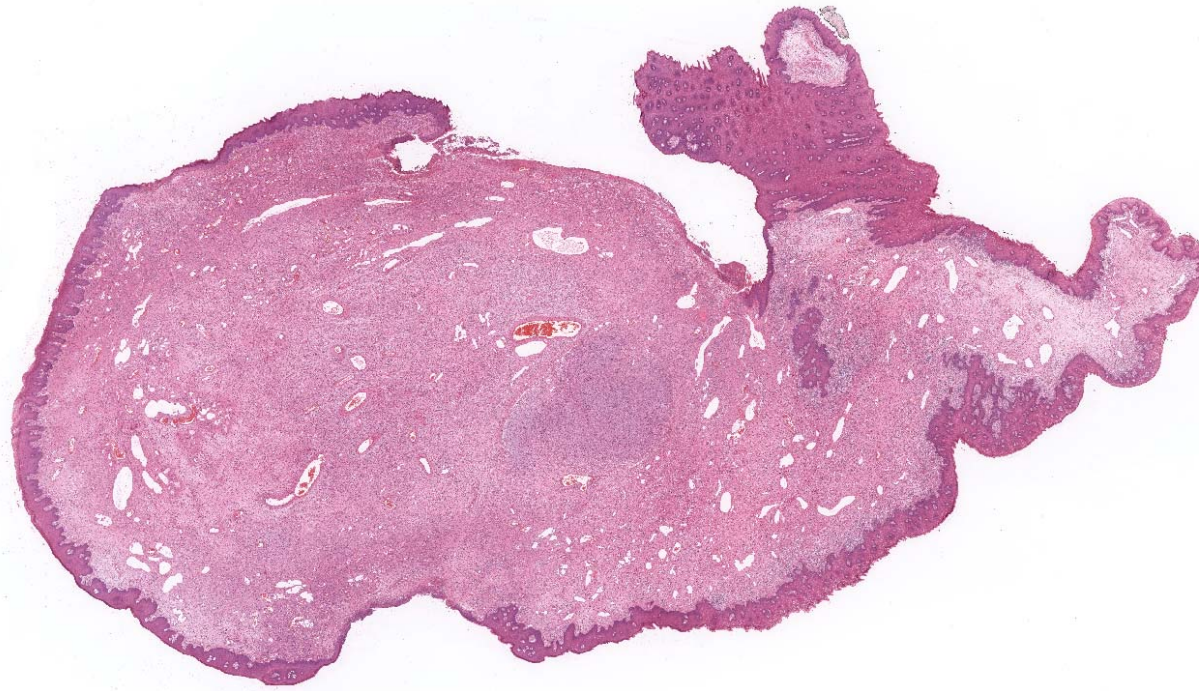


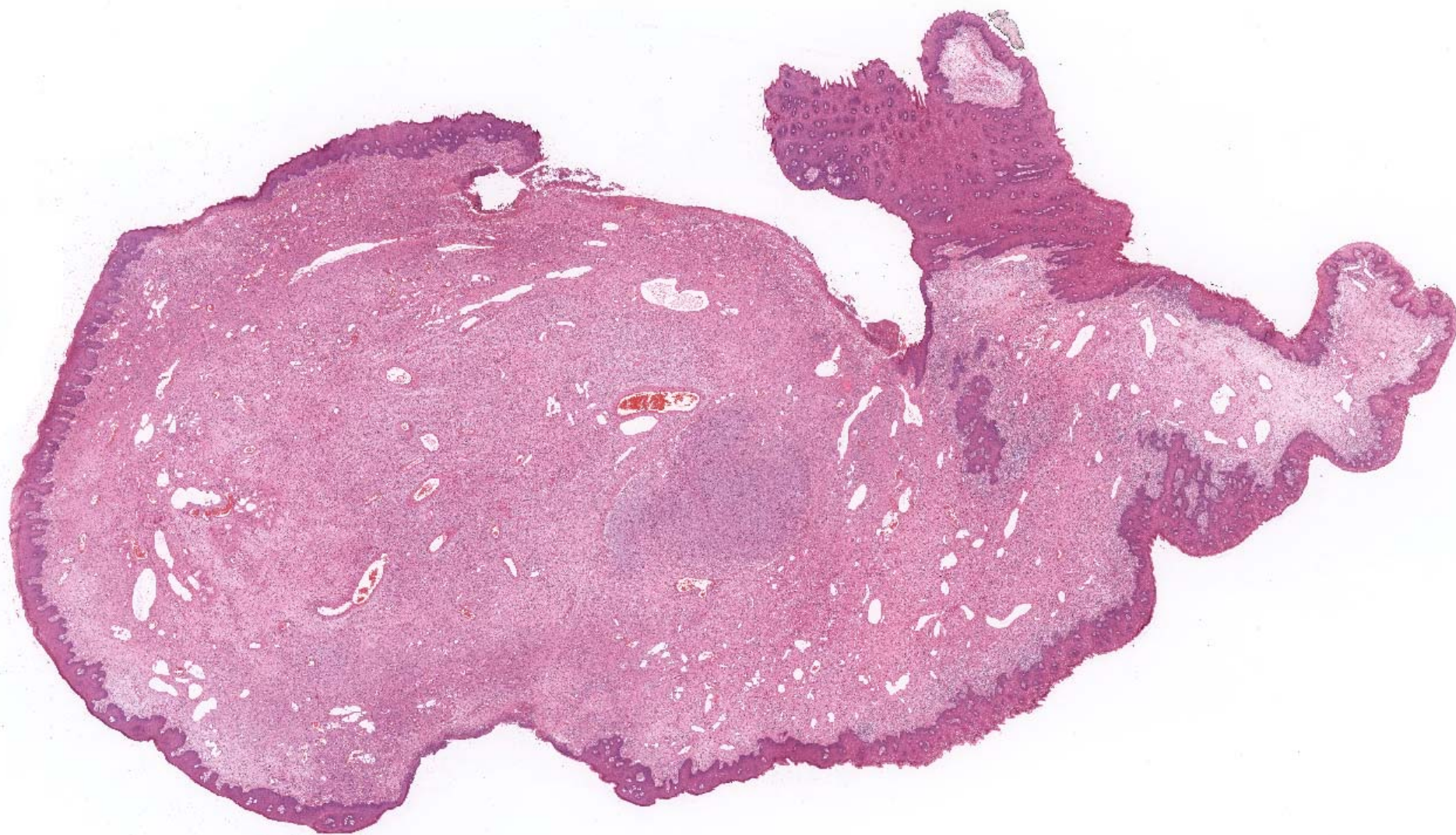
SL-13 Case 5

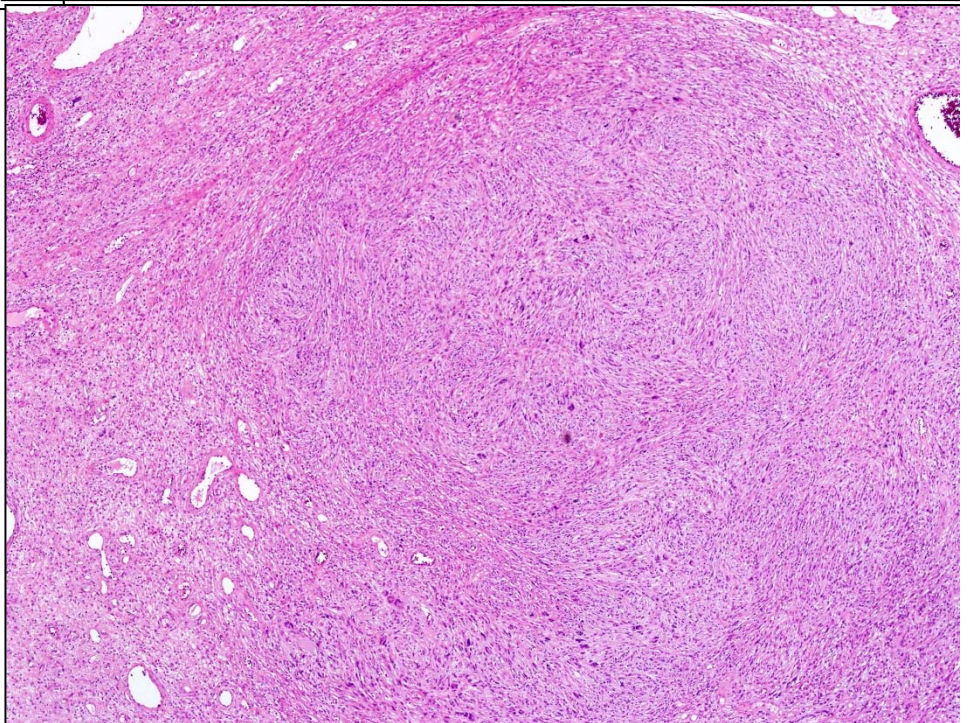
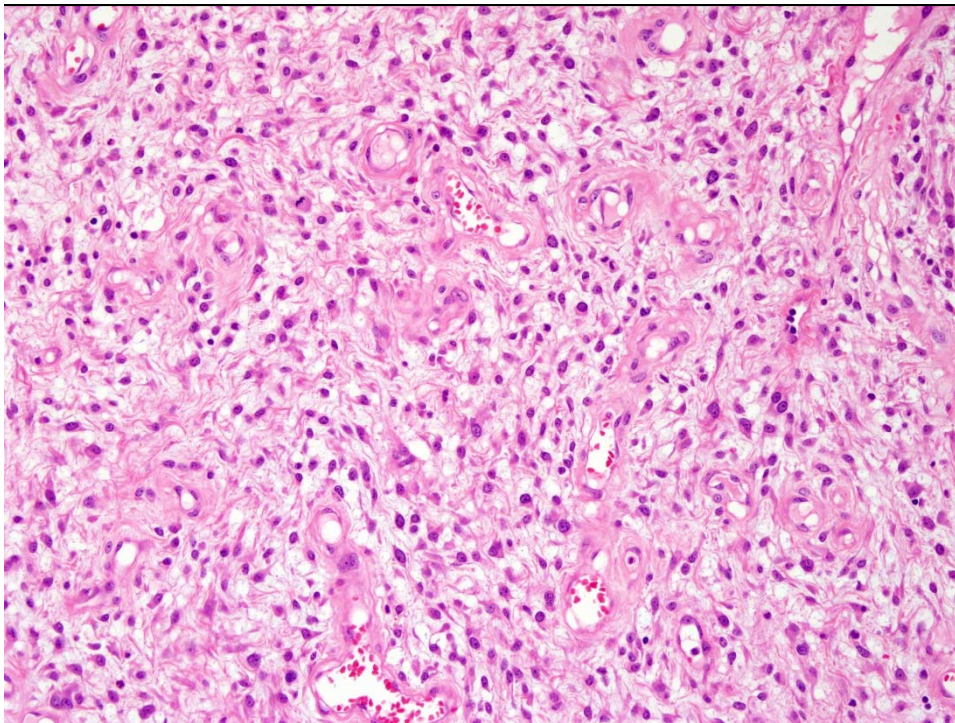
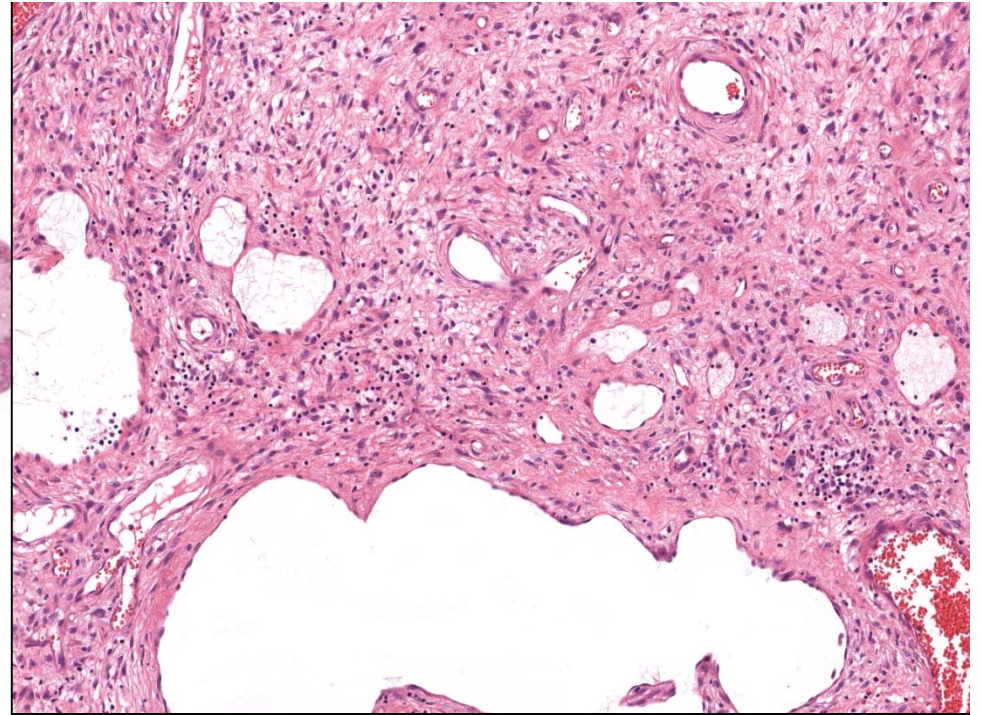
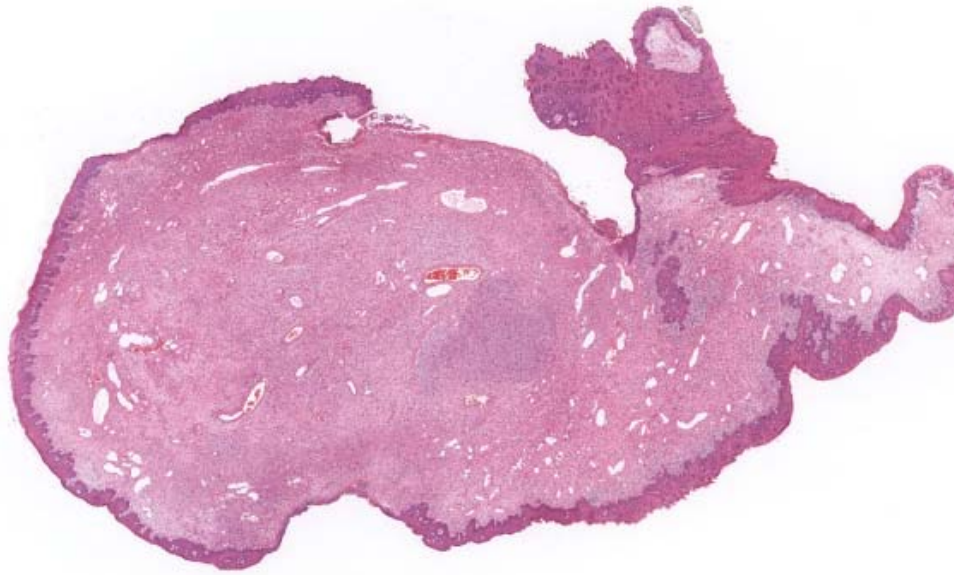


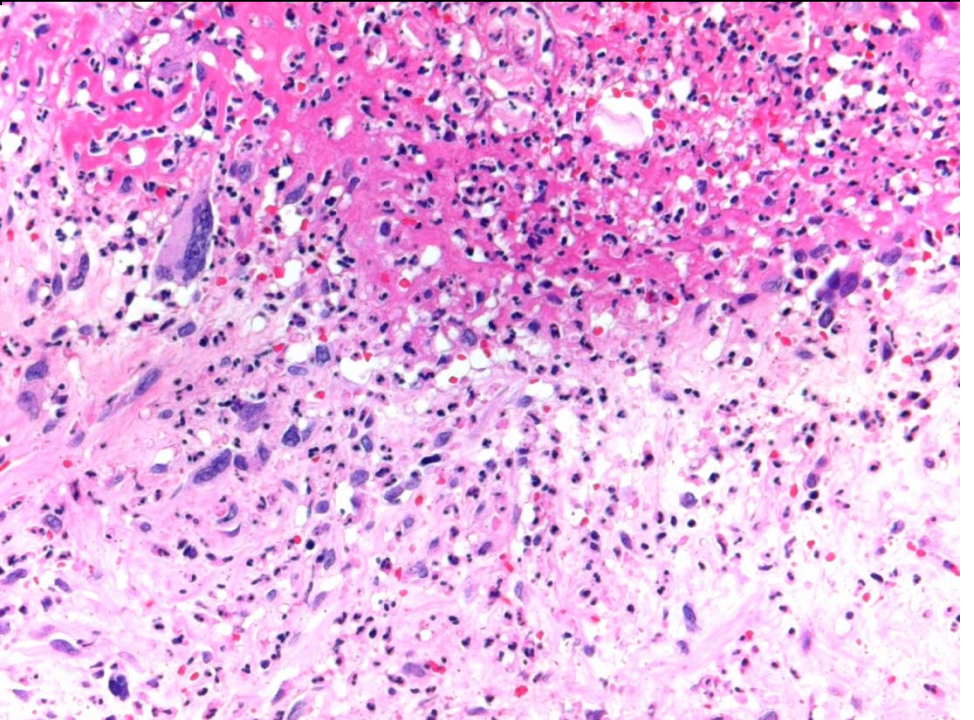
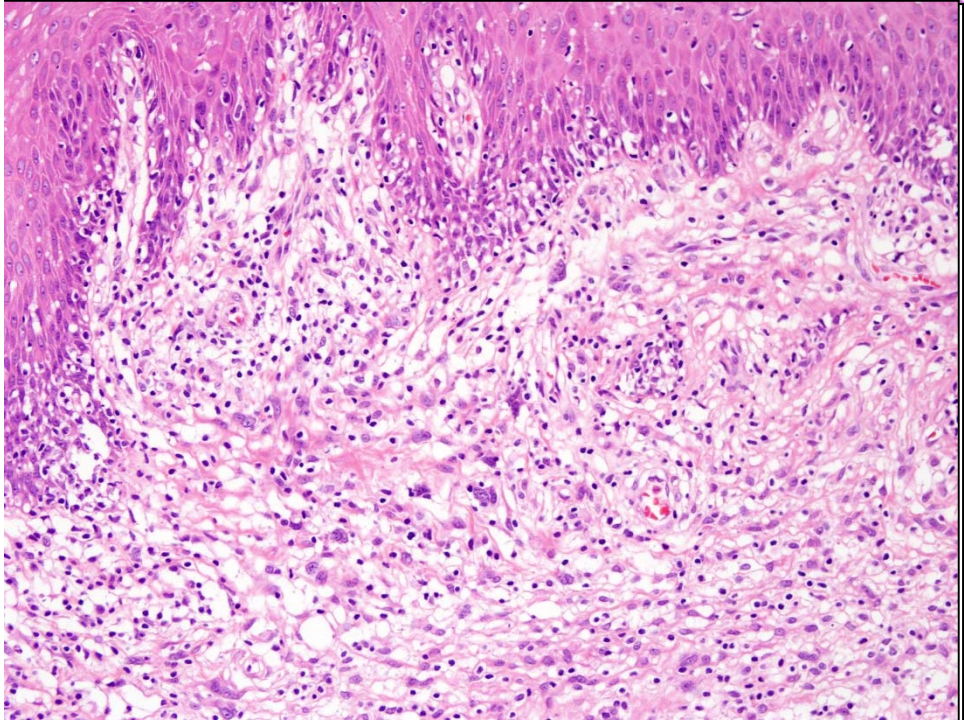
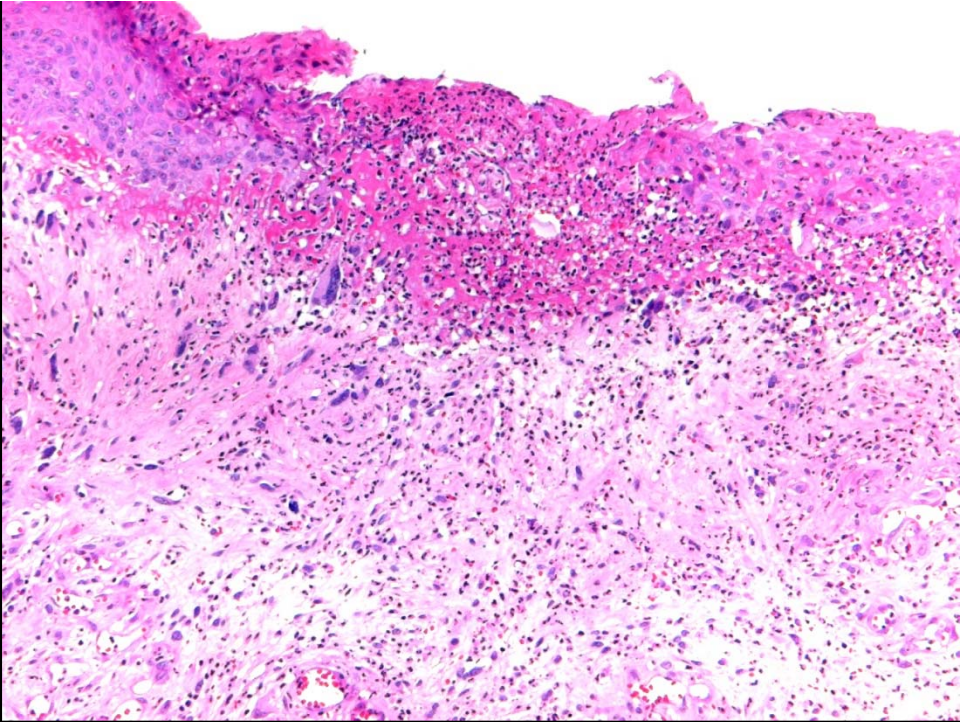
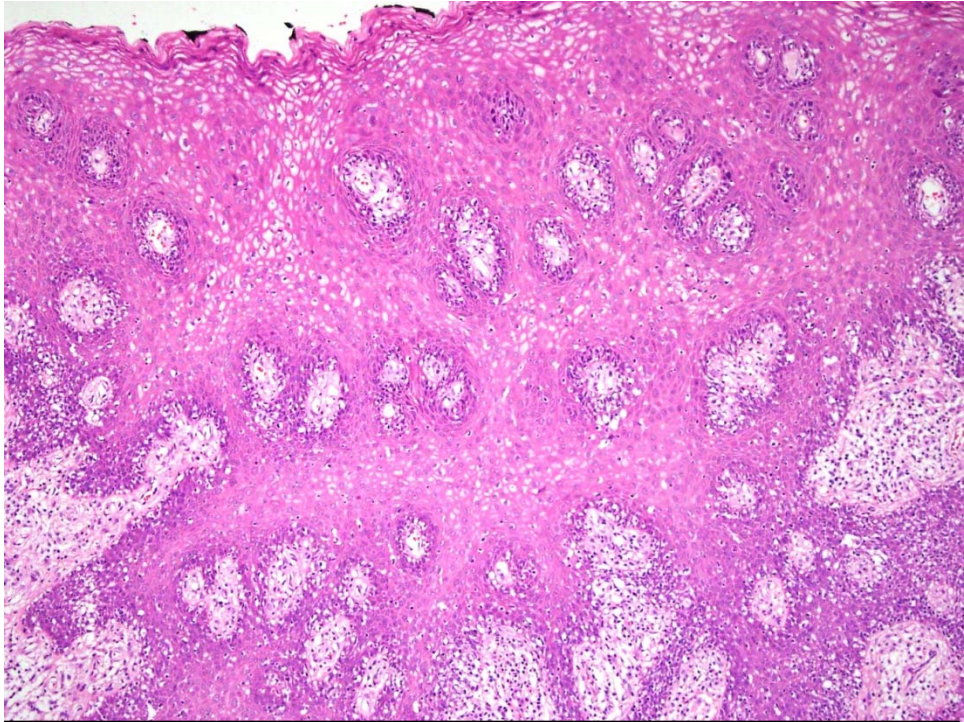
Kyu-Rae Kim, M.D.
Department of Pathology, Asan Medical Center,
University of Ulsan College of Medicine, Seoul, Korea

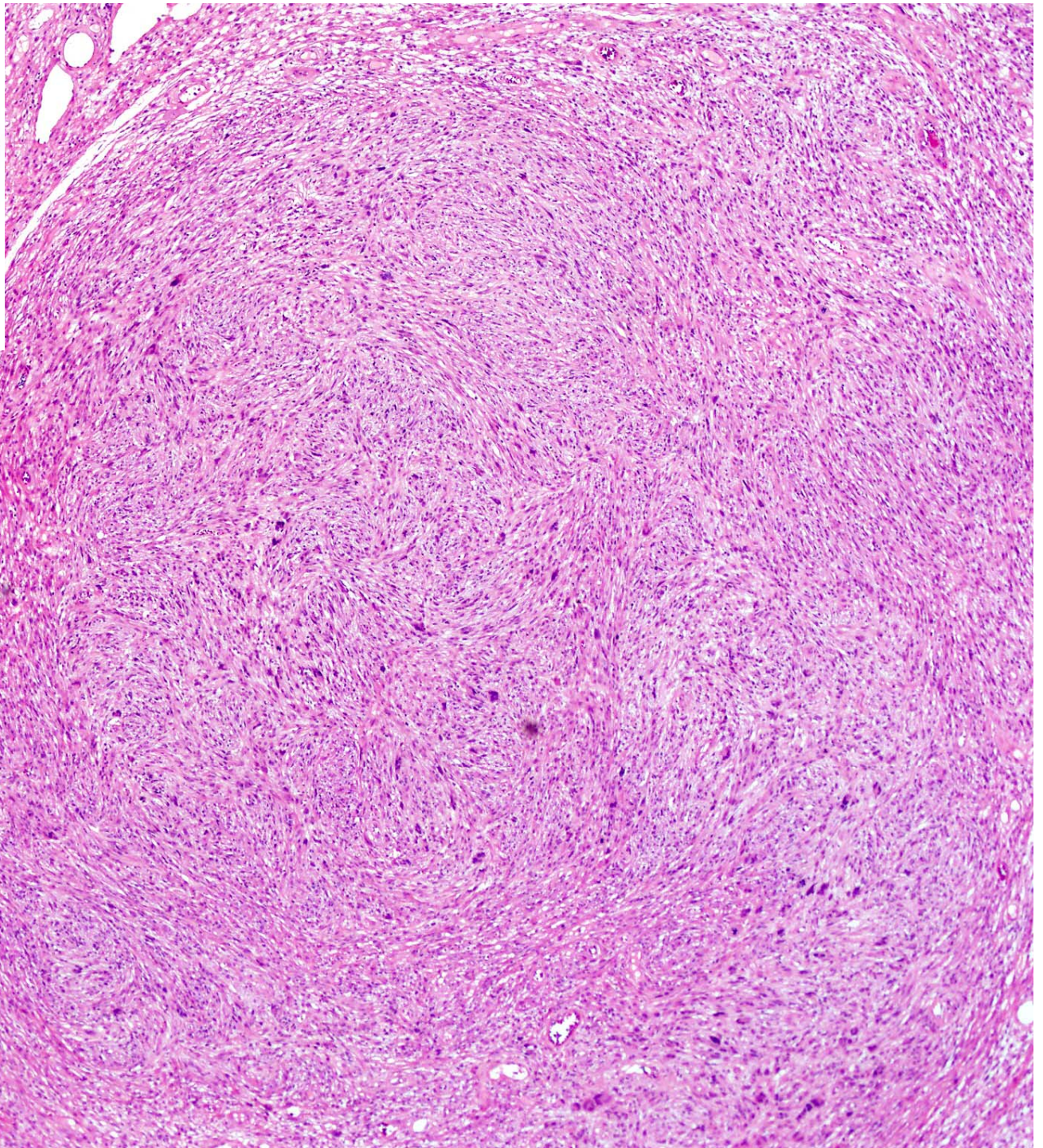
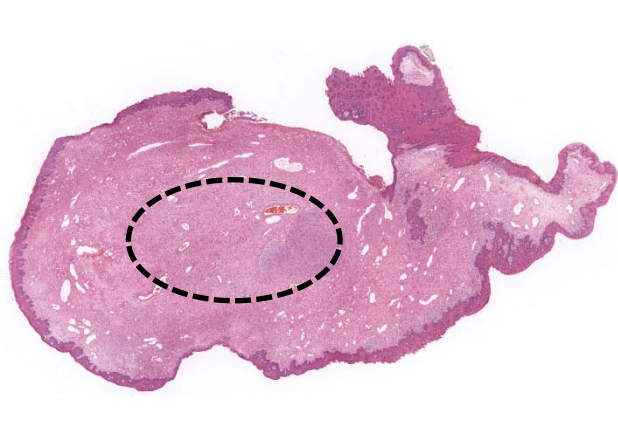
Clinical History

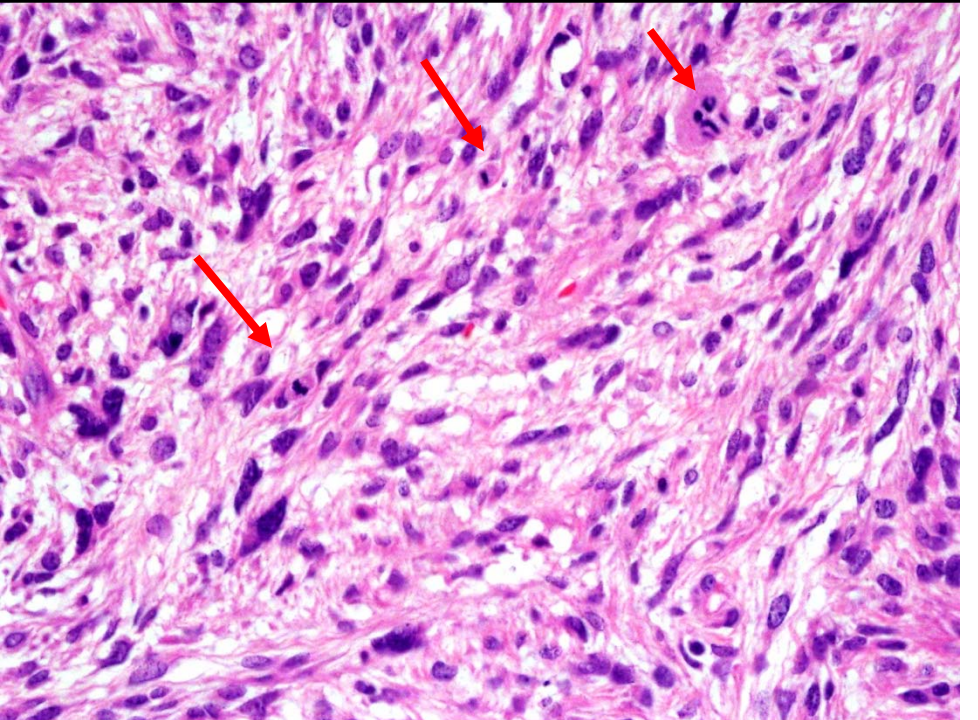
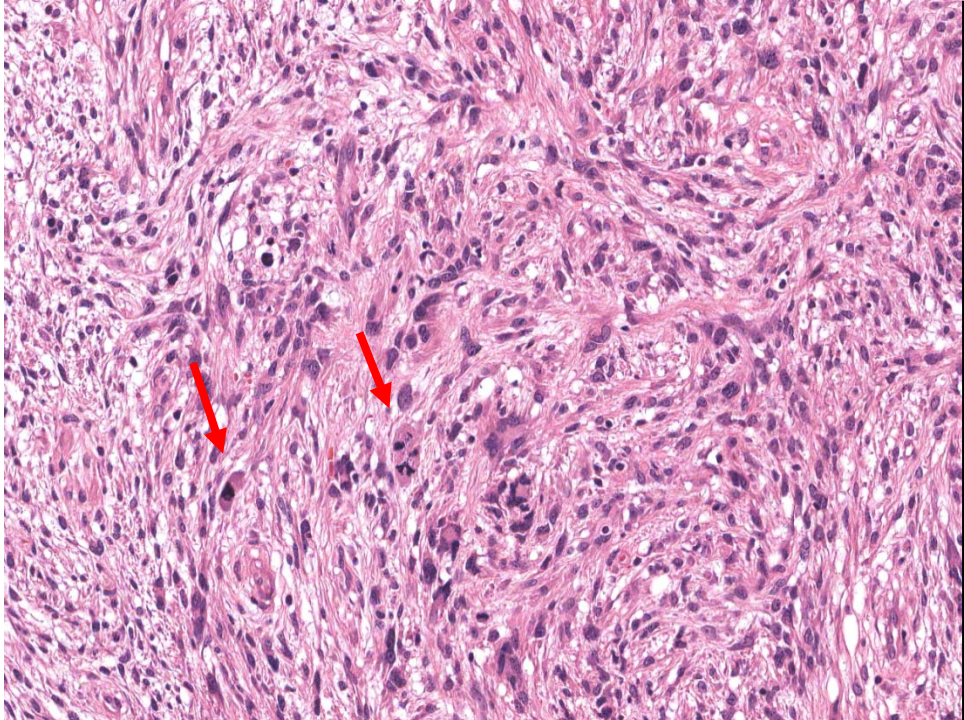
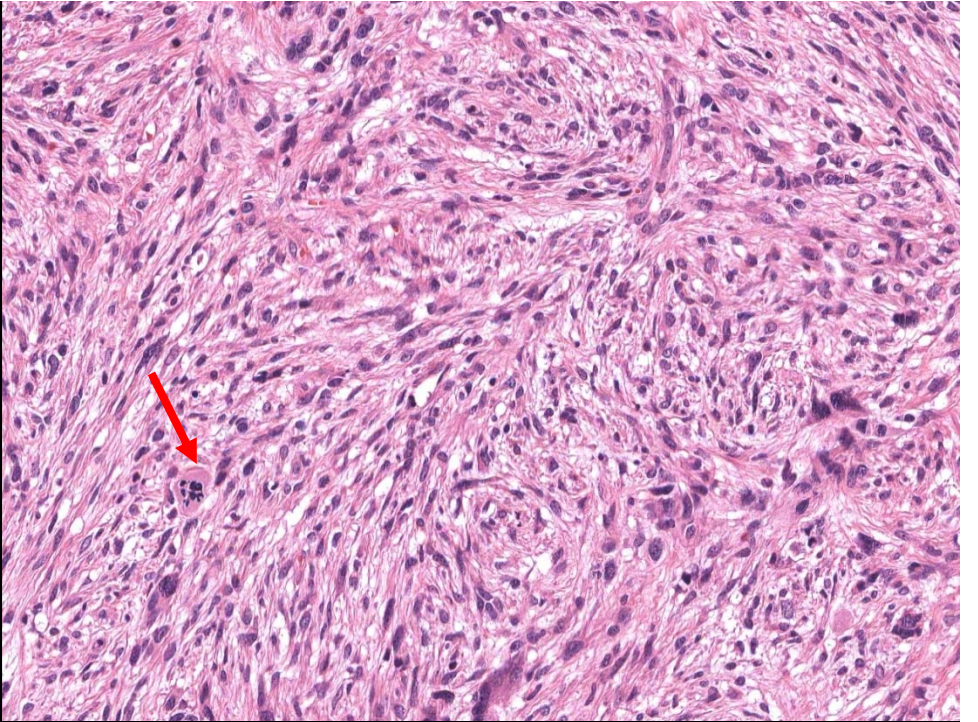
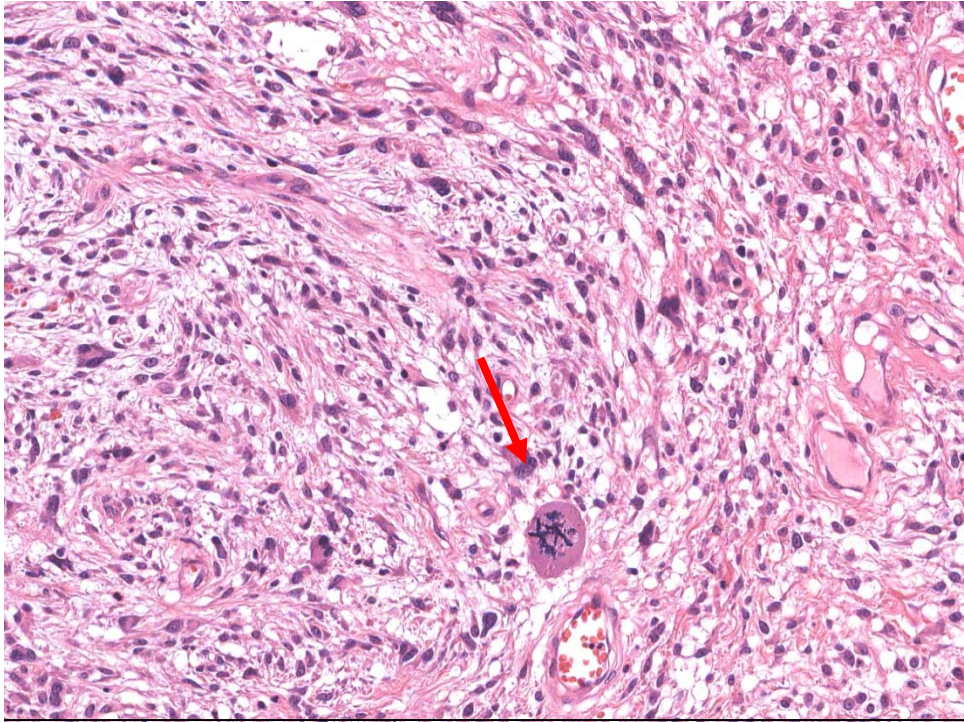
- 30 year old, 1-0-1(0/1)-1(0/1), female.
- 36 gestational weeks.
- Unremarkable past clinical history.
- Incidentally identified 2.5 cm-sized polypoid mass in the vaginal wall
- The vaginal mass completely removed after delivery.



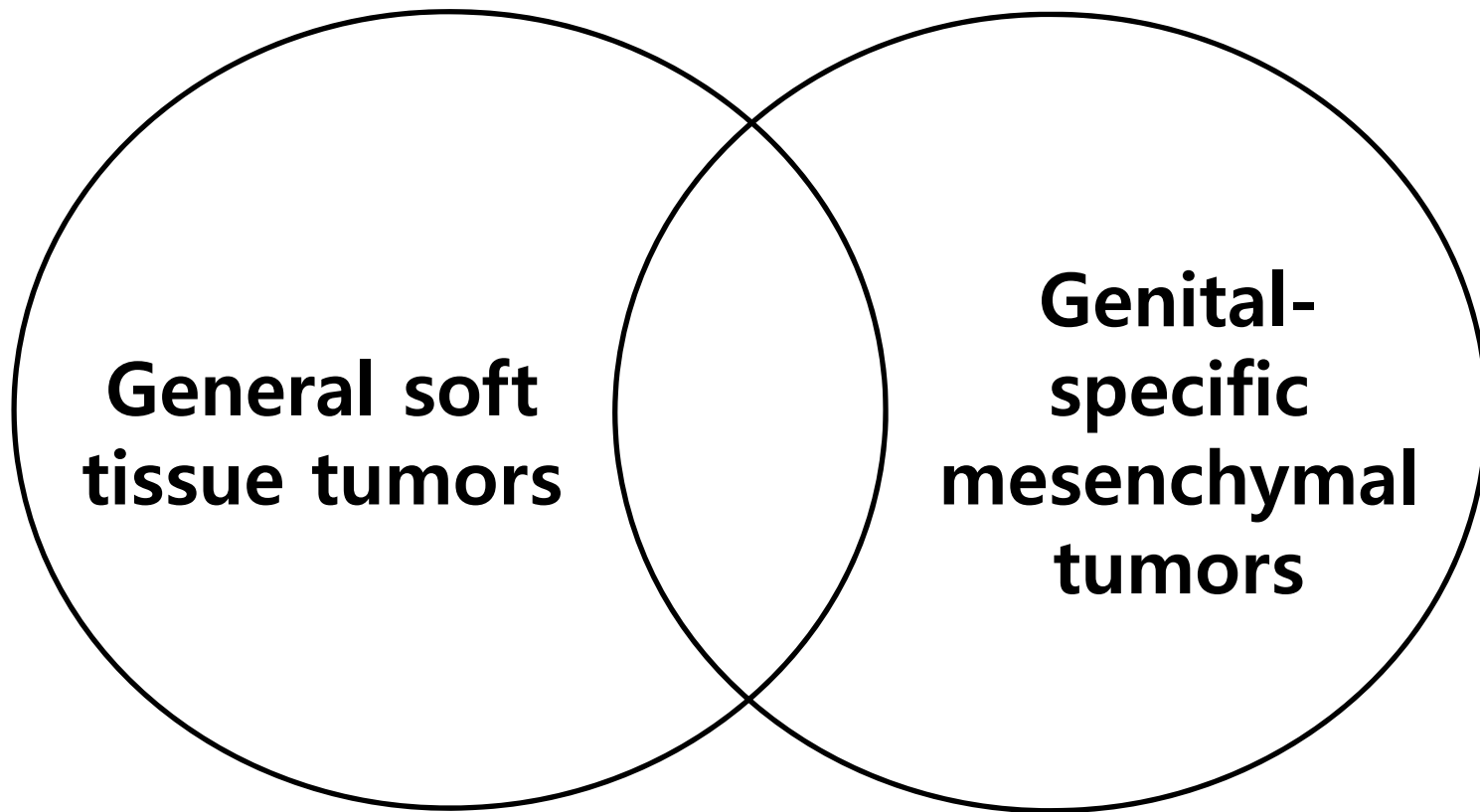








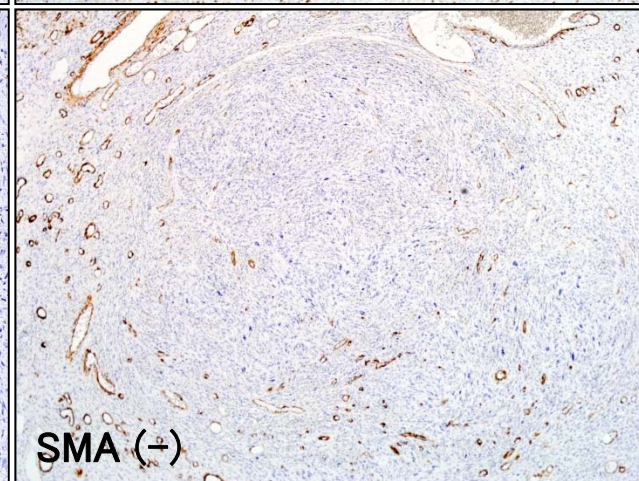
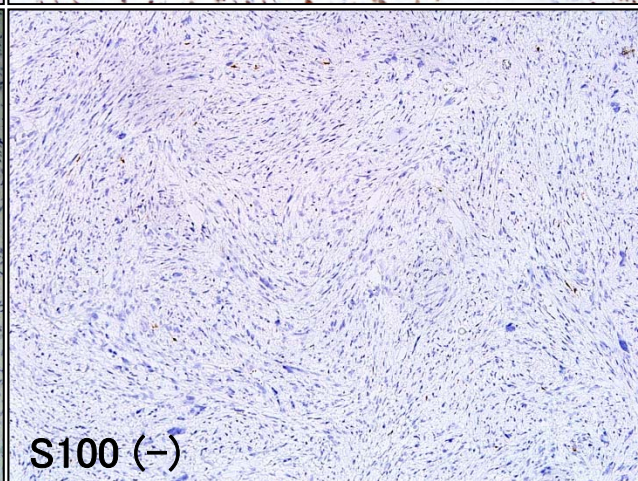
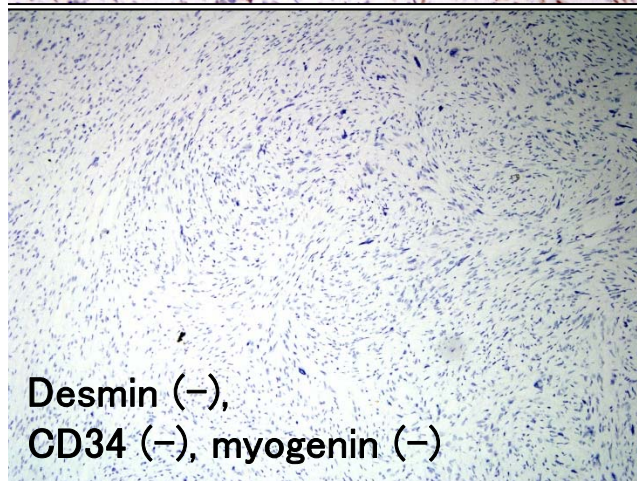
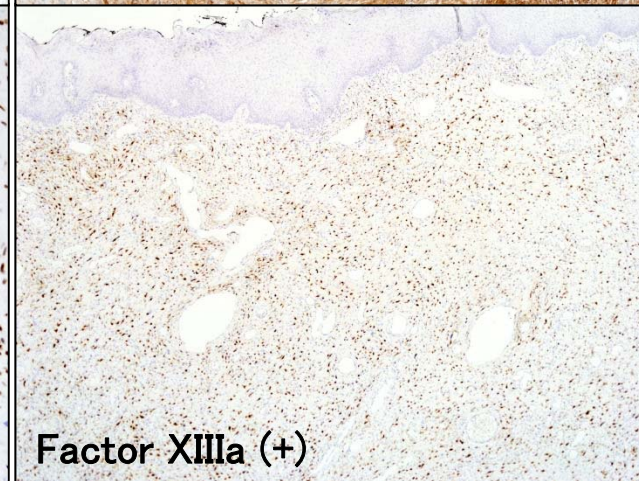
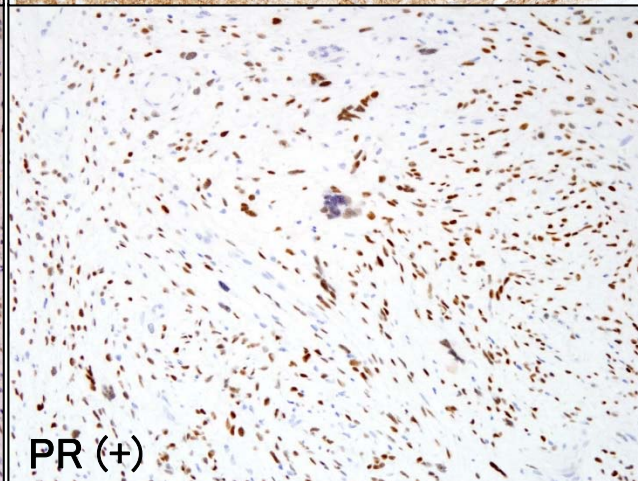
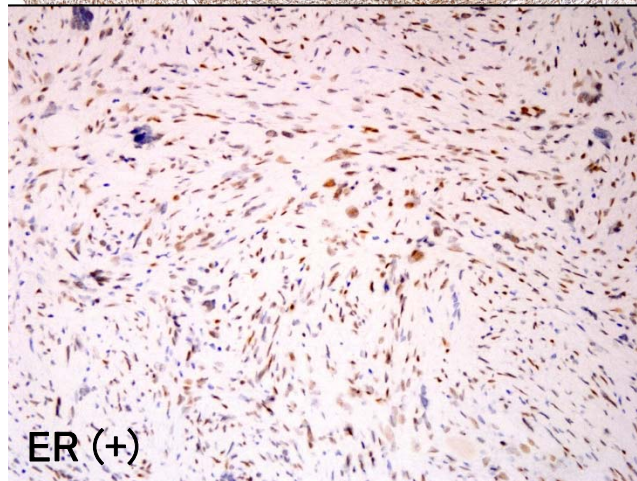
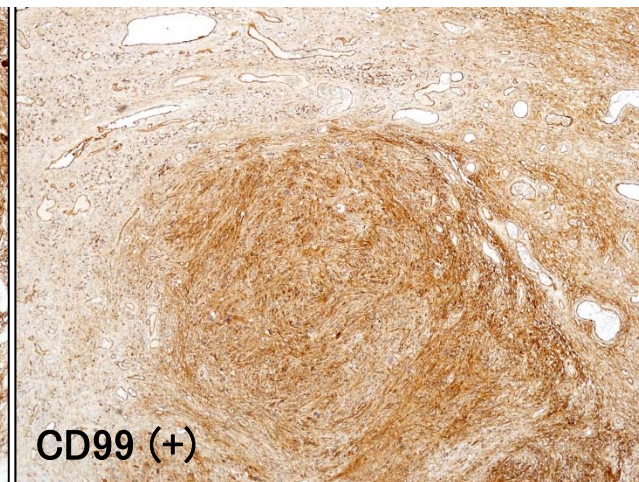
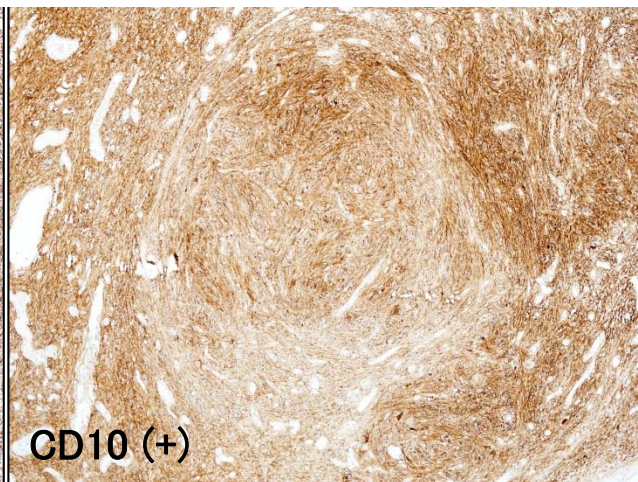
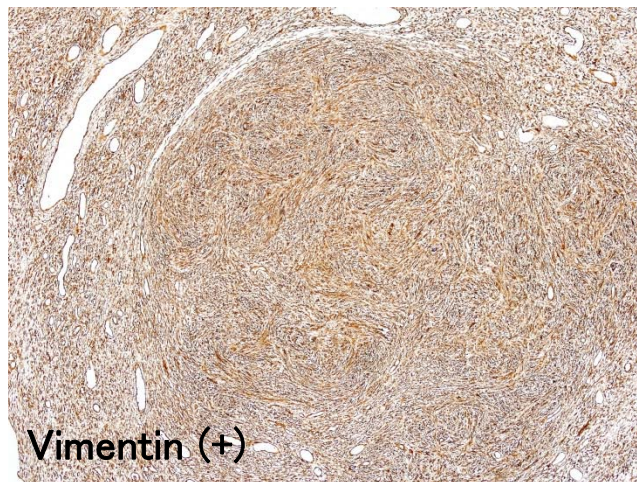
Mesenchymal Lesions of the Vulvovaginal Regions

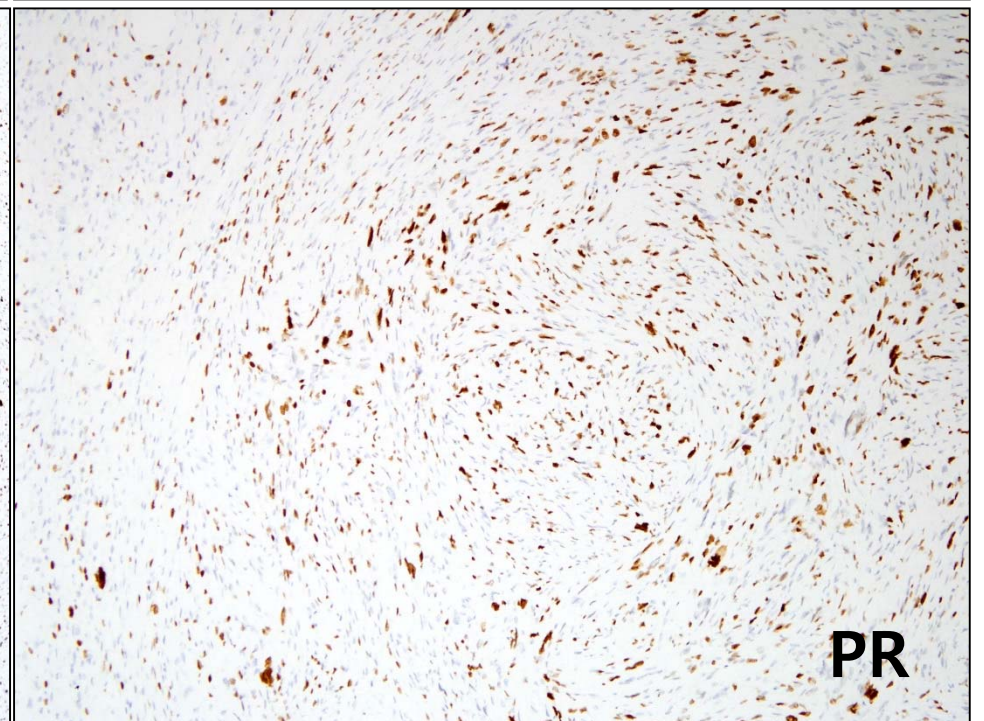
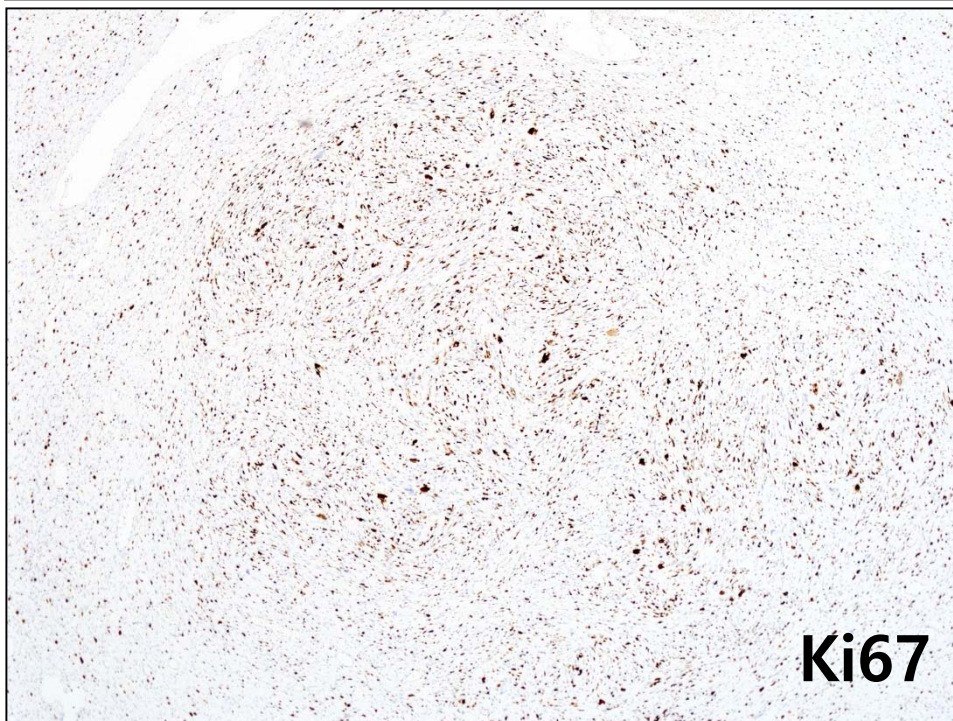
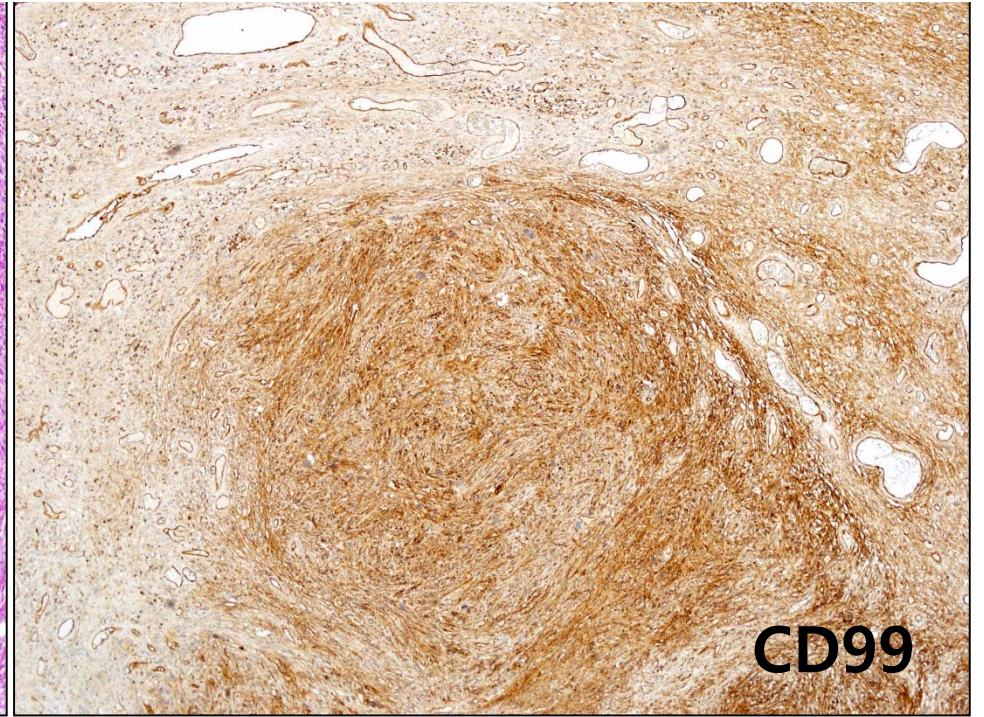
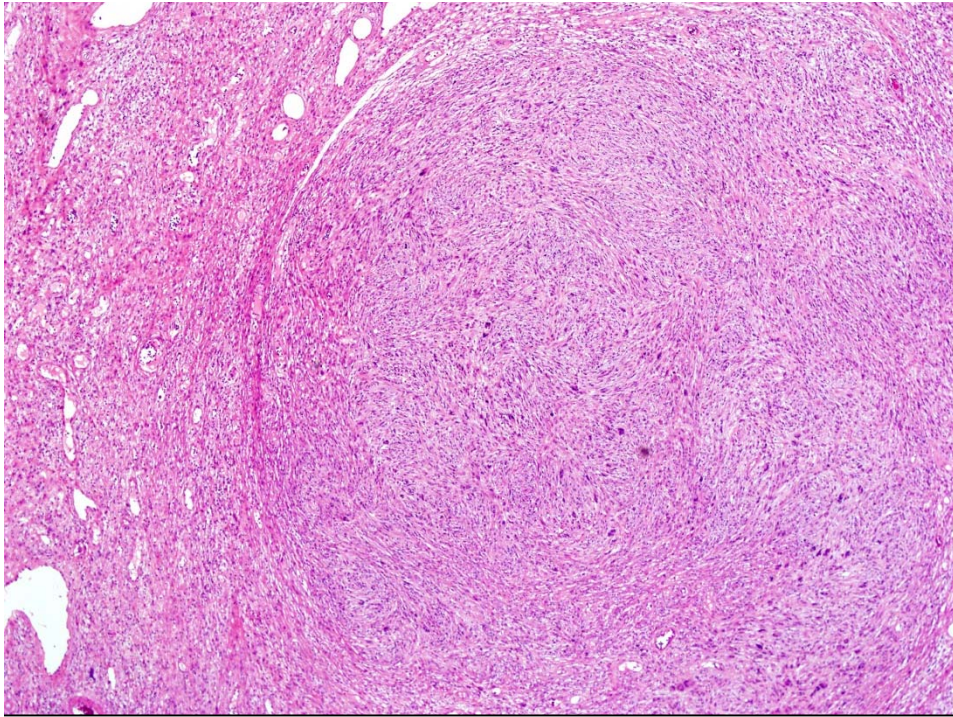


**Striking morphologic and immunophenotypic
similarity !!**

Differential Diagnosis

1. Cellular (pseudosarcomatous) fibroepithelial stromal polyp
2. Superficial (cervicovaginal) myofibroblastoma
3. Cellular angiofibroma
4. Angiomyofibroblastoma
5. Aggressive angiomyxoma
6. Superficial angiomyxoma
7. Botryoid embryonal rhabdomyosarcoma
8. Solitary fibrous tumor
9. Sarcomatoid squamous cell carcinoma



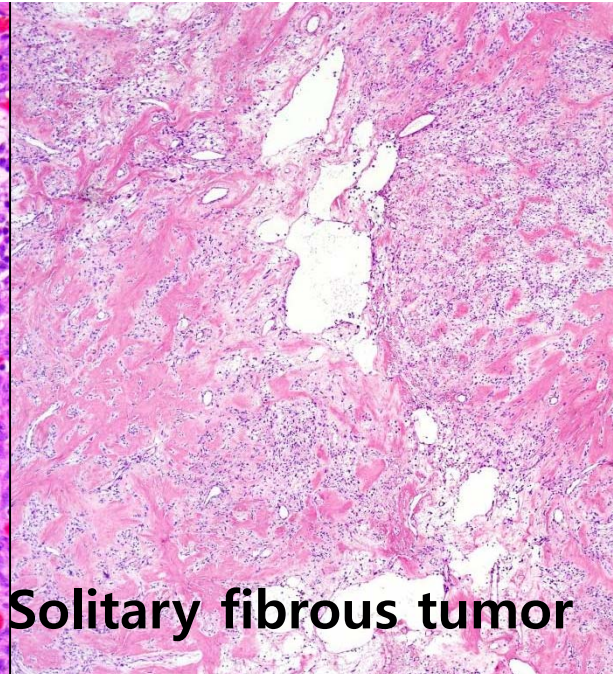
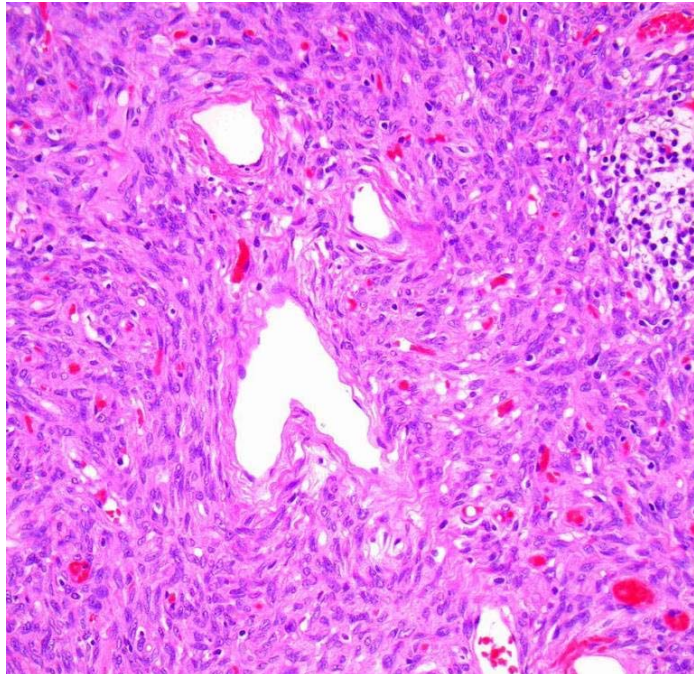


Clinicopathologic Characteristics

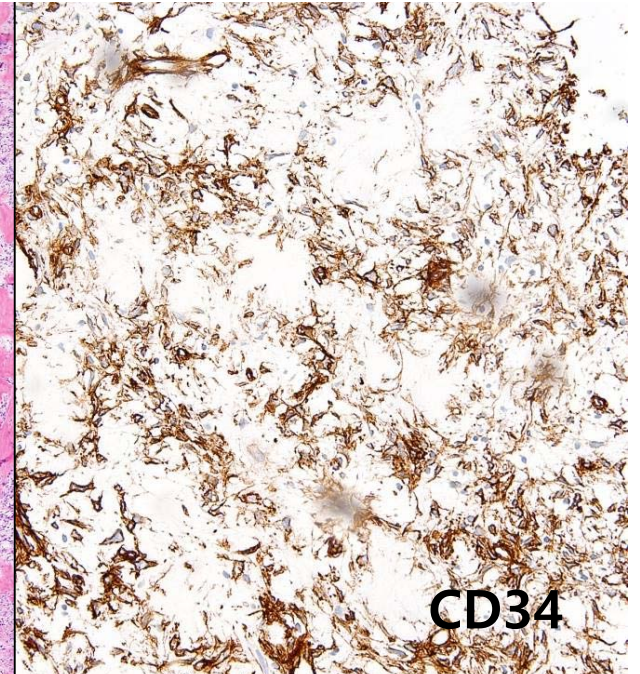
- Pregnant woman
- Polypoid mass in the vaginal wall
- No Grenz zone from overlying epithelium
- Vascularized spindle cell lesion
- Significant nuclear atypia, stromal giant cells, increased mitotic figures including atypical forms
- Fibrocollagenous stroma
- +ve; Vimentin, CD10, CD99, Fac XIIIa, ER & PR

Differential Diagnosis

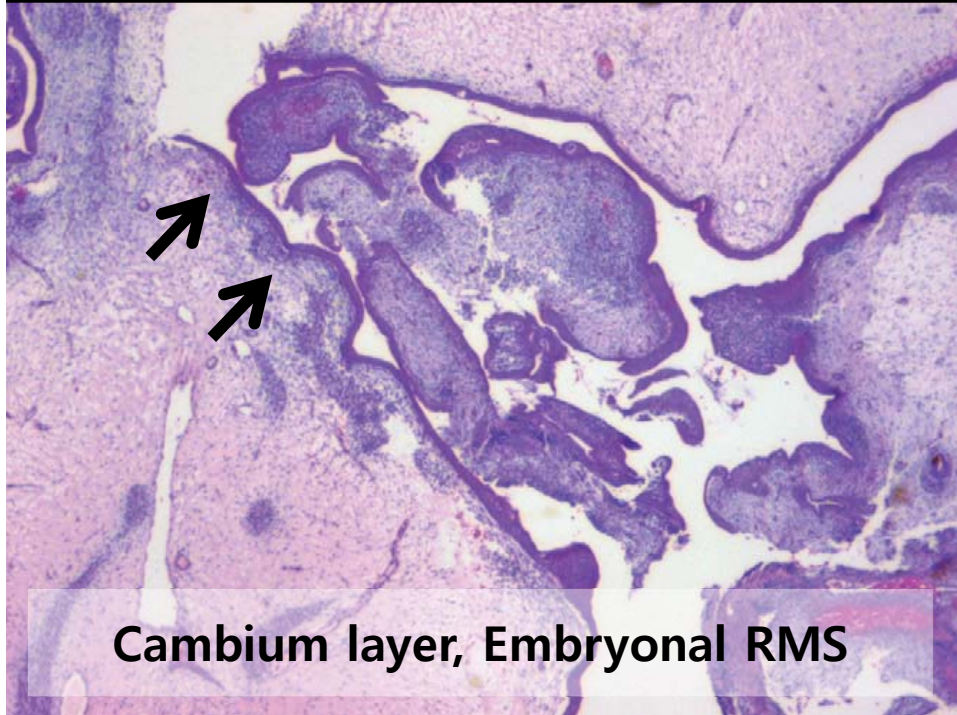
1. Cellular (pseudosarcomatous) fibroepithelial stromal polyp
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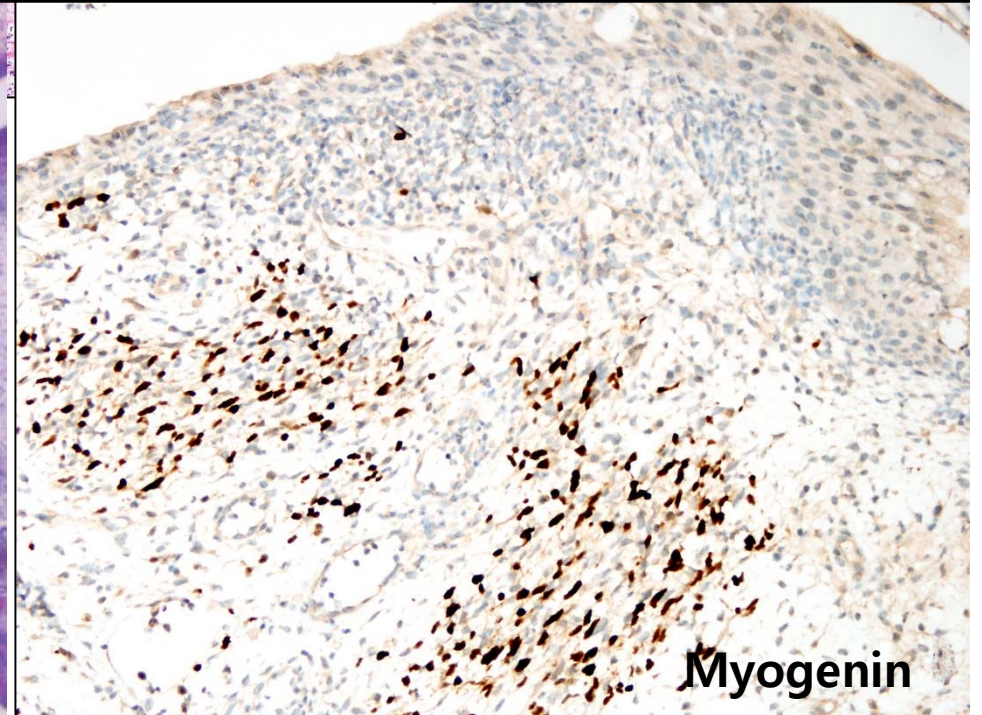
Solitary fibrous tumor



CD34



Cambium layer, Embryonal RMS

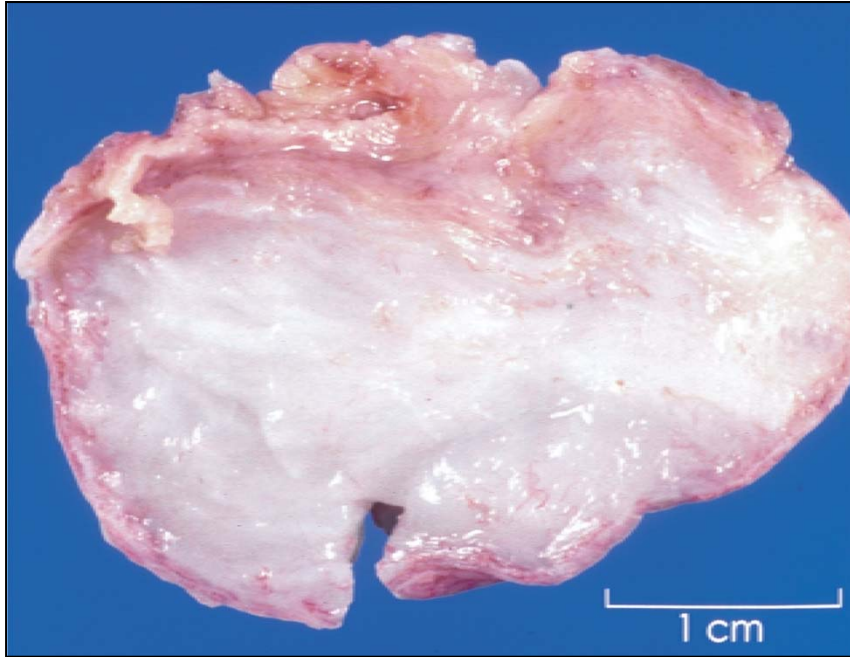


Myogenin

Differential Diagnosis

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4. Angiomyofibroblastoma
5. Superficial angiomyxoma
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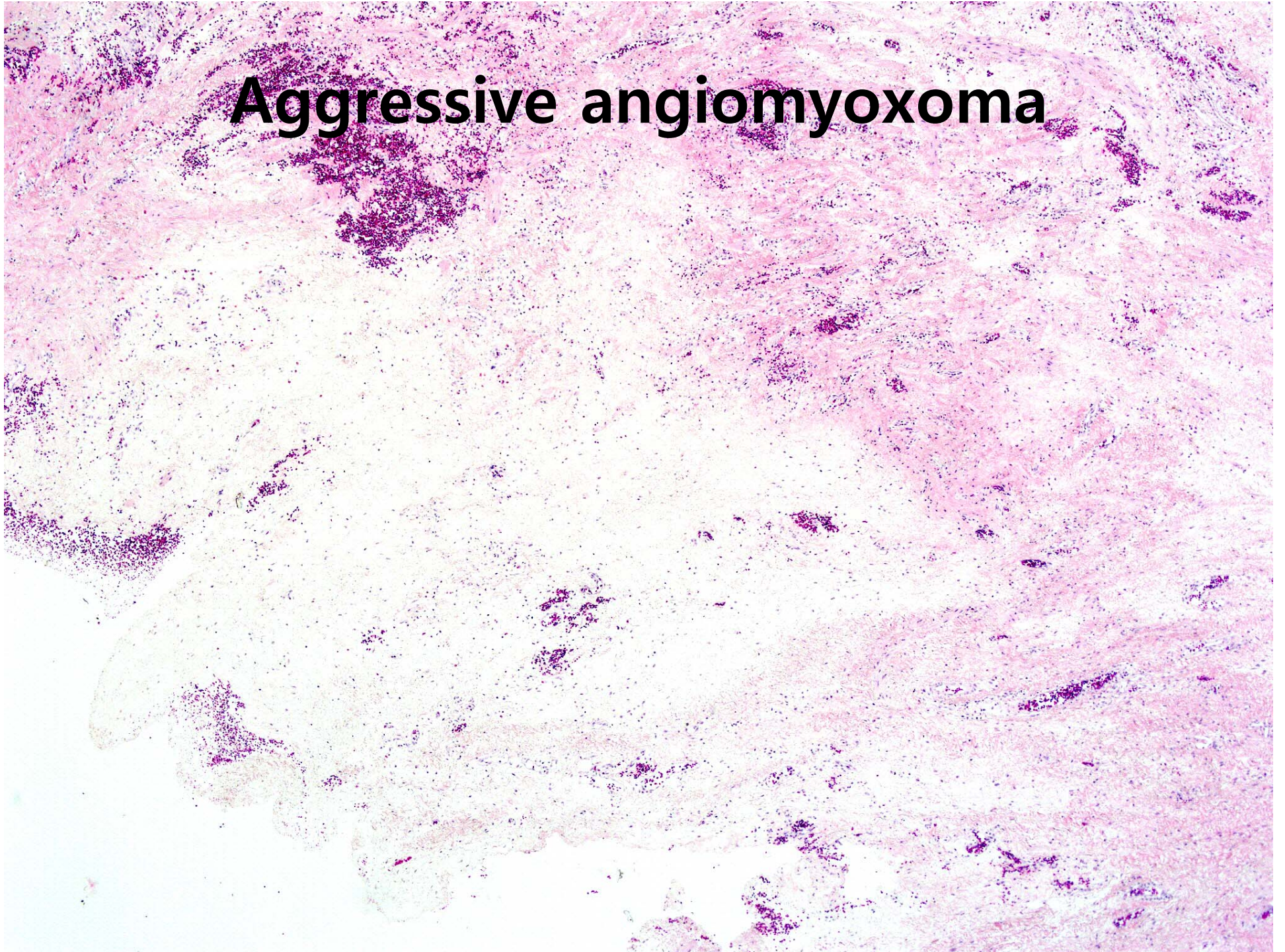
Aggressive angiomyxoma

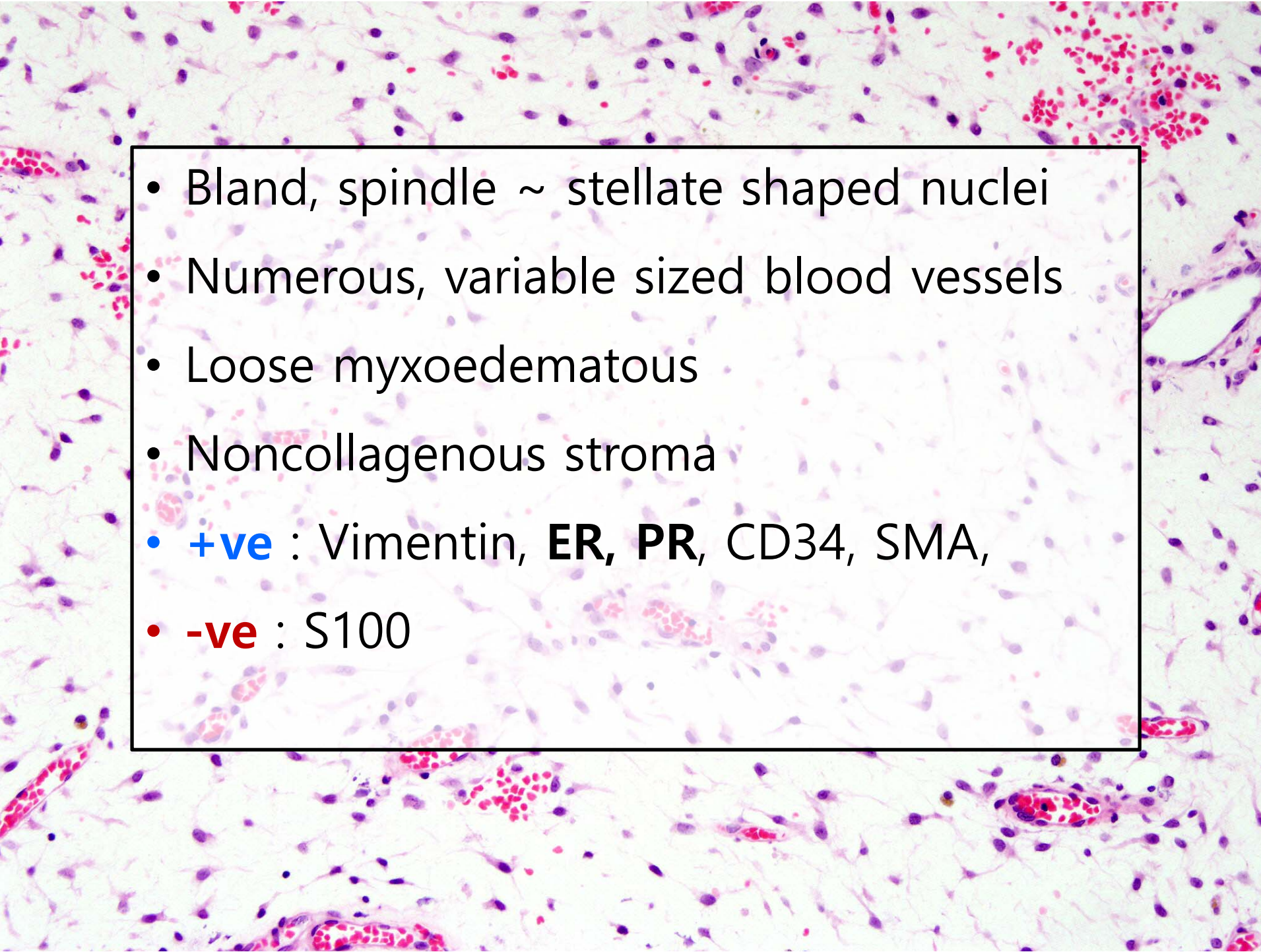


Adopted from Nielsen GP, Int J Gyn Pathol 2001

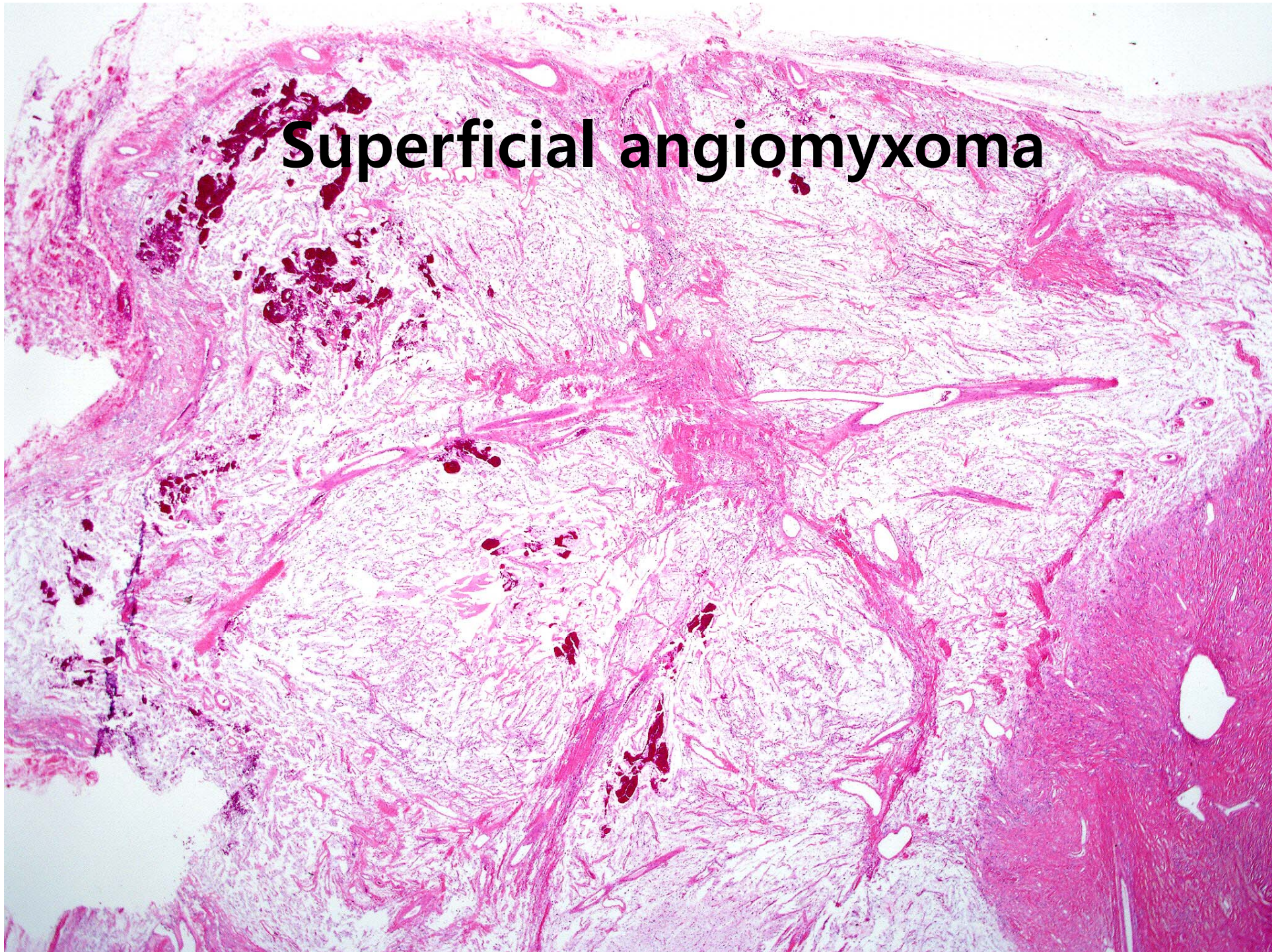
- Deep soft tissue mass of the vulvovaginal region, perineum and pelvis
- Reproductive age >> menopause
- Local infiltrative growth
- Poorly circumscribed
- Frequent local recurrence 30-40%
- Metastasis rare

Aggressive angiomyxoma



- 
- A histological slide showing a tissue sample with numerous spindle-shaped cells and variable-sized blood vessels. The cells have bland, spindle to stellate shaped nuclei. The stroma is loose and myxoedematous, and noncollagenous. The slide is stained with hematoxylin and eosin (H&E), showing purple nuclei and pink cytoplasm/extracellular matrix.
- Bland, spindle ~ stellate shaped nuclei
 - Numerous, variable sized blood vessels
 - Loose myxoedematous
 - Noncollagenous stroma
 - **+ve** : Vimentin, **ER**, **PR**, CD34, SMA,
 - **-ve** : S100

Superficial angiomyxoma

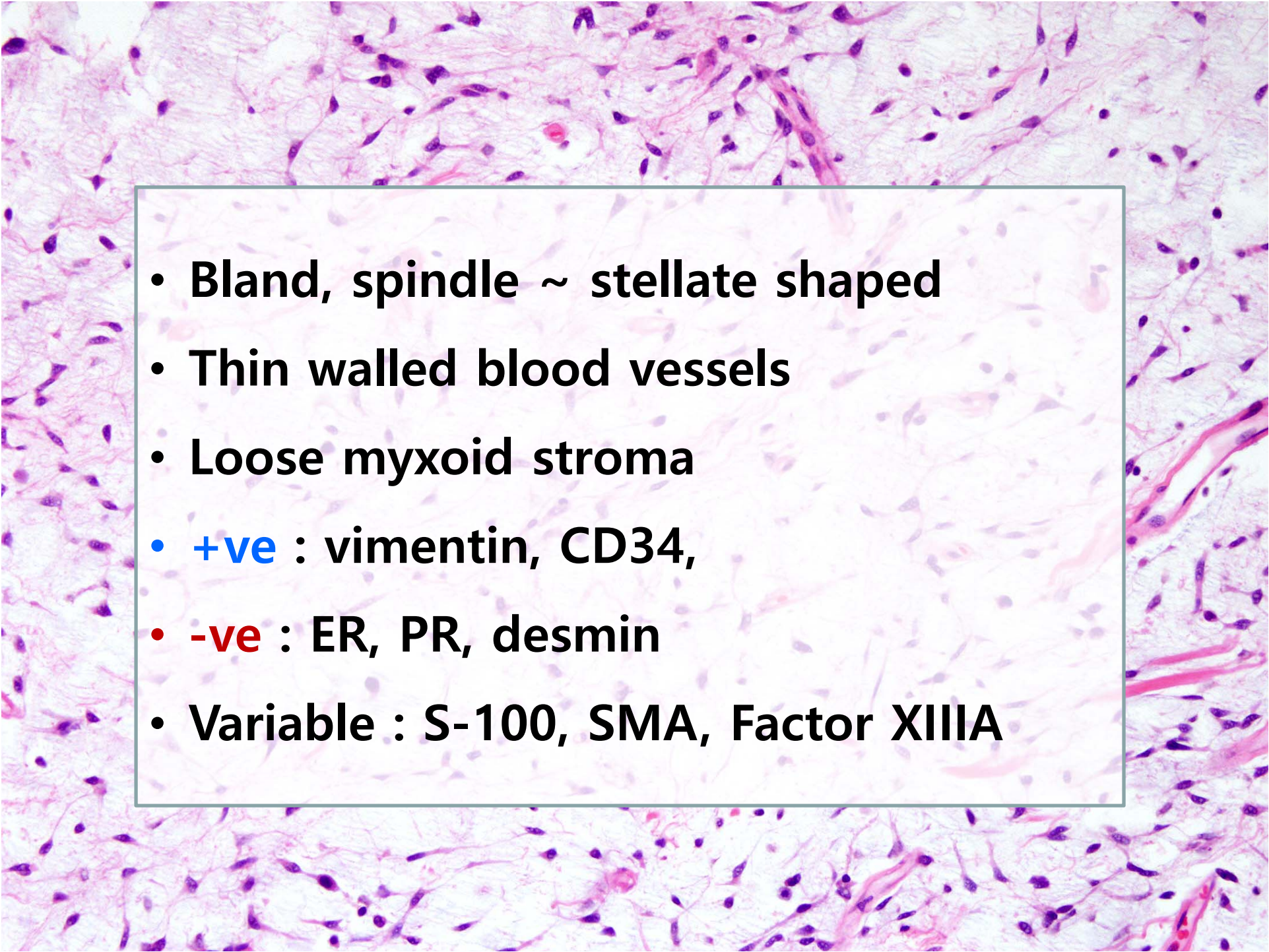


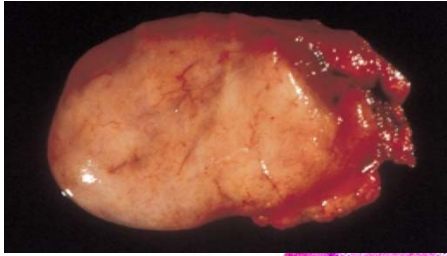
Superficial angiomyxoma



- Slow growing painless mass
- Superficial SQ /soft tissue layer
- Trunk, limb, H&N,
- Vulva, groin, scrotum & vagina

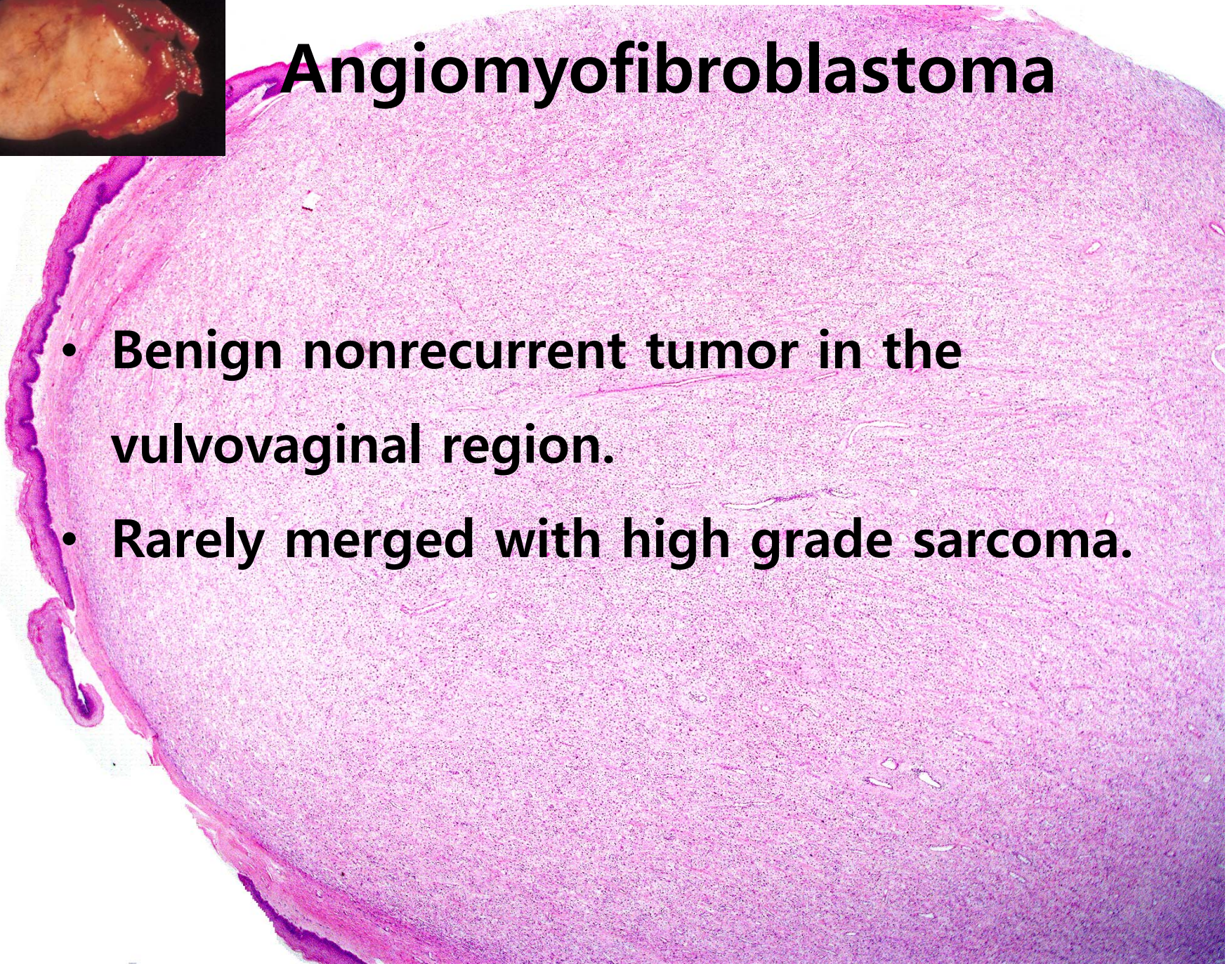
Adopted from HS Kim et al. J Cutan Pathol 2010

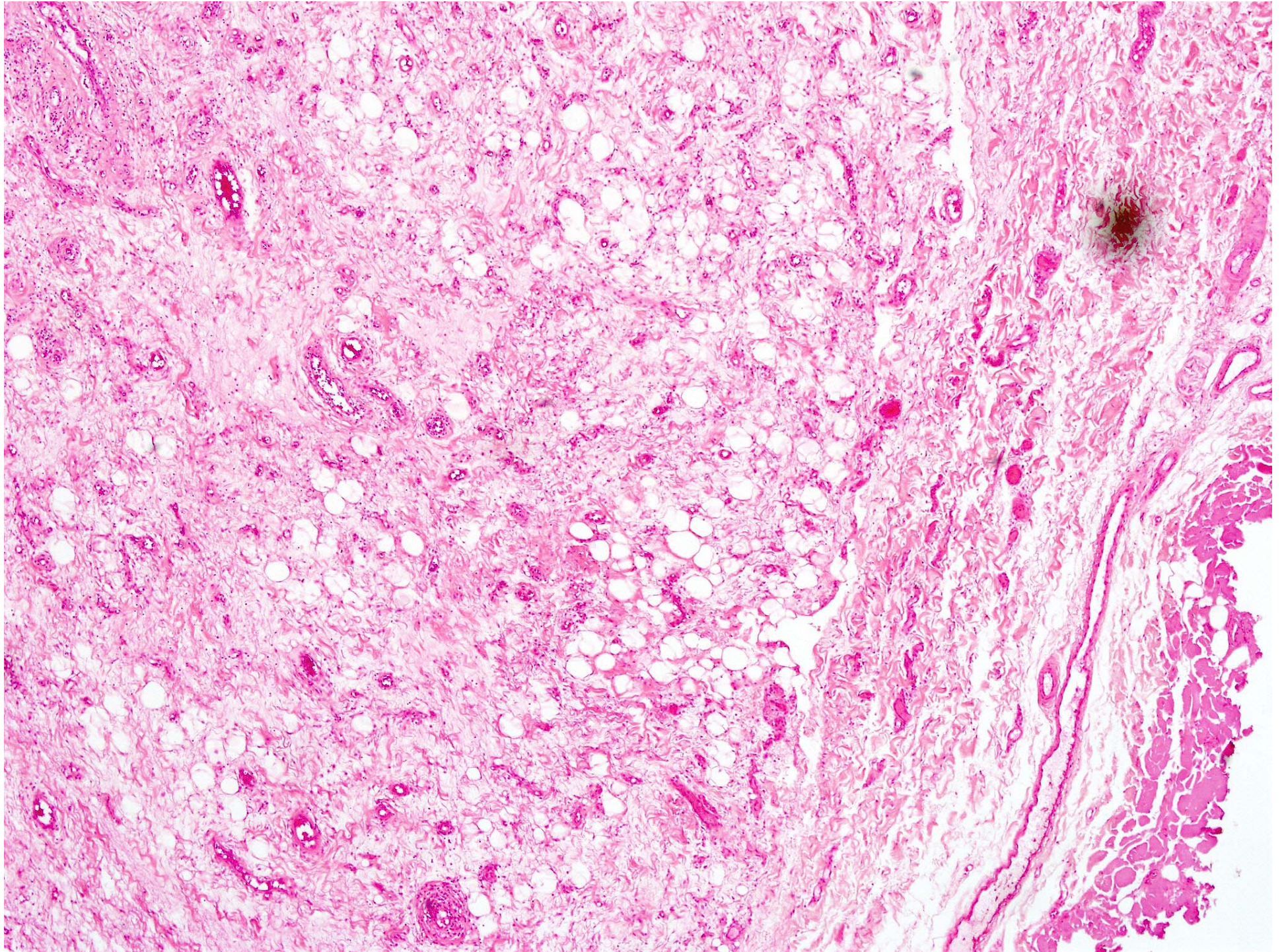
- 
- A histological slide showing a tissue sample with numerous spindle-shaped cells. The cells have elongated, tapering nuclei and are embedded in a loose, myxoid stroma. Some cells are arranged in small clusters, while others are more isolated. The overall appearance is characteristic of a soft tissue tumor, such as a myxoid sarcoma or a low-grade spindle cell sarcoma.
- Bland, spindle ~ stellate shaped
 - Thin walled blood vessels
 - Loose myxoid stroma
 - +ve : vimentin, CD34,
 - -ve : ER, PR, desmin
 - Variable : S-100, SMA, Factor XIIIa

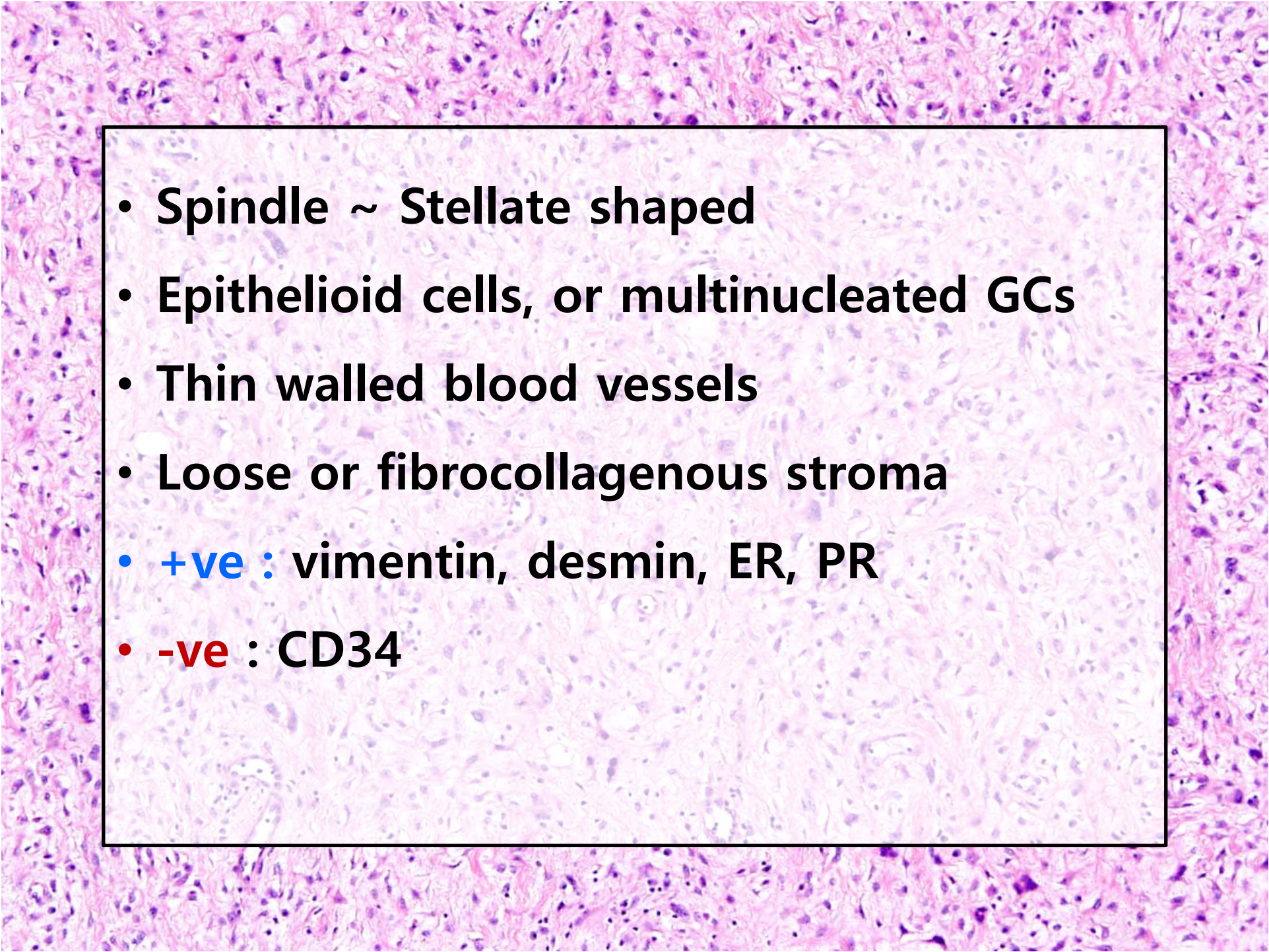


Angiomyofibroblastoma

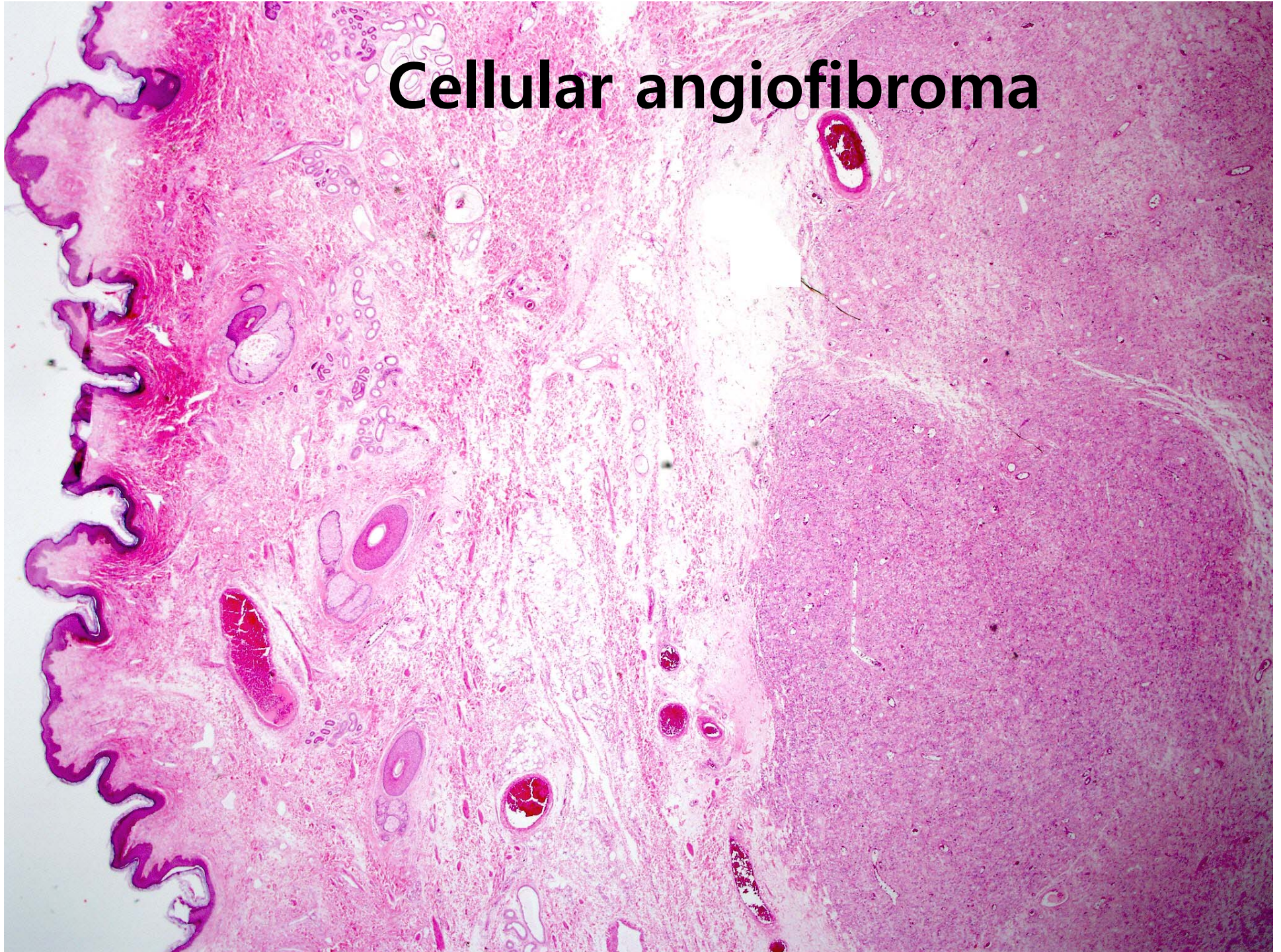
- Benign nonrecurrent tumor in the vulvovaginal region.
- Rarely merged with high grade sarcoma.

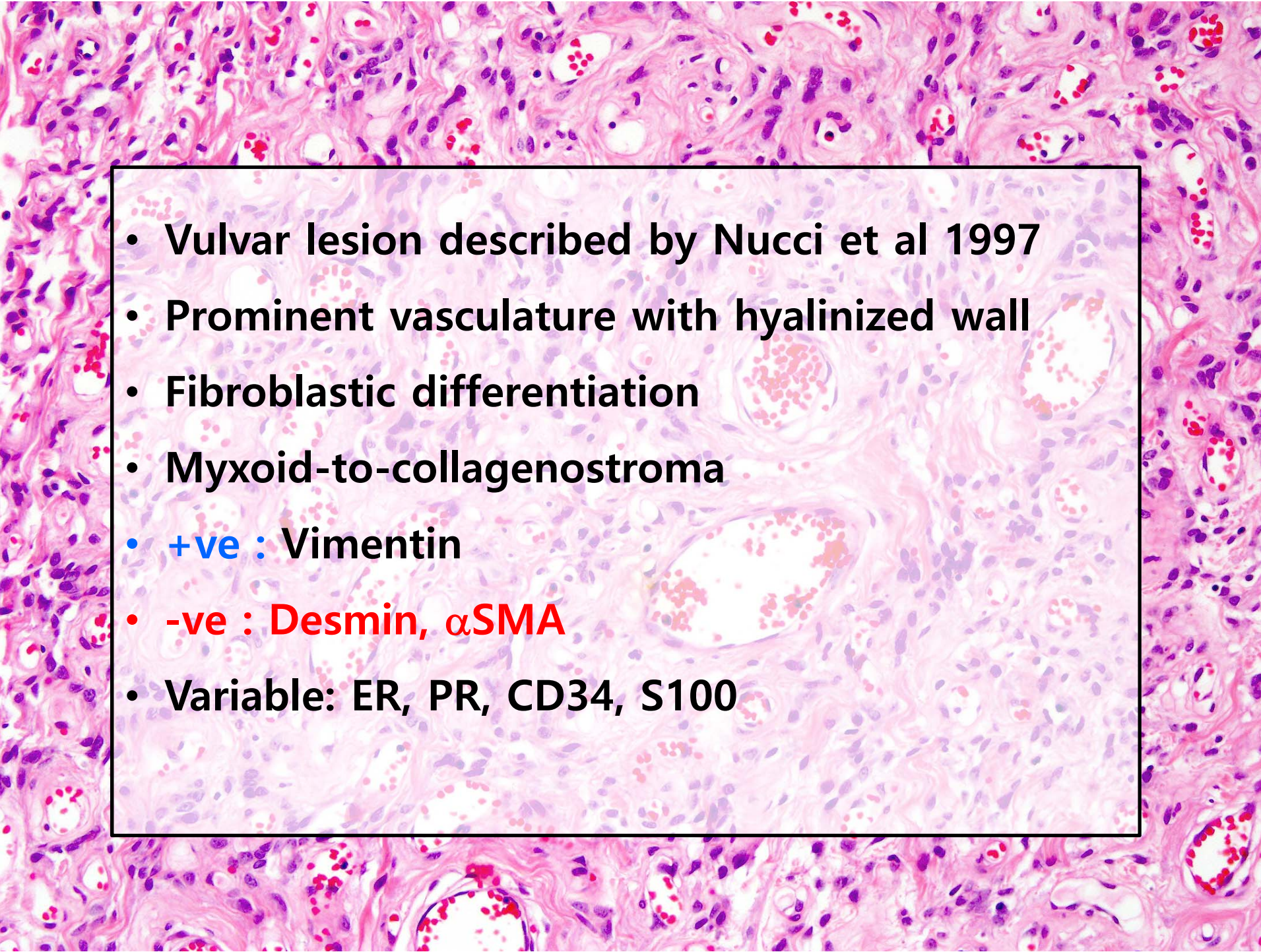




- 
- **Spindle ~ Stellate shaped**
 - **Epithelioid cells, or multinucleated GCs**
 - **Thin walled blood vessels**
 - **Loose or fibrocollagenous stroma**
 - **+ve : vimentin, desmin, ER, PR**
 - **-ve : CD34**

Cellular angiofibroma



- 
- Vulvar lesion described by Nucci et al 1997
 - Prominent vasculature with hyalinized wall
 - Fibroblastic differentiation
 - Myxoid-to-collagenostroma
 - +ve : Vimentin
 - -ve : Desmin, α SMA
 - Variable: ER, PR, CD34, S100

Superficial (cervicovaginal) myofibroblastoma

- Cervix, vagina, and vulva
- Nodular polypoid painless mass
- Perimenopausal or postmenopausal women
- Overlapping features with FSP and AMFB
- Moderately cellular, finely collagenous
- Bland-appearing spindle ~stellate shaped cells below the epithelium.
- Desmin (75-100% +), ER/PR (80-100%), CD34 (50-85%,+), SMA (1-5% +)

Cellular angiofibroma: another mesenchymal tumour with 13q14 involvement, suggesting link with spindle cell lipoma and (extra)-mammary myofibroblastoma

Maggiani F. et al. 2007; 5: 410-2

Genetic overlap among cellular angiofibroma, spindle cell lipoma, and (extra) mammary myofibroblastoma

Monoallelic deletion FOX1A1/13q14 loci

Arise from a common stromal precursor cell, which undergoes (myo)fibroblastic or adipocytic differentiation

Differential Diagnosis

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4. Angiomyofibroblastoma
5. Superficial angiomyxoma
6. Aggressive angiomyxoma
7. Botryoid embryonal rhabdomyosarcoma
9. Sarcomatoid squamous cell carcinoma

Cellular pseudosarcomatous fibroepithelial stromal polyp

Nucci MR et al. Am J Surg Pathol 2000. 24: 231-40

- Synonym: Mesodermal stromal polyp
Pseudosarcoma botryoides
- Of 65 usual fibroepithelial stromal polyps in the vulvovaginal region :
marked hypercellularity (n=33), marked cellular pleomorphism (n=21), frequent mitoses >10/10 HPFs (n=12), atypical mitoses (n=14)

Cellular (pseudosarcomatous) fibroepithelial stromal polyp

Clinical Findings

- Age : 16-75 yrs (median 32 yrs)
- Premenopausal : 64% (n=21)
- Site : vagina (18), cervix (7), vulva (8)
- Pregnancy (14/33), Tamoxifen (3/33), oral progesterone (1/33)
- Follow up :
NED (30/33),
Local non-destructive recurrence (3/33)

Evidences Suggesting Hormone-Related lesion

- Frequent during pregnancy.
- Associated with HRT in postmenopausal.
- Extremely uncommon before the menarche.
- Spontaneous regression following delivery.
- ER (+) and PR (+) in stromal cells.
- Intensity of PR(+) correlated with Ki67 proliferating index.

Diagnosis

**Cellular (pseudosarcomatous)
fibroepithelial stromal polyp of the
vaginal wall**

Follow-up result

No evidence of recurrence for 9 years.

Differential Points

Layers	Superficial	SAM, SMF, AMFB, FSP, CAF
	Deep	AAM
Margin	Well-demarcated	SAM, SMF, AMFB, FSP, CAF
	Infiltrative	AAM
Stroma	Myxoid/hypocellular	AAM, SAM
	Fibro-collagenous /celluar	CAF, FSP, SMF
	Mixed	AMFB
Differentiation	Fibroblastic Myofibroblastic	
Blood vessels	Vascularity, size, hyalinization ..etc	
Location	Cervicovaginal vs. Vulva	
IHC patterns	CD34, vimentin, S100, Desmin	

Take Home Message

- Many tumors have hybrid or overlapping histologic features between the classic types.
- Immunophenotypes are widely variable in a same type of mesenchymal lesions in the lower female genital tract.
- Thus, clinical and histological characteristics are most important in the diagnosis.

A close-up photograph of a person's hand holding a dark green pencil over a light blue card. The card is placed on a brown cardboard envelope, which is resting on a grey surface. The card has the words "Thank You" written in a black cursive font. A faint circular stamp is visible above the text. The pencil has a barcode and the number "4 005401 193081" printed on it.

Thank You