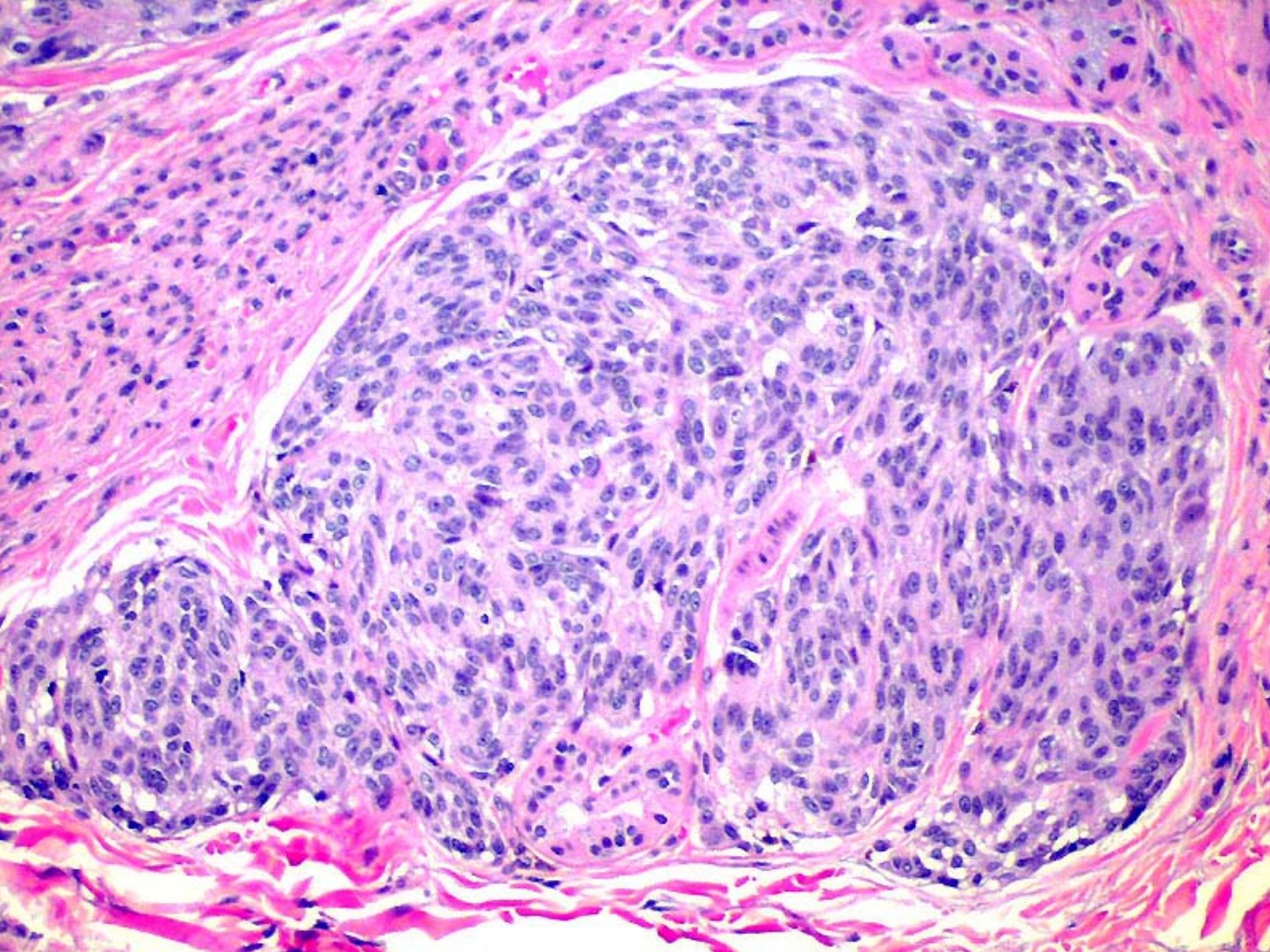


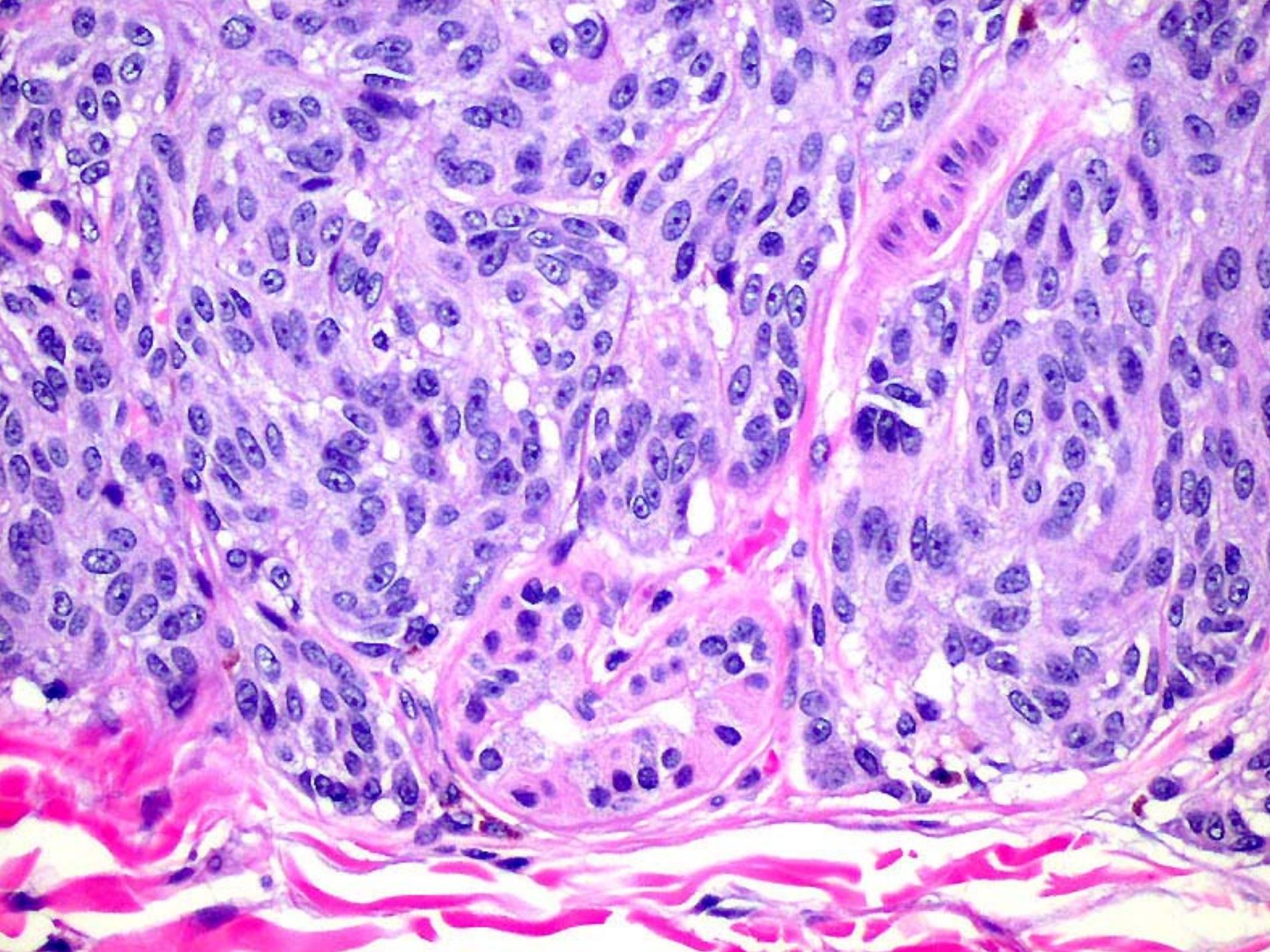


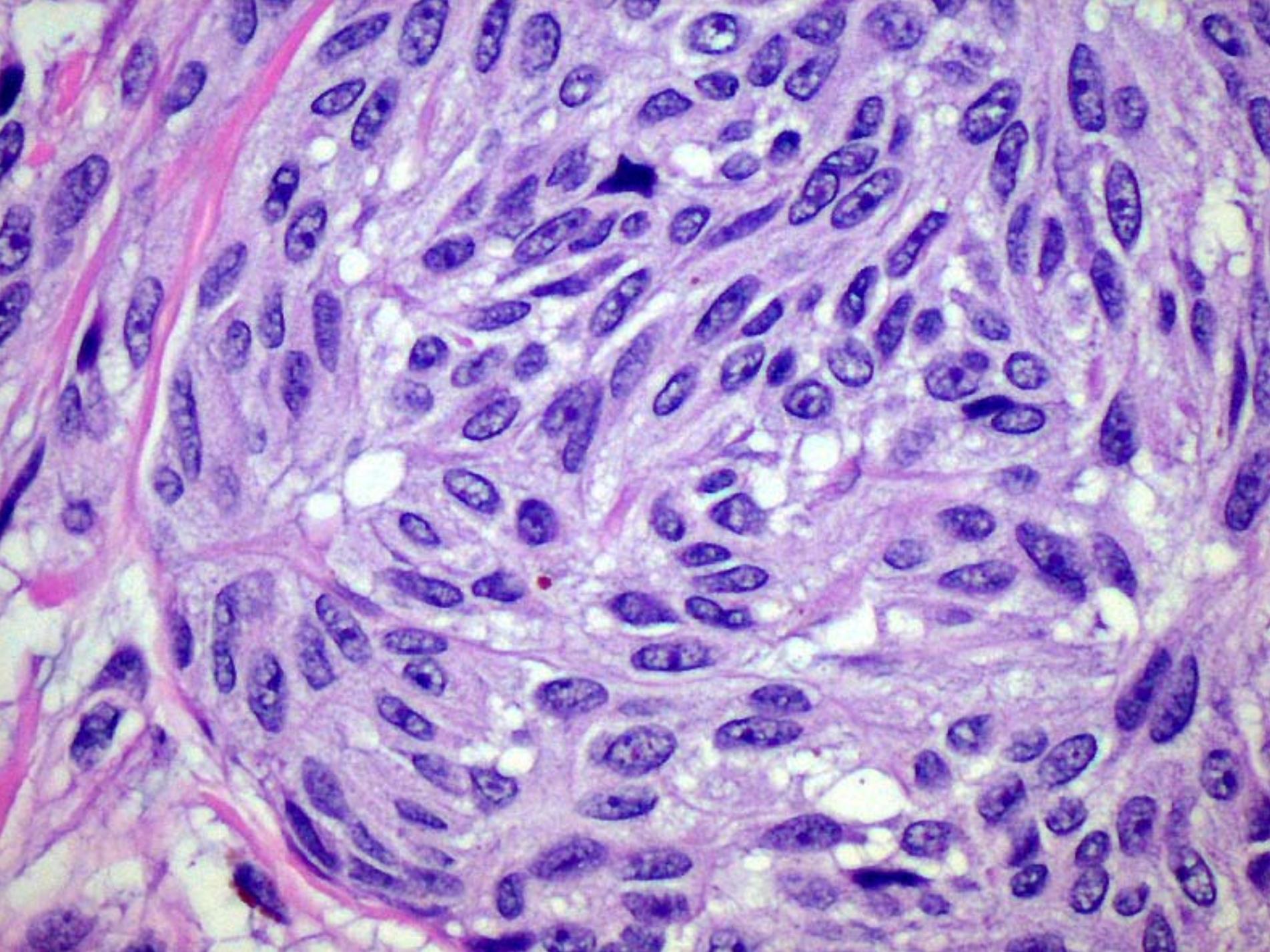






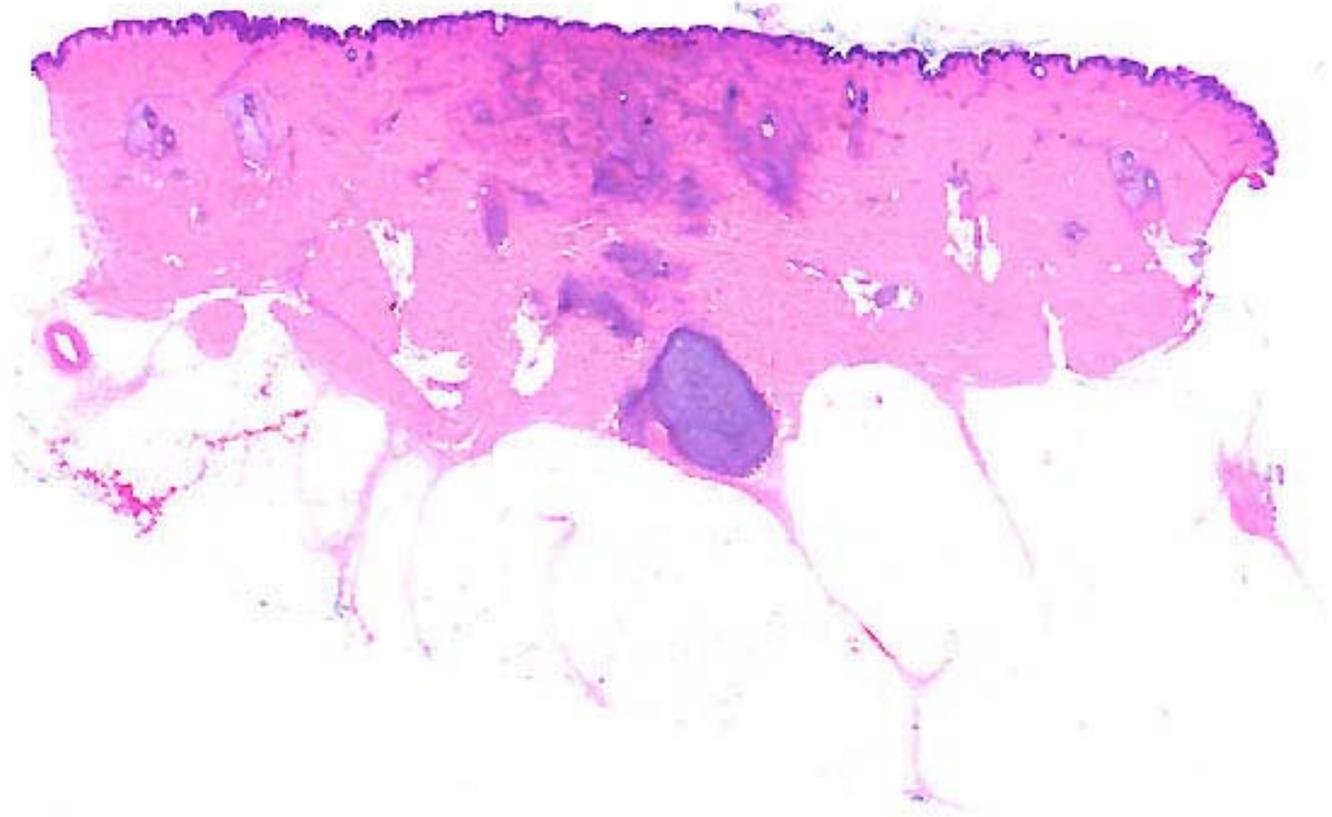


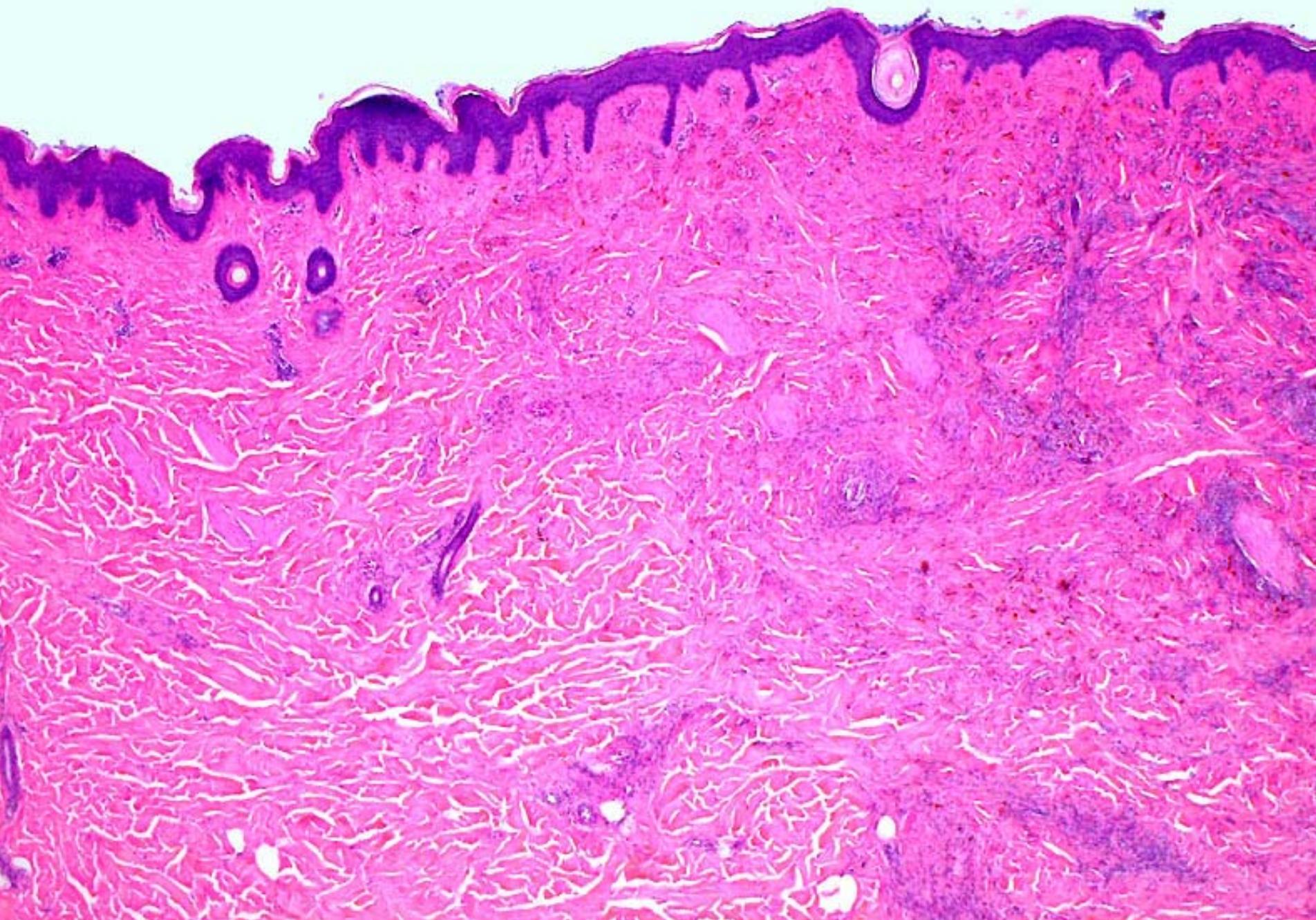


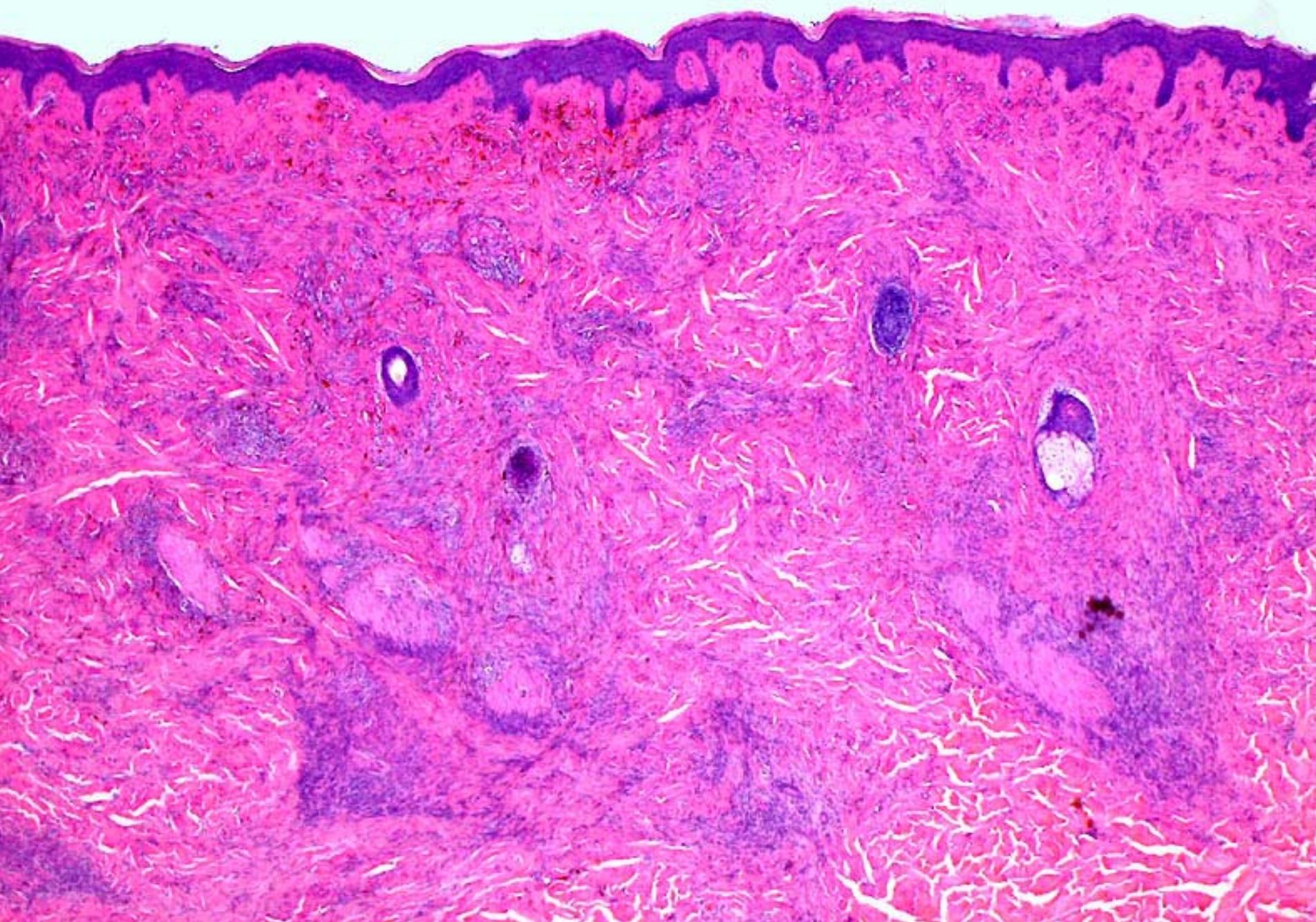


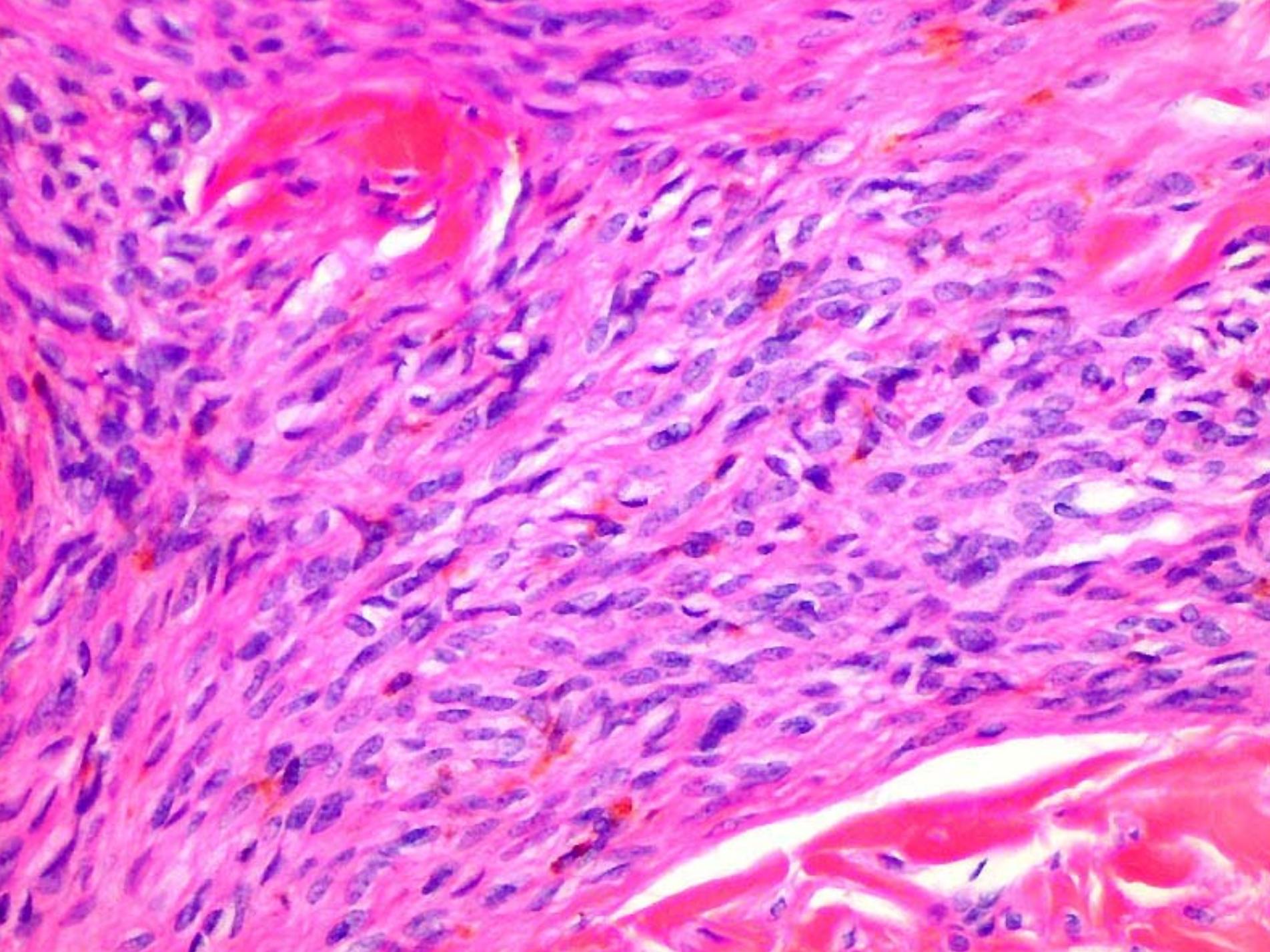
# Cellular Nodule in CMN

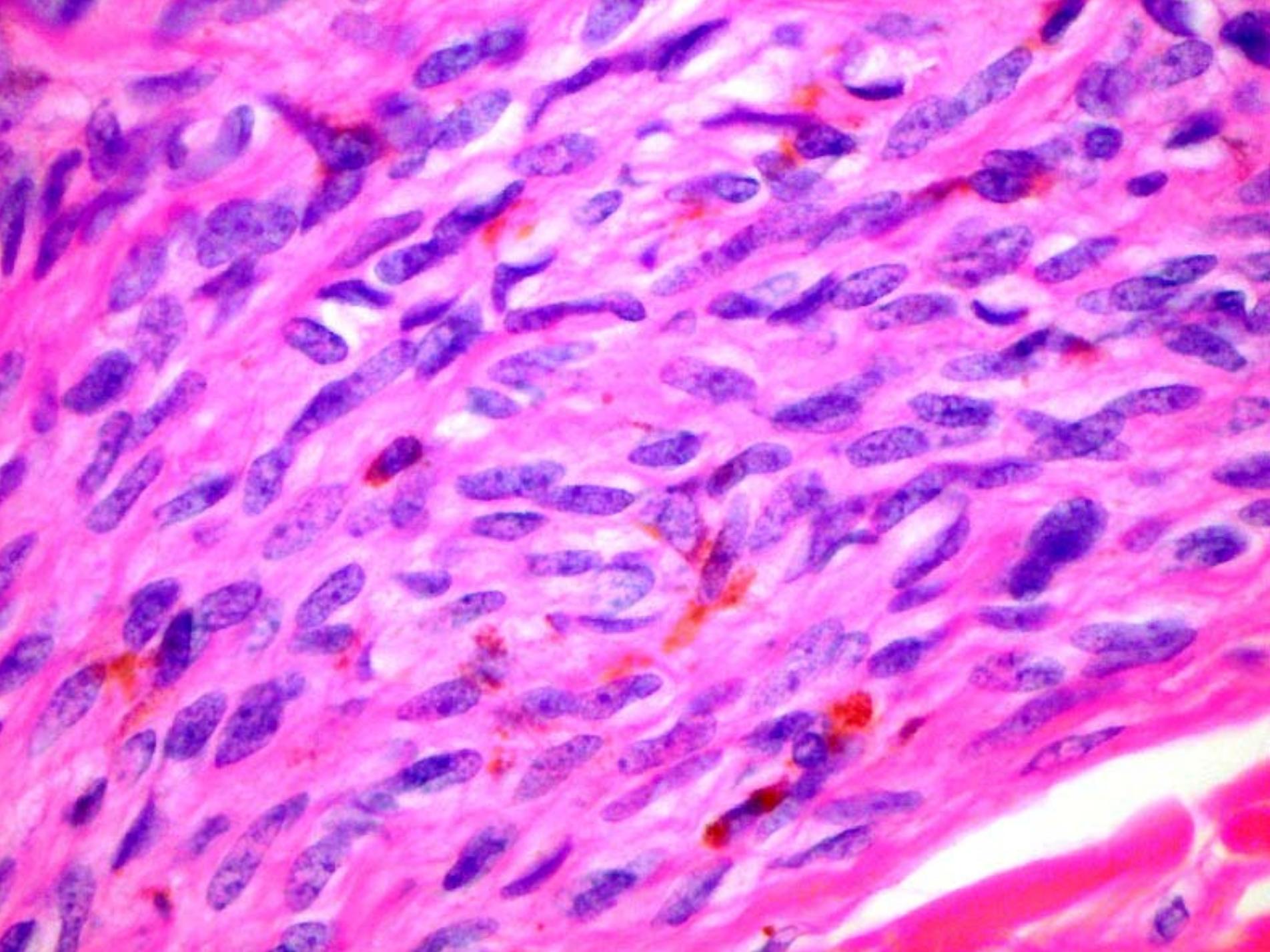
<b>Cellular Nodule</b>	<b>Melanoma</b>
<b>&lt;5mm</b>	<b>&gt;5mm</b>
<b>Mitoses rare and always typical</b>	<b>Mitoses common with atypical MF</b>
<b>No Necrosis</b>	<b>Necrosis may be present</b>
<b>Circumscribed and symmetric</b>	<b>Not circumscribed and asymmetric</b>

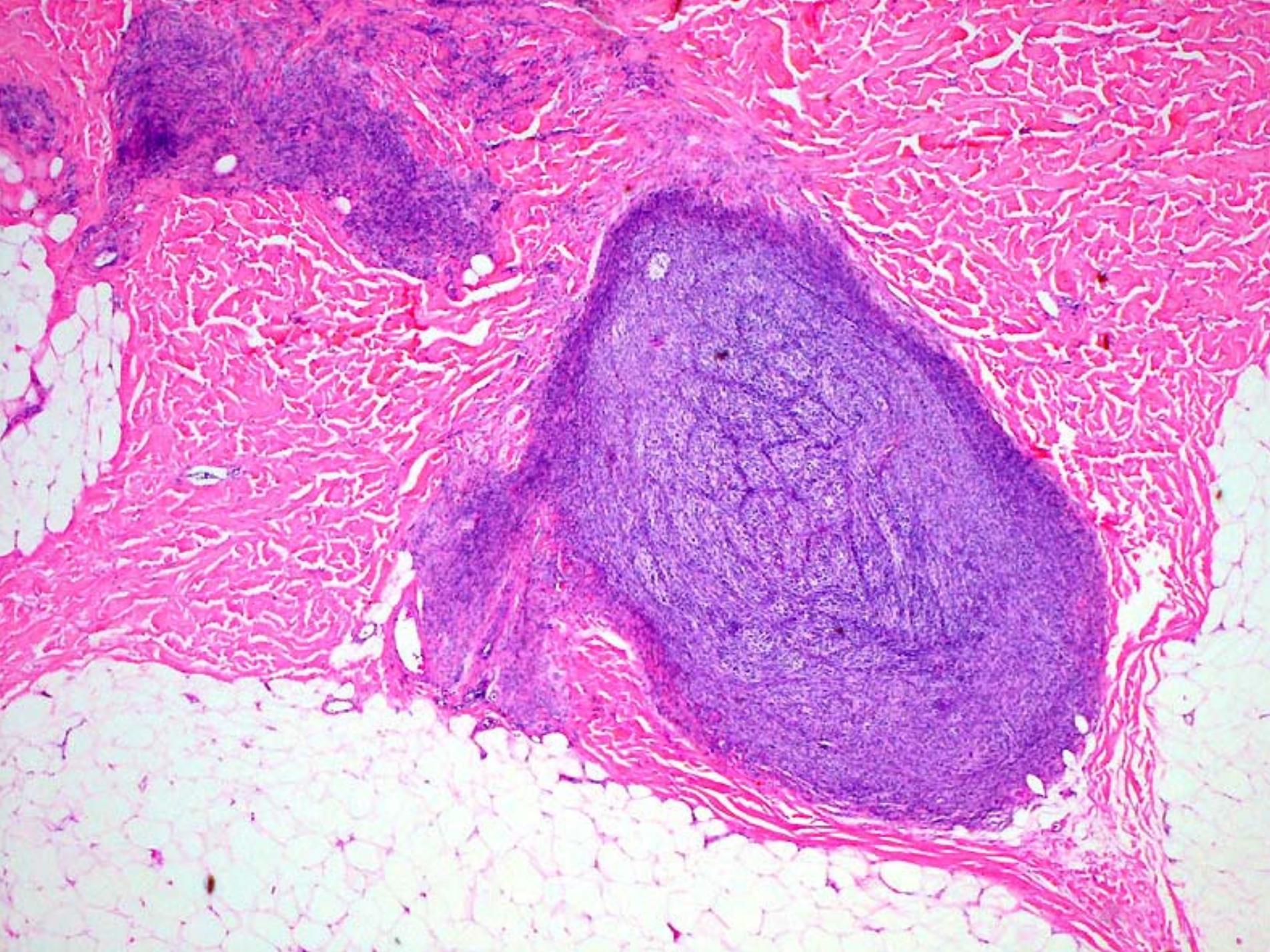


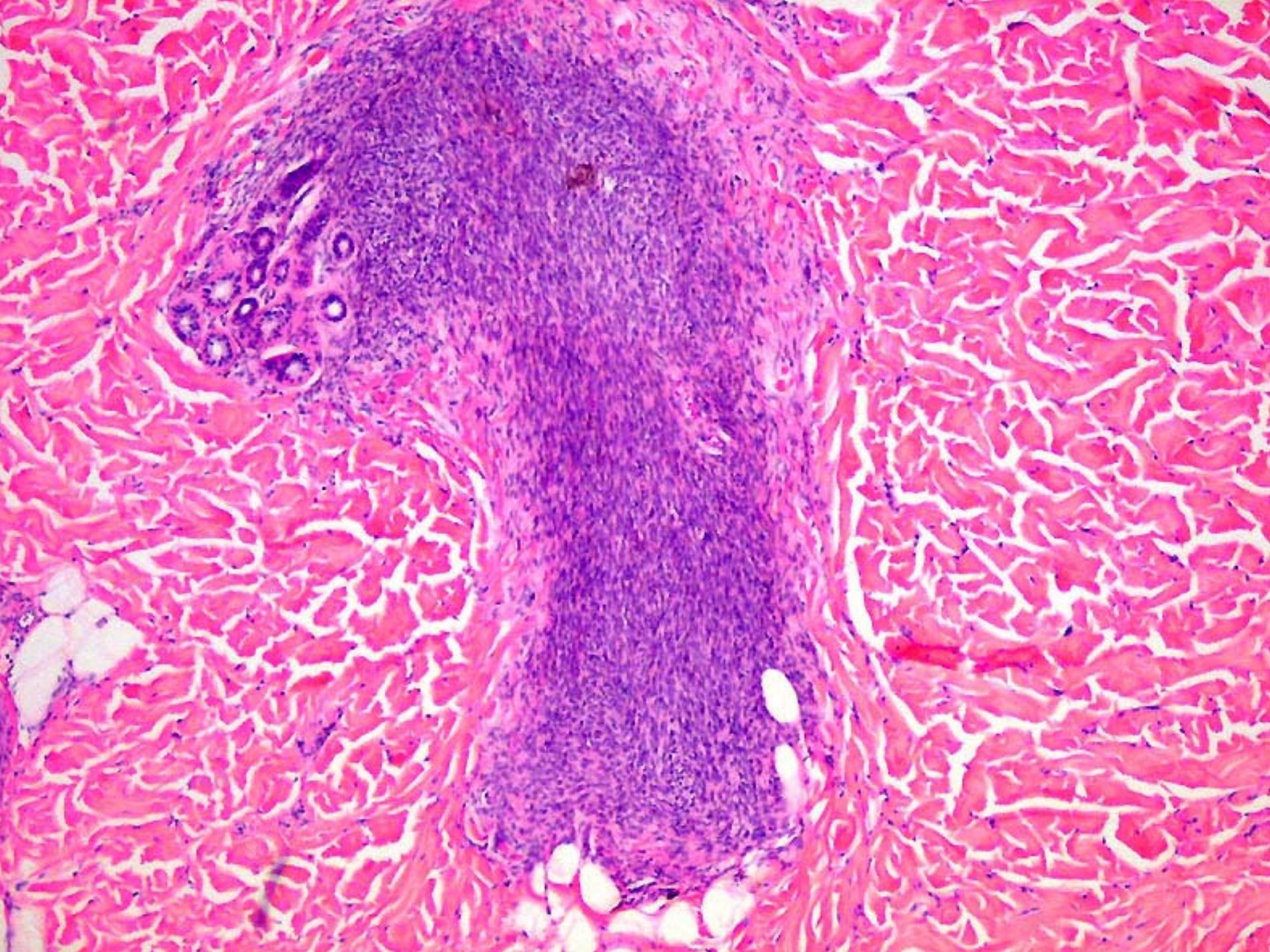


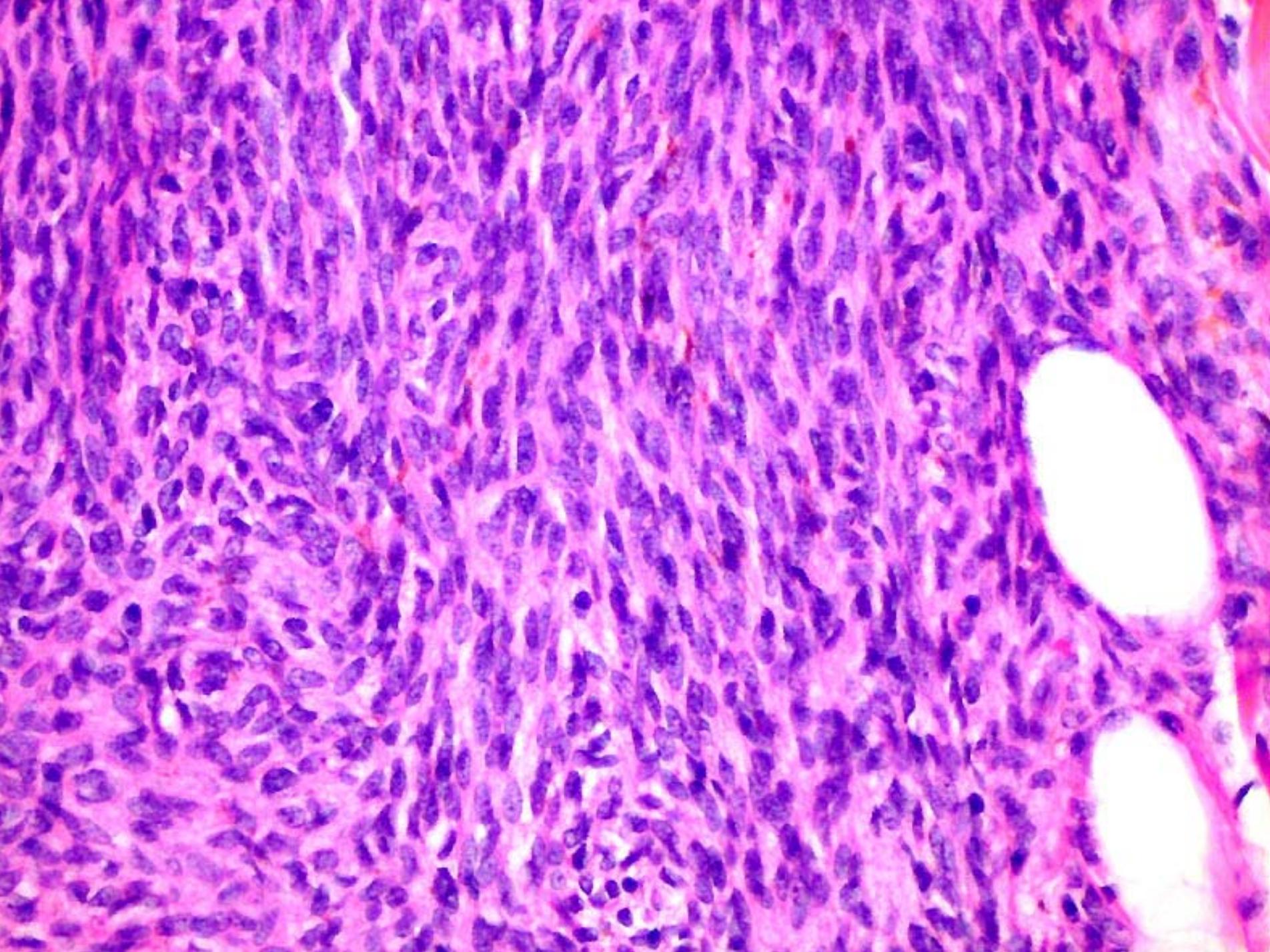


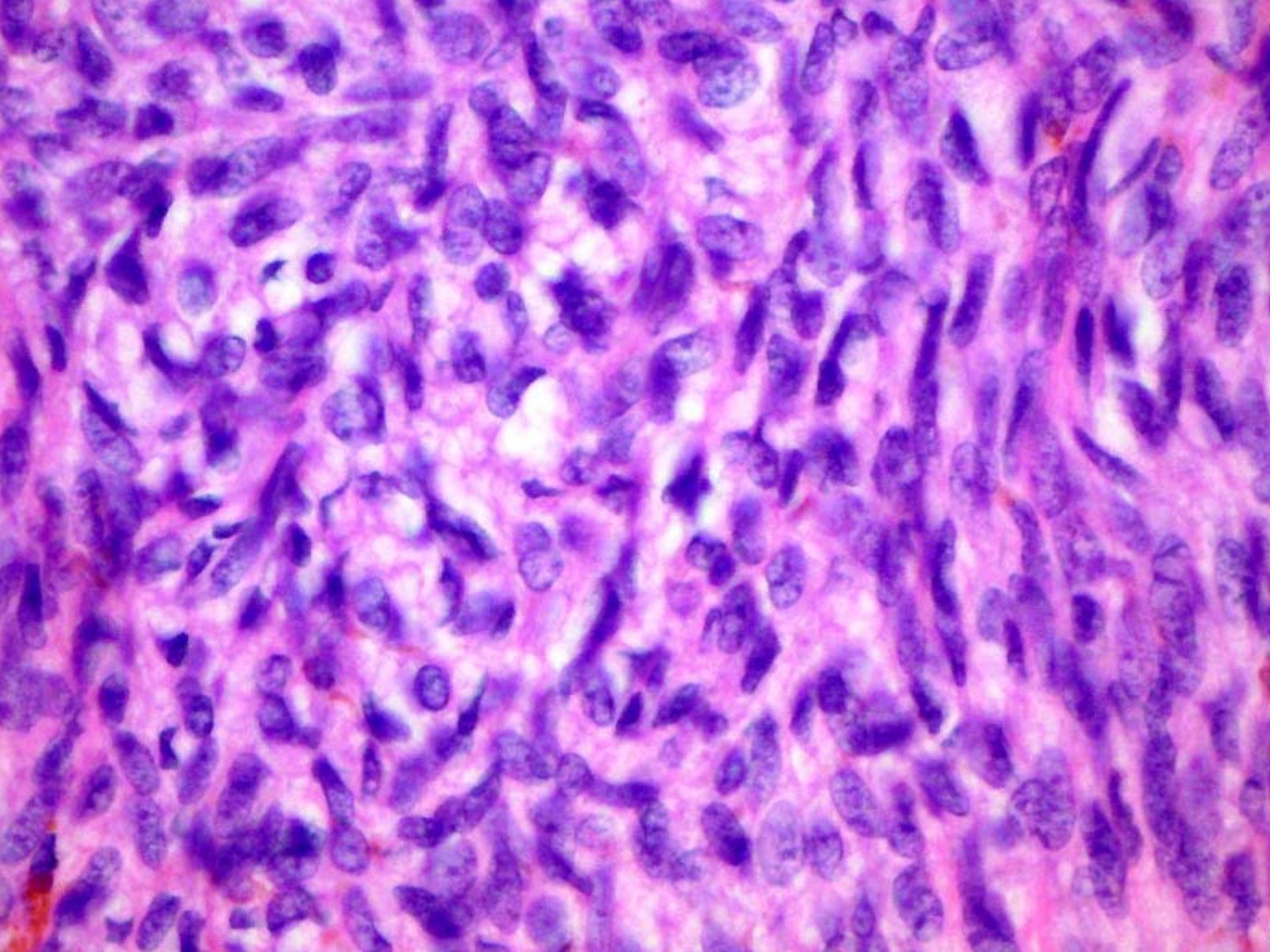












**Cellular Nodule  
in  
Congenital Blue Nevus**

# Studies

Study	Melanoma Risk
Swerdlow, etal. 1995	1046:1 >5% BSA (n=33) None in <5% BSA (n=232)
Rhodes 1982	5-15% overall risk for all CN 5-20% for large CN  Histology of nevi with melanoma
Sahin, etal. 1998	None for intermediate sized nevi 6.7 yrs f/u Av. age 25.5 yrs
Shpall, etal. 1994	164-2000:1 to age 75 yrs (Blacks only)

**CN <4% of BSA not  
significant risk for melanoma**

# Melanoma in LCMN

- 50% < 5 yrs
- 70% < 10 yrs

# Treatment



# Issues

- Risk of melanoma
- Risk of NCM
- Follow up
- Psychological impact of scars

# Surgical Removal

- Impossible to completely remove all melanocytes
- Excision lowers but does not eliminate melanoma risk
- Removal secondary concern in NCM-first get MRI

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