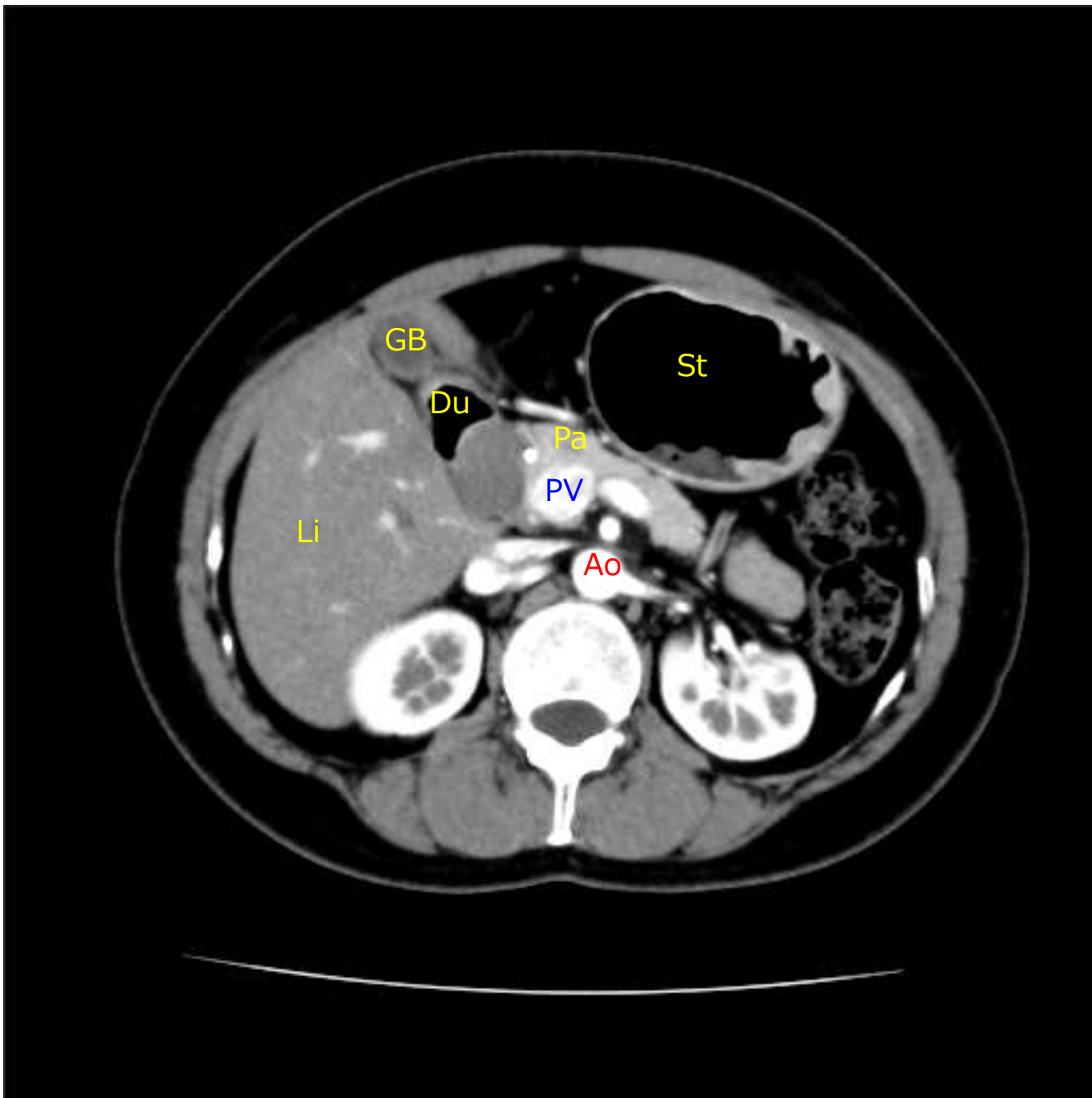
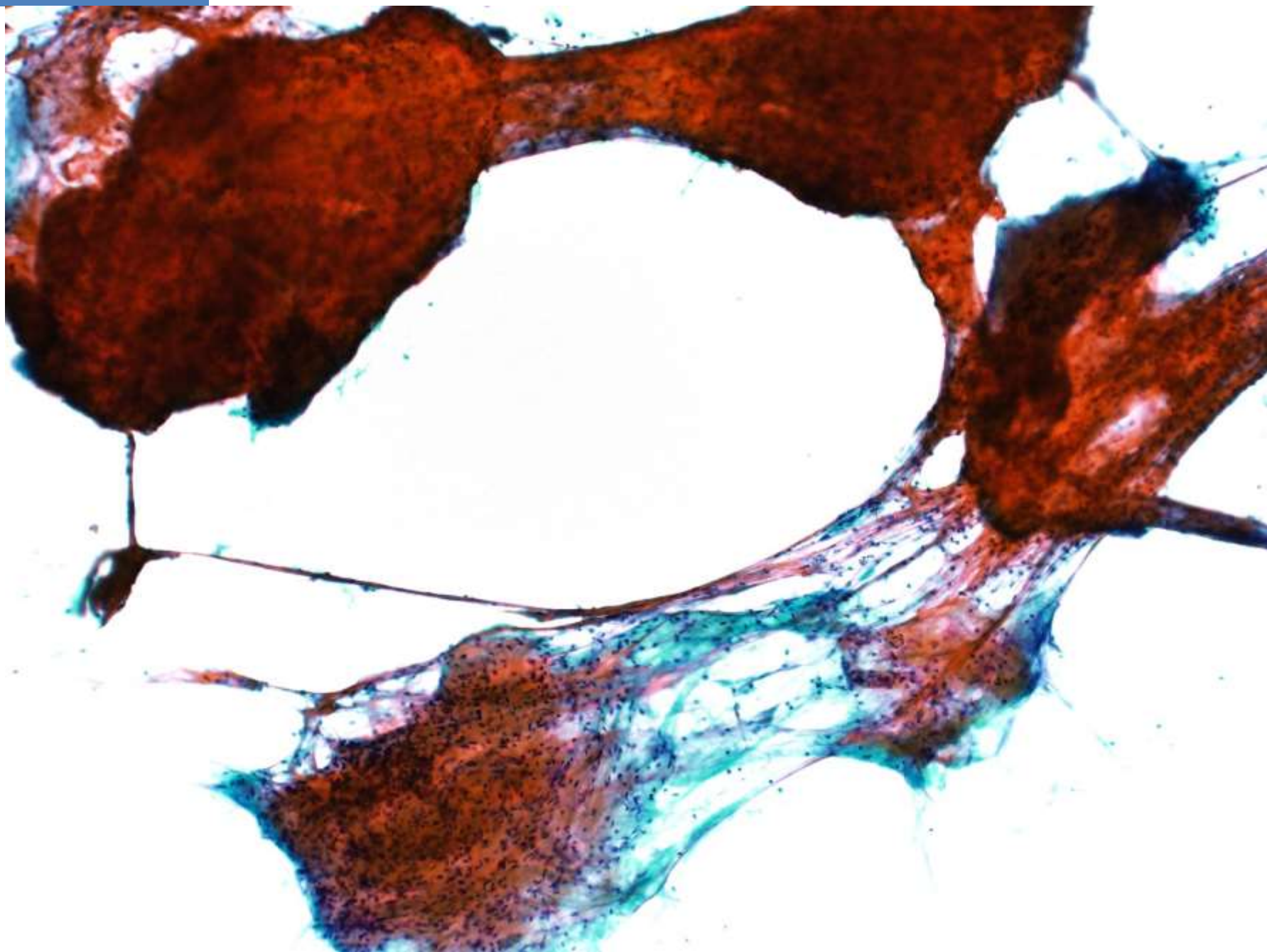


40代、女性。

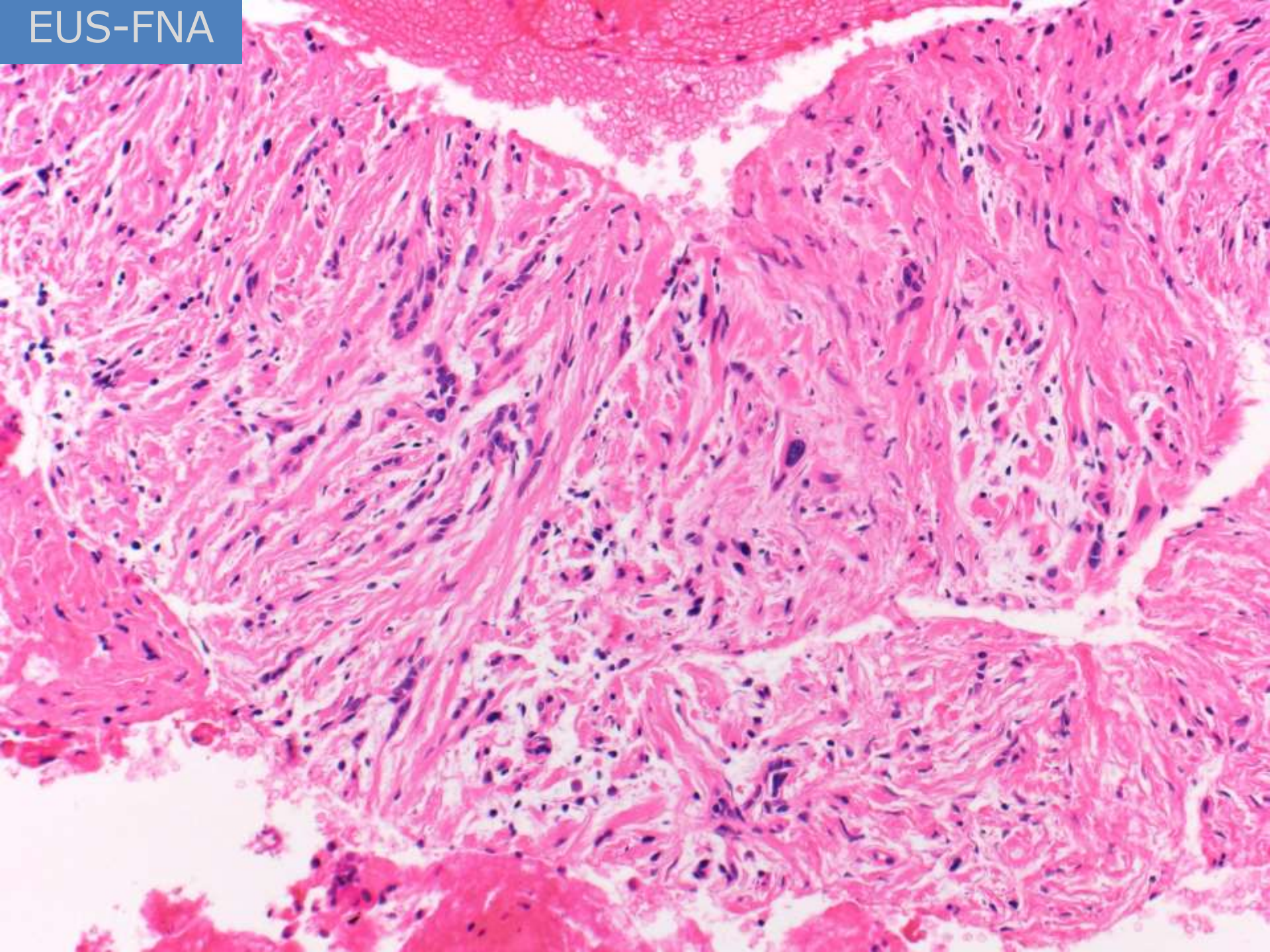
既往歴：子宮筋腫にて子宮全摘術

現病歴：検診の上部消化管造影検査で異常を指摘された。  
内視鏡を施行したところ、胃には著変なかったが、  
十二指腸にSMTを指摘された。



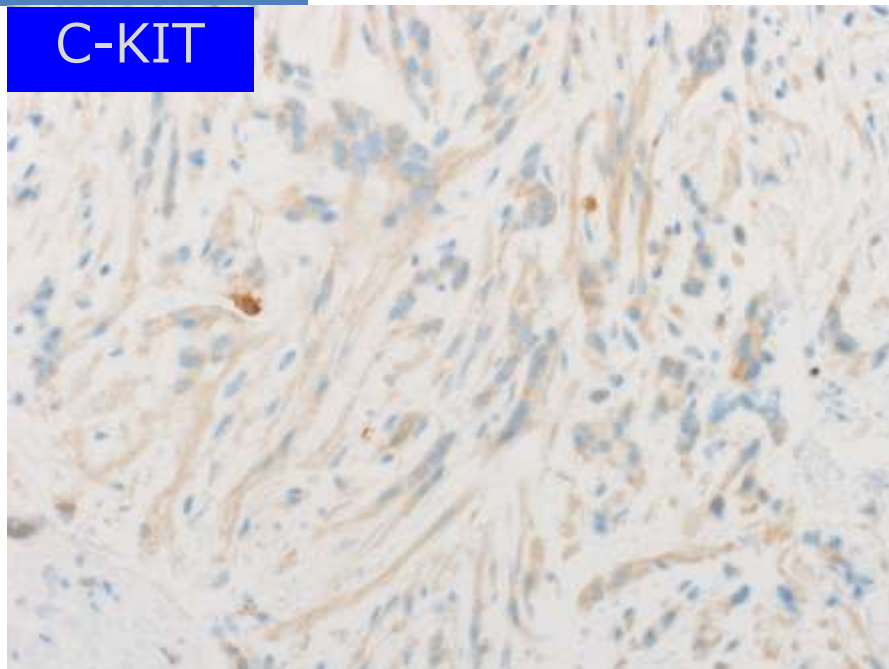


EUS-FNA

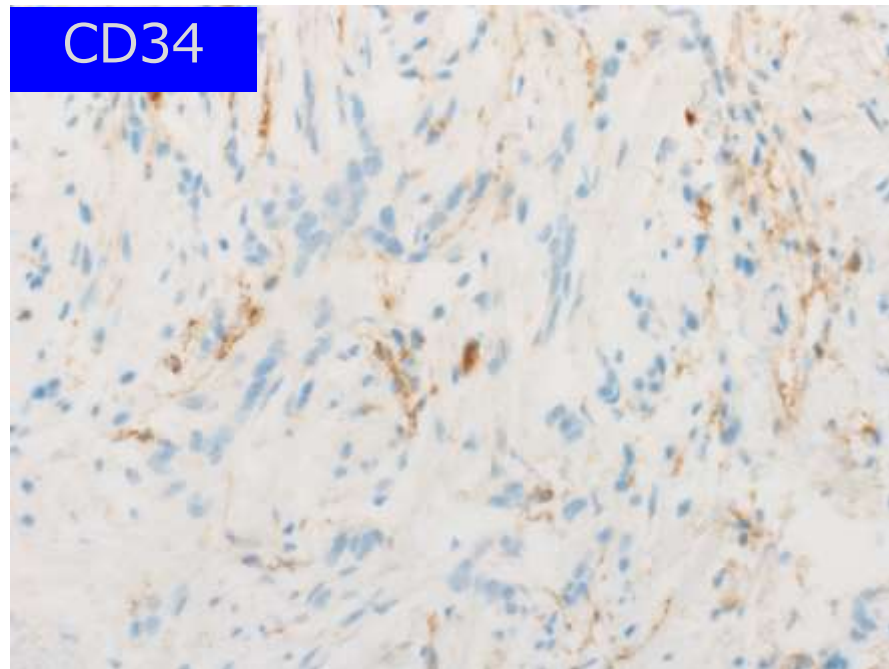


# EUS-FNA

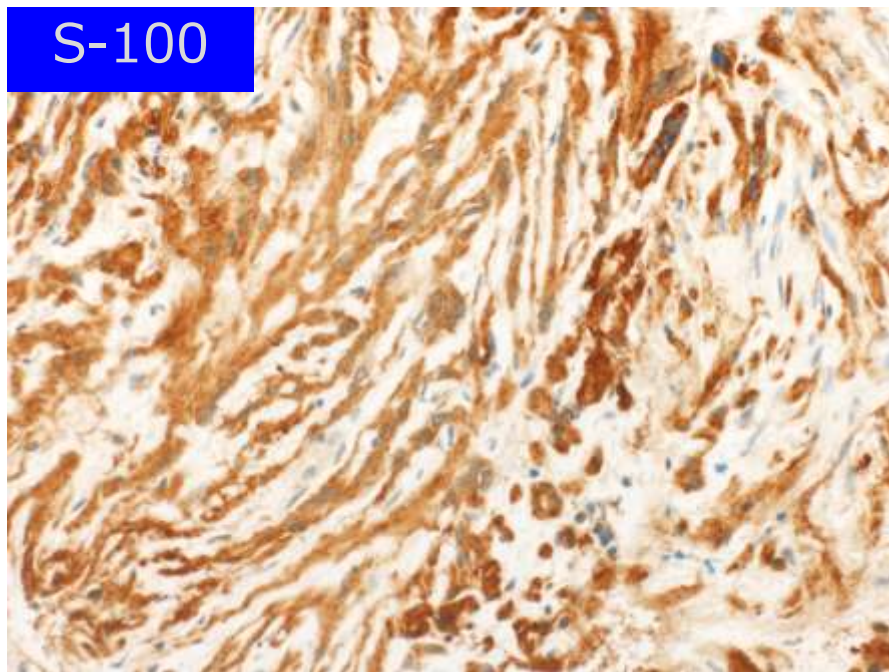
C-KIT



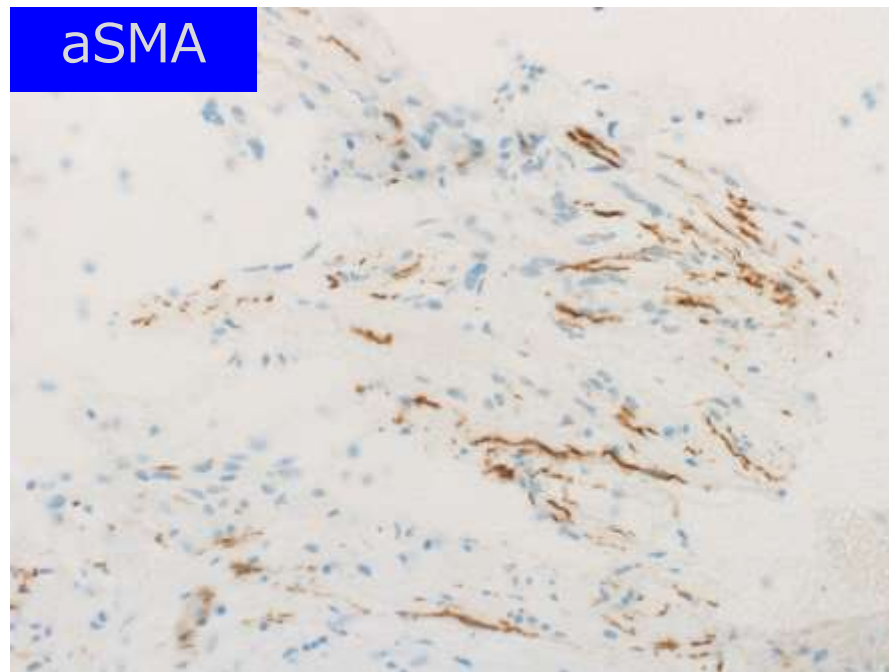
CD34



S-100



aSMA



EUS-FNA: ClassⅢ s/o GIST

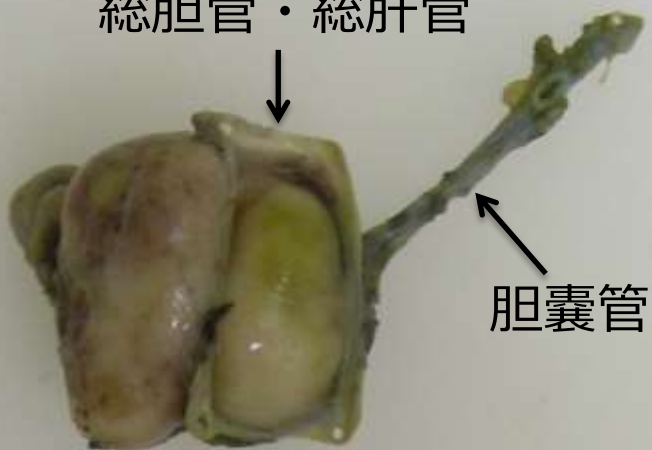
術前診断GIST。十二指腸部分切除術の予定にて開腹された。

腫瘍は十二指腸球部にはわずかに接しているのみ  
→筋層を切らずに容易に剥離できた。

腫瘍は総肝管・総胆管に固着していて剥離困難  
→腫瘍と肝外胆管が合併切除された。

# 切除材料

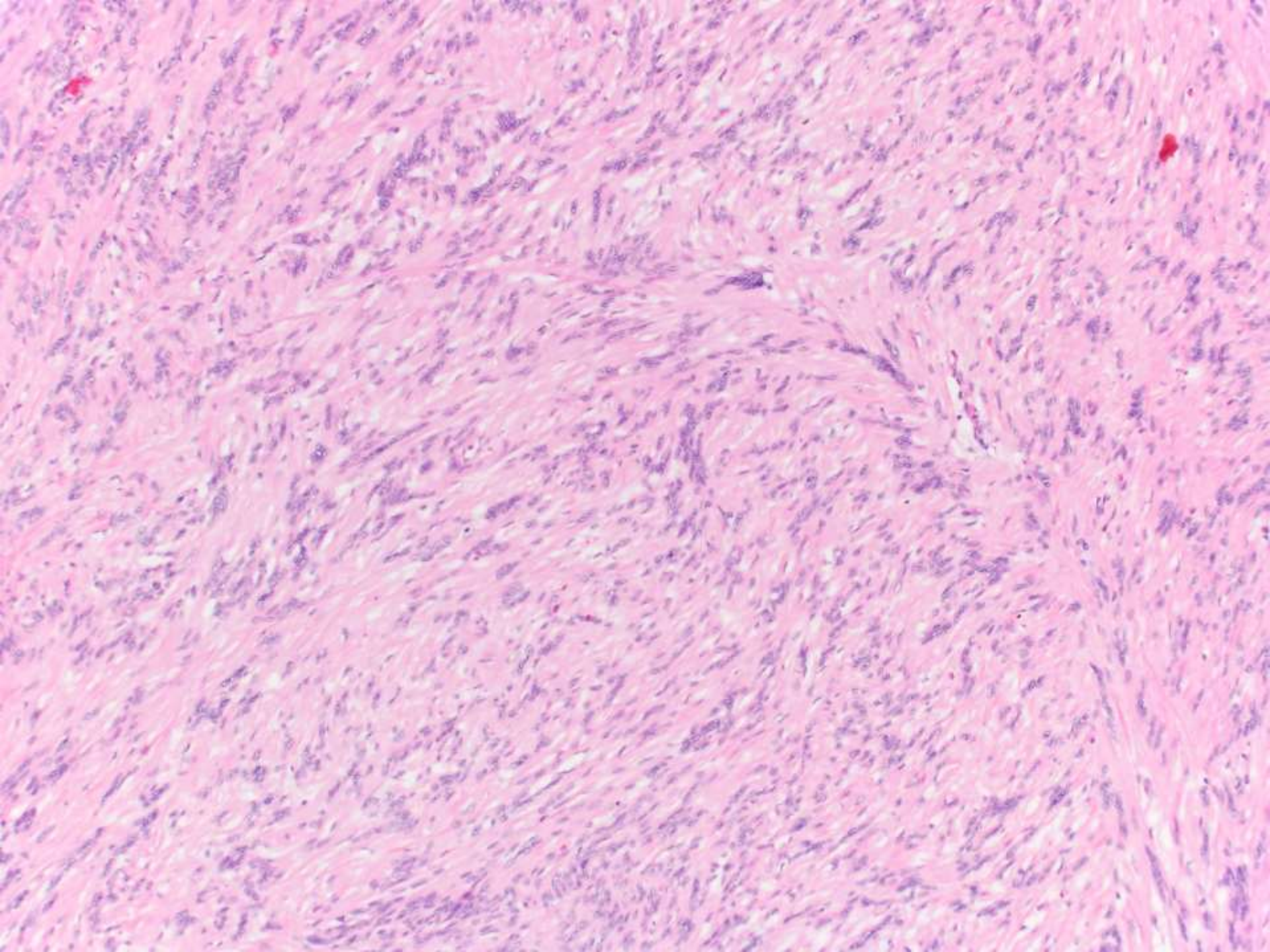
総胆管・総肝管



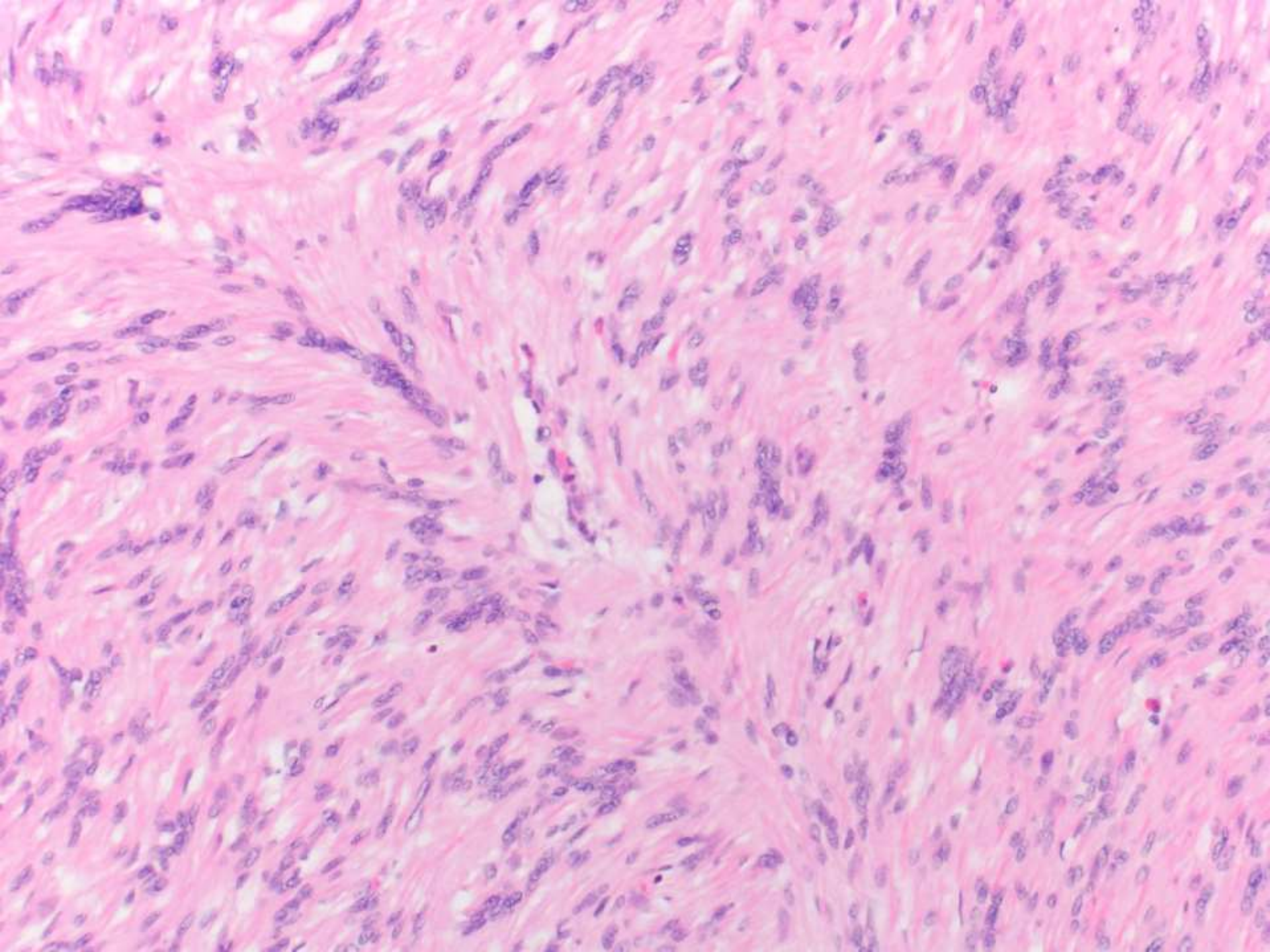
総胆管・総肝管

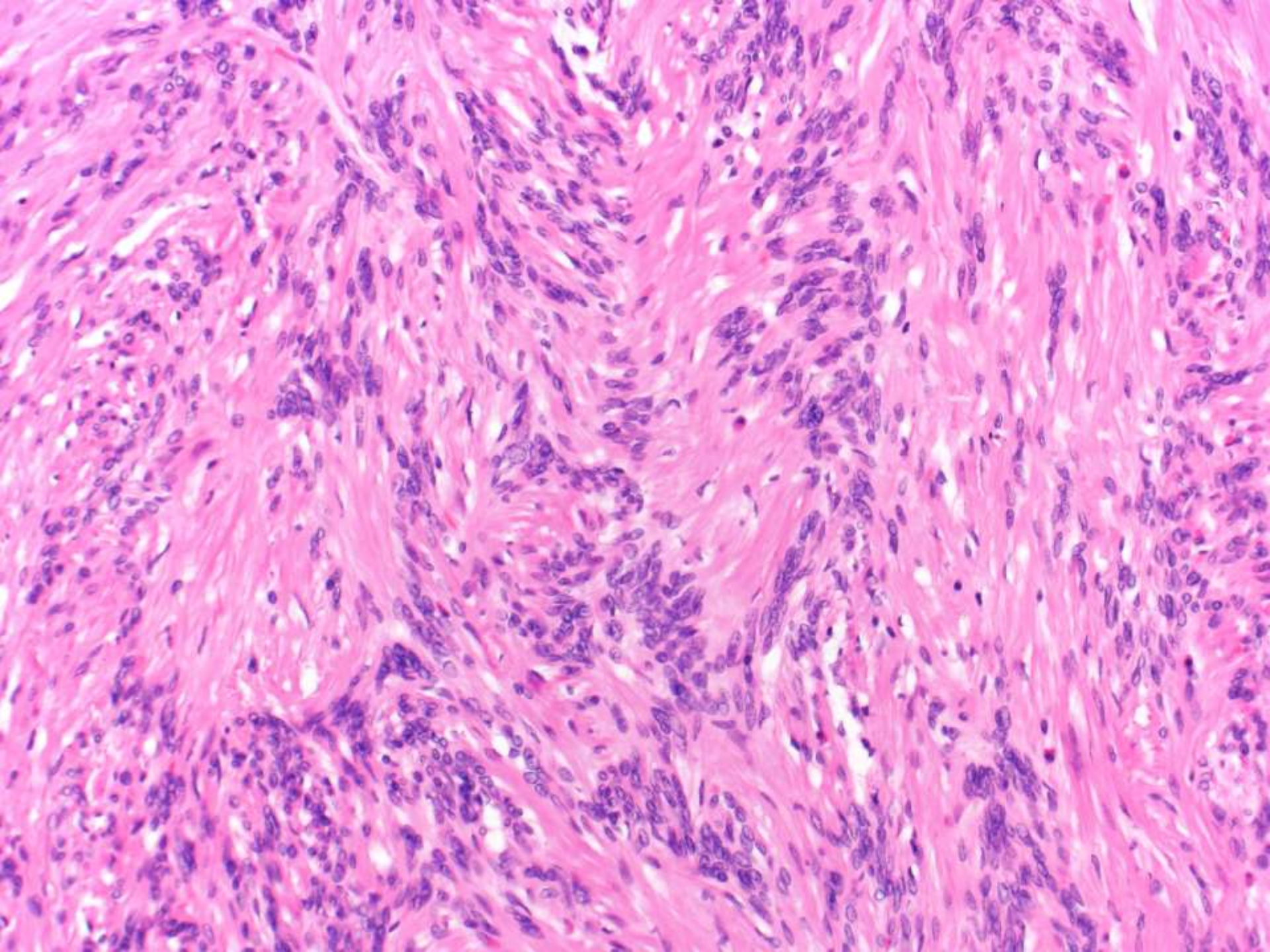


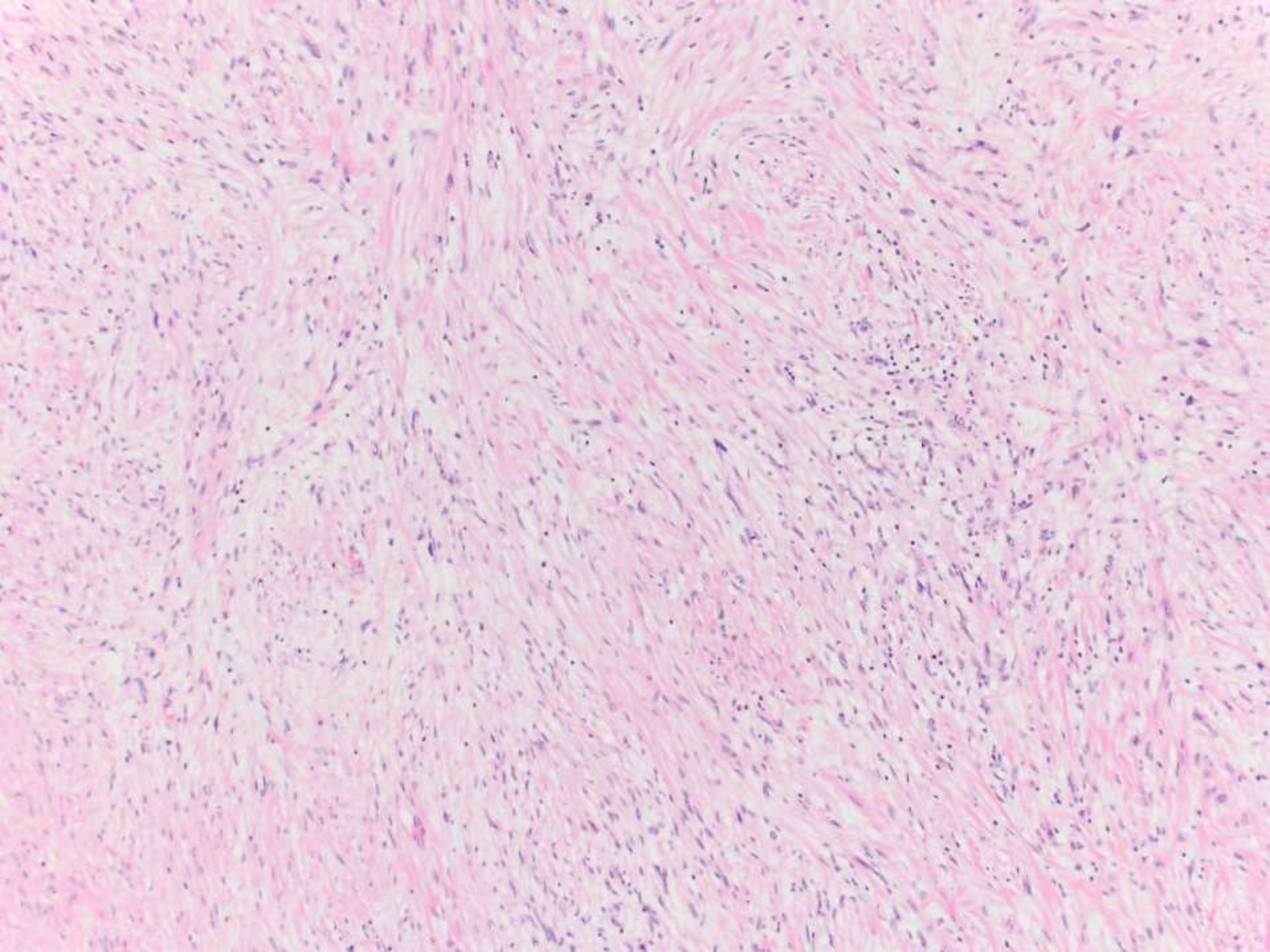
腫瘤：33x30x19mm

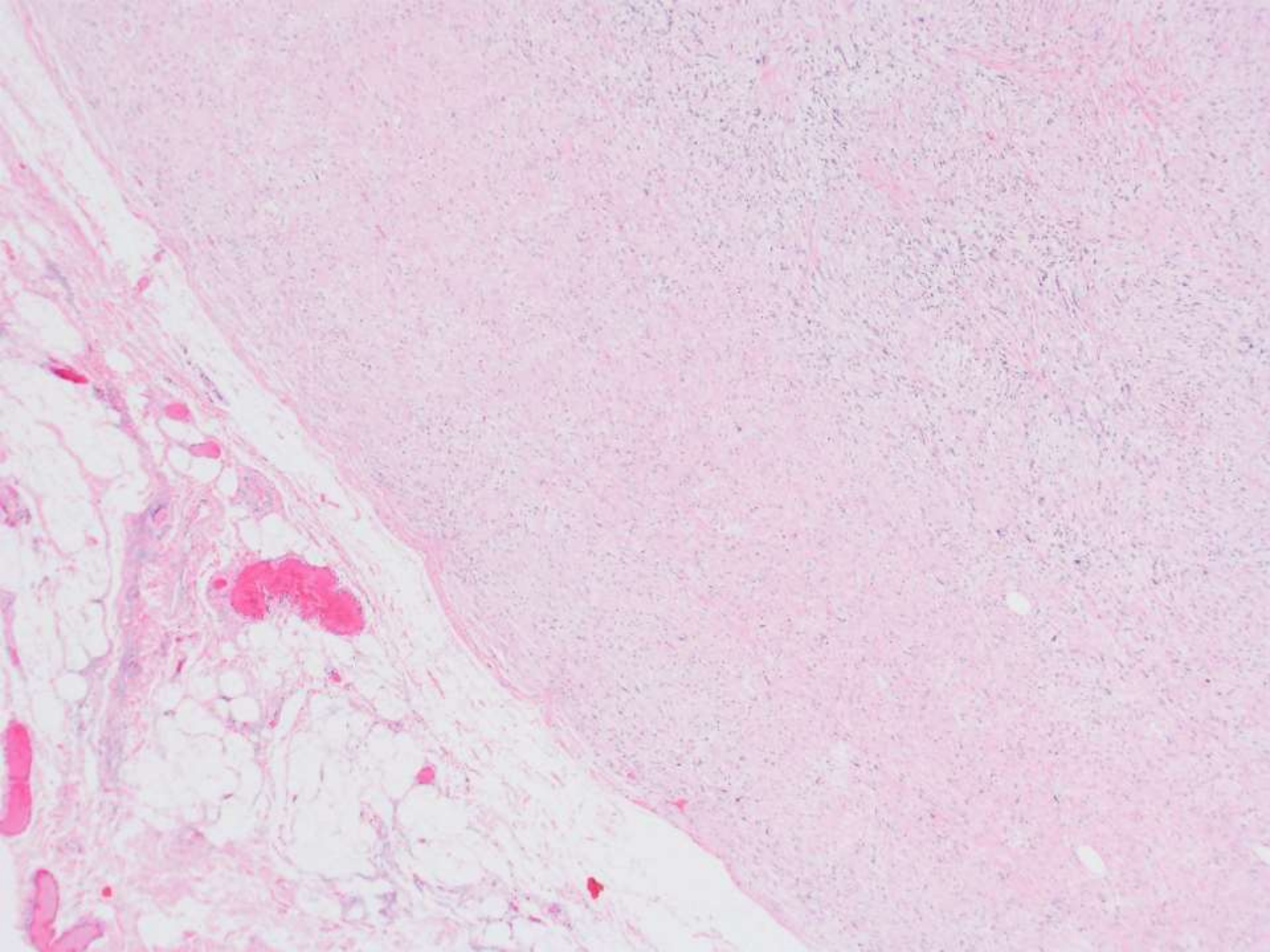


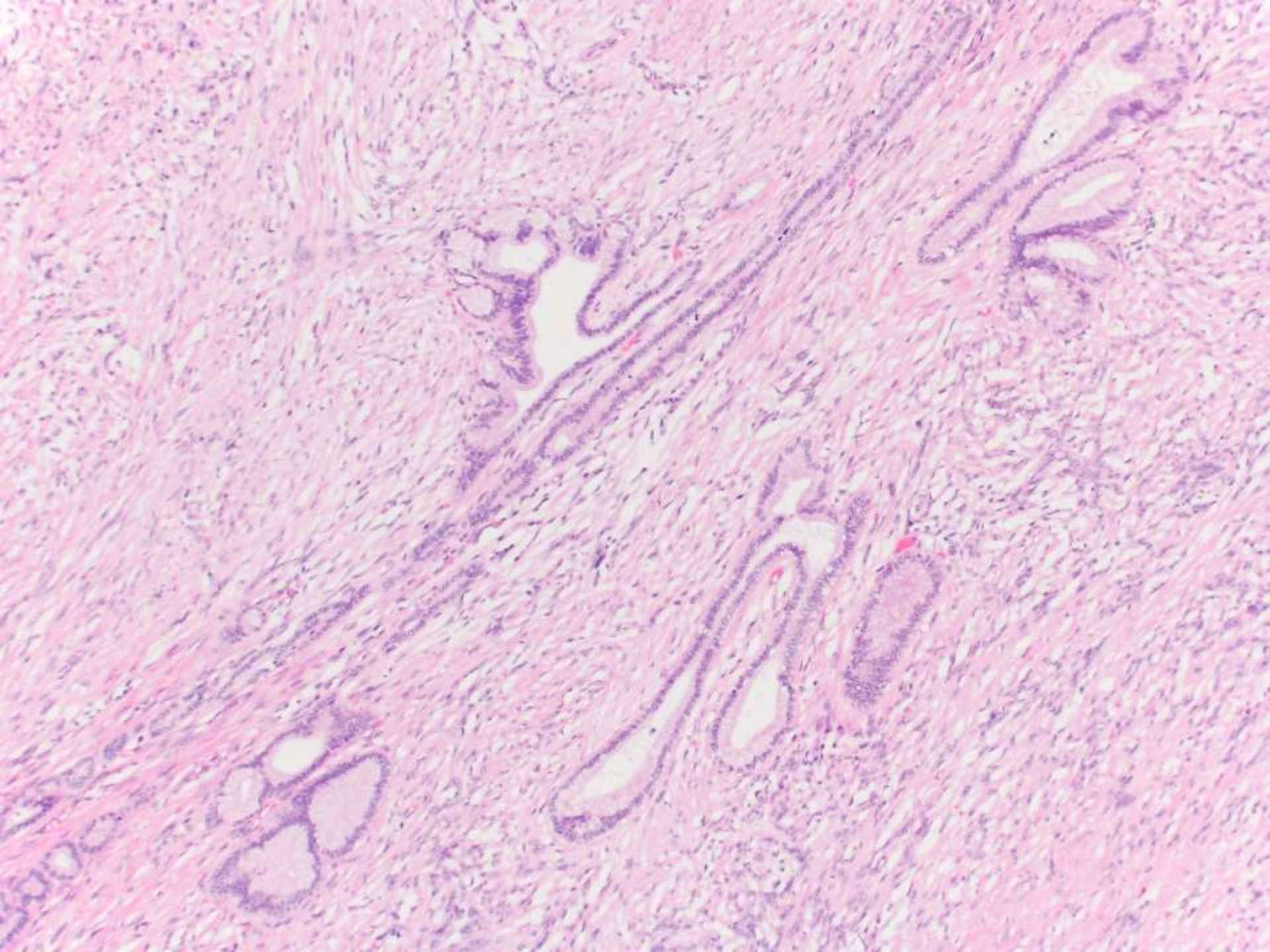






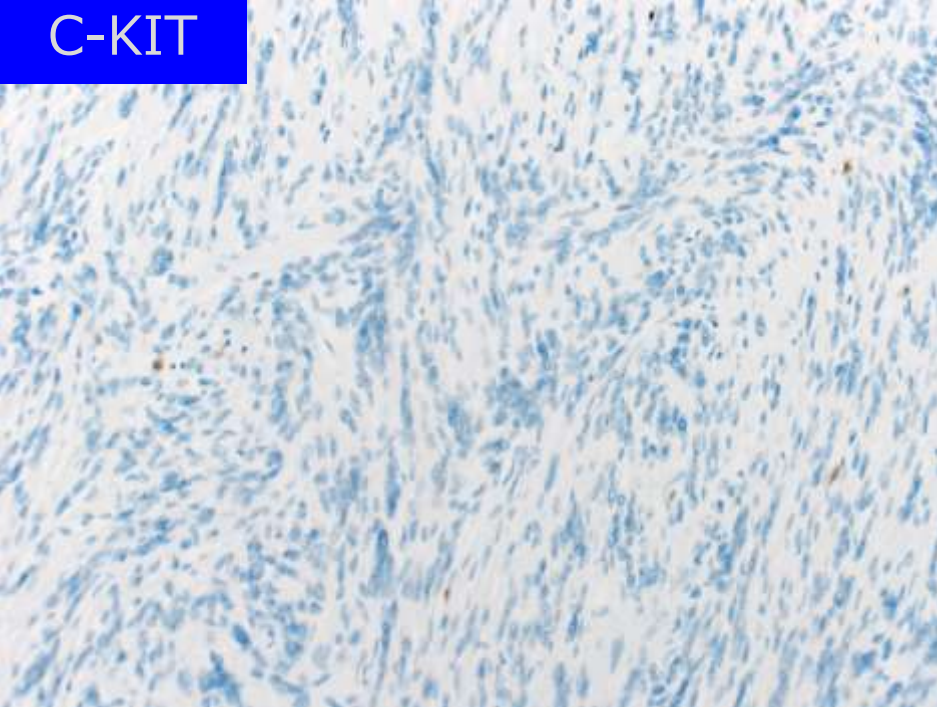




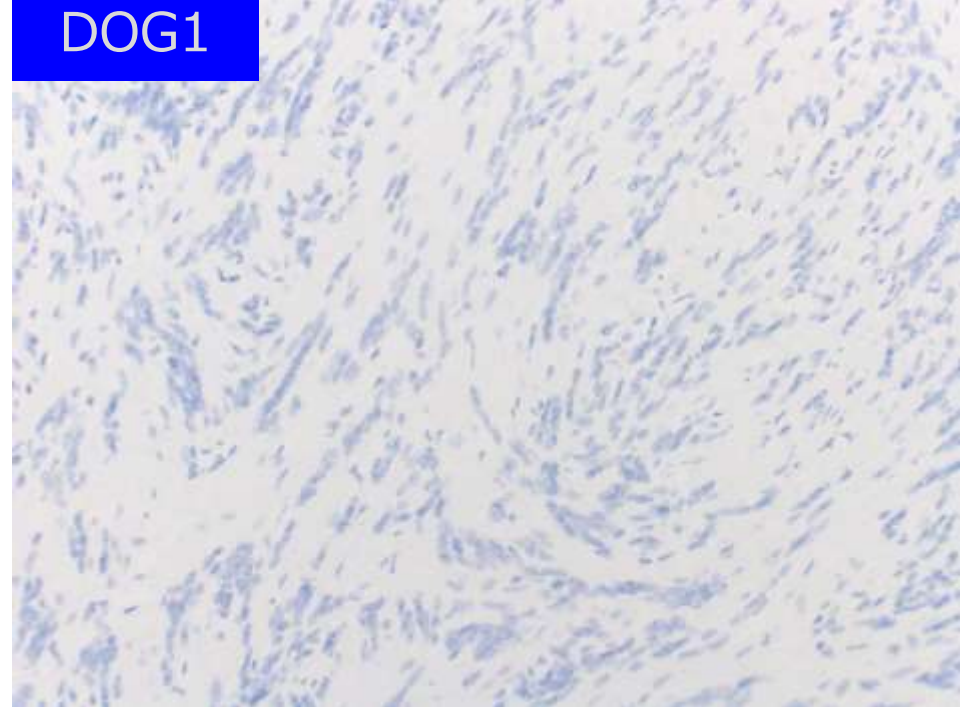


- GIST
- Leiomyoma/Adenomyoma
- Angiomyolipoma
- Schwannoma
- Neurofibroma
- Desmoid

C-KIT



DOG1



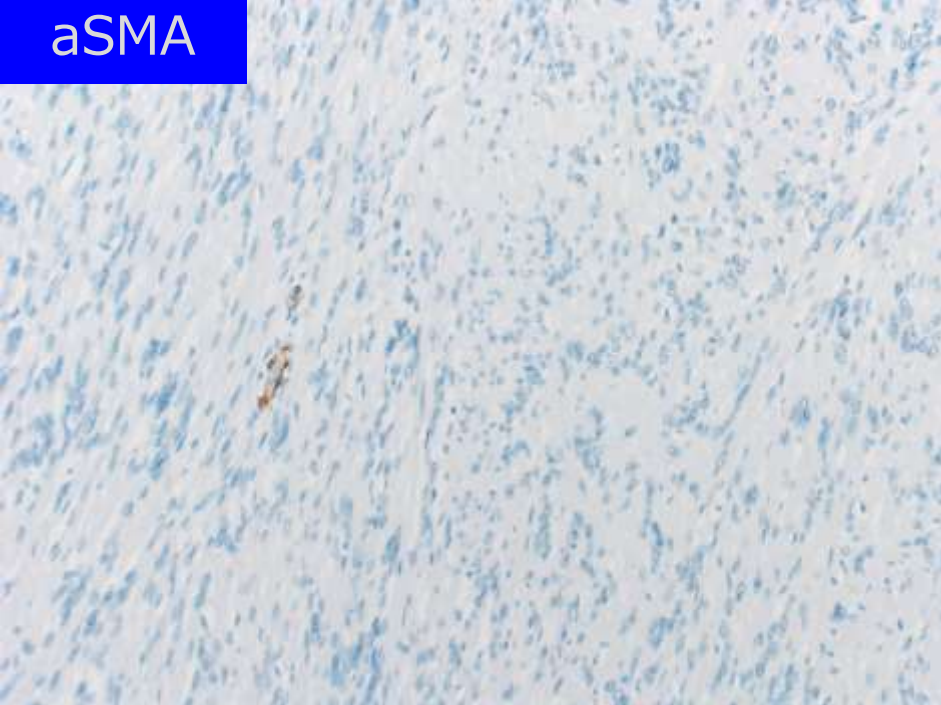
### Immunohistochemistry according to GIST genotype

	genotype				
	KIT mutation	PDGFRA mutation	BRAF mutation	SDHB, C,D mutation	SDHA mutation
KIT	+	-	+	+	+
DOG1	+	+	+	+	+
SDHB	+	+	+	-	-
SDHA	+	+	+	+	-

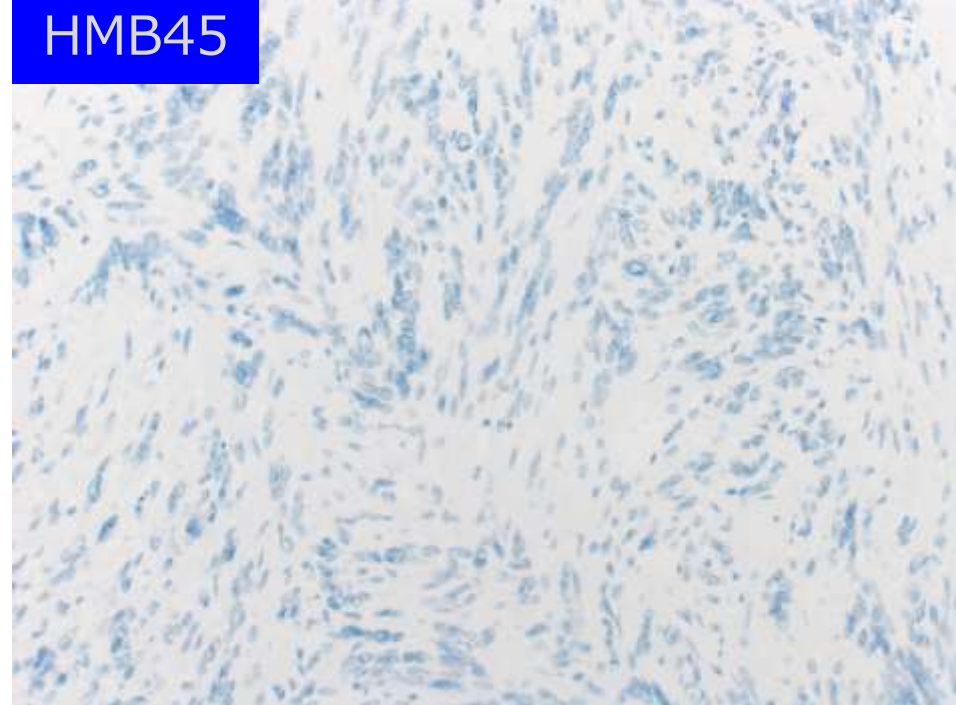
Entinger&Weiss Soft tissue tumors 6<sup>th</sup> ed. p578

C-KIT (-), DOG1 (-) → GISTはどのgenotypeとしても否定的

aSMA



HMB45

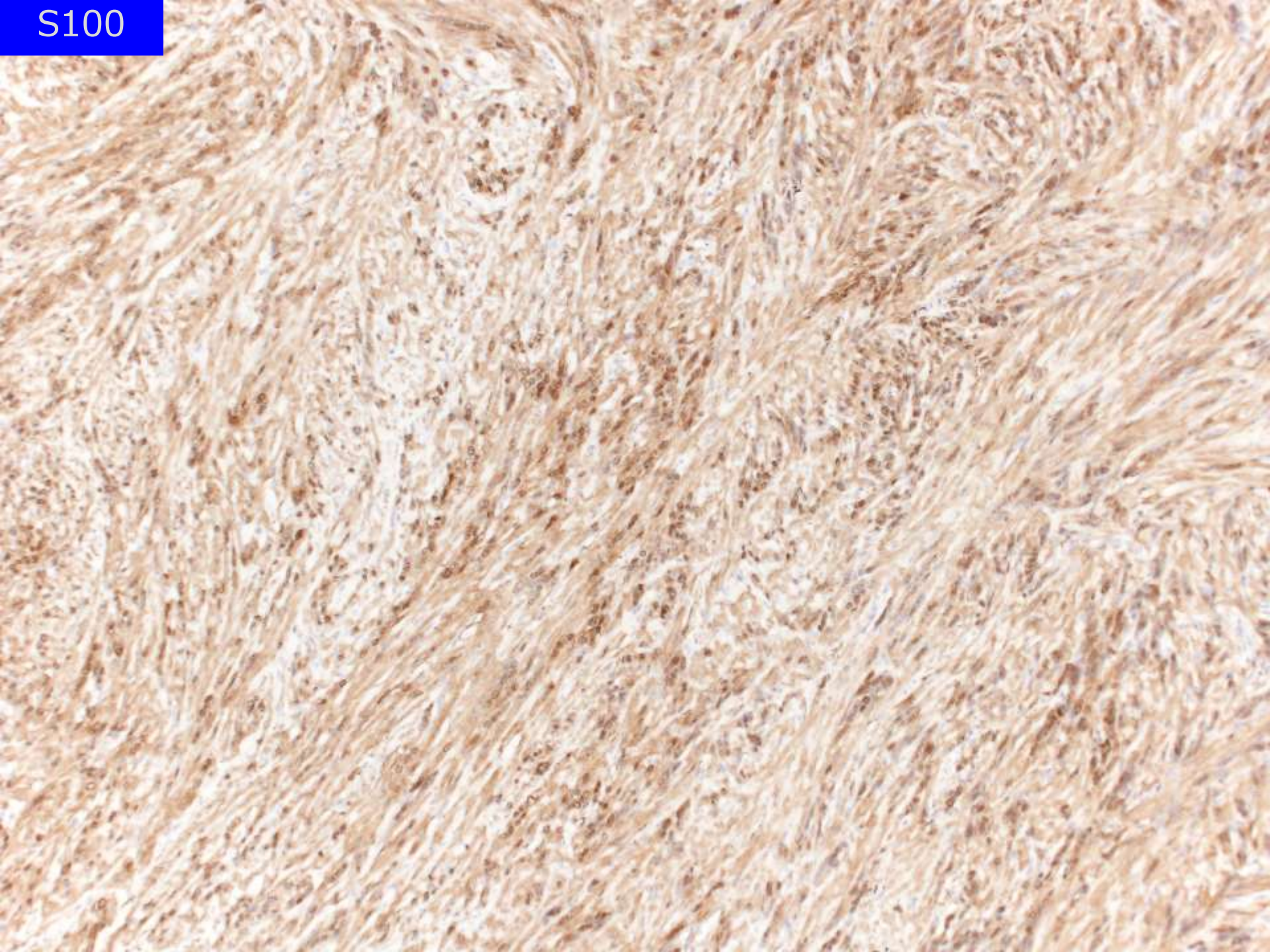


aSMA (-) → Leiomyoma, Adenomyoma 否定的  
Desmoid 否定的

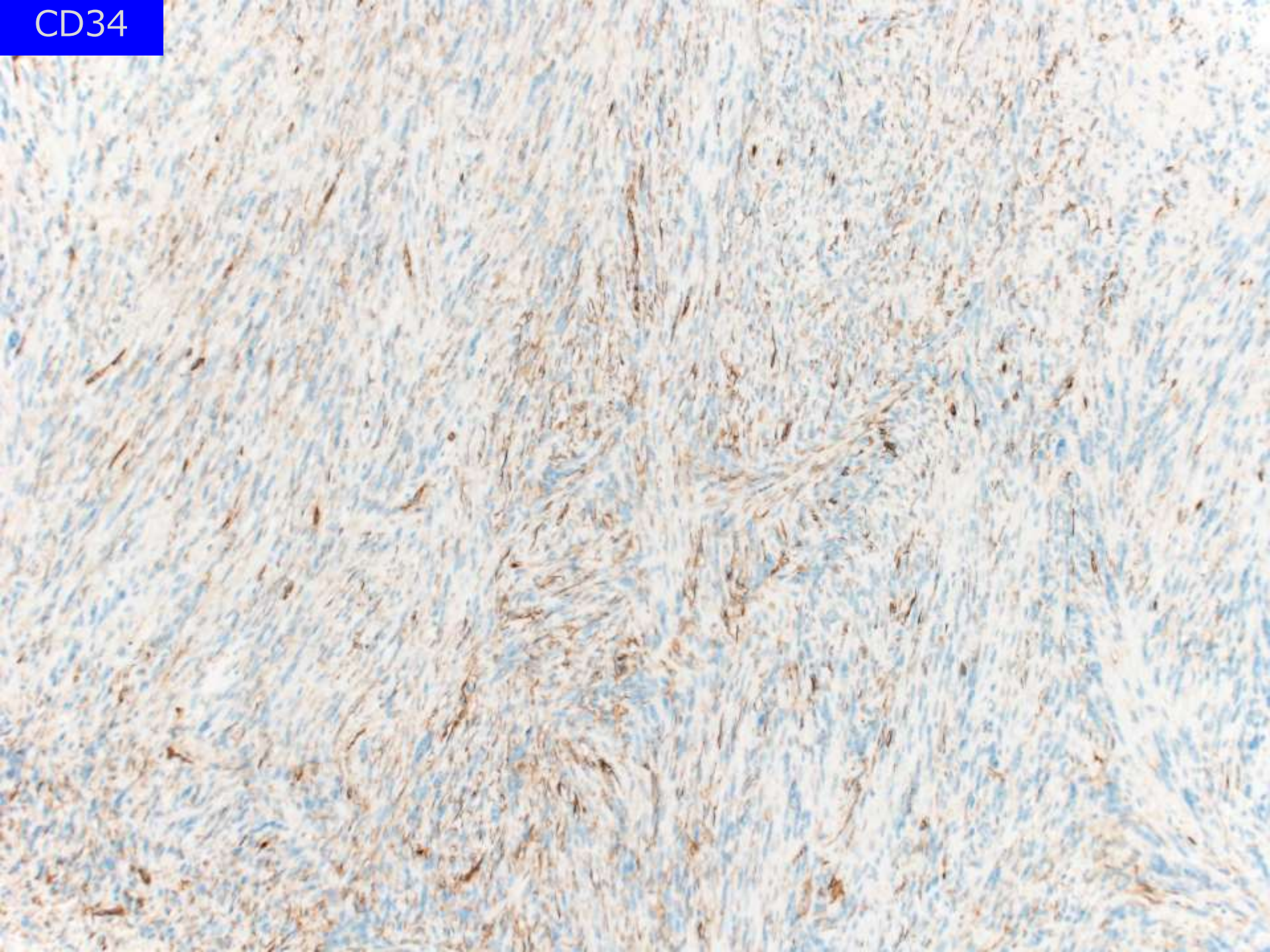
HMB45 (-) → Angiomyolipoma 否定的



S100



CD34



# Neurofibroma ? Schwannoma?

## Differential diagnosis between intraneural neurofibroma and Schwannoma

Differential feature	Neurofibroma	Schwannoma
Relationship to nerve of origin	Fusiform mass involving the whole nerve	Forms an eccentric mass around the nerve
Circumscription	Encased in nerve	Usually encapsulated
Nuclear palisading	Absent	Often present
Myxoid stroma	Often present	Rarely present
Vascular dilatation	Not prominent	Often present
Xanthoma cells	Absent	Often present
S100 protein in cellular areas	Variable numbers of cells positive	All cells positive
CD34	Positive cells in the cellular area	Cellular Schwann cell areas negative
Neurofilament	Residual axons positive	Positive elements usually absent

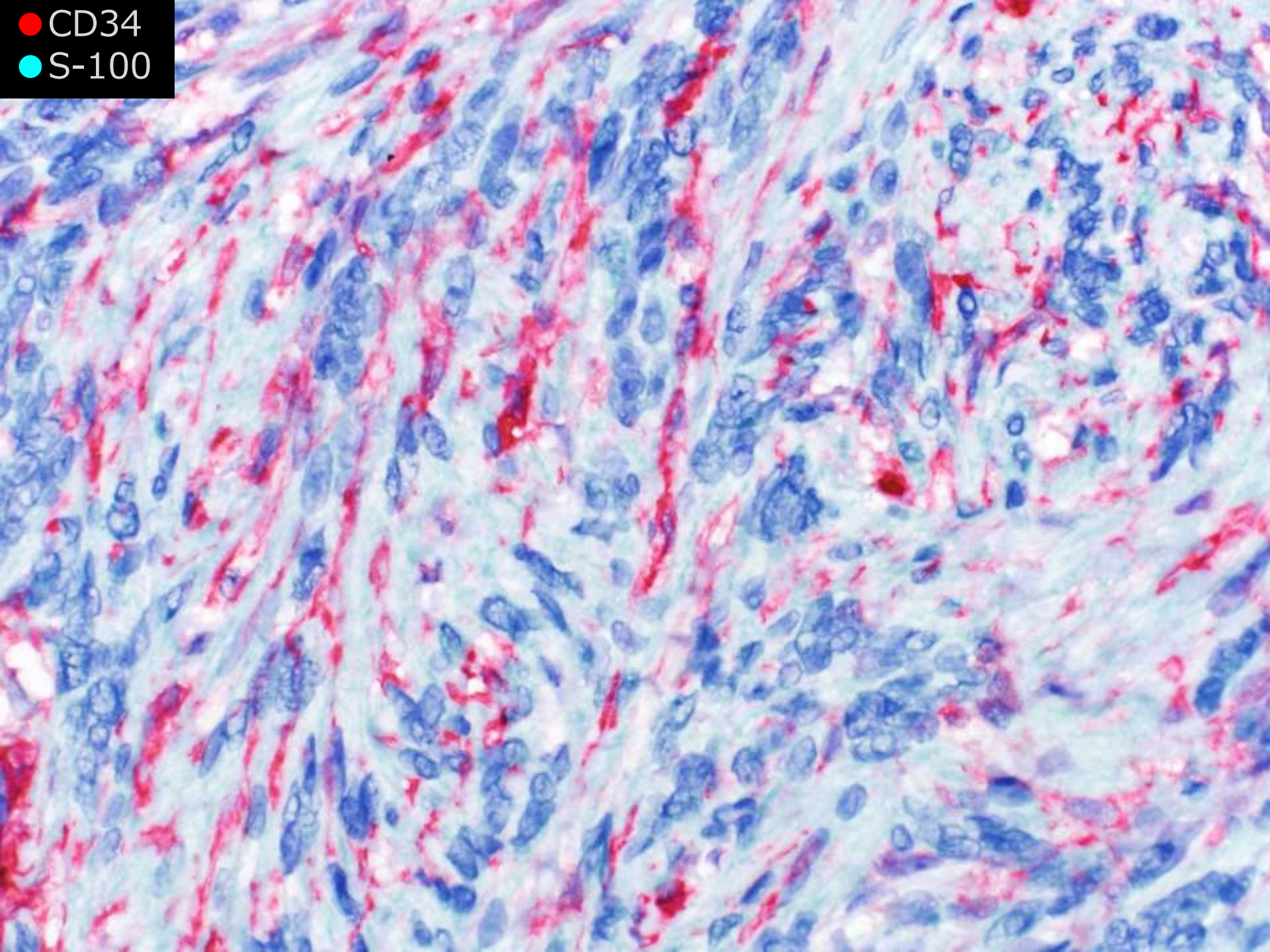
Miettinen Modern soft tissue pathology 1<sup>st</sup> ed. p678

電顕では、

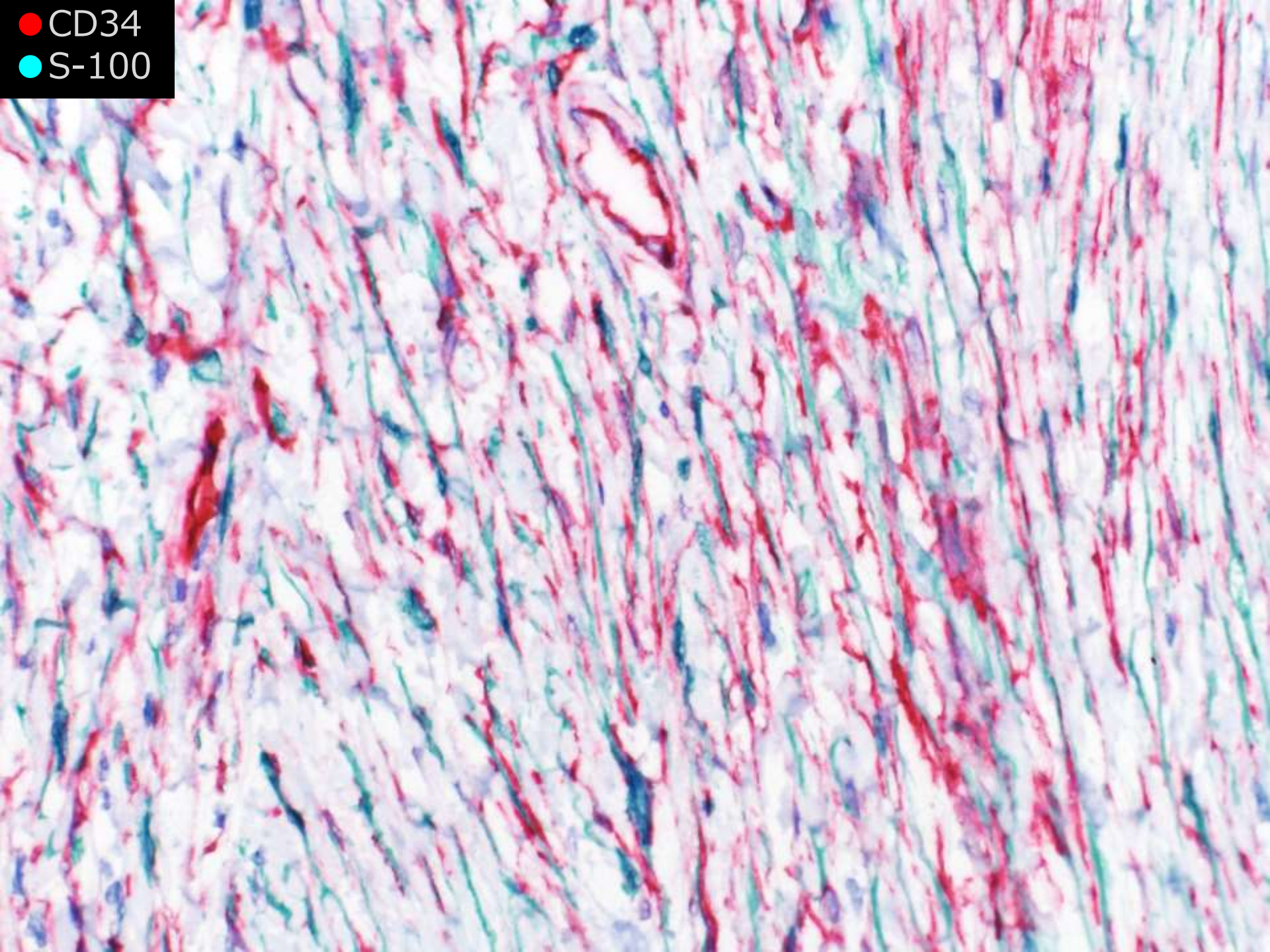
Neurofibroma は Schwann細胞とfibroblast の混在から成り、  
Schwannoma はほぼ Schwann細胞のみから成る。

Enzinger&Weiss's Soft tissue tumors 6<sup>th</sup> ed. p820

● CD34  
● S-100

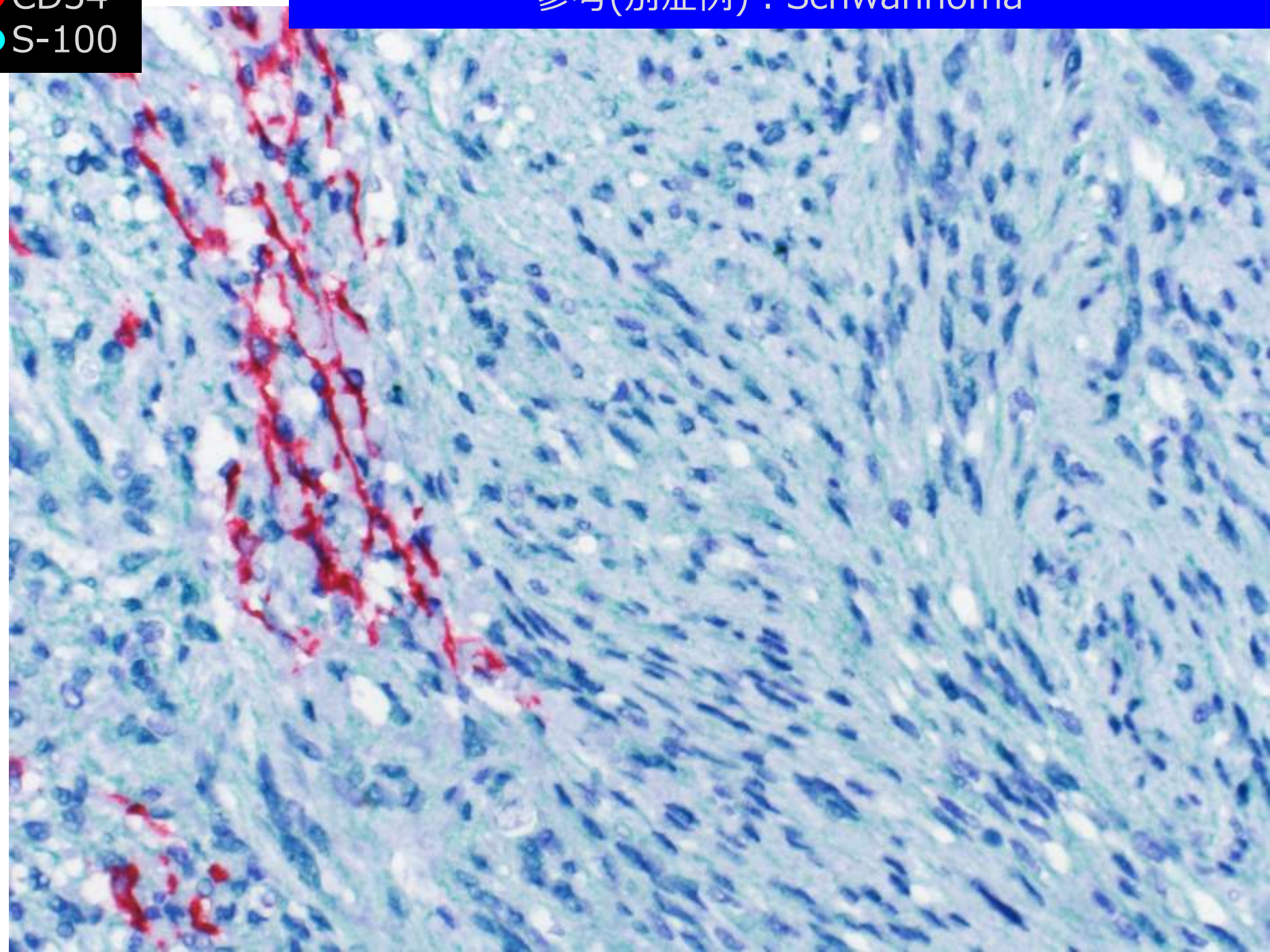


● CD34  
● S-100



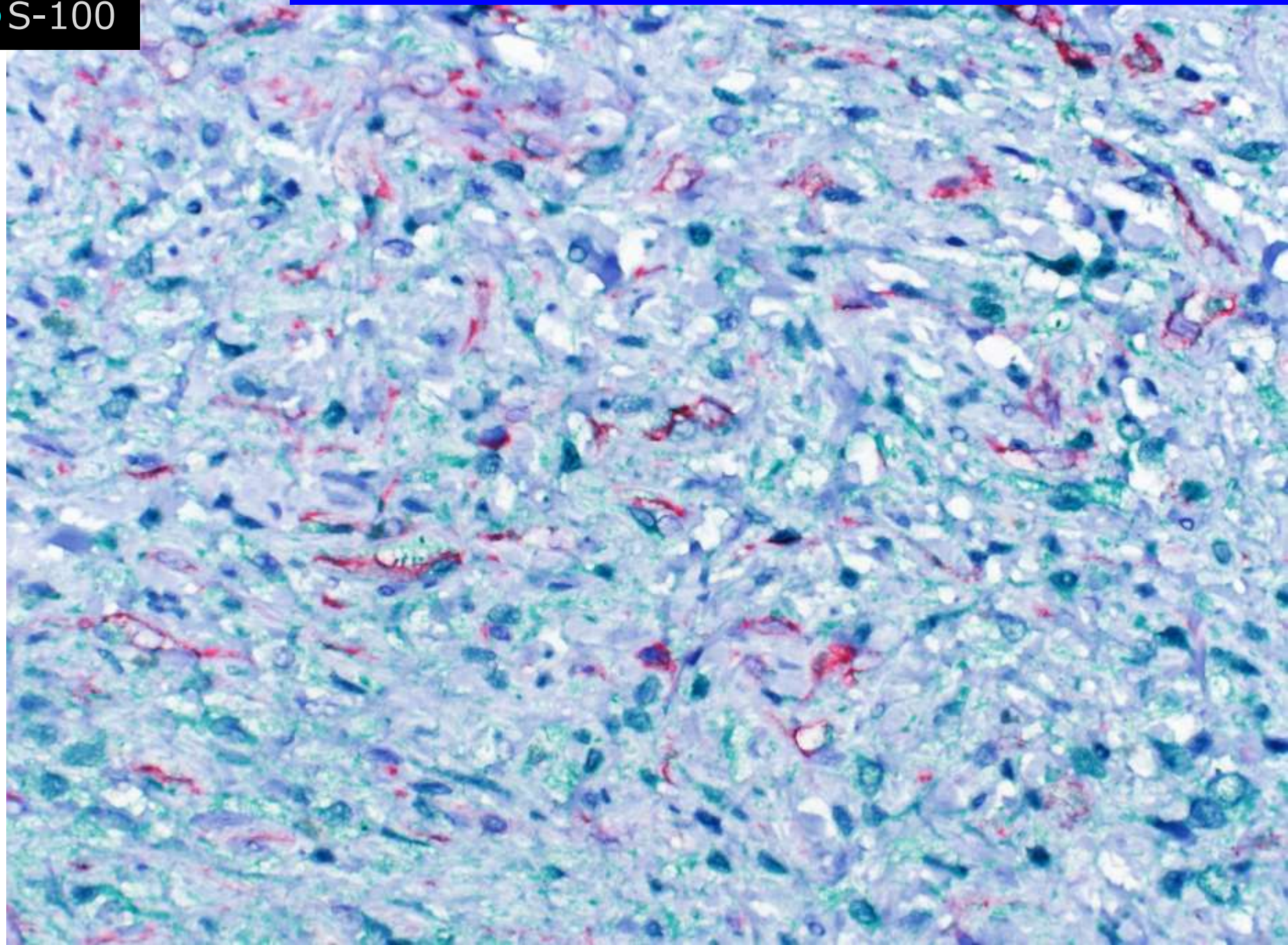
● CD34  
● S-100

参考(別症例) : Schwannoma



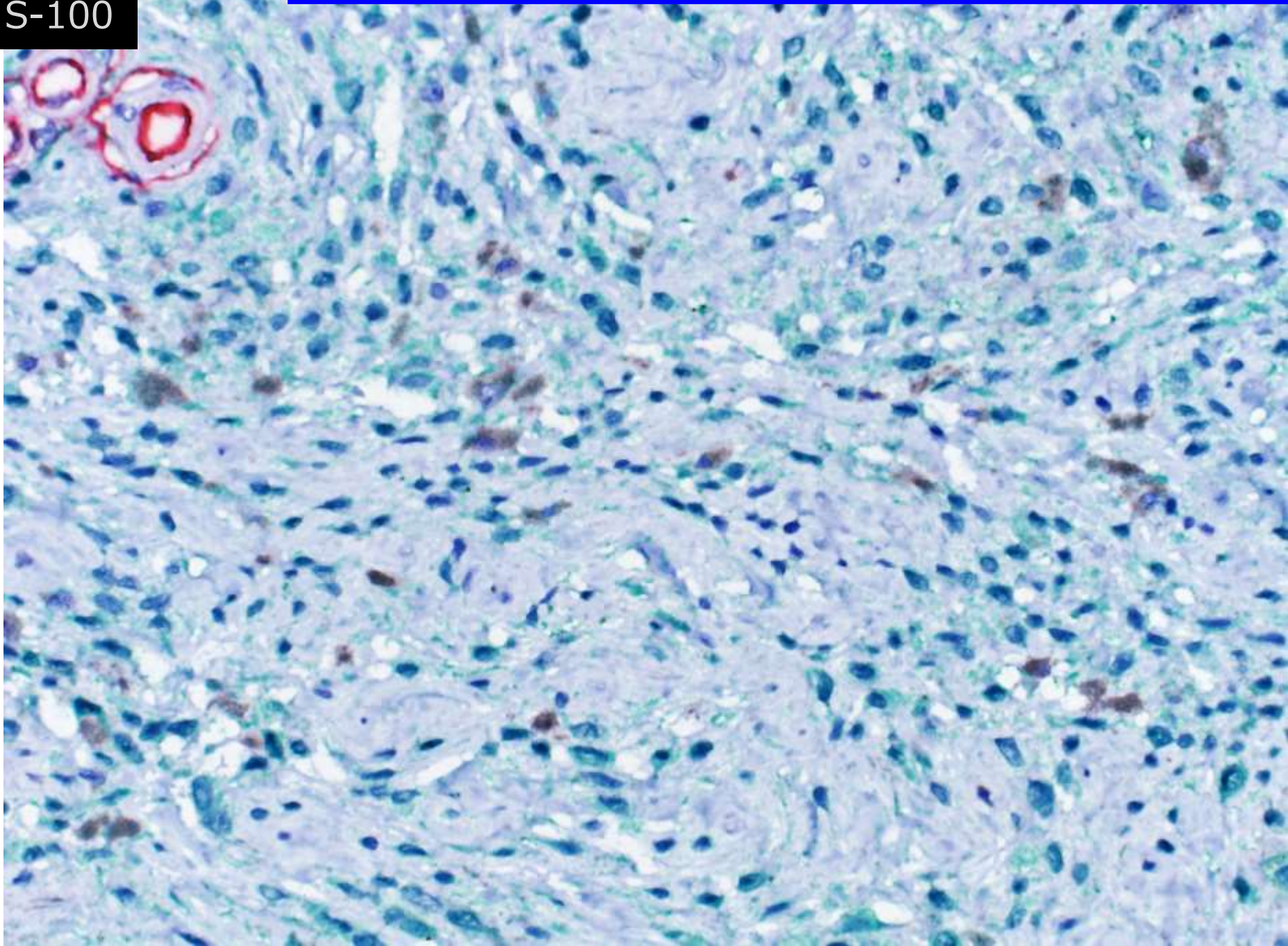
● CD34  
● S-100

参考(別症例) : Neurofibroma



● CD34  
● S-100

参考(別症例) : Neurofibroma





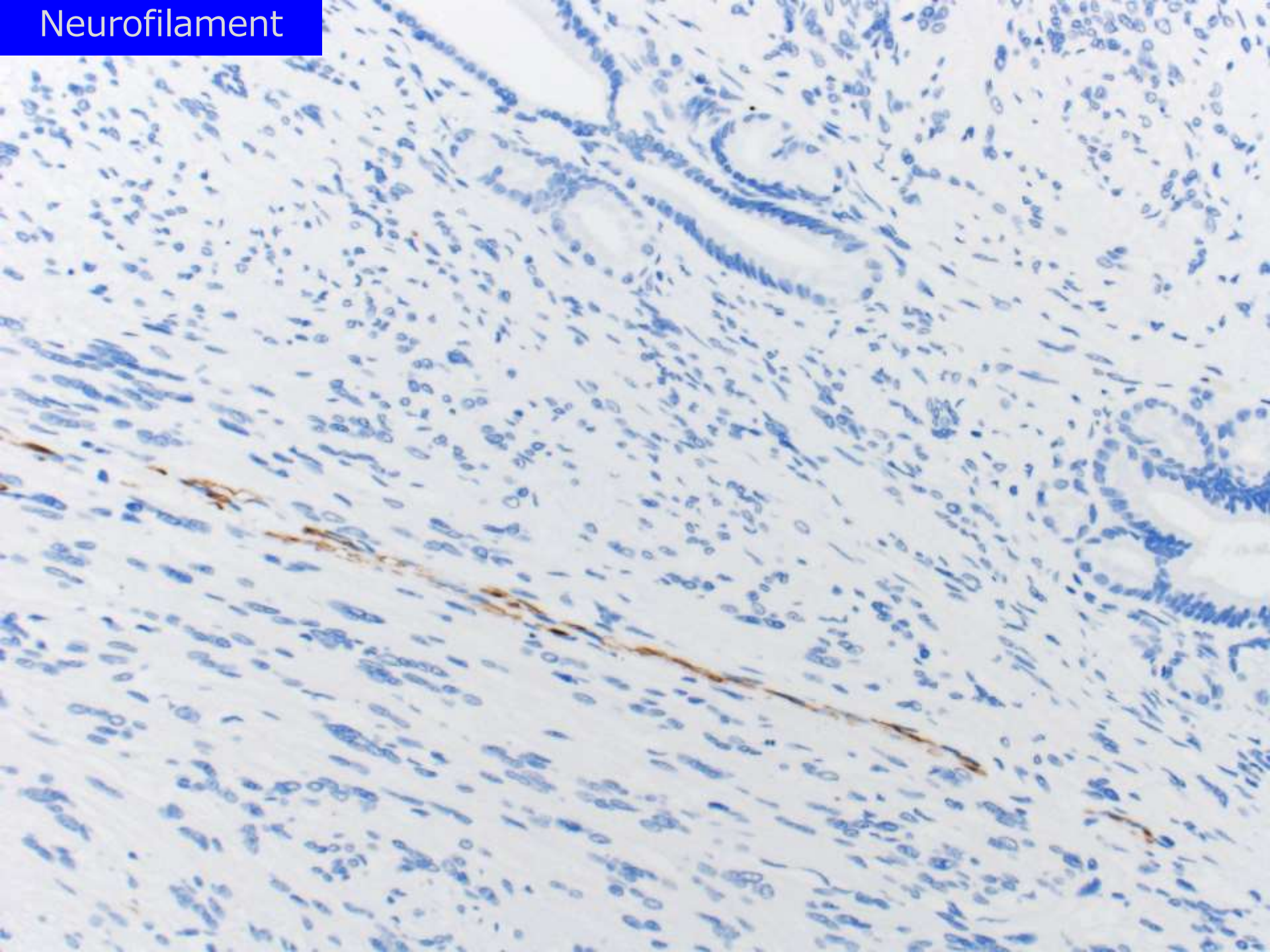
# Neurofibroma ? Schwannoma?

## Differential diagnosis between intraneural neurofibroma and Schwannoma

Differential feature	<b>Neurofibroma</b>	<b>Schwannoma</b>
Relationship to nerve of origin	Fusiform mass involving the whole nerve	Forms an eccentric mass around the nerve
Circumscription	Encased in nerve	Usually encapsulated
Nuclear palisading	Absent	Often present
Myxoid stroma	Often present	Rarely present
Vascular dilatation	Not prominent	Often present
Xanthoma cells	Absent	Often present
S100 protein in cellular areas	Variable numbers of cells positive	All cells positive
CD34	Positive cells in the cellular area	Cellular Schwann cells areas negative
<b>Neurofilament</b>	<b>Residual axons positive</b>	<b>Positive elements usually absent</b>

Miettinen Modern soft tissue pathology 1<sup>st</sup> ed. p678

Neurofilament



概して、Neurofibroma は Schwannoma様の成分を有することがあるが、Schwannoma は Neurofibroma 様の成分を有することはない。

Miettinen Modern soft tissue pathology 1<sup>st</sup> ed. p677

Cellular neurofibroma は SchwannomaのAntoni A areaに似ていることもあるが、2つのareaが明瞭に分かれていないし、被膜に覆われていない。

Enzinger&Weiss's Soft tissue tumors 6<sup>th</sup> ed. p798

演者診断

Neurofibroma

## Bile duct neurofibroma; Literature review

Age Sex	Genetic background	Symptoms	Size	Reference
61 M	NF1	obstructive jaundice		Meyer GW Ann Int Med (1982) 97:722
32 F	sporadic	obstructive jaundice	15x10x10mm	Li FY Digest Dis Sci (2005) 50:1166
28 F	sporadic	upper abdominal pain, fever	30mm	Béchade P Gastrointest Endosco (2010) 72:895
47 F	sporadic	obstructive jaundice	25x20mm	Ray S Am J Surg (2011) 202:e1
65 F	sporadic	obstructive jaundice		Jiang L Dig Liver Dis (2011) 43:85
70 F	sporadic	obstructive jaundice		Rosa AD Ann R Coll Surg Engl (2013) 95:e14
36 M	sporadic	obstructive jaundice	20x20mm	Guo H J Med Ultrasonics (2016) 43:537
46 F	sporadic	no symptoms	33x30x19mm	Present case

\* これら以外に、非英語などで参照できないもの8件

\* 日本語での報告なし

胆管の Schwannomaの報告例もほぼ同数

肝外胆管に発生した Neurofibroma とと思われる症例である。

Neurofibroma は S-100陽性の Schwann細胞と CD34陽性の fibroblast の混在からなる。同じ細胞が S-100とCD34の両方に染まるのではない。

Schwannoma は基本的に S-100陽性の Schwann細胞のみからなるが、被膜や血管周囲では CD34陽性のfibroblast も混在している。

胆管の Neurofibroma は非常にまれで、これまでに15例程度の報告があるのみである。

NF1合併例よりも孤発例が多く、圧倒的に女性に多い。閉塞性黄疸で発症することが多い。