Surgical Pathological Correlation

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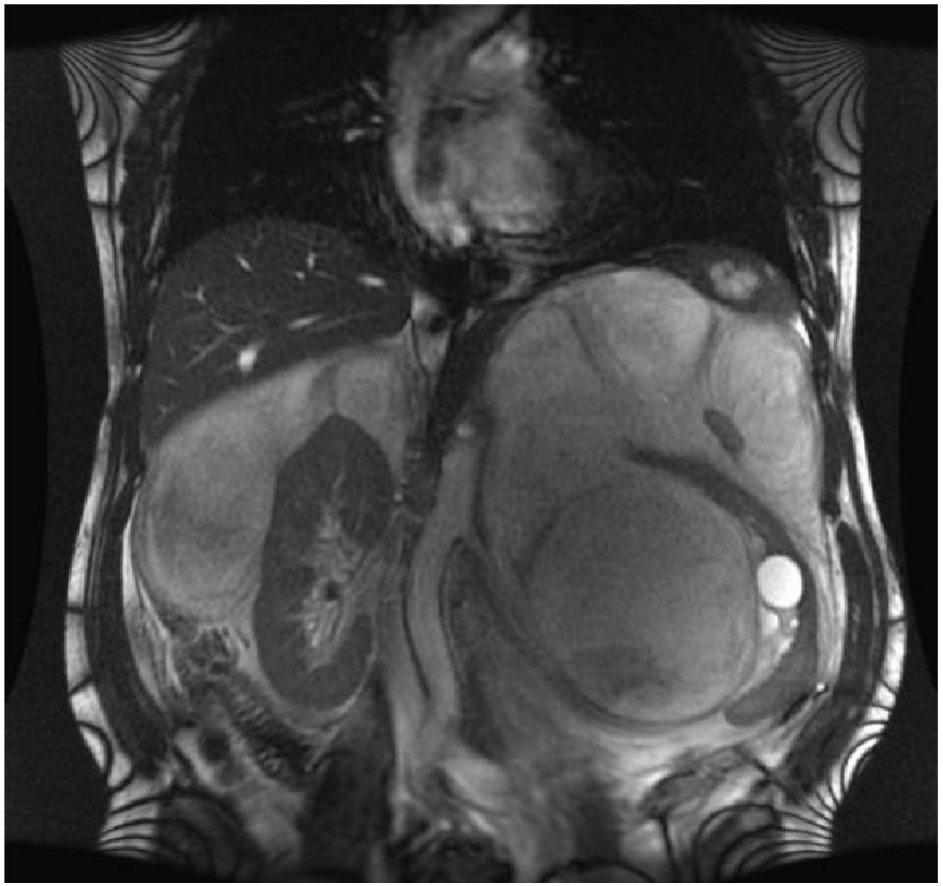
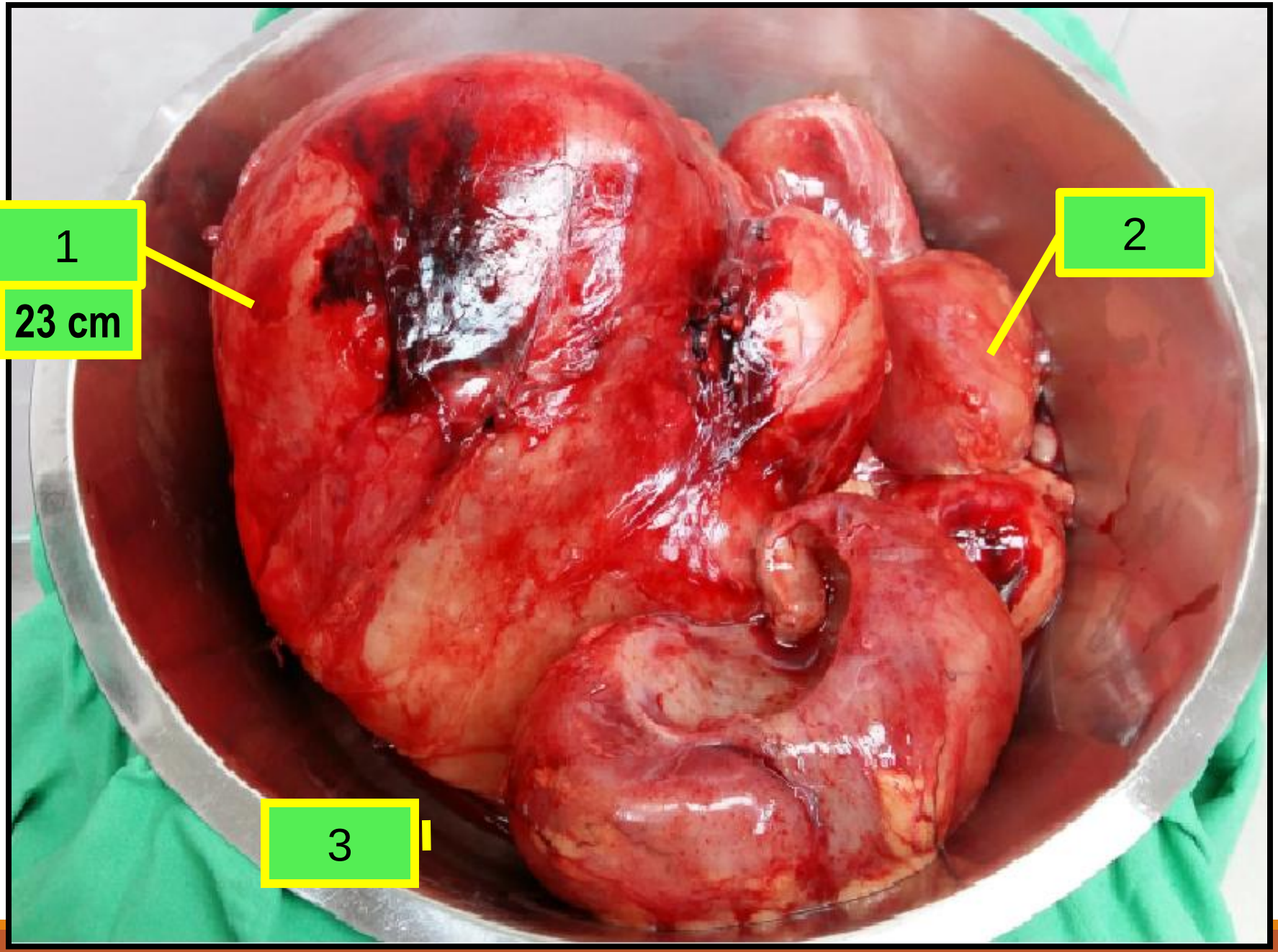
Clinical Manifestation

- Age: 51 years-old
- Sex: male
- History: SLE with medication
- MRI Image finding:
  Bilateral retroperitoneal tumors, with left kidney invasion, and suspect spleen invasion

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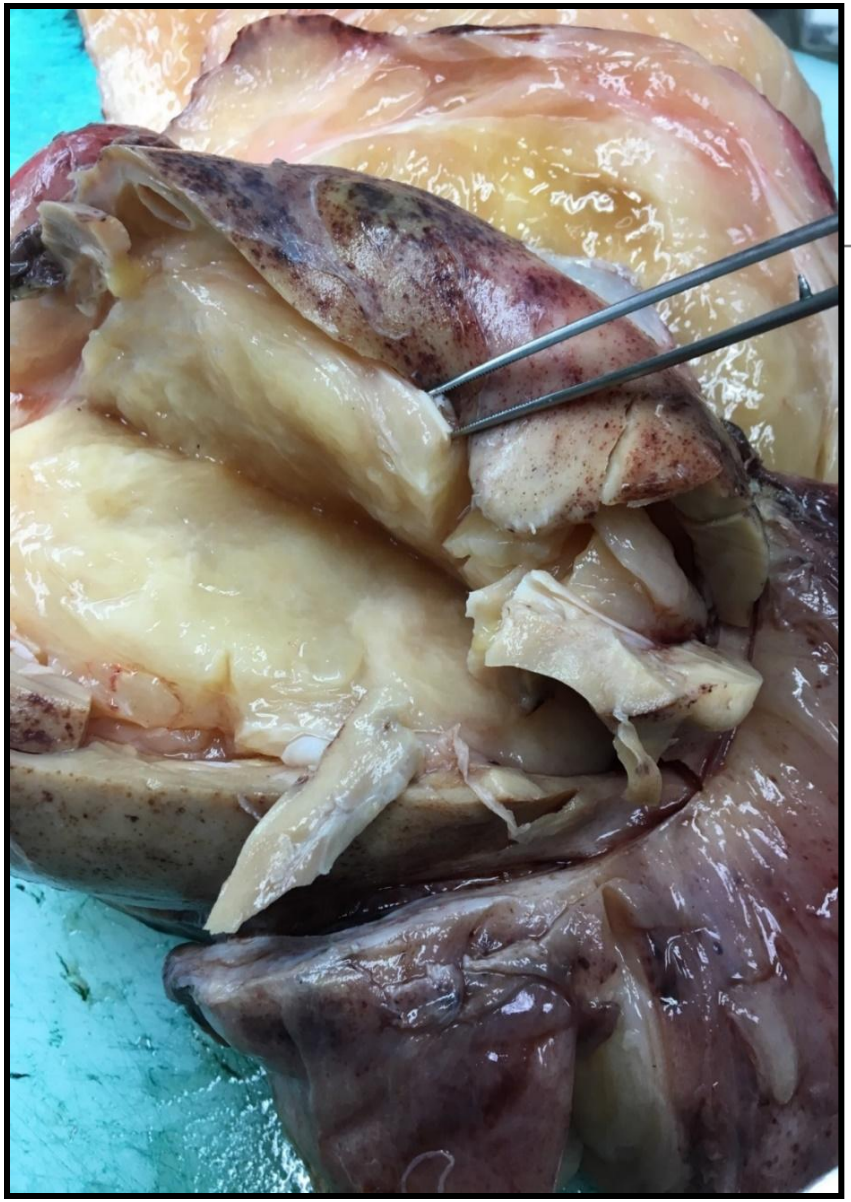
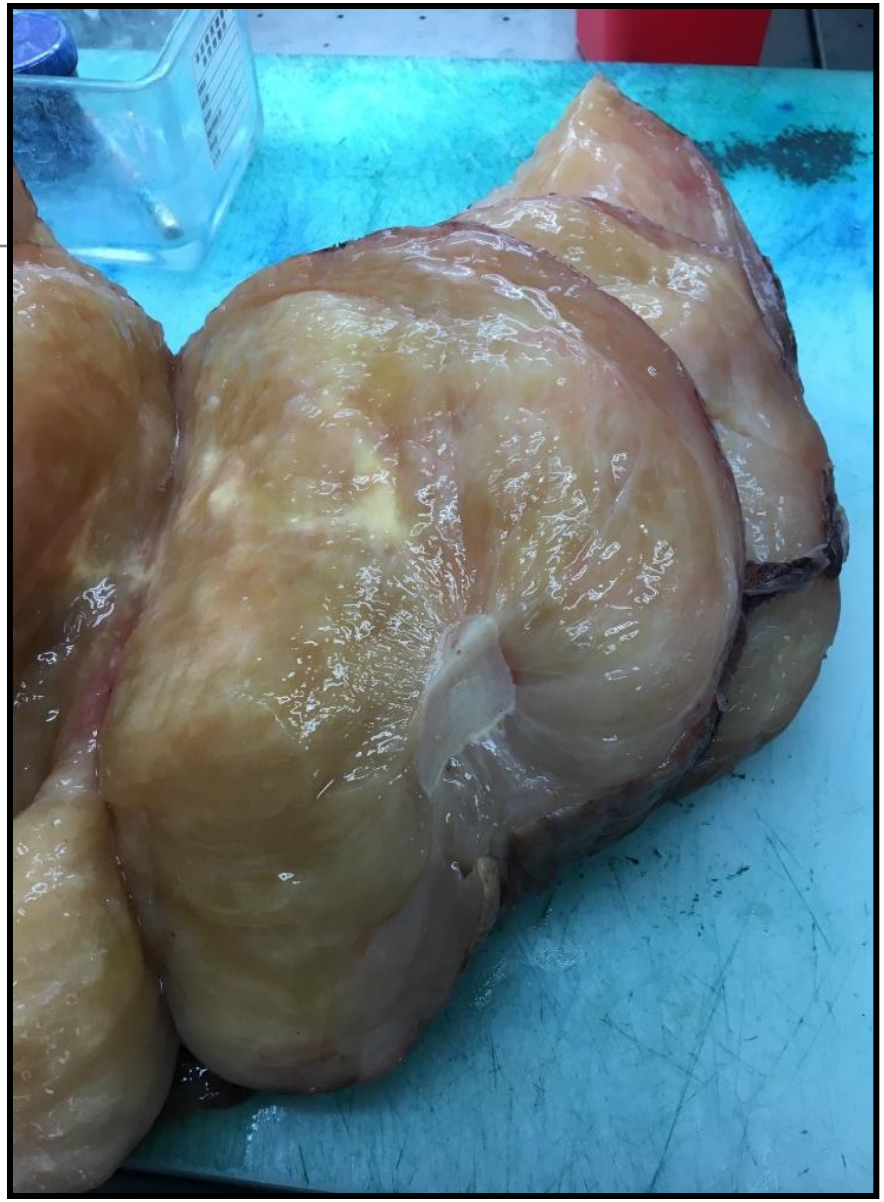
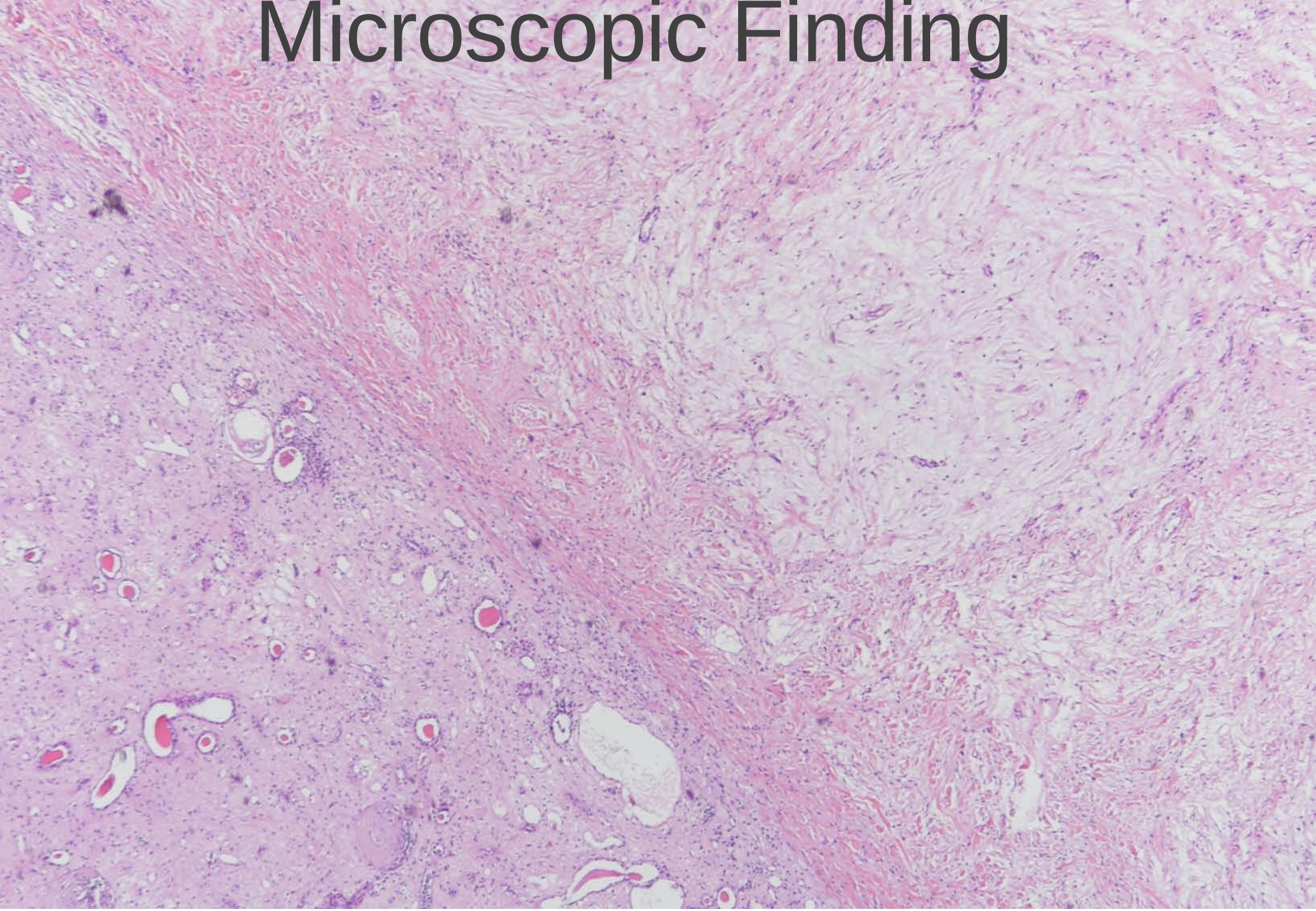
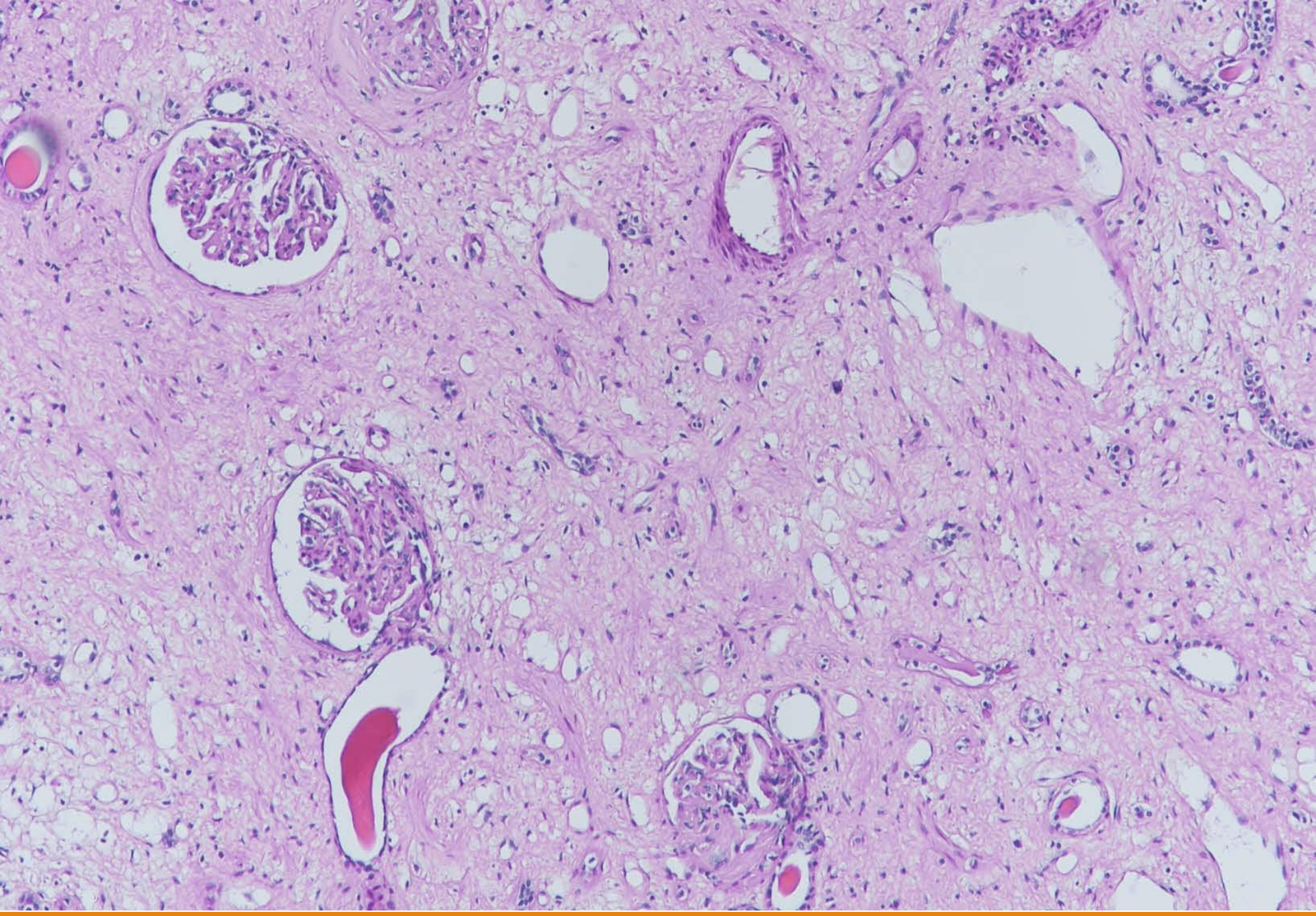
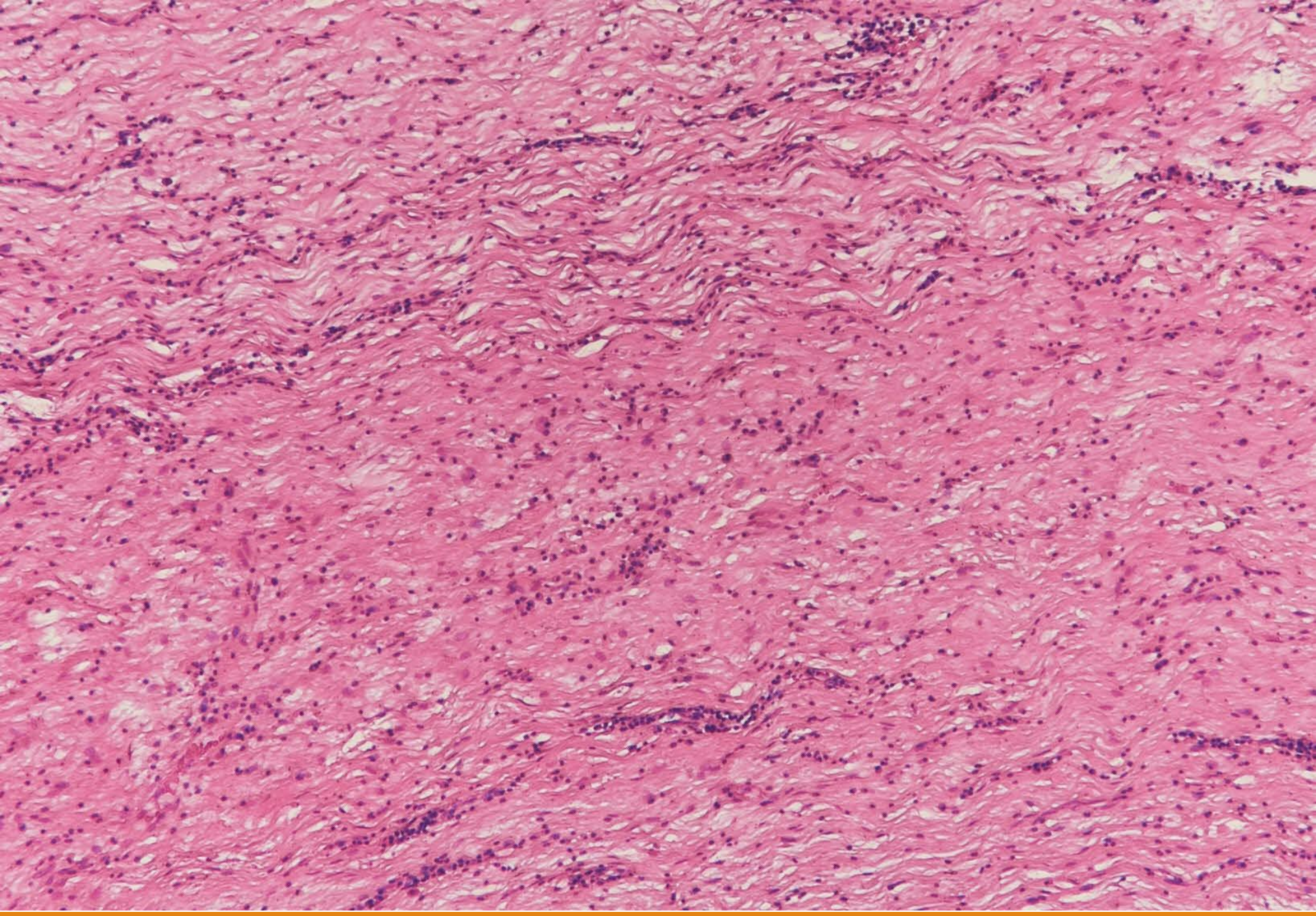


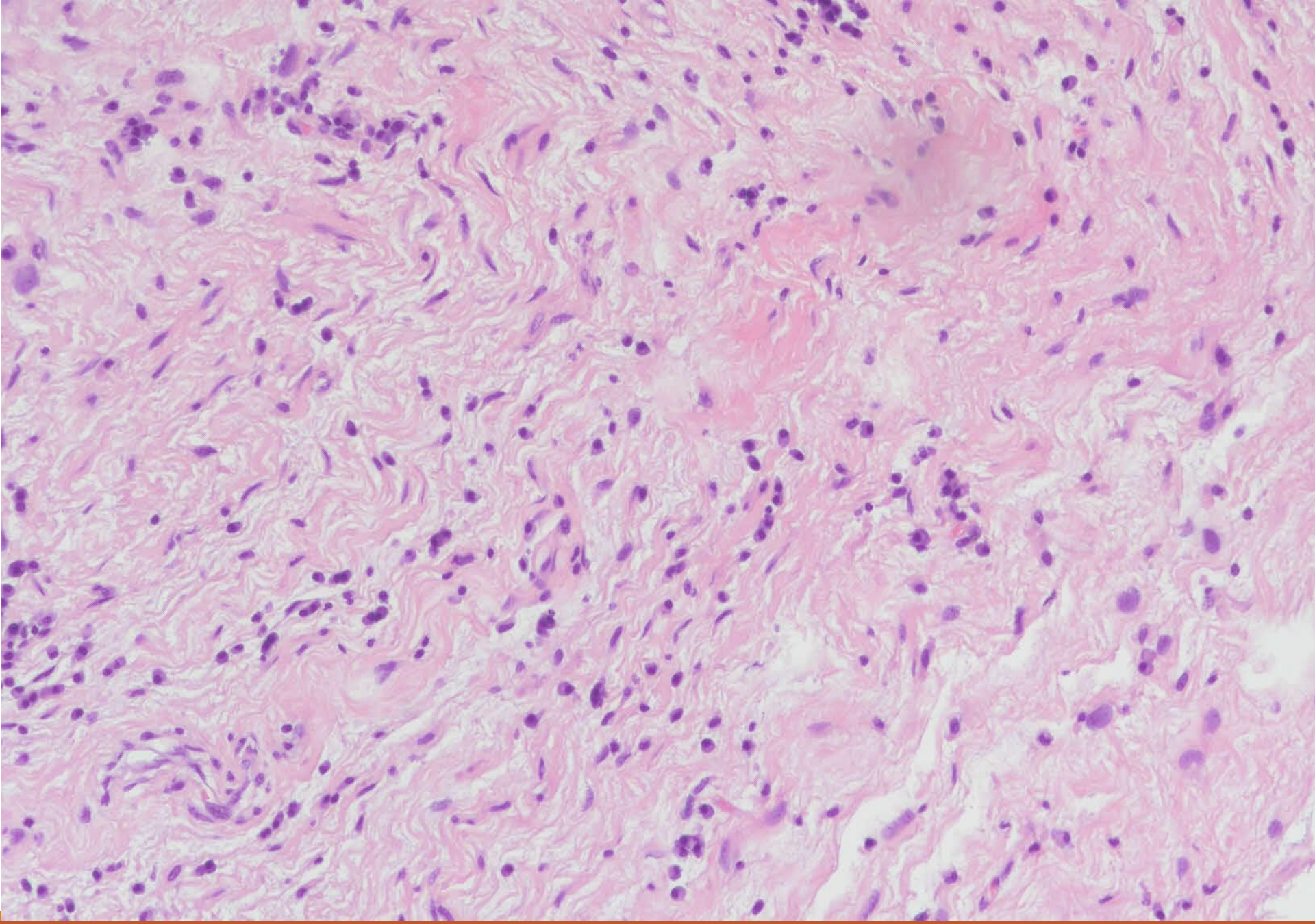


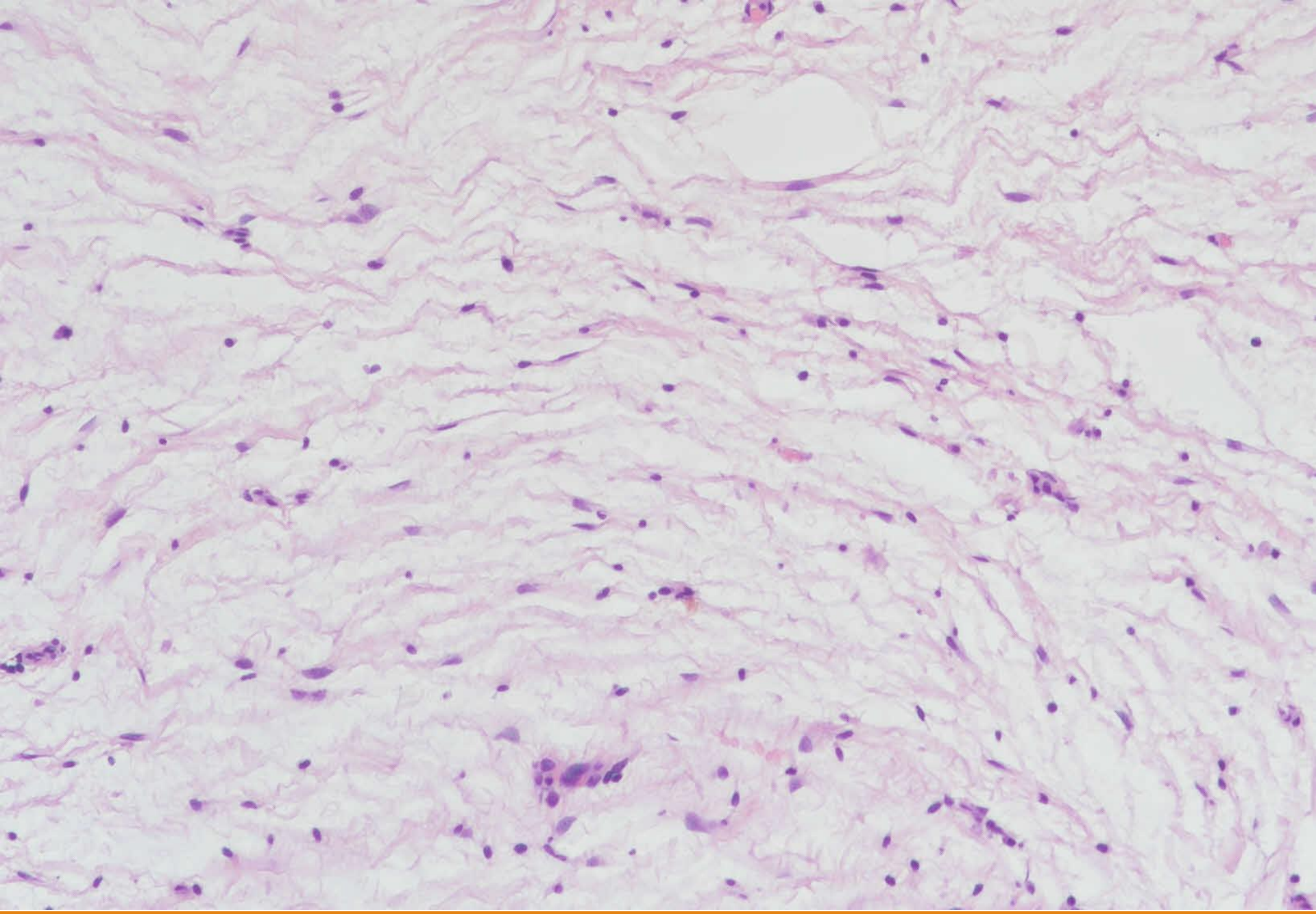
Microscopic Finding











Summary of Microscopic Finding

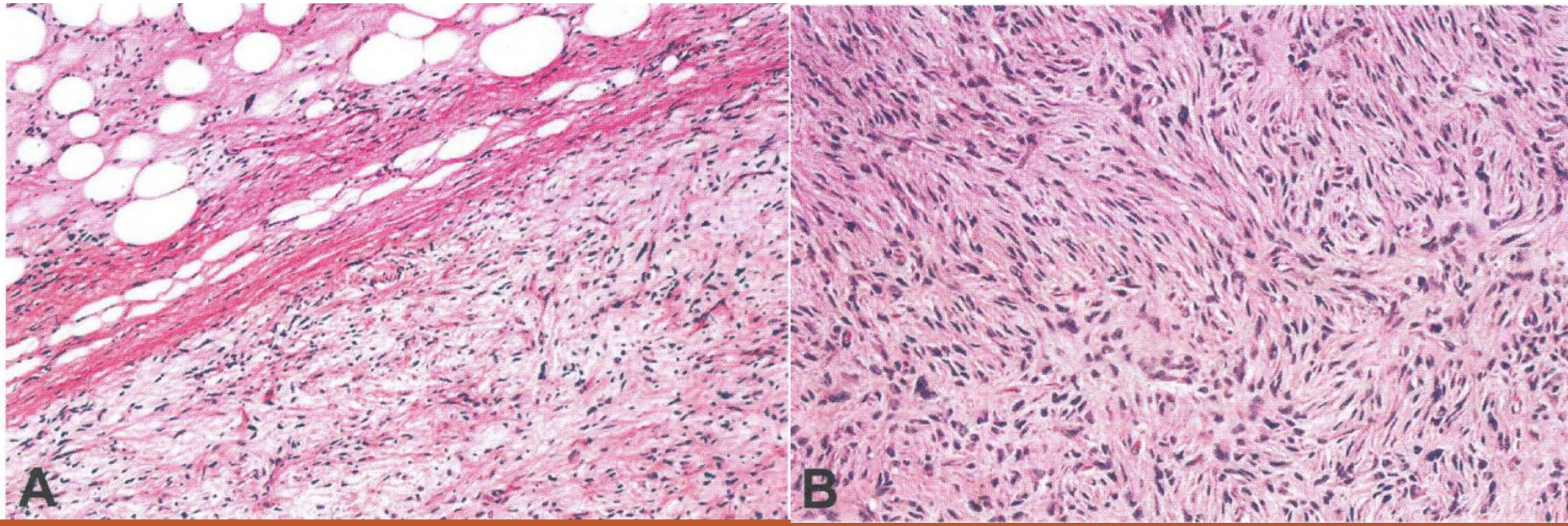
- Hypocellular growth
- Myxoid and collagenous background
- In vague fascicles bundle or lamellar arrangement
- Thin and elongated cells
- No mitotic figure, no cellular atypia, no necrosis
- Foci of perivascular inflammatory cells infiltrates

Differential Diagnosis

- Dedifferentiated liposarcoma
- Low grade fibromyxoid sarcoma
- Solitary fibrous tumor
- Inflammatory myofibroblastic tumor
- Soft tissue perineurioma

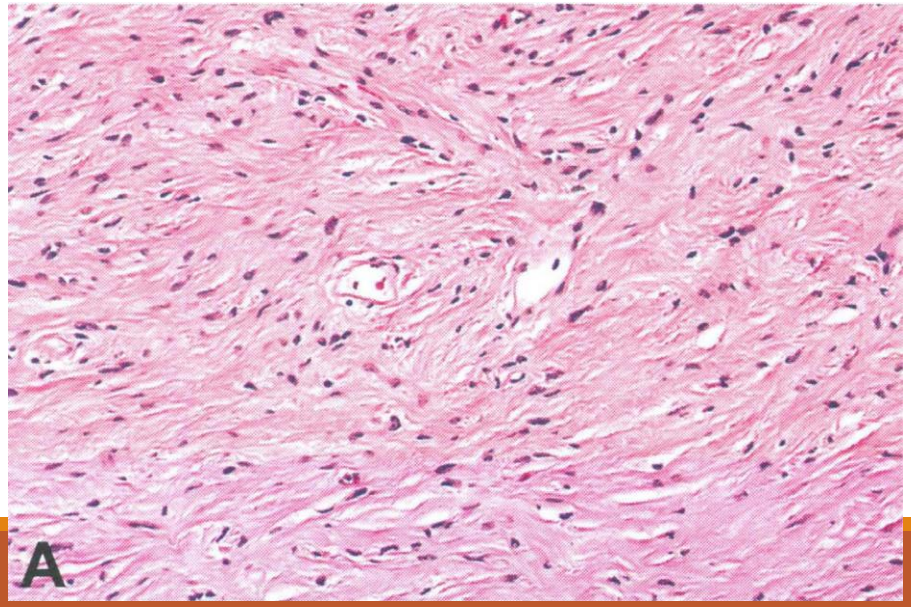
Dedifferentiated Liposarcoma

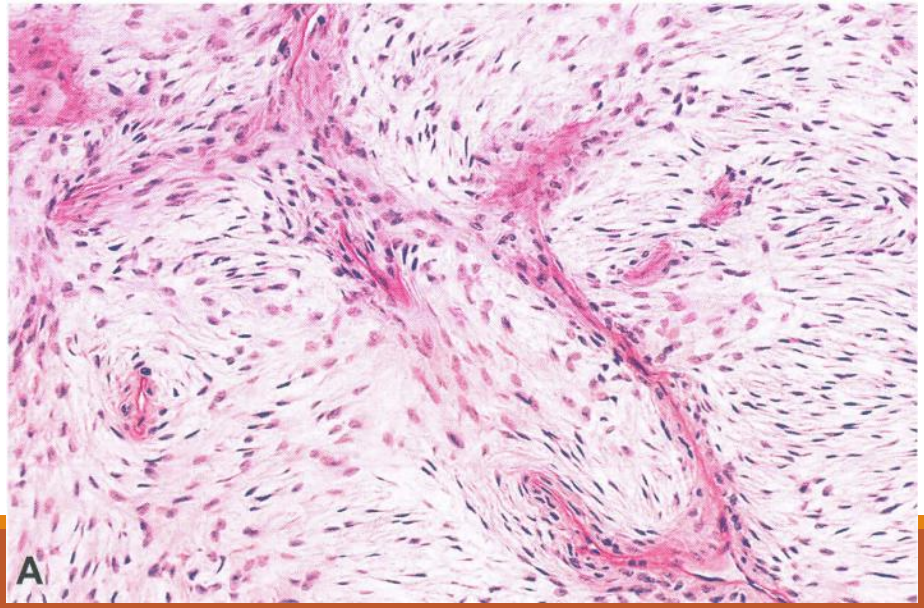
- Dedifferentiated from atypical lipomatous tumor/well-differentiated liposarcoma
- Retroperitoneum : somatic = 5 : 1
- May be no lipogenic content
- Immunostains: MDM2(+) and/or CDK4(+)



Low Grade Fibromyxoid Sarcoma

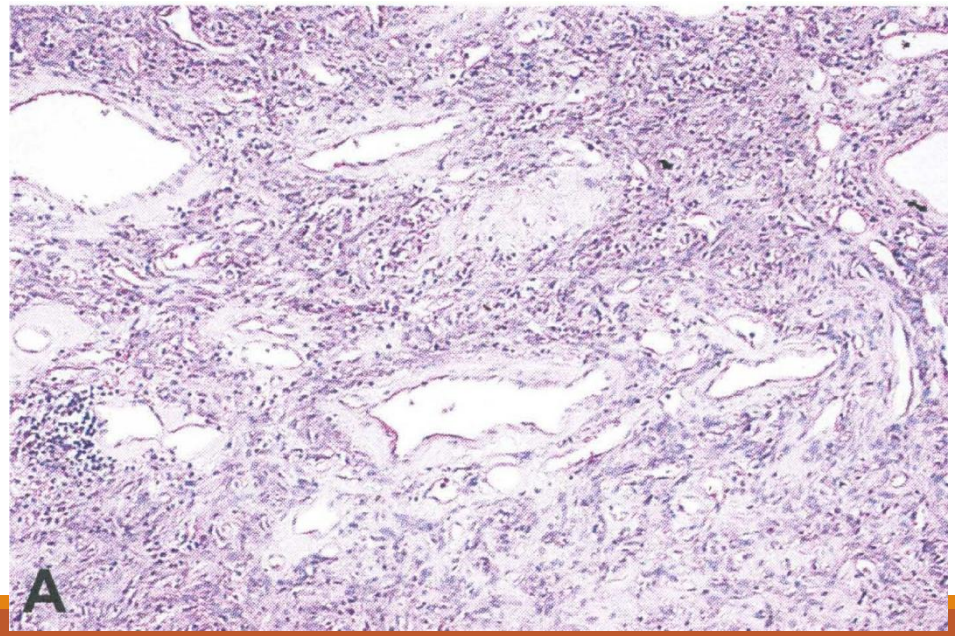
- Typically at extremities or trunk
- Admixed heavily collagenized hypocellular areas and cellular myxoid nodule
- Short fascicular and whorling growth with arcades of small vessel
- Immunostains: MUC-4(+). Genetic: FUS-CREB3L2 or FUS-CREB3L1

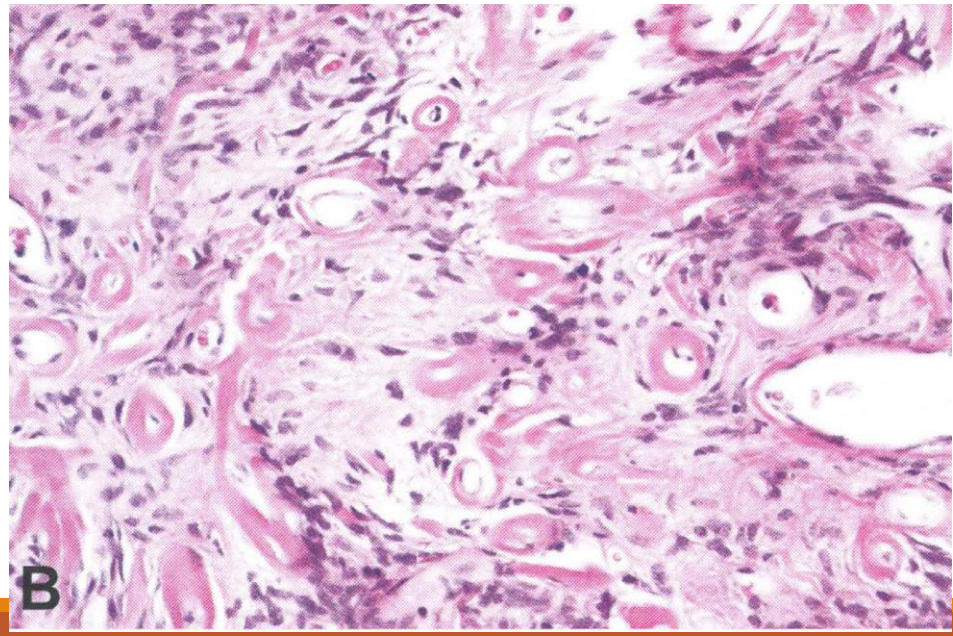




Solitary Fibrous Tumor

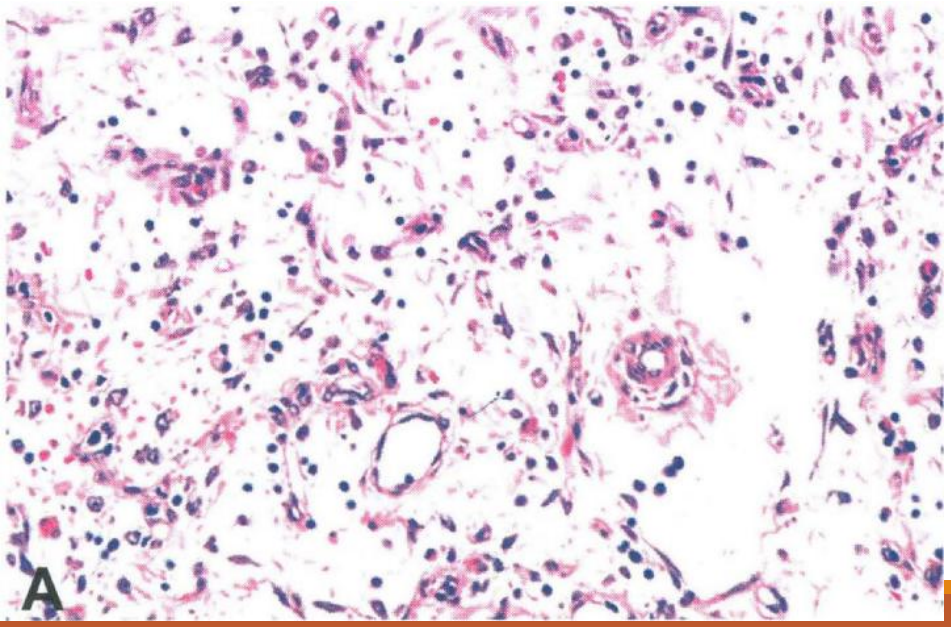
- May present at any location. Many on serosal side.
- “Patternless” architecture separated by thick collagen and staghorn-like vessel
- Immunostains: CD34(+), STAT-6(+)

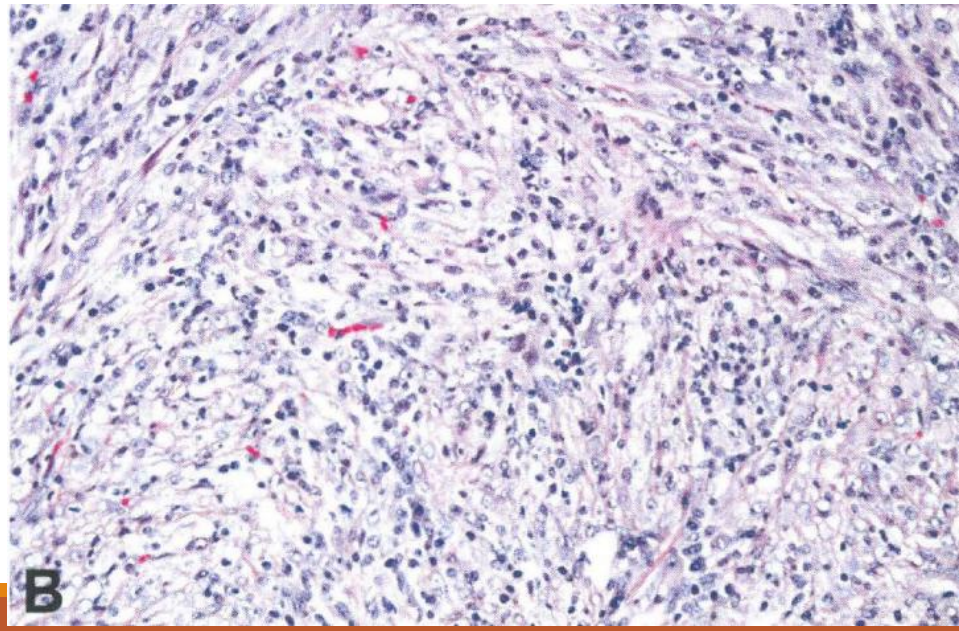




Inflammatory Myofibroblastic Tumor

- Primarily children and young adult
- Throughout whole body, predominant abdominal cavity
- Spindle cells in myxoid background with abundant inflammatory cells infiltrates
- Immunostains: ALK(+)



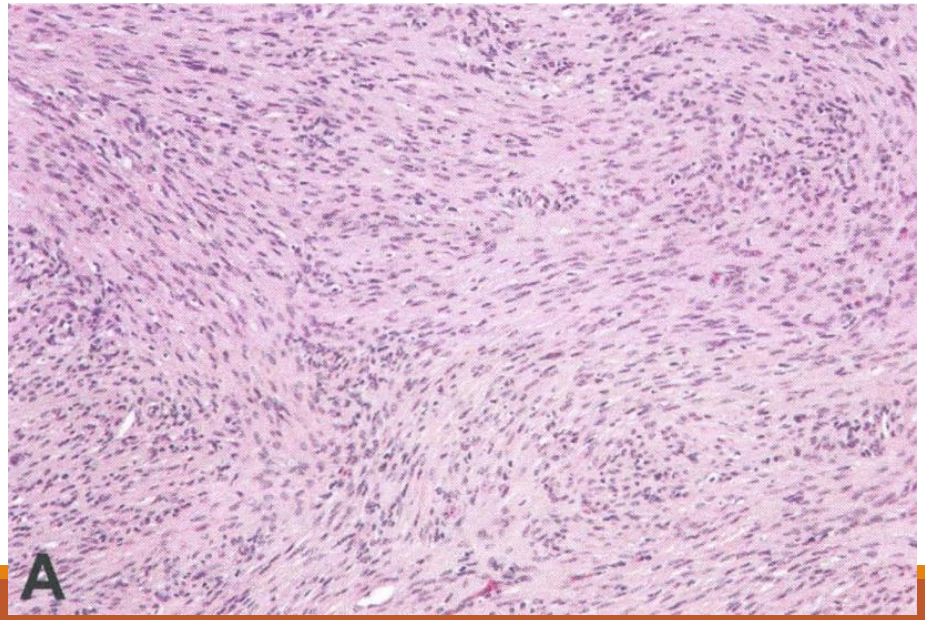


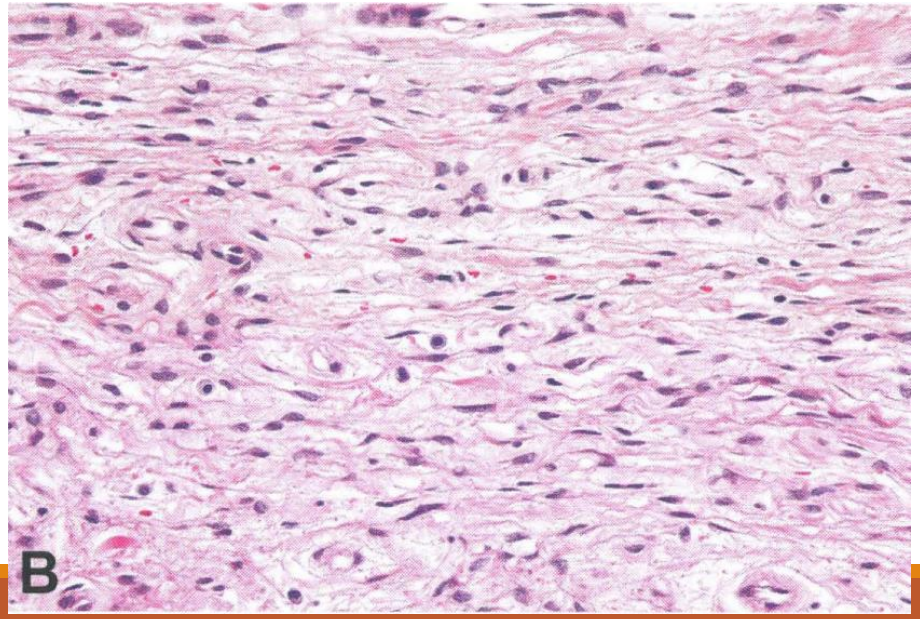
Differential Diagnosis

- Dedifferentiated liposarcoma → MDM2(-), CDK4(-), MDM2 FISH(-)
- Low grade fibromyxoid sarcoma → MUC-4(-)
- Solitary fibrous tumor → although CD34(+), STAT6(-)
- Inflammatory myofibroblastic tumor → ALK(-), ROS1(-)
- Soft tissue perineurioma

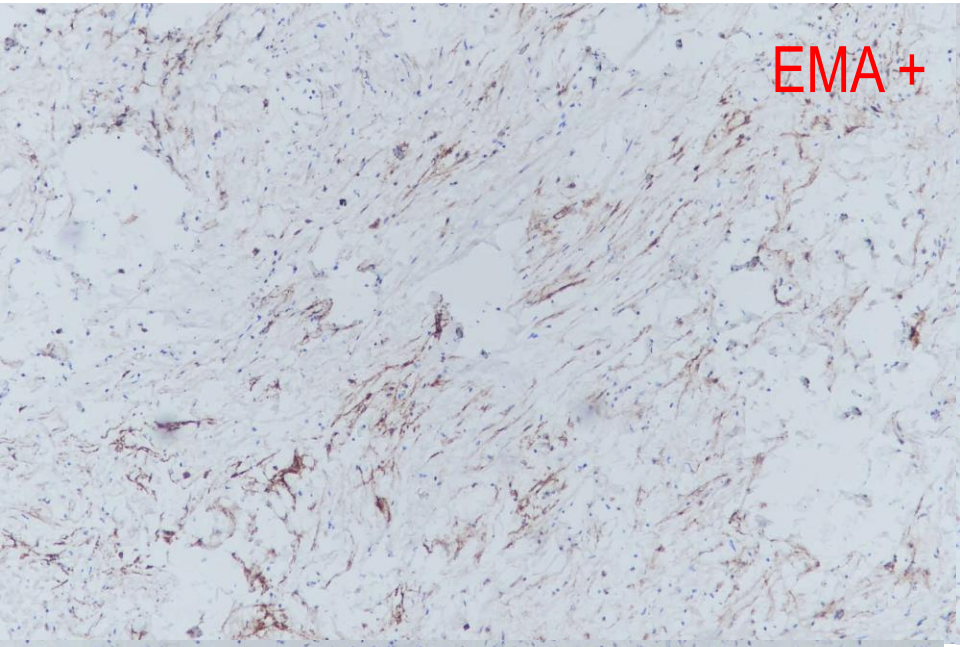
Soft Tissue Perineurioma

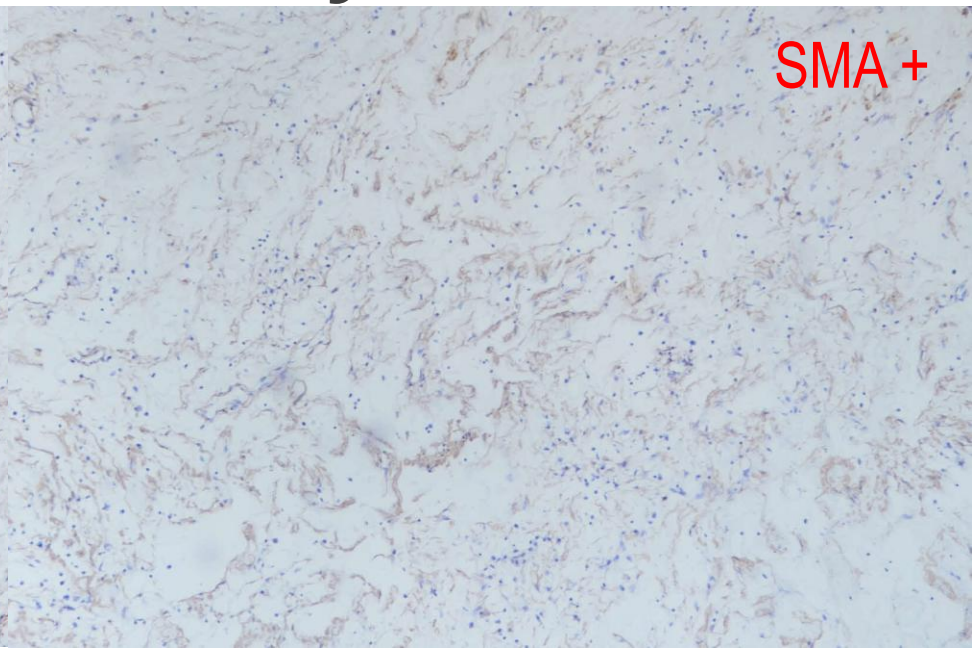
- More common in limbs and trunk, retroperitoneum rarely
- In storiform, fascicles, lamellar arrangement
- Slender to plump cells with wavy nuclei
- Could be collagenous, or myxoid, or mixed background
- Immunostains: EMA(+), GLUT-1(+), Claudin-1(+), S-100(-)

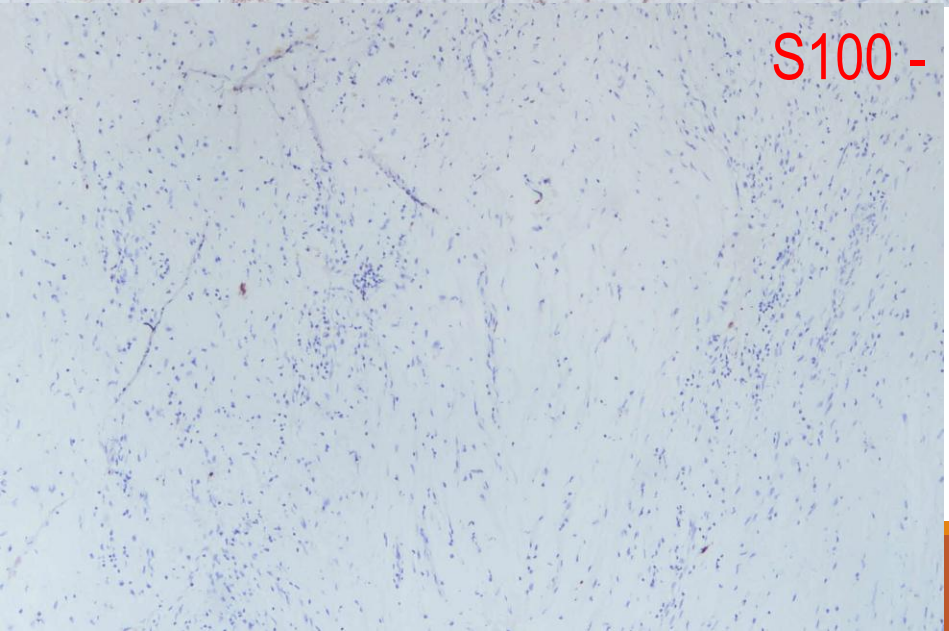




Immunohistochemistry







Immunostains:CD34(diffuse positive)

(外院) GLUT-1(negative)

Claudin-1(focal positive)

Pathologic Consultation

- The section show hypocellular growth of bland elongated cells in vague parallel bundles with myxocollagenous stroma abutting the renal and perihilar fat tissue. No significant nuclear atypia or mitotic figure is seen.

- The results of IHCs in from your hospital are as follows: MDM2 (-) and CDK4 (-), not supportive of low-grade dedifferentiated liposarcoma, MUC4 (-, aruging against LGFMS), EMA (+), S100 (-), ALK (-, arguing against IMT), STAT6 (-, arguing against SFT).

Pathologic Consultation

- The IHCs of my lab reveals as folllows: CD34 (diffuse +), EMA(+), GLUT1 (-), claudin-1(focal +), ROS1 (arguing against IMT).

- There is no amplification of MDM2 by FISH, arguing against low-grade dedifferentiated liposarcoma.

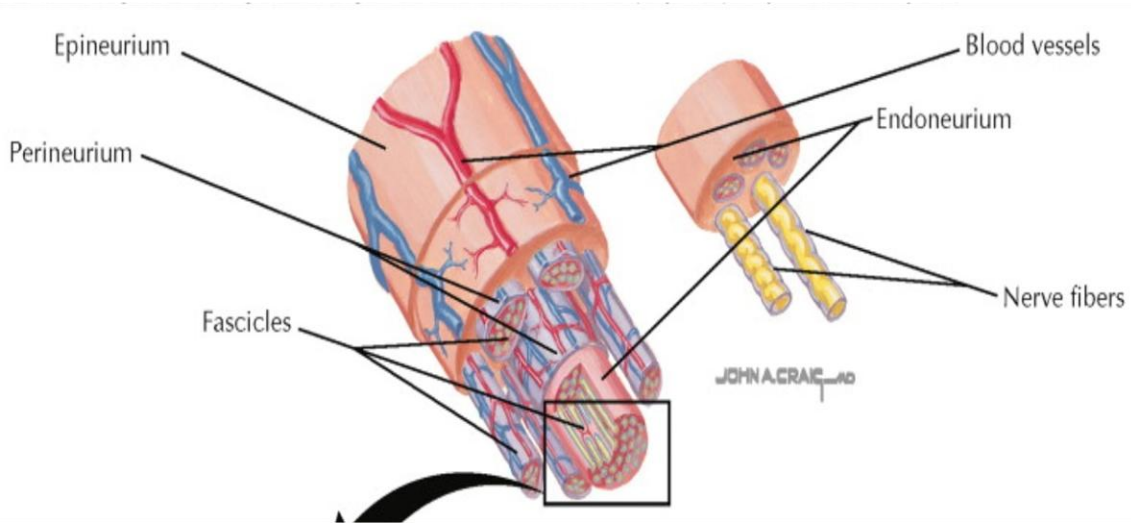
- According to above histopathological findings and IHCs results (CD34, EMA and claudin-1), perineurioma is favored. Follow up is recommended.

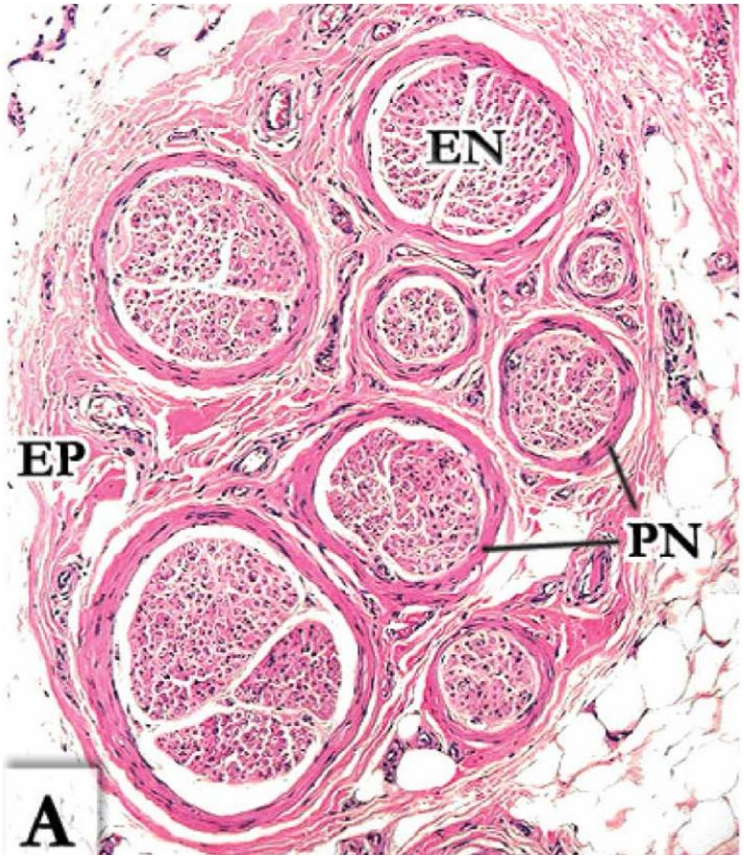
Diagnosis

- Soft tissue perineurioma, involved by retroperitoneum, left kidney and spleen hilum, status post retroperitoneal tumor removal, left nephrectomy, splenectomy, enterolysis and right ureteral double J placement
- Systemic lupus erythematosus with medical control

Discussion

Perineurium





- **Schwann cells: S-100(+)**
- **Perineurial cells: Claudin-1(+), GLUT-1(+), S-100(-)**

Netter essential histology

The Normal and Neoplastic Perineurium: A Review
Adv Anat Pathol2008;15:147–164

Perineurioma

- Proposed in 1978 by ultrastructure, by *Lazarus SS, Trombetta LD.*
- More common in limbs and trunk, rarely in head and neck, retroperitoneum
- Intraneural perineurioma: “Pseudo-onion”. Concentric proliferation of perineurial cells leading to sausage-like swelling of nerve.
- Extraneural perineurioma : “Soft tissue perineurioma ”. Not associated with nerves.
- Sclerosing perineurioma: Bland epithelioid cells embedded in densely collagen
- Reticular perineurioma: Spindled cells arranged in a reticular pattern

Colonic perineurioma (benign fibroblastic polyp): case report and review of the literature

Ref.	No of cases	Mean age in years (Range)	F:M	Distal to transverse colon	Mean size (Range) in mm	Endoscopic description	No of cases with polyps elsewhere in colon
[1]	14	62 (37-84)	8:6	13/14	5.1 (2 – 15)	'polyps'	10/14
[10, 11]	5	65 (52-77)	3:2	4/5	3 (2 – 4)	NS	4/4
[8]	9	51 (35-72)	7:2	7/8	6.8 (2-30)	Small sessile polyps	5/9
[7]	4	66 (58-71)	2:2	3/4	9.8 (6-15)	Two sessile polyps depicted	3/4
[2, 5, 12, 15]	60	60 (36-84)	30:30	52/60	3.4 (1-8)	Flat/sessile to round/ pedunculated	21/60
[9]	4	59 (47-80)	3:1	4/4	3.8 (3-5)	Sessile polyps	3/4
[13]	1	54	0:1	NS	5	NS	NS
[14]	1	50	0:1	1/1	6	Polypoid lesion	0/1
[3]	29	64 (43-84)	23:6	26/28	2.7 (3-9)	NS	19/29
[4]	20	58 (44-87)	9:11	17/20	5.1 (3-15)	NS	15/20
[16]	9	62 (45-84)	5:4	4/9	4 (NS)	NS	6/9
[17]	1	51	1:0	1/1	15	'Submucosal lesion'	0/1
Current case	1	42	0:1	0/1	5	Pedunculated polyp	0/1
Total	158	60 (35-87)	F:M = 1.3	132/155 85%	4.1 (1 – 30)		86/156 55%

Colonic perineurioma (benign fibroblastic polyp): case report and review of theLiterature van Wyket al. Diagnostic Pathology (2018) 13:16

Case Report:
Bilateral Renal Myxoid Perineuriomas

- 25-year-old man with large bilateral renal masses
- Bilateral perineurioma are diagnosed
- Surgical excision
- No recurrence at 2 years follow-ups

Bilateral Renal Myxoid Perineuriomas

Am J Med Sci. 2012 Mar;343(3):265-6

Soft Tissue Perineurioma: Analysis from 81 Cases

- 81 cases of soft tissue perineuriomas between 1994 and 2003 from the authors' consult files

- Material and methods: Based on previously published histologic criteria:
  ➔ spindle cells with curved or wavy nuclei and thin elongated cytoplasmic processes arranged in lamellae, bundles, loose whorls, and storiform patterns

Soft Tissue Perineurioma: Clinicopathologic Analysis of 81 Cases Including Those With Atypical Histologic Features

Am J Surg Pathol 2005;29:845–858

Soft Tissue Perineurioma: Analysis from 81 Cases

- Soft tissue perineurioma is regarded as a benign diagnosis, so many patients did not return for follow-up. (41/81 returned OPD)
- Only two tumors recurred (5%).
  - One of primary excision margins is positive margin.
  - The other one is unknown at the primary excision.
- No tumor metastasized.

Soft Tissue Perineurioma: Clinicopathologic Analysis of 81 Cases Including Those With Atypical Histologic Features

Am J Surg Pathol 2005;29:845–858

Soft Tissue Perineurioma: Analysis from 81 Cases

- 14 of 81 (17%) soft tissue perineuriomas show **atypical features**:
  - 12 cases scattered pleomorphic cells, 1 show an abrupt transition to a markedly hypercellular area and mild cytologic atypia, 1 show diffuse infiltration of adjacent skeletal muscle
- Only one atypical perineurioma experienced recurrence.
  - The patient is well after excision again

Soft Tissue Perineurioma: Clinicopathologic Analysis of 81 Cases Including Those With Atypical Histologic Features

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Malignant peripheral nerve sheath tumor with perineural cell differentiation

- Described as **Malignant peripheral nerve sheath tumor(MPNST) with perineural cell differentiation,** or **Malignant perineurioma**
  - MPNST exhibit immunohistochemical characteristics of perineural cells, but not schwann cells features
- Microscopically, similar to benign perineuriomas as storiform, whorled architecture with marked fascicular areas, but present **atypical cells** and **mitoses** and **the presence of infiltrative margins**
- The prognosis of perineural MPNST appears to be more favorable than that of conventional MPNST.

1. The Normal and Neoplastic Perineurium: A Review
Adv Anat Pathol 2008;15:147–164

2. Perineurial malignant peripheral nerve sheath tumor (MPNST): a clinicopathologic, immunohistochemical, and ultrastructural study of seven cases
Am J Surg Pathol. 1998 Nov;22(11):1368-78

Reviewing Perineurioma

年度	病理號	位置	診斷
2015	04-34487	Colon	Perineurioma
2015	04-42041	Colon	Hybrid hyperplastic polyp and perineurioma
2016	05-44211	Colon	Hybrid hyperplastic polyp and perineurioma
2017	06-23184	Colon	Perineurioma
2016	05-01377	T8/9 spinal tumor	Perineurioma
2016	05-44922	Thumb	Perineurioma

Outpatient Follow Up

- Remove right double-J tube
- Crea:1.08
- Arrange abdominal CT three months later

Take Home Message

- Soft tissue perineurioma is an uncommon tumor, which is originated from perineurium
- It develops mostly in the extremities and trunk and rarely in the retroperitoneum, head and neck.
- Perineurioma has characteristic microscopic findings and immunostains
- Perineurioma is regarded as a benign entity with rare recurrence and no metastasis during follow-up examinations in several studies.

Thanks for your listening !