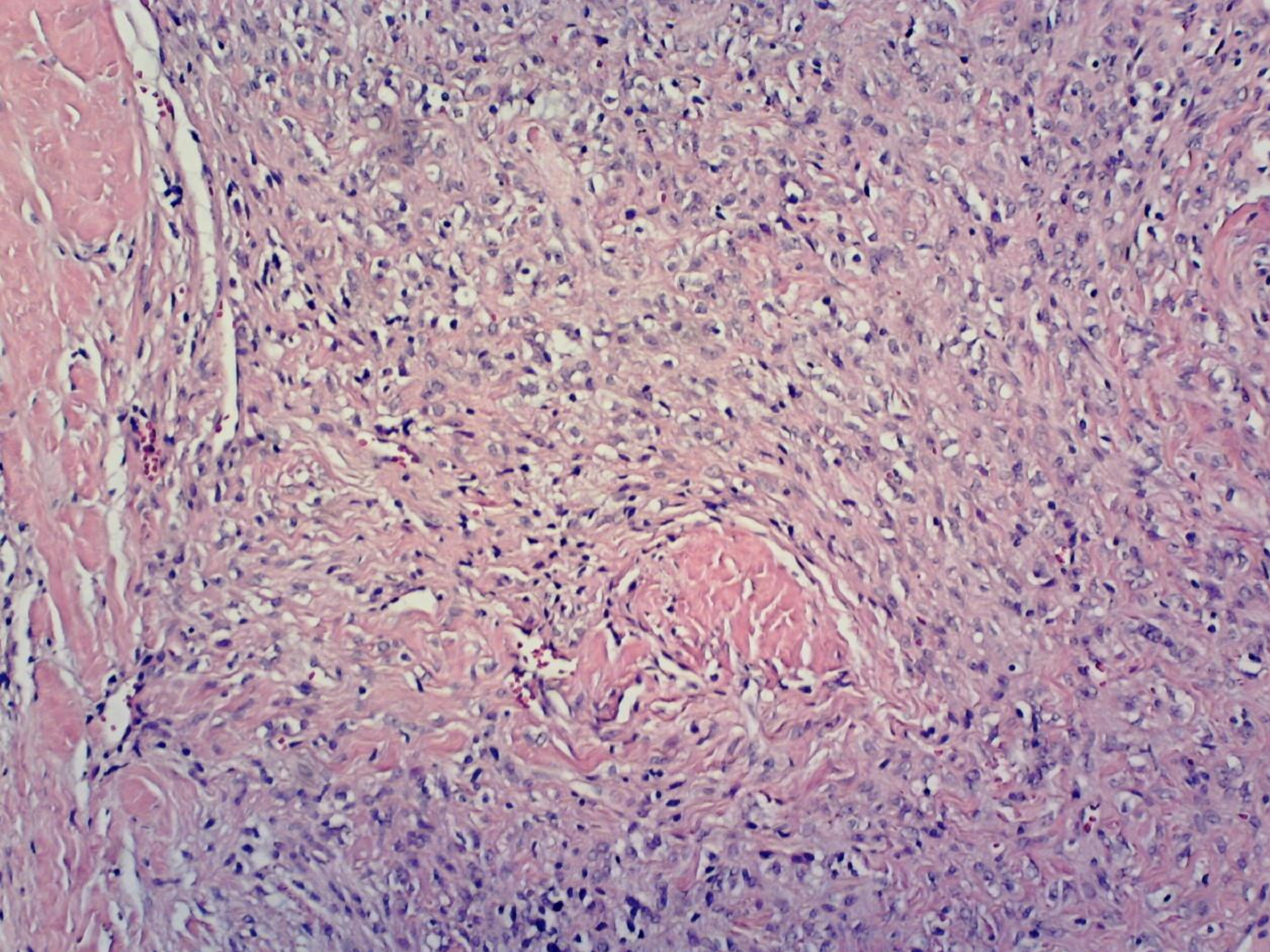
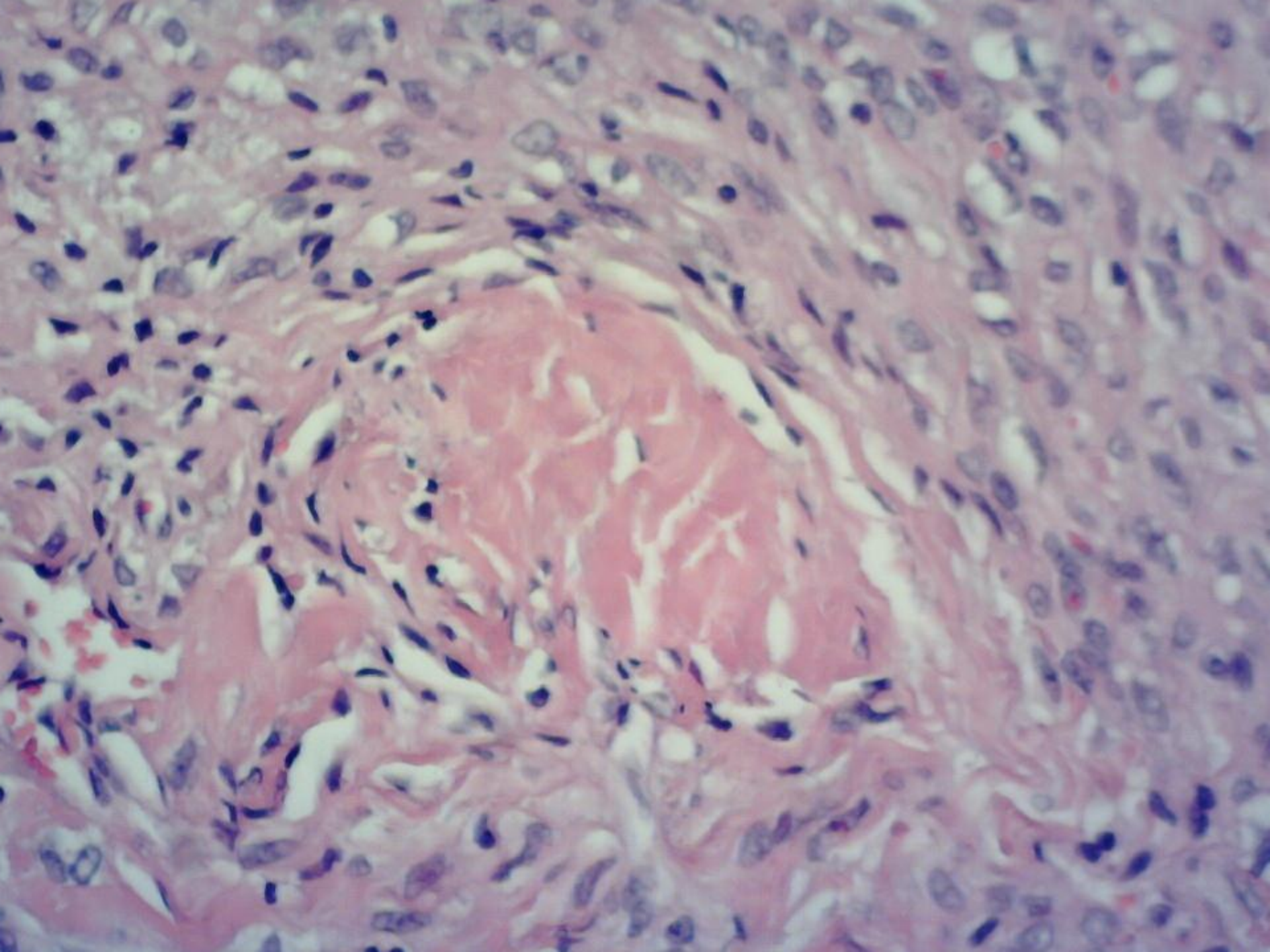
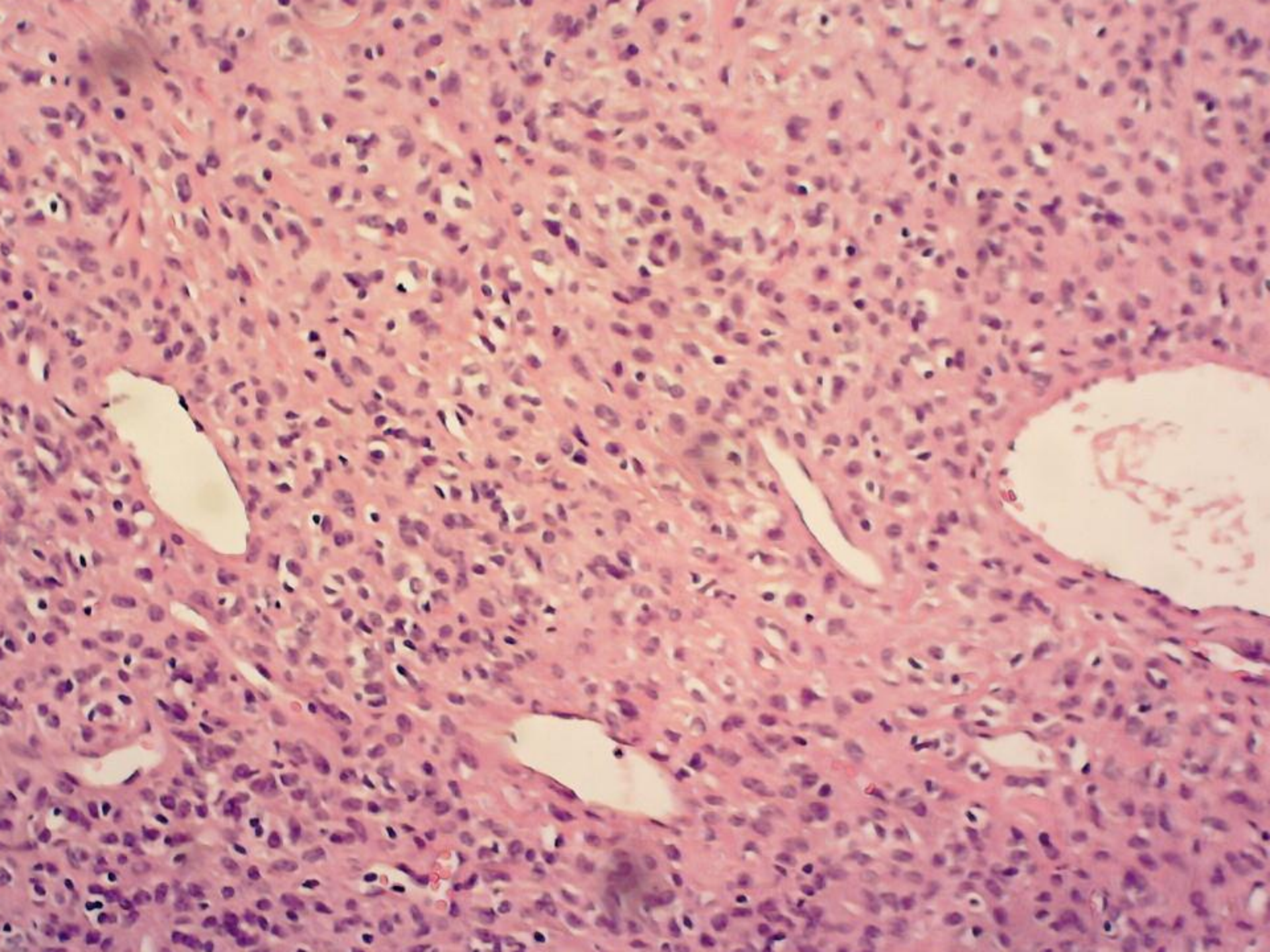
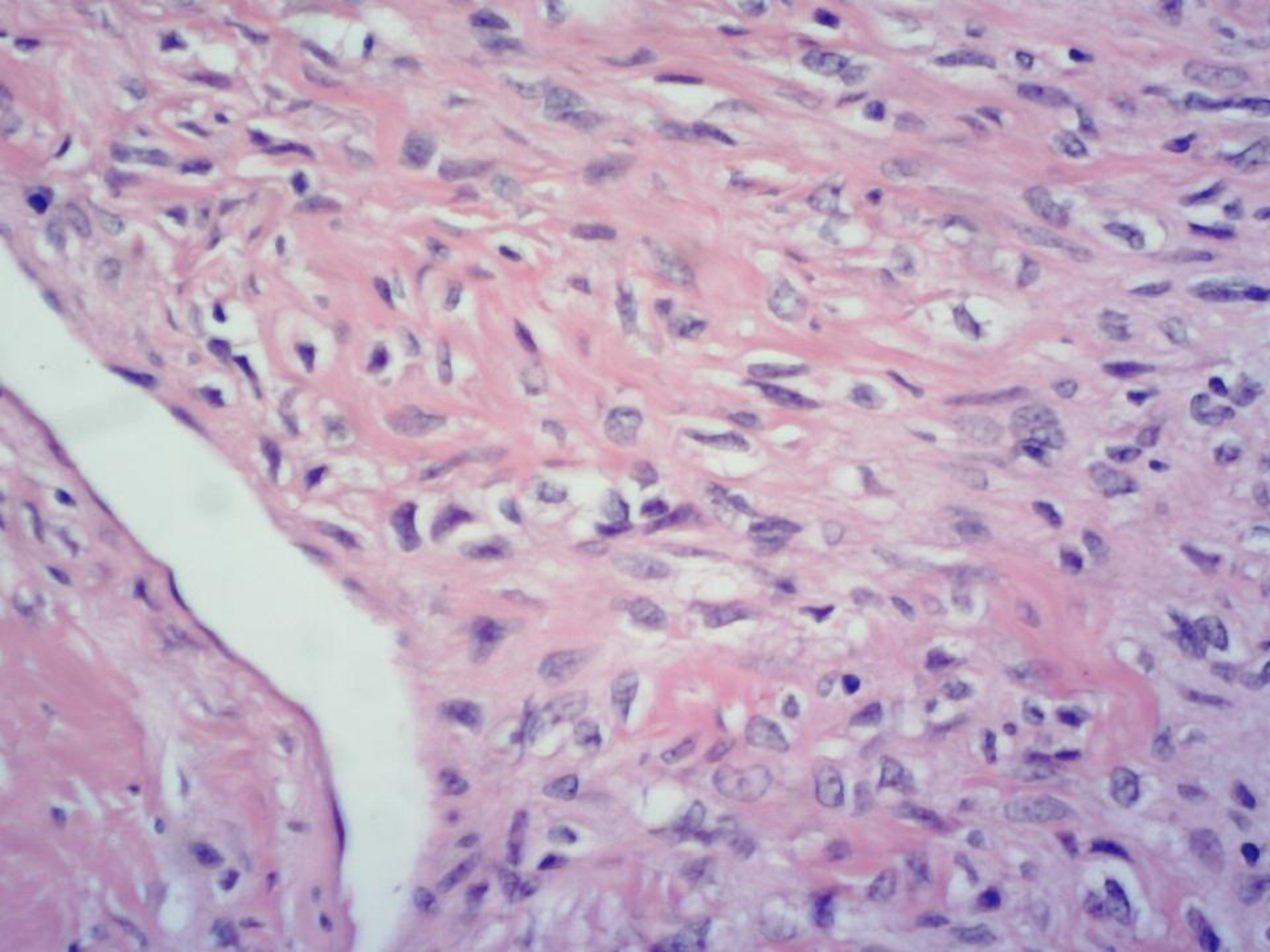
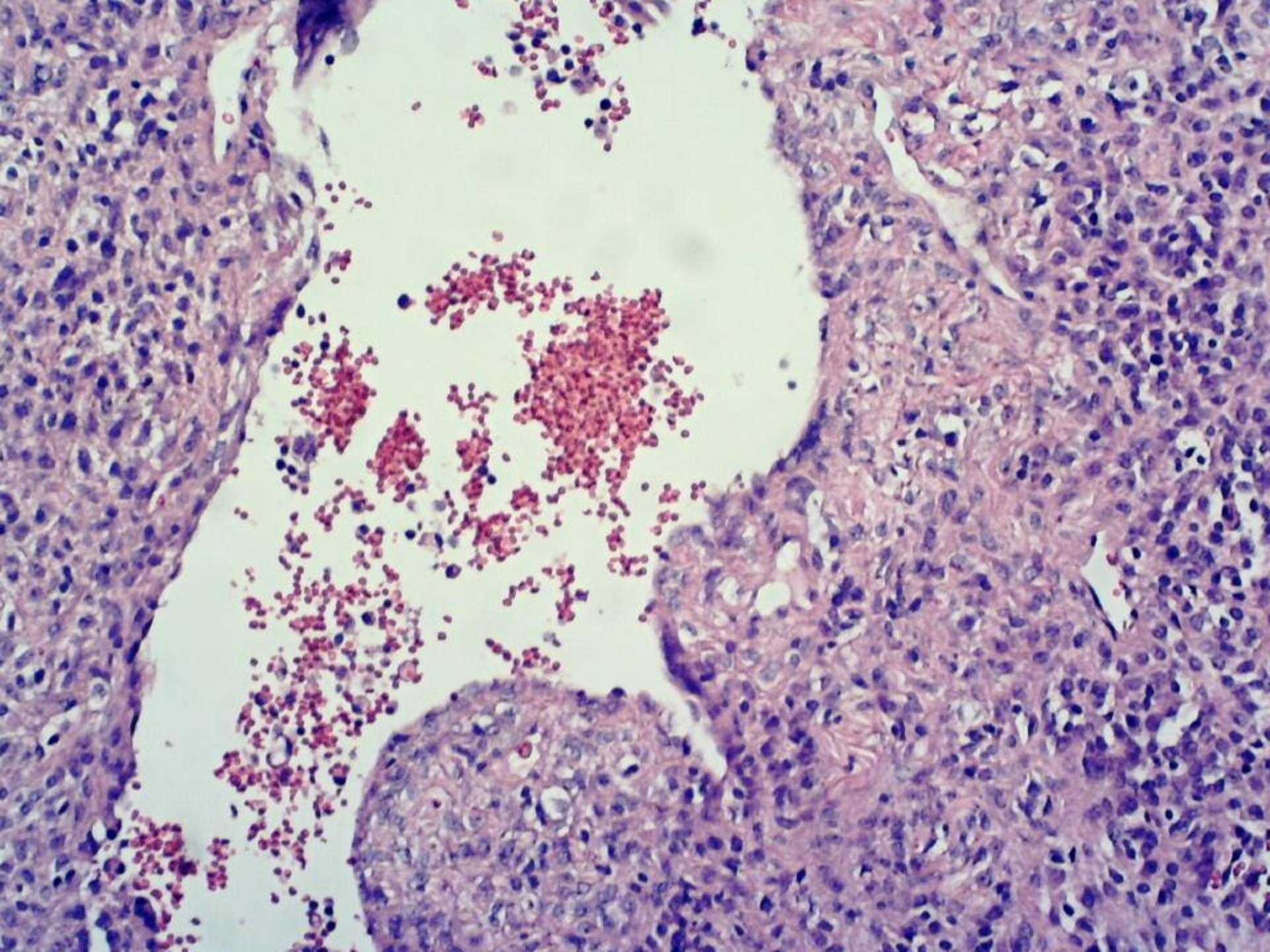
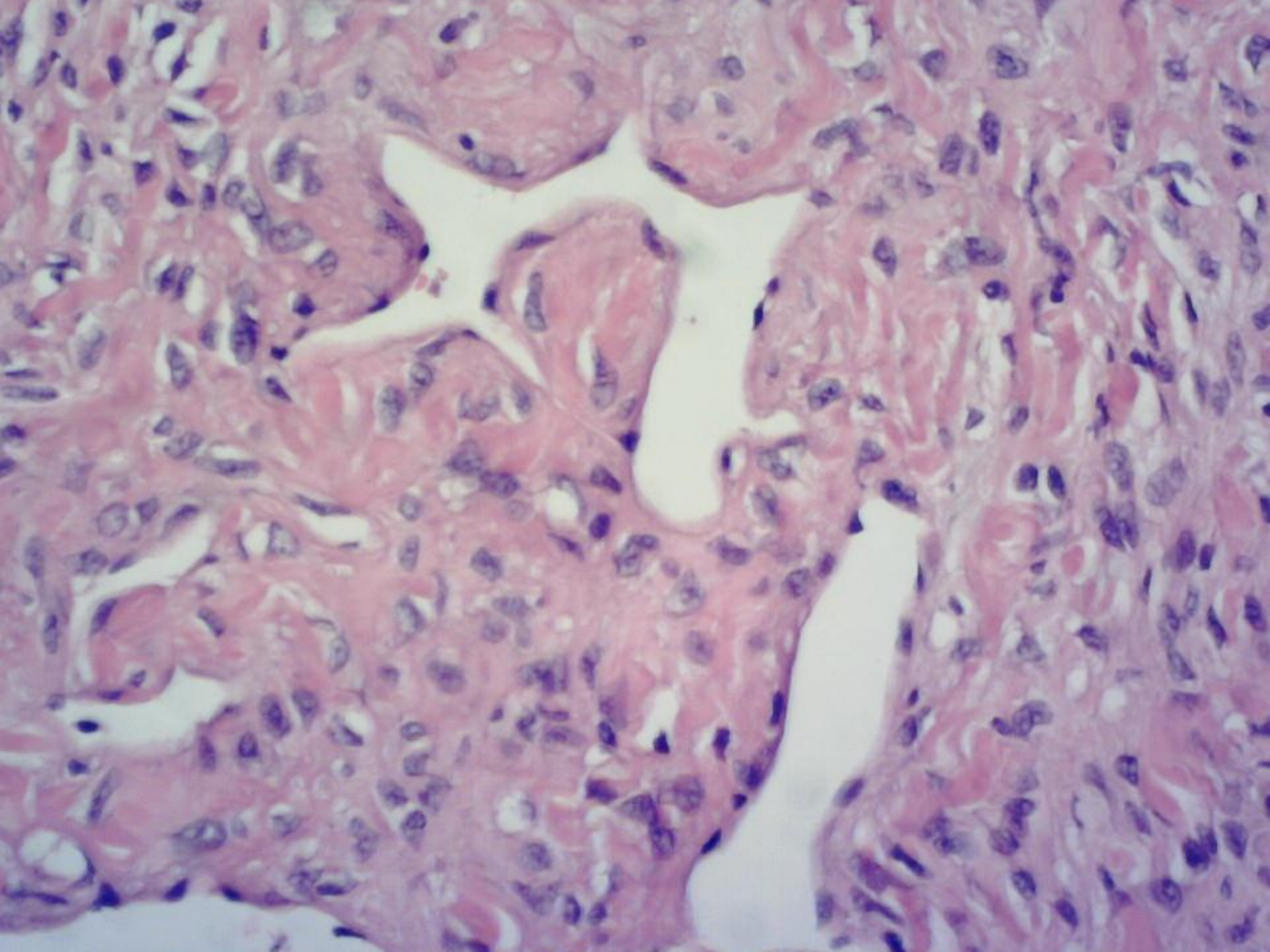


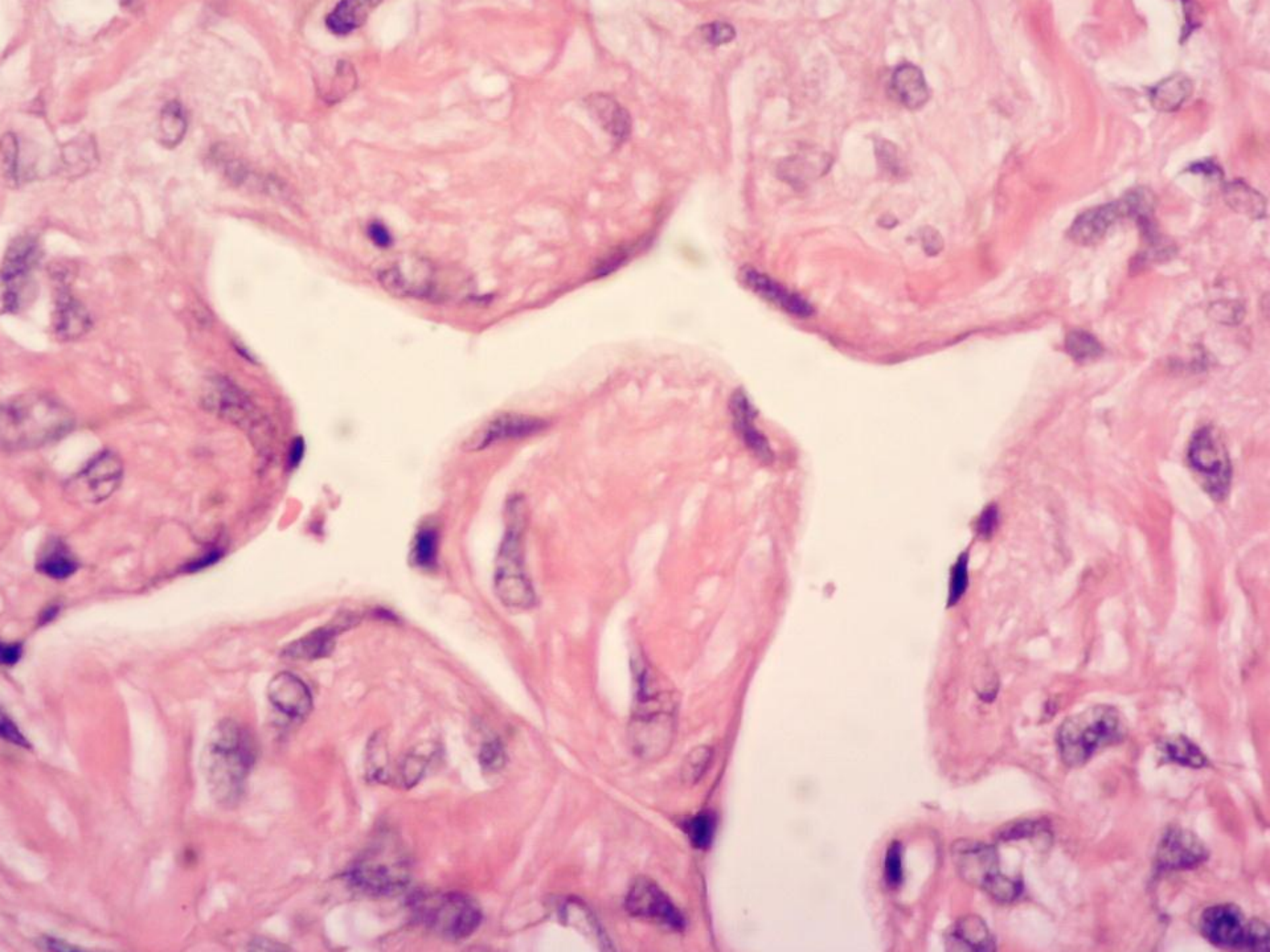


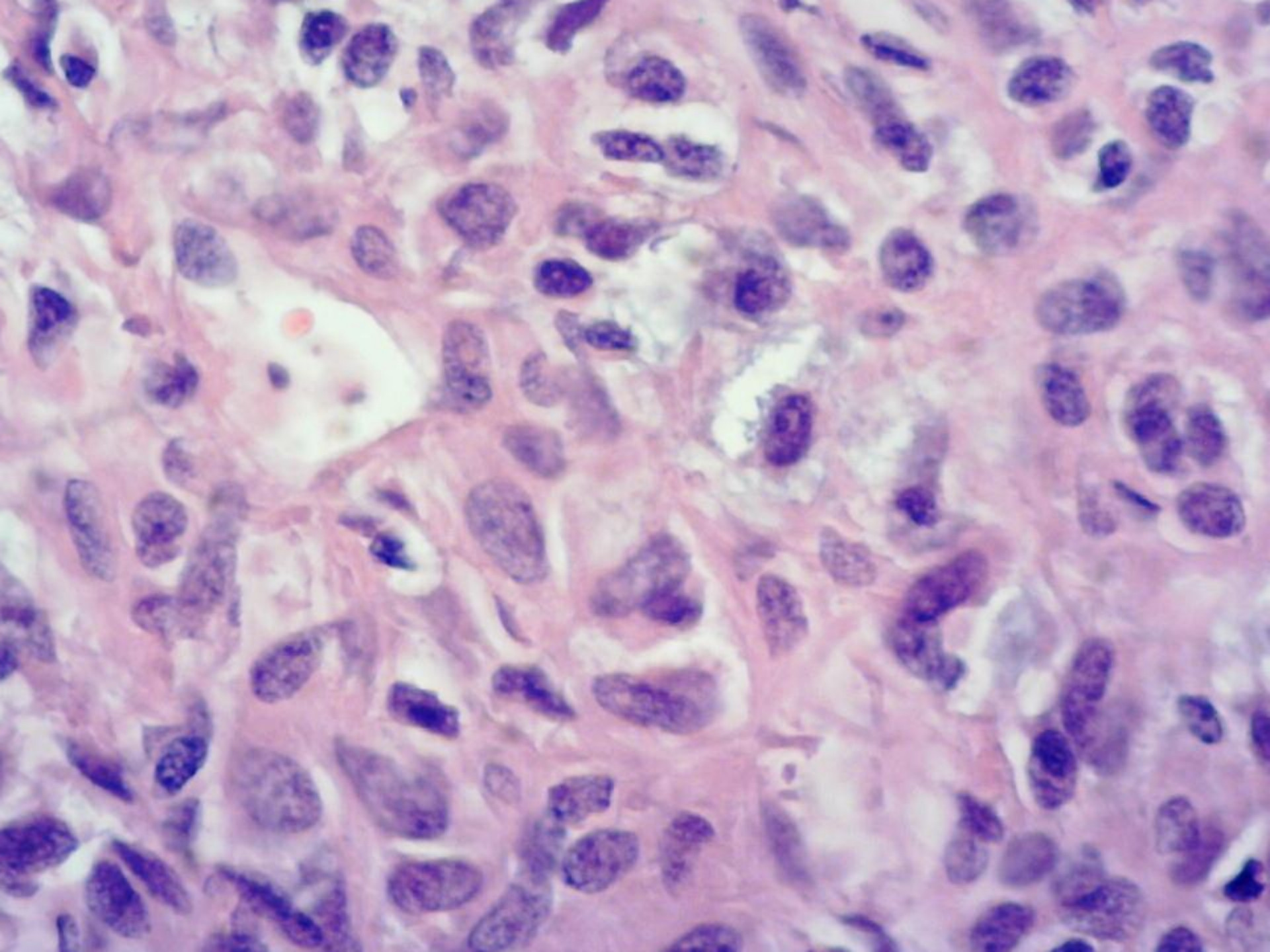


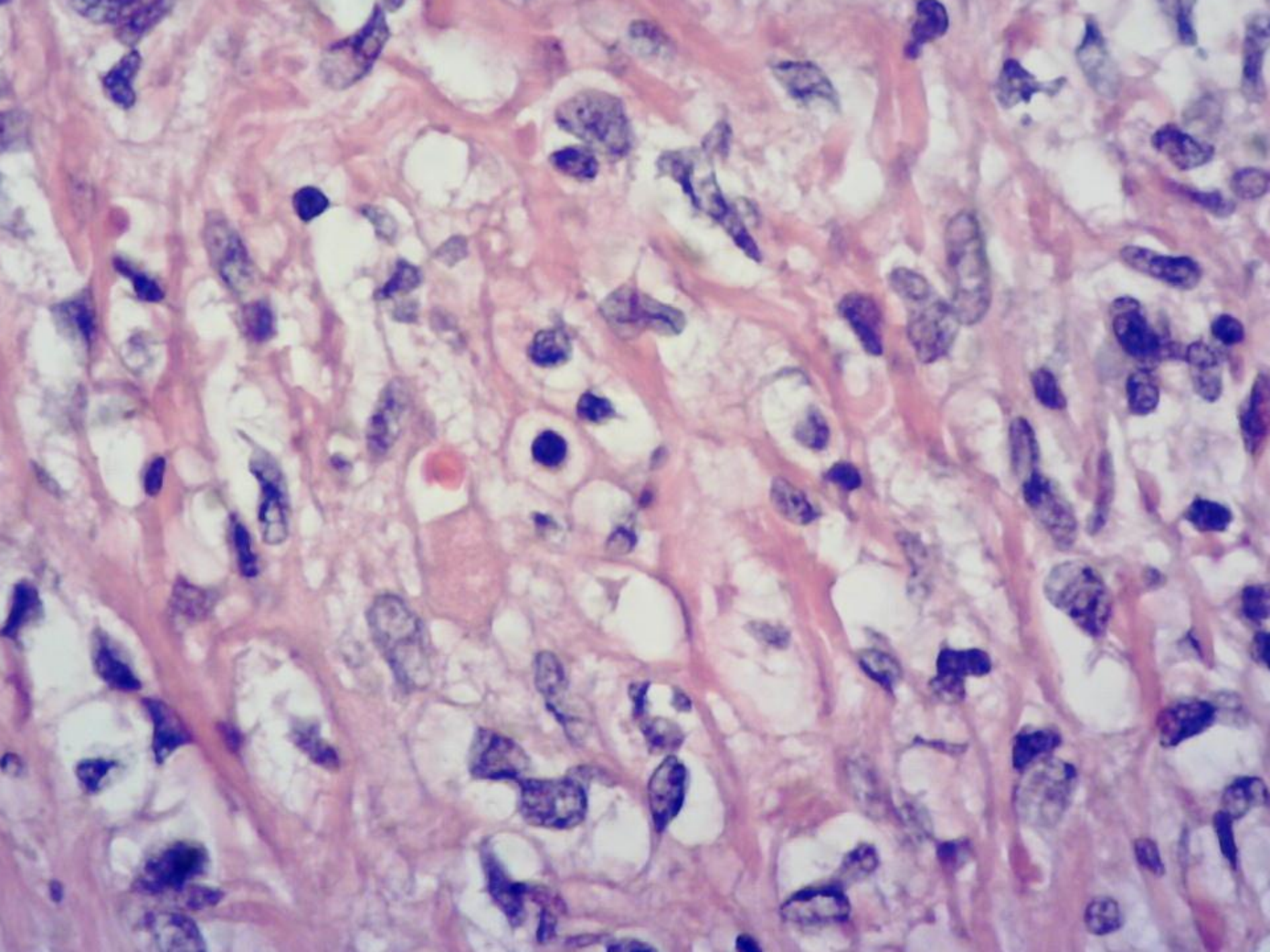


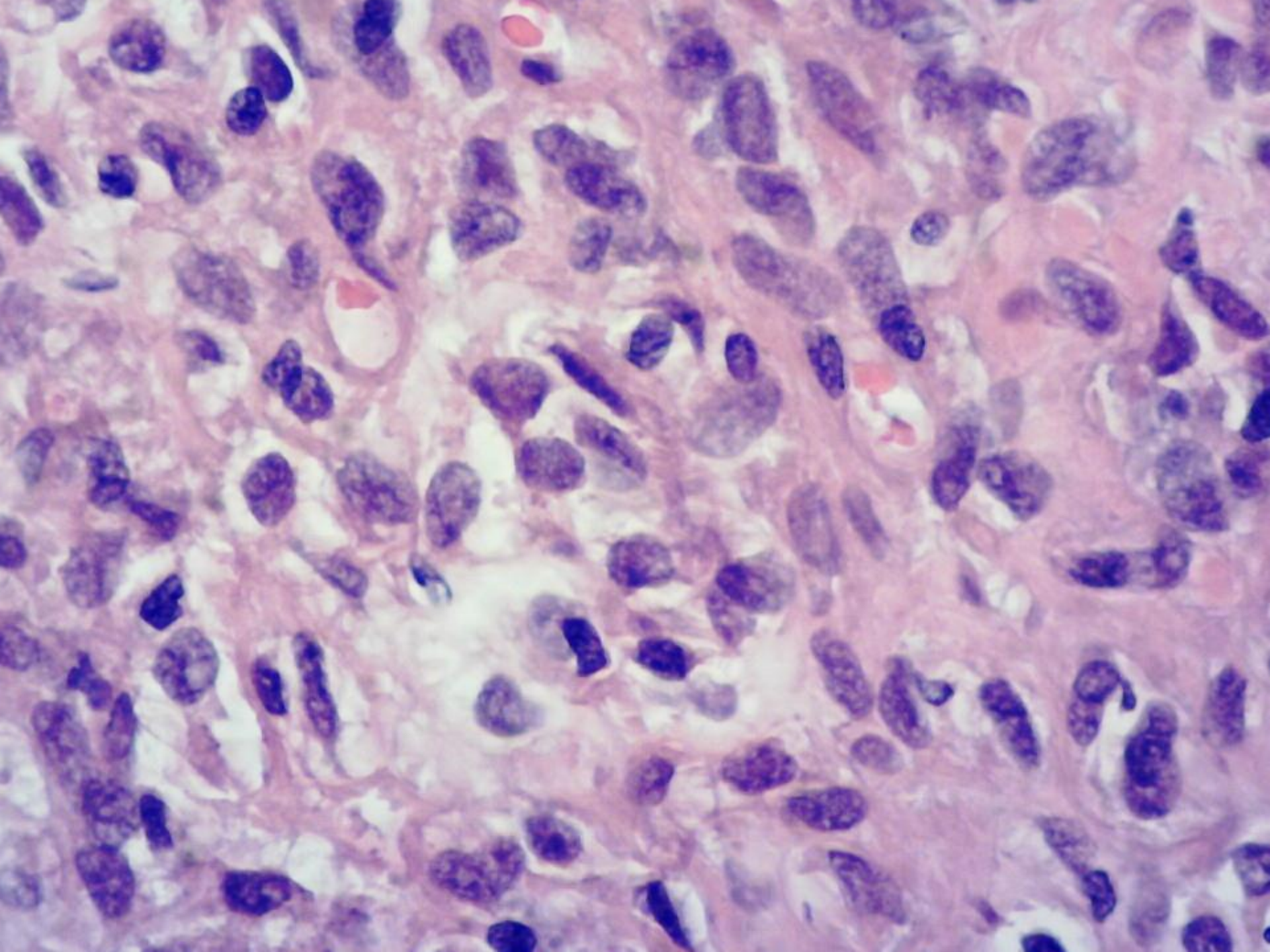












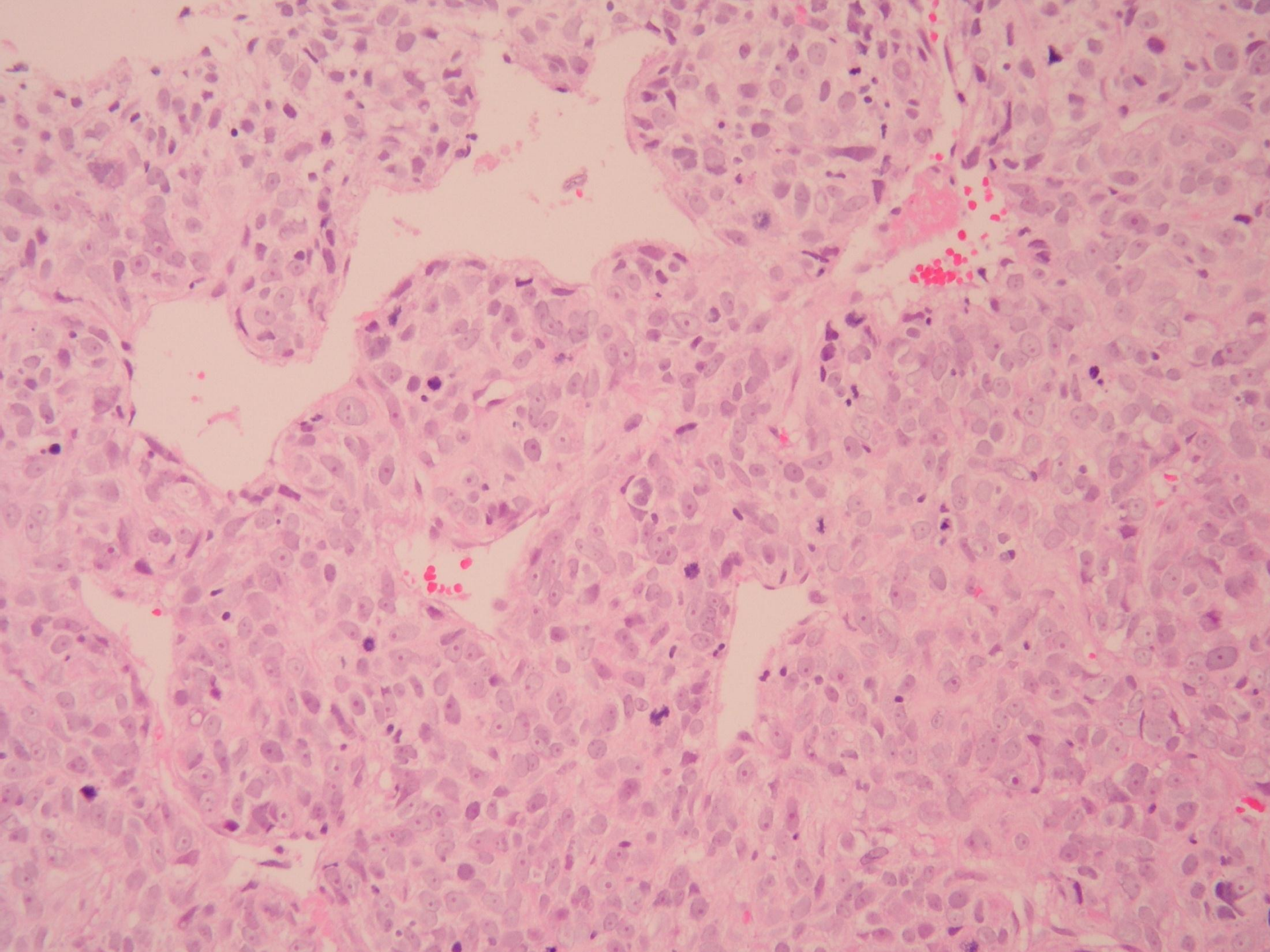
Soft tissue, neck, posterior, wide resection, solitary fibrous tumor.

The specimen submitted consists of many tissue fragments measuring up to 5x4x2.8 cm in size fixed in formalin.

Grossly,a light tan and well defined mass surrounded by muscle is seen. The cut surface is homogeneous and firm.

Representative sections are taken and A1-4 and B-E

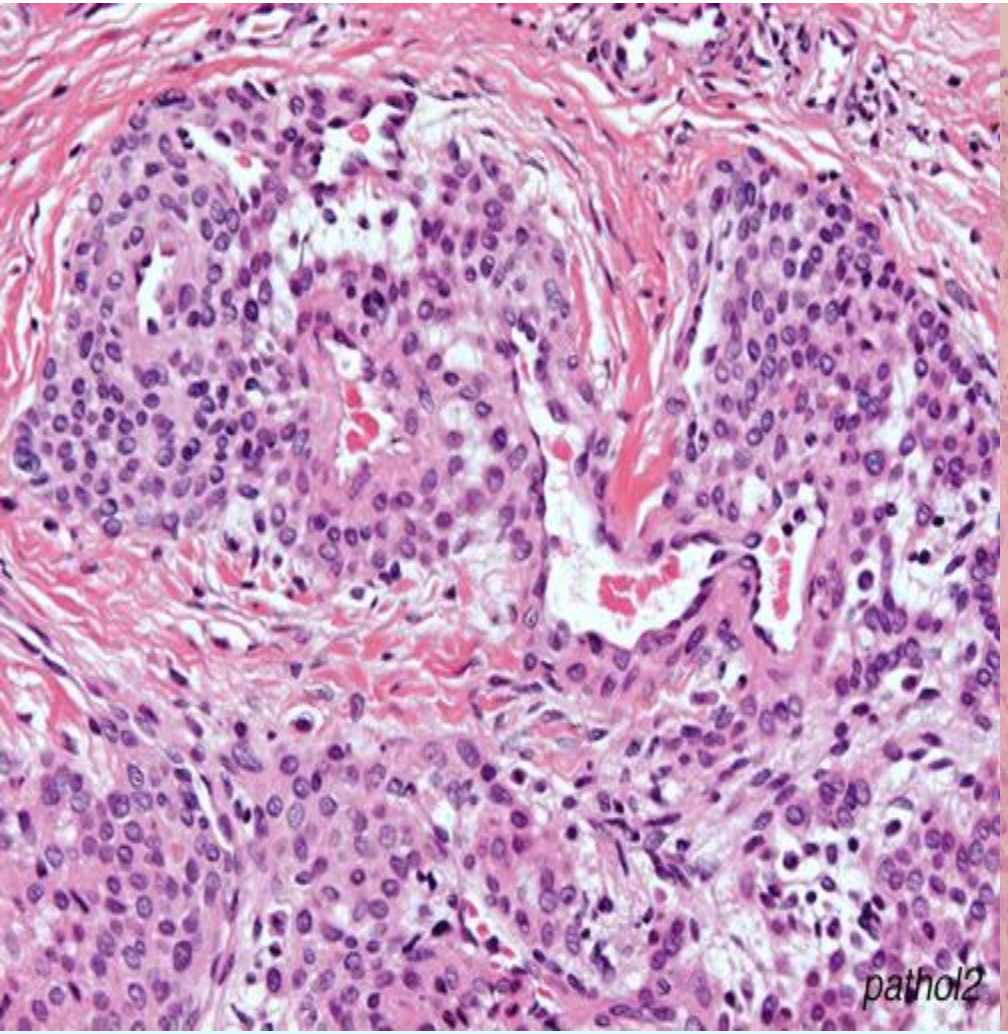
Microscopically, oval to spindl-shaped cells with concentric perivascular growth and exhibiting a characteristic well-developed staghorn branching vascular pattern are seen. Immunoreactivity for Bcl-2 and CD34, while no reactivity for S-100,SMA,EMA and desmin.MIB-1 is less than 5%.A solitary fibrous tumor is considerd.

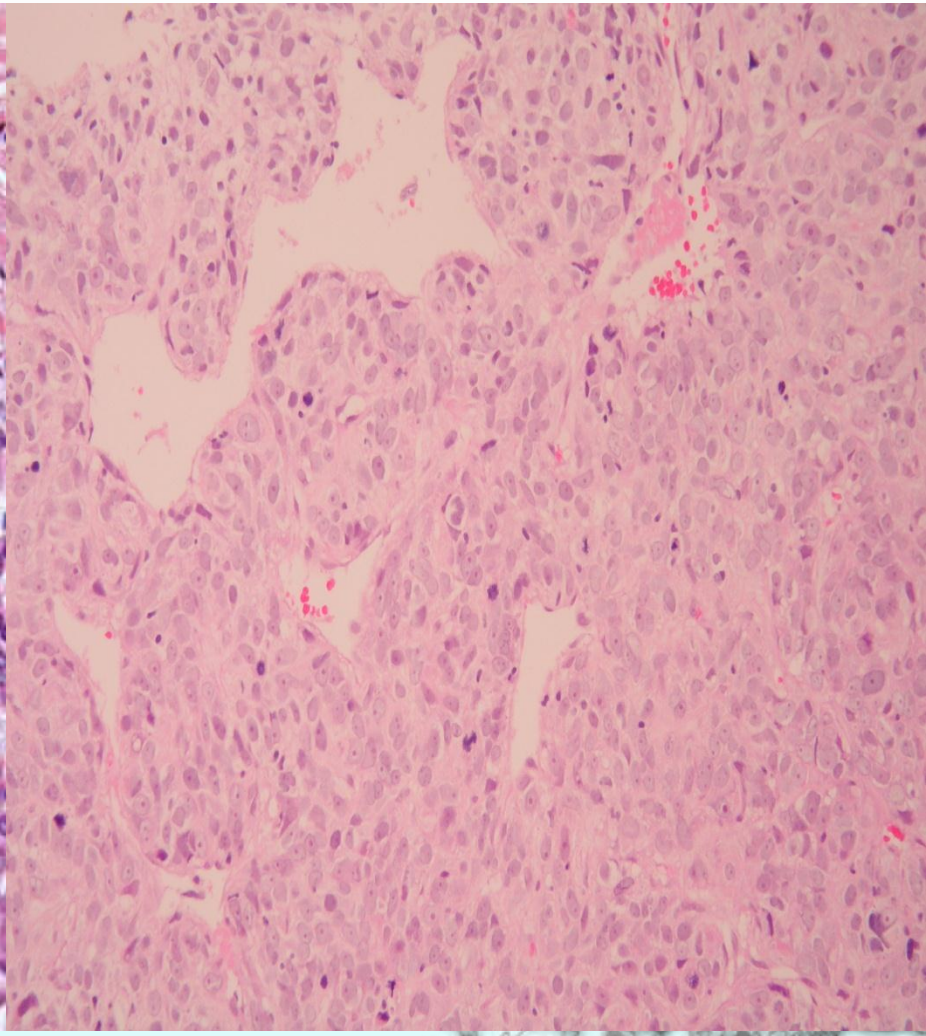


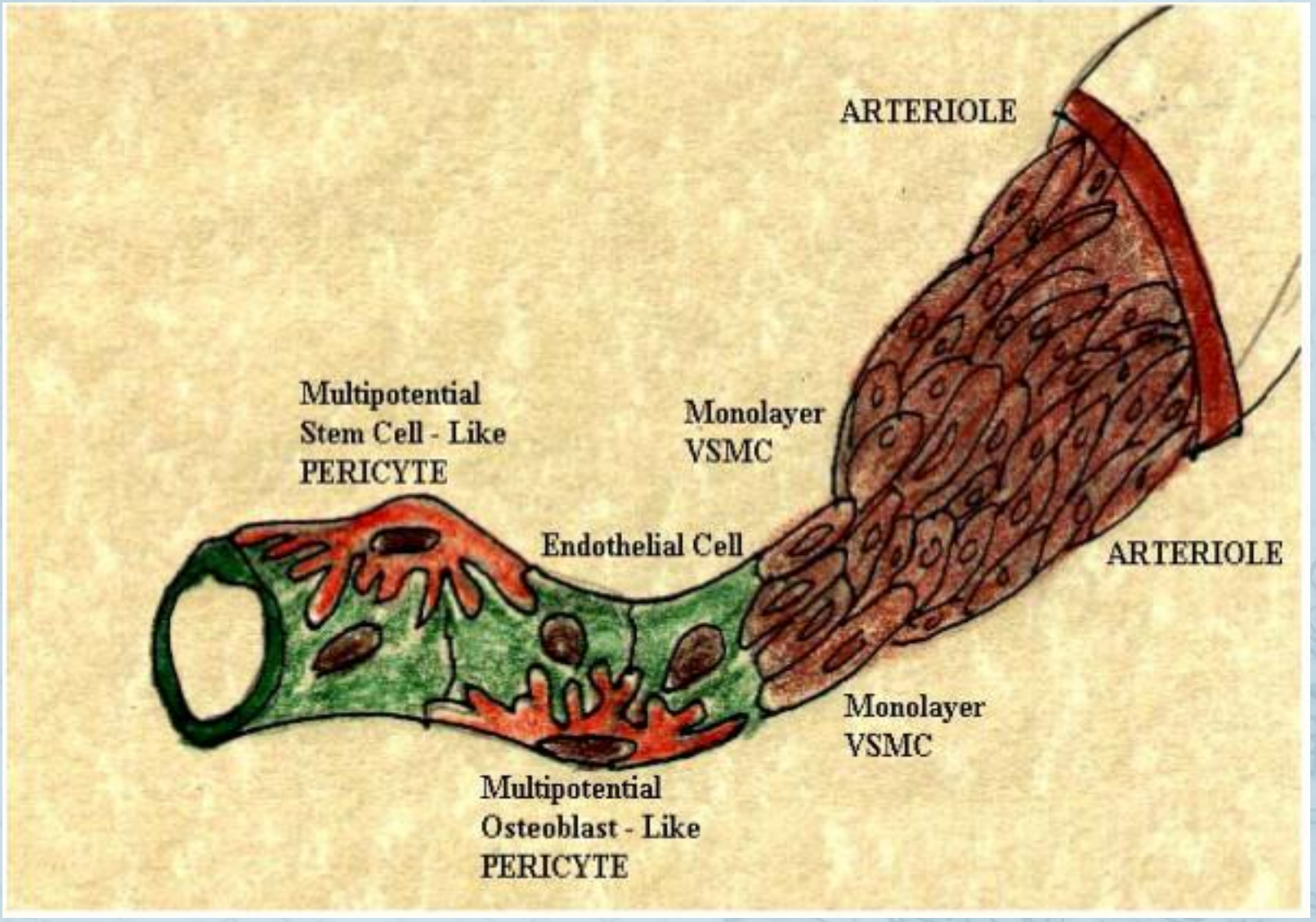
Perivascular Neoplasms

Perivascular neoplasms comprise traditionally glomus tumor and hemangiopericytoma (HPC). Whereas glomus tumor represents a well-defined entity, the existence of HPC as a separate entity has been questioned because a number of neoplasms of different lines of differentiation are characterized by a HPC-like vascular growth pattern.

Armed Forces Institute of Pathology, 2001:371-385


pathol2




ARTERIOLE
Multipotential
Stem Cell - Like
PERICYTE
Monolayer
VSMC
Endothelial Cell
ARTERIOLE
Monolayer
VSMC
Multipotential
Osteoblast - Like
PERICYTE

Pericytes in the Microvasculature

It has been speculated that pericytes represent pluripotent cells that may differentiate into smooth muscle cells, adipocytes, and osteoblasts.

Cardiovasc Res. 1996;32:687-698.

Perivascular Neoplasms

Glomus tumor
Hemangiopericytoma (HPC)
- glomangiopericytoma
- myopericytoma
- solitary fibrous tumour
- mesenchymal chondrosarcoma
- monophasic synovial sarcoma
- endometrial stromal sarcoma

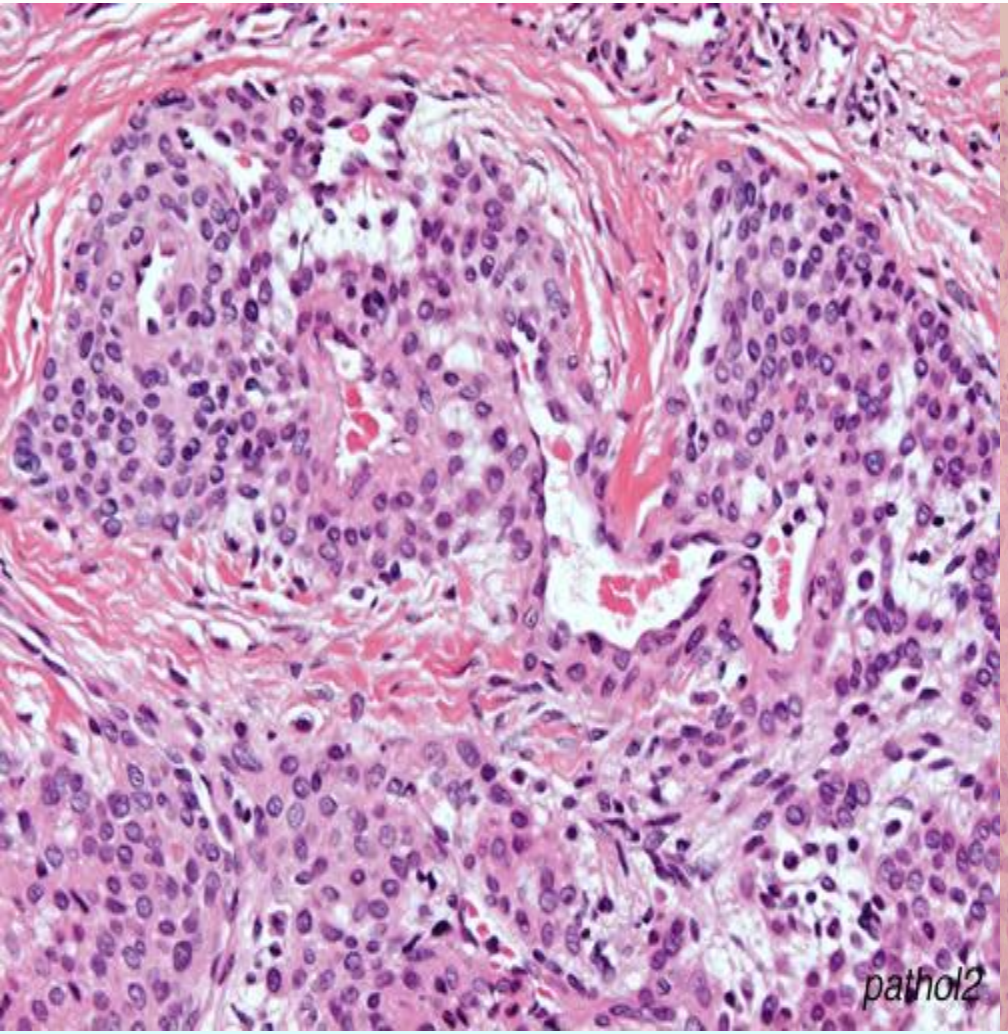
Glomus tumour

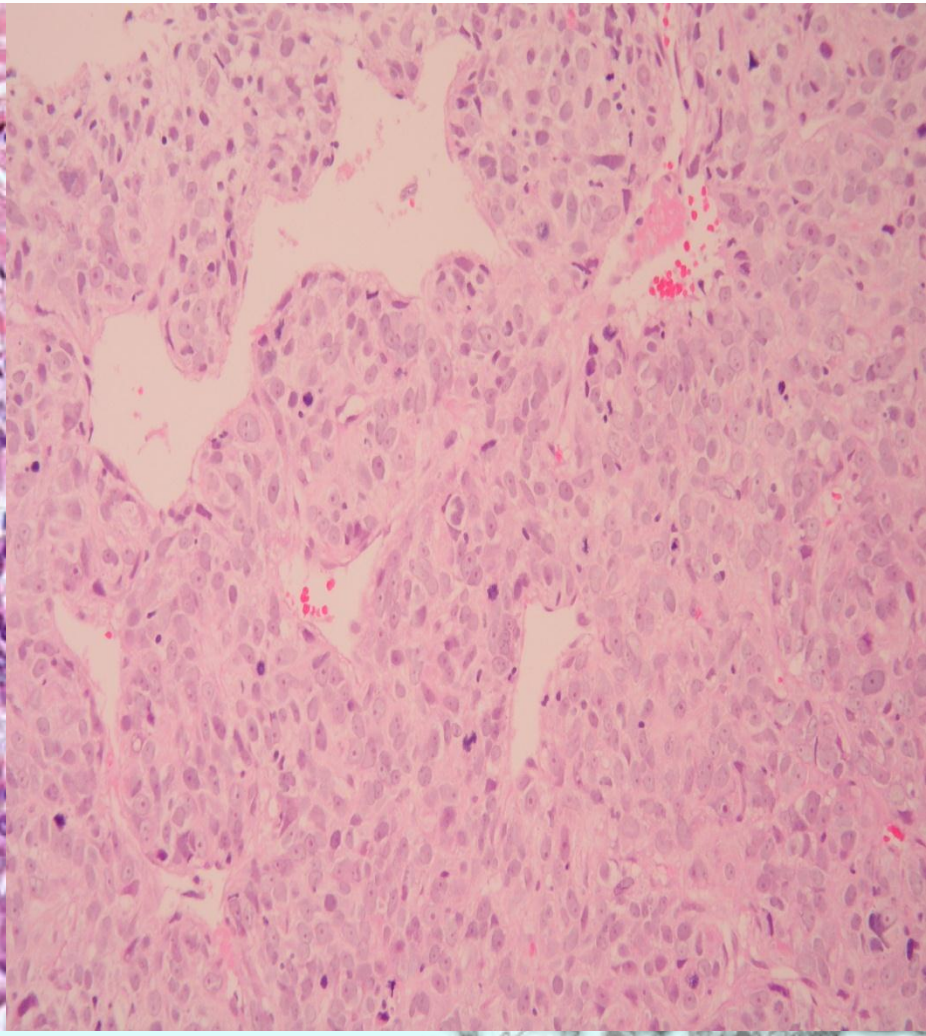
Immunohistochemistry

Smooth muscle actin	40/40[1], 31/34[2]
Desmin	2/32[1], 0/34[2]
Calponin	5/11[1], 25/30[2]
h-Caldesmon	7/12[1], 27/31[2]
Collagen type IV (investing individual cells)	8/8[1], 31/34[2]
Vimentin	14/14[1], 34/34[2]
Laminin	30/33[2]
Cytokeratin	0/34[1]
Cytokeratin 18	0/34[2]

Glomus tumour

S-100	0/34[1], 0/34[2]
CD34	5/27[1], 31/34[2]
CD117	0/30[2]
chromogranin	0/30[2]
synaptophysin	0/30[2]
CD20	0/34[2]
CD45 (LCA)	0/34[2]
HMB-45	never


pathol2



Myopericytoma

Immunohistochemistry

SMA	47/47[2]
h-caldesmon	29/32[2]
Desmin	3/33[2]
S-100	negative[2]
MNF116	negative[2]
AE1/AE3	negative[2]
EMA	negative[2]

Glomangiopericytoma

When the lesional cells in a myopericytoma show morphological differentiation towards glomus cells, by having distinct cytoplasmic borders and central round nuclei, the term glomangiopericytoma has been proposed.

Am. J. Surg. Pathol. **1998; 22; 513–525**

Toxicol Pathol OnlineFirst, published on July 22, 2008 as doi:10.1177/0192623308320804

TABLE 1.—Differential diagnosis of perivascular tumors.

Tumor	Myopericytoma	Hemangiopericytoma	Myofibroma	Glomus tumors	Smooth muscle tumors[a]
Diagnostic feature	Concentric perivascular growth composed of oval to spindle-shaped cells	Branching vascular spaces surrounded by round to spindle-shaped cells	Biphasic growth composed with myofibroblastic fascicles and hemangiopericytoma-like appearance	Perivascular growth composed of cuboidal cells with abundant cytoplasms and distinct cell borders	Fascicular growth composed of elongated spindle-shaped cells with cigar-shaped nuclei
Immunohistochemistry					
aSMA	+	−	+	+	+
Desmin	−/+	−	−/+	−	+

a. Including angioleiomyomatous tumor and leiomyoma/leiomyosarcoma.

‘Haemangiopericytoma'

The entity 'haemangiopericytoma' encompassed several different tumour types that shared a common non-distinctive architectural pattern. From the myofibromatosis or myopericytoma,

Many of the reported cases of metastatic haemangiopericytoma probably represent cases of monophasic synovial sarcoma, solitary fibrous tumour, mesenchymal chondrosarcoma and low-grade endometrial stromal sarcoma.

Histopathology. 41(5):450-460, November 2002.

Perivascular Neoplasms'

Glomus tumor
Hemangiopericytoma (HPC)
- glomangiopericytoma
- myopericytoma
- solitary fibrous tumour
- mesenchymal chondrosarcoma
- monophasic synovial sarcoma
- endometrial stromal sarcoma

Solitary fibrous tumour and haemangiopericytoma: evolution of a concept

In practice, any HPC-like lesion can be allocated to one of these categories, leaving the ill-defined 'haemangiopericytoma' category empty.

Histopathology. 48(1):63-74, January 2006.

Epidemiology

Most cases occur in adults with a median age of 45-50 years. It is less common in infants and children. Soft-tissue solitary fibrous tumors represent only about 1-2% of all soft-tissue tumors.

Sarcoma. 2012;2012:690251

Solitary fibrous tumor: A pathological enigma and clinical dilemma

Solitary fibrous tumors are ubiquitous rare spindle cell neoplasms, most commonly arising from the pleura..

In 1931 Klemperer and Rabin first documented the occurrence of a distinctive localized pleural based tumour and proposed a submesothelial cell origin.

Later, based on tissue culture experiments, Stout and Murray claimed derivation from mesothelial cells. This controversy is reflected in the variety of synonyms used for solitary fibrous tumors in the past including localized fibrous tumor, localized fibrous mesothelioma, solitary fibrous mesothelioma, fibrous mesothelioma, subserosal fibroma and submesothelial fibroma.

With the advent of immunohistochemistry a fibroblastic origin, occasionally with myofibroblastic differentiation, is firmly established. This is further reinforced by the description of solitary fibrous tumors in extrathoracic sites devoid of mesothelial cells. Whilst now considered to be derived from mesenchymal cells, the histiogenesis has been the subject of debate

Arch Pathol 1931;11:385-412.
Arch Pathol 1942;34:951-64.

Solitary fibrous tumour

Solitary fibrous tumor was first described in 1870 by Wagner and further established in 1931 by Klemperer and Rabin as a pleural neoplasm.

Three classical clinical forms of this entity are recognized:

- Pleural solitary fibrous tumor
- Soft-tissue solitary fibrous tumor
- Meningeal HPC.

Histopathologic spectrum:

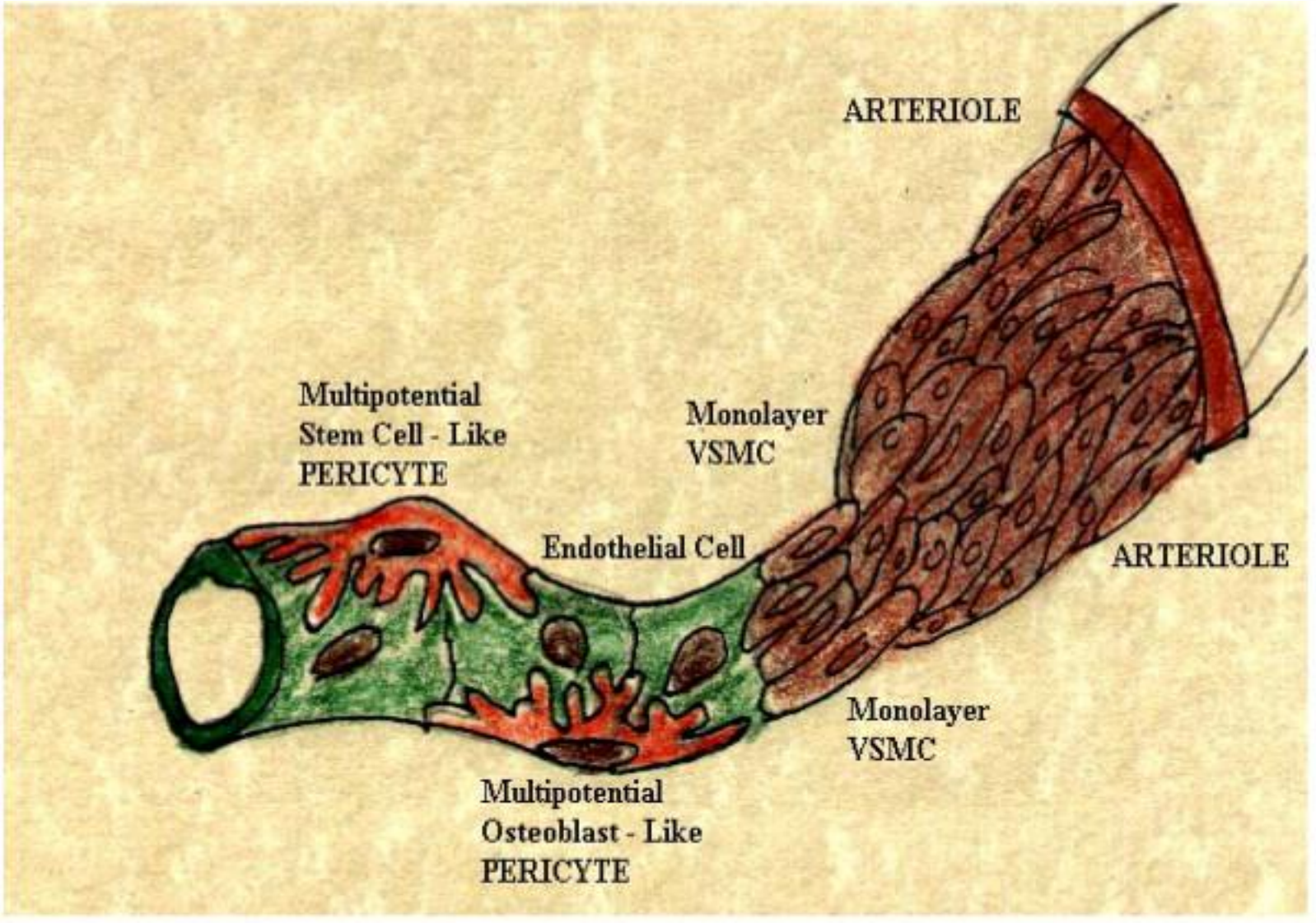
- On one end, a fibrous form is characterized by hyalinized, thick-walled vessels with opened lumina and strong CD34 reactivity.
- On the other end, a cellular form, representing the conventional HPC, has branching, thin-walled vessels and focal or absent CD34 reactivity.

Histopathology. Jan 2006;48(1):63-74.
Sarcoma. 2012;2012:69025
Cancer. Oct 15 2002;95(8):1746-51

Solitary fibrous tumor: A pathological enigma and clinical dilemma

Microscopically solitary fibrous tumors are characterised by hypocellular collagen rich areas alternating with a proliferation of uniform elongated spindled cells in a haphazard distribution. Imunohistochemistry is extremely useful in establishing the diagnosis, no more so than CD34. CD34 is a myeloid progenitor cell antigen which is also positive in endothelial cells and some mesenchymal cells, including subsets of fibroblasts. It is no coincidence that since the description of CD34 expression in solitary fibrous tumors there has been a flurry of case reports in a wide range of sites.

Human Path 1995;26:440-9.


ARTERIOLE
Multipotential
Stem Cell - Like
PERICYTE
Monolayer
VSMC
Endothelial Cell
ARTERIOLE
Monolayer
VSMC
Multipotential
Osteoblast - Like
PERICYTE

Solitary fibrous tumour and haemangiopericytoma: evolution of a concept

Immunohistochemically, SFTs, especially the fibrous form, commonly express CD34 (80–90% of cases) and CD99 (70%). Bcl-2 (30%), epithelial membrane antigen (EMA) (30%) and smooth muscle actin (SMA) (20%) may occasionally be expressed. They are usually negative for S100 protein, desmin and cytokeratins. Cellular forms of SFT tend to be less frequently positive for CD34

Histopathology. 48(1):63-74, January 2006.

Frequent Expression of bcl-2 Protein in Solitary Fibrous Tumors

The distinction of solitary fibrous tumors from histologically similar neoplasms is often difficult because they rarely occur at a variety of extrapleural sites. CD34 immunoreactivity has recently been recognized to be an adjunct for the diagnosis of solitary fibrous tumors. However, it is now known that CD34 staining is not entirely specific for this entity. We evaluated 23 solitary fibrous tumors and 54 other spindle cell tumors often considered in the differential diagnosis for immunoreactivity using monoclonal antibodies directed against bcl-2 protein, which protects cells from apoptosis and CD34.

Jpn J Clin Oncol 1998;28:86-91

Frequent Expression of bcl-2 Protein in Solitary Fibrous Tumors

The patients with solitary fibrous tumors comprised 11 men and 12 women, ranging in age from 35 to 85 years (mean, 57.6 years). Fourteen tumors arose in the pleura, four in the retroperitoneum, three in the superficial soft tissue and one each in the mediastinum and uterine cervix. Nineteen of 23 solitary fibrous tumors (83%), irrespective of tumor site, demonstrated diffuse cytoplasmic staining for bcl-2 protein, bcl-2 immunoreactivity was also observed in five of seven neurofibromas (71%), eight of 10 synovial sarcomas (80%) and one of three spindle cell lipomas (33%). CD34 immunoreactivity was present in all but one solitary fibrous tumor (96%), seven of seven neurofibromas (100%), three of three spindle cell lipomas (100%), five of five dermatofibrosarcomas (100%), three of three hemangiopericytomas (100%) and two of seven malignant fibrous histiocytomas (29%). To date, most of the pleural and extrapleural cases have not shown aggressive features. We suggest that bcl-2 protein can be used together with CD34 in the diagnosis of solitary fibrous tumor to distinguish this entity from other spindle cell neoplasms.

Jpn J Clin Oncol 1998;28:86-91

Extrapleural solitary fibrous tumour

Immunohistochemistry

CD34	95% (78/82)[1], 12/12[21], 7/9[22], 16/16[18], 3/3[25]
bcl-2	96% (72/75)[1], 12/12[18]
CD99	31/31[1], 12/12[21], 8/12[18]
vimentin	19/19[10], 12/12[21], 13/13[18], 3/3[25]
Factor XIIIA	10/10[18]
muscle-specific actin	0/5[10]
desmin	0/5[10]
SMA	2/12[18]
S-100	0/10[10]
CD68	0/10[10]
cytokeratin	0/19[10], 0/5[10]

Solitary (localised) fibrous tumour

Immunohistochemistry

Summating results from various papers, positivity is seen in[1]:

	pleural	extra-pleural
CD34 (My10 may be more sensitive than QBEND10[8])	92% (196/213), 10/10[9]	95% (78/82)
bcl-2	96% (81/84)	96% (72/75)
CD99	10/11, 8/10[9]	31/31
CD10	5/5	
vimentin	91% (170/186)	19/19[2]
NSE	36% (10/28)	
muscle-specific actin	18% (12/66)	
desmin	12% (20/168): staining is focal	
SMA	4% (5/132): staining is focal	

Solitary (localised) fibrous tumour

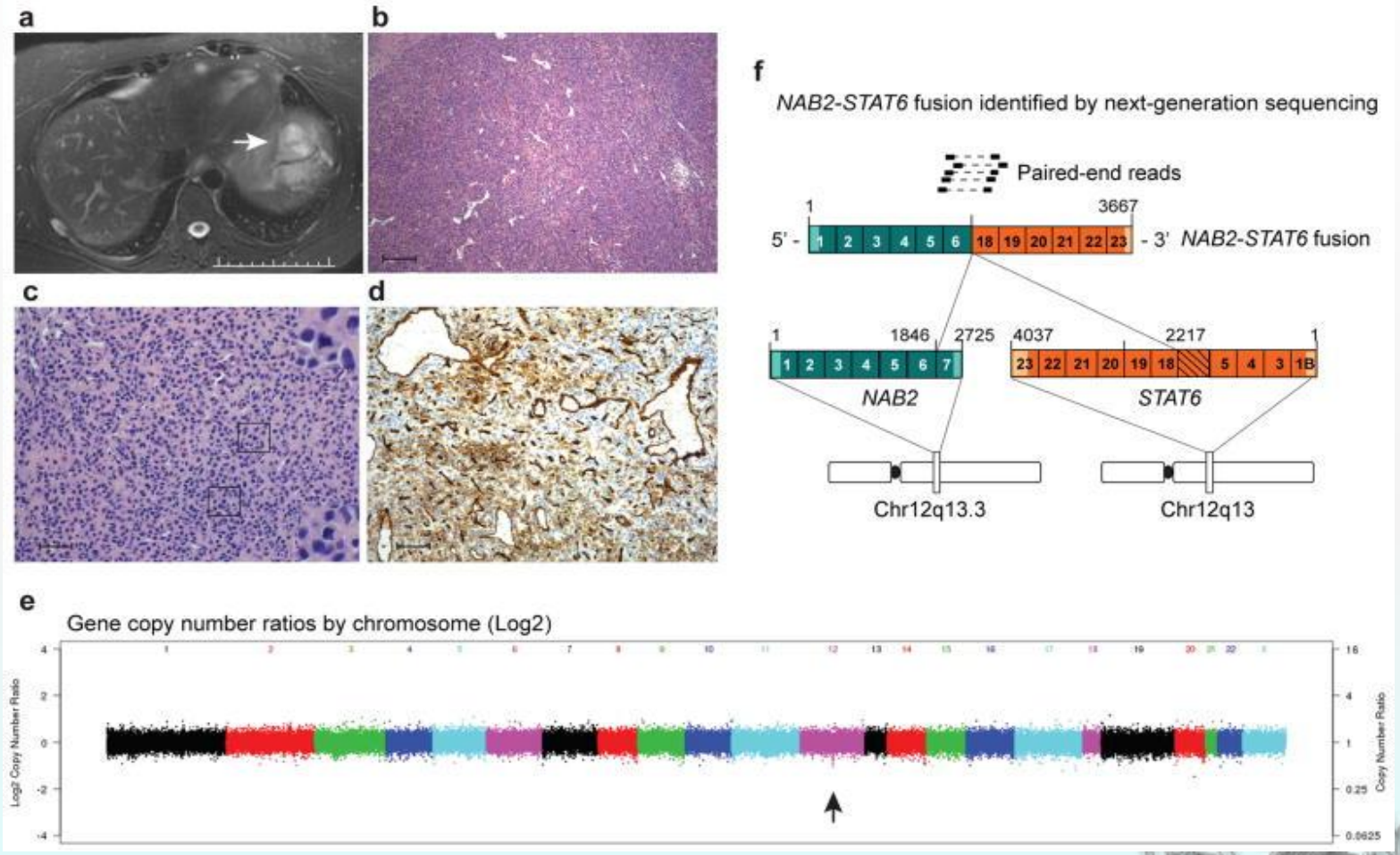
S-100	1% (1/107)	
cytokeratin	2% (4/248)	0/19[2]
EMA	negative[8]	
Calretinin	negative[8]	
neurofilament	0% (0/33)	

No positive staining has been reported for factor VIII-related antigen, EMA, CEA, alpha-1-antitrypsin or calretinin

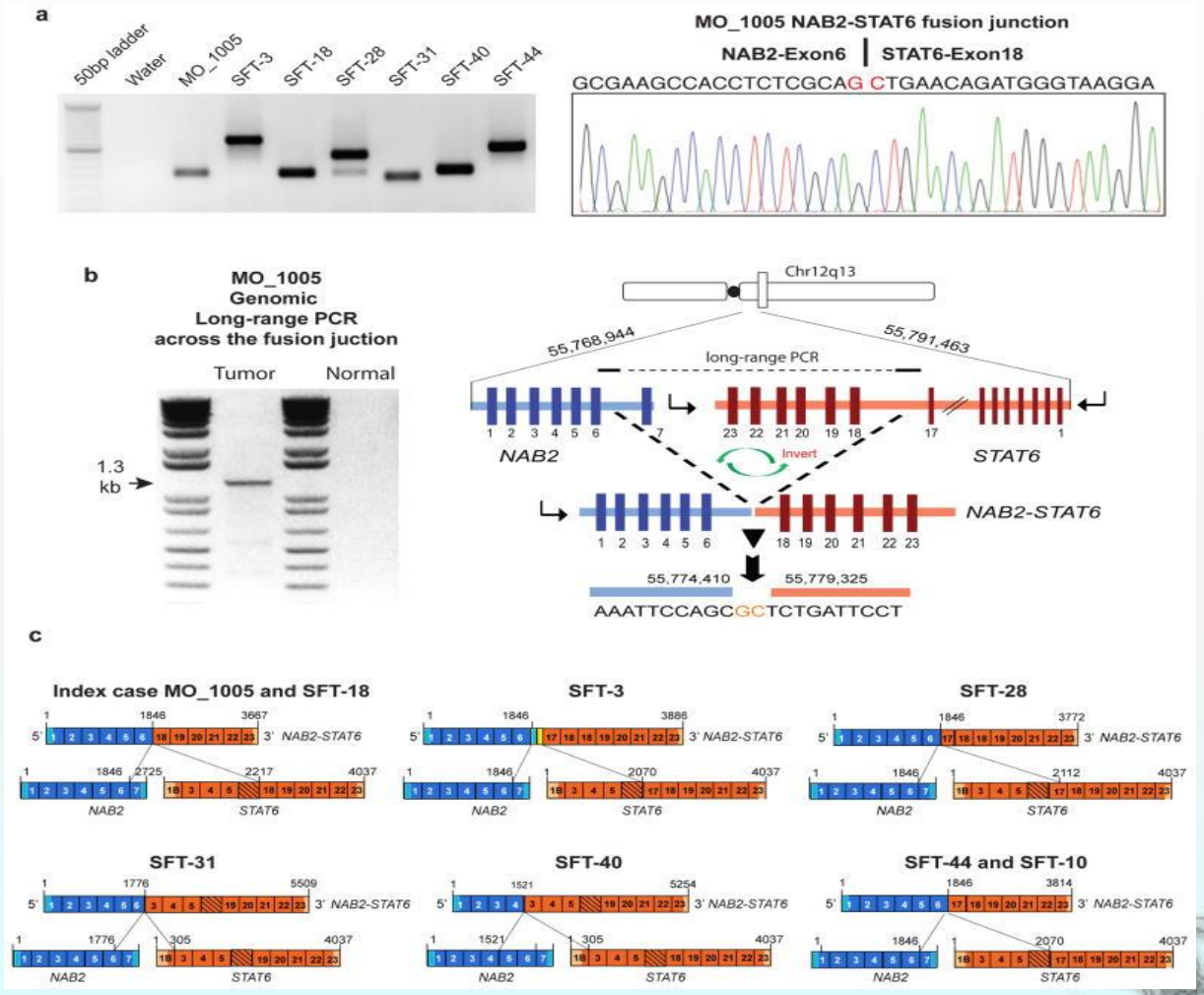
Molecular Biology

Recurrent somatic fusions of the two genes, NAB2 (NGFI-A–binding protein 2) and STAT6, located at chromosomal 12q13 region have been identified in solitary fibrous tumors.

Nat Genet. 2013 Feb; 45(2): 180–185.



Nat Genet. 2013 Feb; 45(2): 180–185.



Nat Genet. 2013 Feb; 45(2): 180–185.

Solitary fibrous tumor: A pathological enigma and clinical dilemma

England et al used high cellularity, mitotic activity (more than four mitotic figures per 10 high-power fields), pleomorphism, hemorrhage and necrosis as criteria for distinguishing tumors with a favourable course from those that have the propensity for recurrence, local invasion and metastatic spread. Unfortunately biological behaviour does not always correlate with atypical histological features.

Am J Surg Pathol 1989;13:640-58.

Atypical and malignant solitary fibrous tumors in extrathoracic locations: evidence of their comparability to intra-thoracic tumors.

Abstract

Solitary fibrous tumor (SFT), first described as a pleural lesion, has been reported at numerous extrathoracic sites over the past 10 years. About 10% to 15% of intrathoracic SFTs are histologically or clinically malignant, but such cases have very rarely been described at other locations.

Among 92 cases of extrathoracic SFT in our files, we identified 10 that either had recurred (2 cases) or had a least one atypical histologic feature (8 cases).

The ten tumors occurred in five men and five women, 32 to 81 years old (median 56), measured 1.9 cm to 20 cm (median 11.5 cm), and were located in the abdomen/pelvis (4 cases), retroperitoneum (3 cases), groin, trunk, and upper arm. Nuclear atypia (8 cases), markedly increased cellularity (6 cases), areas of necrosis (4 cases), and greater than 4 mitoses/10 HPFs (3 cases) were seen in addition to the typical histologic features of SFT. Six tumors had at least two of these atypical histologic features. Nine cases were positive for CD34, six were positive for O-13, and one was focally positive for smooth muscle actin. Eight were excised completely. Subsequent follow-up revealed tumor relapse in eight cases (follow up 6-180 months, median 24). Four patients had local recurrence at 12 to 168 months. Distant metastasis developed at 1 to 6 years in five cases with spread to lung (2 cases), liver (4 cases), and bone. Metastasis or local recurrence developed within 2 years in five patients. To date, no patient has died of their tumor. These findings demonstrate that nuclear atypia, hypercellularity, greater than 4 mitoses/10 HPFs, and necrosis may be seen in up to 10% of extrathoracic SFTs, and are associated with, but are not by themselves predictive of, aggressive clinical behavior. In addition, our findings confirm that the behavior of extrathoracic SFTs is unpredictable, entirely comparable to that of their better known pleural counterparts, and confirm that patients with SFTs in all locations require careful, long-term follow up. It is probably unwise to regard any such lesion as definitely benign.

Am J Surg Pathol. 1998 Dec;22(12):1501-11

Solitary fibrous tumor: A pathological enigma and clinical dilemma

De Perrot et al stratified the risk of recurrence based on histologic and morphologic indicators among 185 reported solitary fibrous tumors of the pleura. Recurrence was observed in 63% of all patients presenting with a malignant sessile lesion but 2% of the patients with a benign pedunculated tumor recurred. The proliferation marker Ki67 has been used to stratify lesions as to their clinical outcome. Positive staining is greater in malignant versus benign tumors but the overlap limits its usefulness. Clearly whilst the site, growth pattern and histological features correctly identify the malignant potential in the majority of cases, there still remains a small subset which behaves in an unpredictable fashion.

Ann Thorac Surg 2002;74:285-93.

Jpn J Clin Oncol 1998;28:86-91.

Soft tissue, neck, posterior, wide resection, solitary fibrous tumor.

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Grossly,a light tan and well defined mass surrounded by muscle is seen. The cut surface is homogeneous and firm.

Representative sections are taken and A1-4 and B-E

Microscopically, oval to spindl-shaped cells with concentric perivascular growth and exhibiting a characteristic well-developed staghorn branching vascular pattern are seen. Immunoreactivity for Bcl-2 and CD34, while no reactivity for S-100,SMA,EMA and desmin.MIB-1 is less than 5%.A solitary fibrous tumor is considerd.

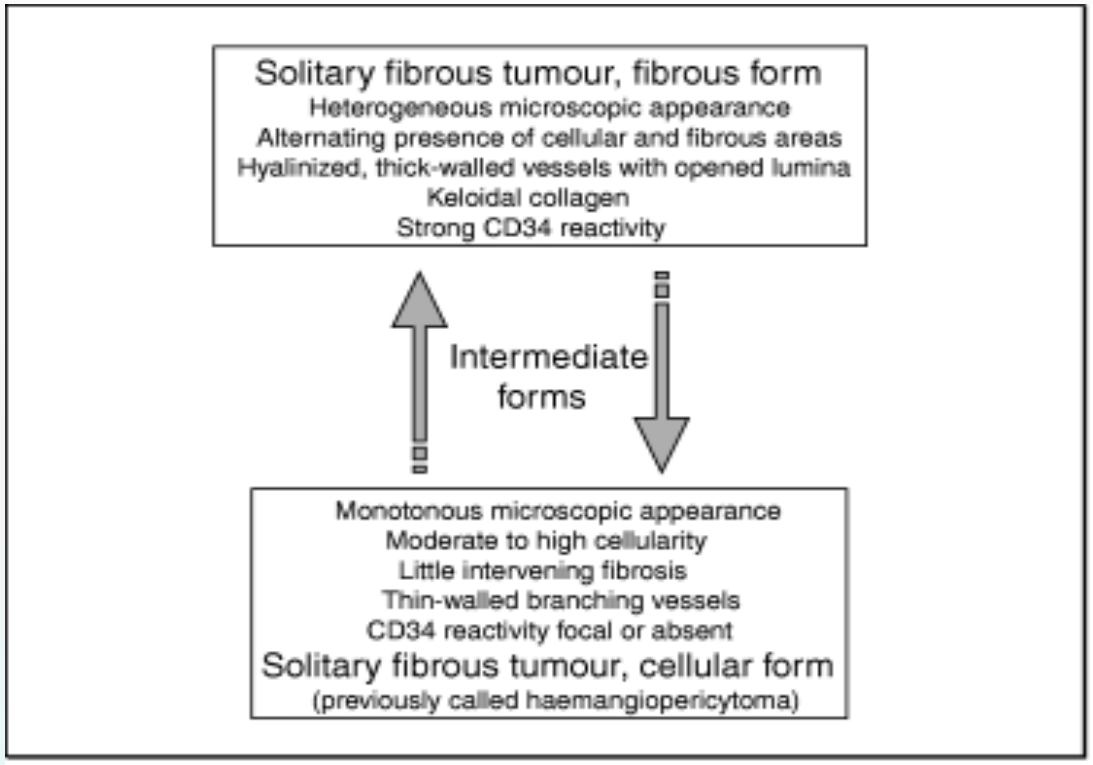
Solitary fibrous tumor: A pathological enigma and clinical dilemma

Most solitary fibrous tumors behave in a benign fashion. When arising from the pleura, 13-23% are classified as malignant in con-

trast to most extrapleural tumors which, with the exception of those of mediastinal origin, have a benign outcome

Histopathology 1997;31:568-76.

Solitary fibrous tumour



Histopathology. 48(1):63-74, January 2006.

Take-home messages

Solitary fibrous tumors are tumors of mesenchymal origin that occur often in the extremities.

The behavior of extrathoracic SFTs is unpredictable, entirely comparable to that of their better known pleural counterparts, and confirm that patients with SFTs in all locations require careful, long-term follow up. It is probably unwise to regard any such lesion as definitely benign.

This tumor typically spreads via hematogenous dissemination, primarily to the lungs, but rarely spreads via the lymphatics.

Metastatic disease is usually the cause of death.

Conventional hemangiopericytoma: modern analysis of outcome.

METHODS: Between July 1982 and February 1998, 62 patients with a diagnosis of primary, recurrent, or metastatic HPC were identified from a prospectively maintained database. The pathology of all cases for which material was available (57 cases) was re-reviewed for histologic confirmation of the HPC diagnosis. Using strict pathologic criteria, including immunohistochemistry and electron microscopy, tumors from 25 of 57 patients qualified for the diagnosis of conventional hemangiopericytoma; those tumors formed the basis of the current report. Survival was determined by the Kaplan-Meier method.

RESULTS: At the time of initial presentation, 19 patients had primary tumors, 3 had locally recurrent disease, and 3 had metastatic disease. The most frequent anatomic sites for HPC were the extremities, the pelvis, and the head and neck, accounting for 80% of the total cases. The median followup (n = 25) was 49 months (range, 1 to 160 months). The two and five year overall survival rates (n = 25) were 93% and 86% respectively. The disease-specific survival was 86% at last followup. Patients undergoing complete resection (n = 16) showed a 100% median survival at 60 months.

CONCLUSIONS: At present, complete tumor resection for patients with conventional HPC is recommended. However, considering the favorable outcome in this disease, the authors caution against performing operations that may potentially cause loss of function or are limb threatening.

Cancer. 2002; 95(8):1746-51

Solitary fibrous tumour and haemangiopericytoma: evolution of a concept

ABSTRACT

Haemangiopericytoma (HPC) was described in 1942 by Stout and Murray as a distinctive soft tissue neoplasm, presumably of pericytic origin, exhibiting a characteristic well-developed 'staghorn' branching vascular pattern. Over the years, it appeared that this growth pattern was a non-specific one, shared by numerous, unrelated benign and malignant lesions, and that HPC was better considered as a diagnosis of exclusion.

Histopathology. 48(1):63-74, January 2006.

Solitary fibrous tumour and haemangiopericytoma: evolution of a concept

The first category corresponds to those non-HPC neoplasms that occasionally display HPC-like features (e.g. synovial sarcoma).

The second category shows clear evidence of myoid/pericytic differentiation and correspond to true HPCs. They generally show a benign clinical course, and include glomangiopericytoma/myopericytoma, infantile myofibromatosis (previously called infantile HPC), and a subset of sinonasal HPCs.

The third category is the solitary fibrous tumour (SFT) lesional group, which includes fibrous-to-cellular SFTs, and related lesions such as giant cell angiofibromas and lipomatous HPCs.

- Histopathology. 48(1):63-74, January 2006.

Hemangiopericytoma: a dying breed? Reappraisal of an 'entity' and its variants.

Hemangiopericytoma represents a rather questionable entity with debatable and temporarily varying diagnostic criteria. Neoplasms called hemangiopericytoma in the past have in common the presence of numerous thin-walled, branching blood vessels and pericellular reticulin fibers

Curr Diagn Pathol. 1994;1:19-23.

Formerly known as HPC have been partitioned into 3 groups:

So-called “true” HPCs, with clear evidence of myoid and pericytic differentiation, include a subset of sinonasal HPC,

subcutaneous infantile myofibromatosis or infantile HPC,

and glomangiopericytoma or myopericytoma.

.

The third group includes conventional solitary fibrous tumor, a fat-forming solitary fibrous tumor or lipomatous HPC, and a giant-cell–rich variant of solitary fibrous tumor or giant cell angiofibroma.

Many lesions have HPC-like features and are sometimes miscategorized

Perivascular Neoplasms'

Glomus tumor
Hemangiopericytoma (HPC)
- glomangiopericytoma
- myopericytoma
- solitary fibrous tumour
- mesenchymal chondrosarcoma
- monophasic synovial sarcoma
- endometrial stromal sarcoma

The differential diagnosis of malignant myopericytoma

Myofibromatosis or myopericytoma, Haemangiopericytoma

Solitary fibrous tumour,

Monophasic synovial sarcoma,

Mesenchymal chondrosarcoma

Low-grade endometrial stromal sarcoma.

Leiomyosarcoma,

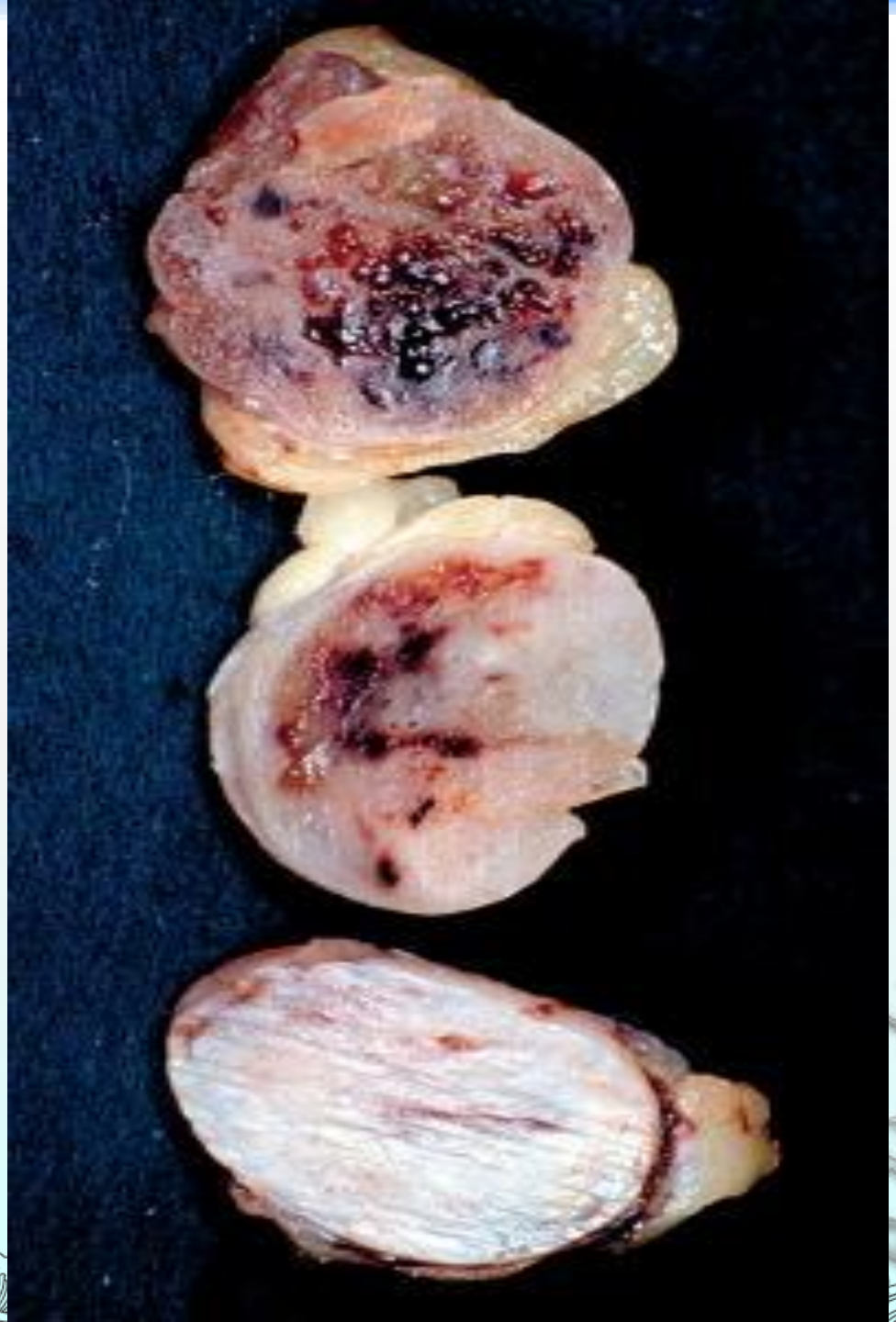
Malignant peripheral nerve sheath tumour.

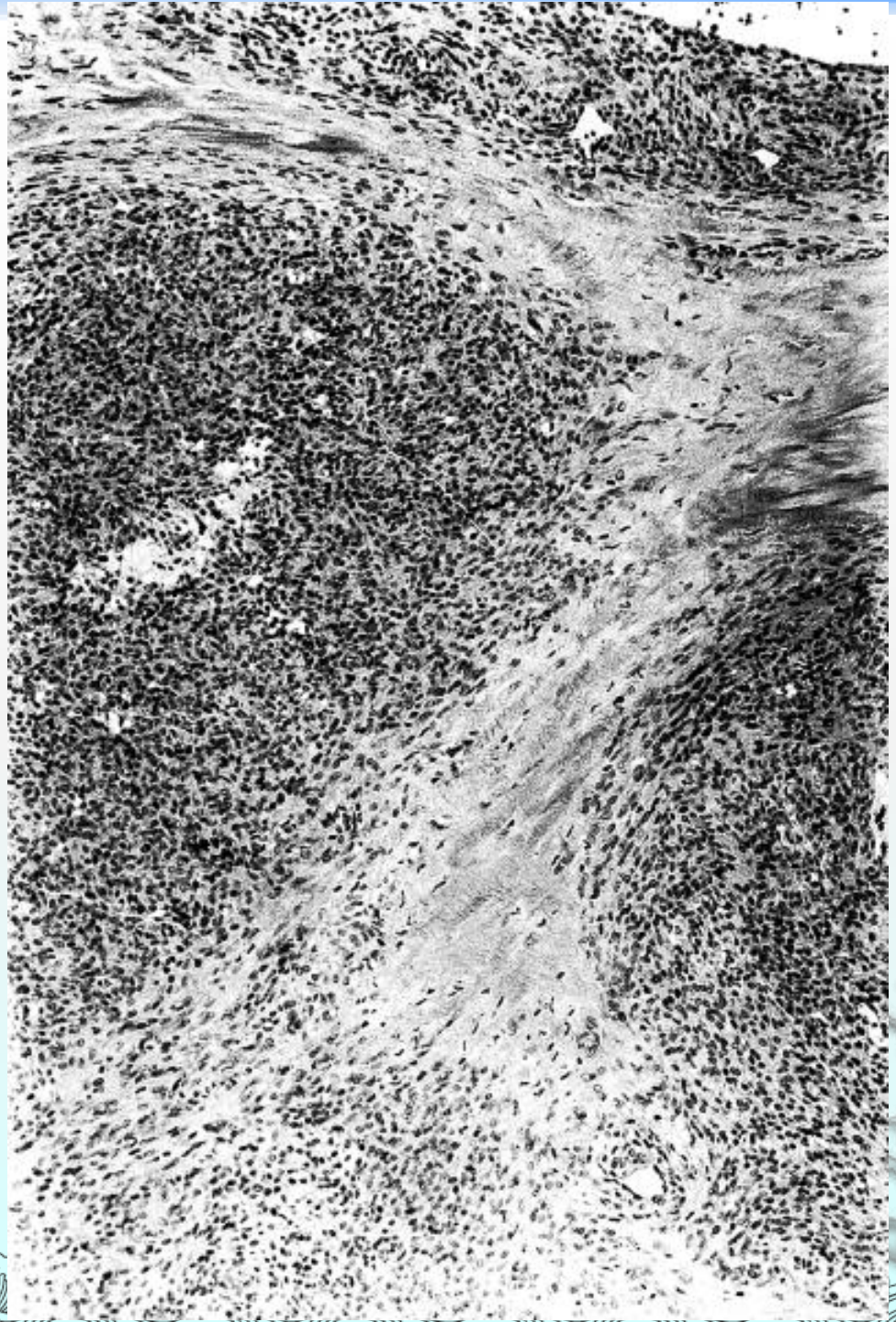
Histopathology. 41(5):450-460, November 2002.

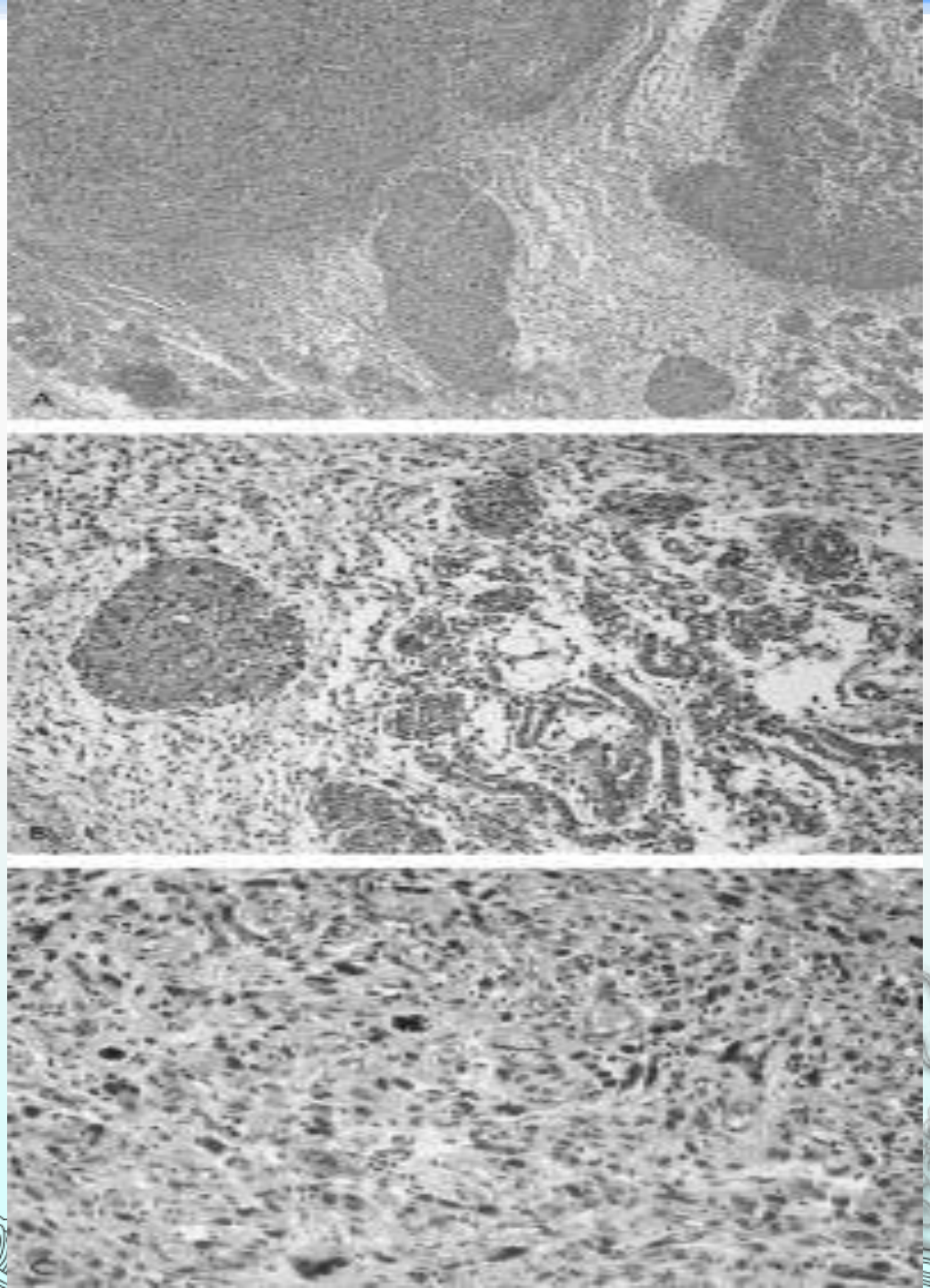
Malignant myopericytoma: expanding the spectrum of tumours with myopericytic

Methods and results: Five cases of malignant myopericytoma were identified in the authors' consultation files. Tumours arose in three females and two males (median age 67 years, range 19–81 years) on the neck, arm, thigh and foot. One patient presented with disseminated metastases. One patient had a prior history of multiple benign myopericytomas in the same location. Four patients developed metastases and three died within 1 year.

Histopathology. 41(5):450-460, November 2002.



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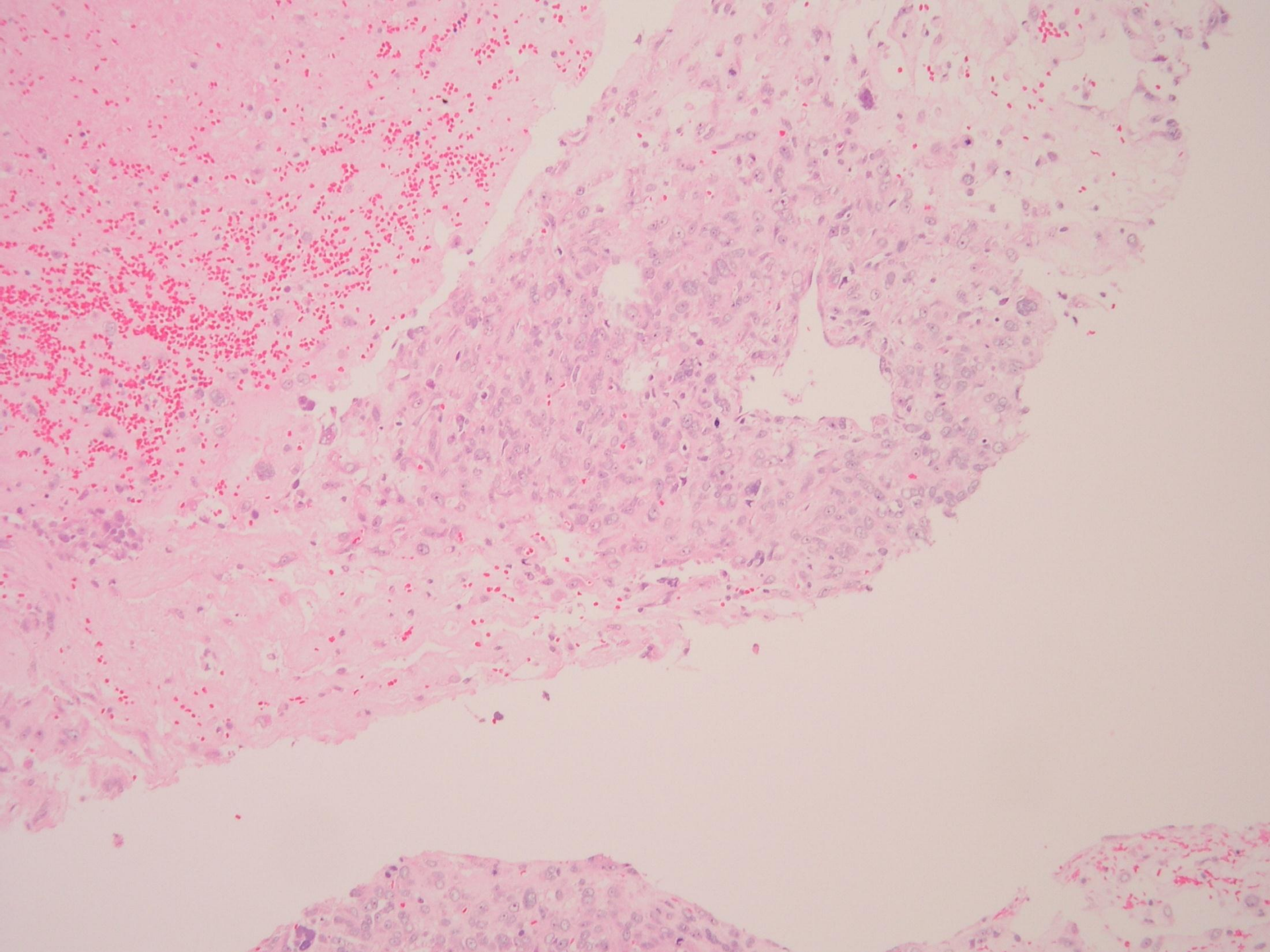

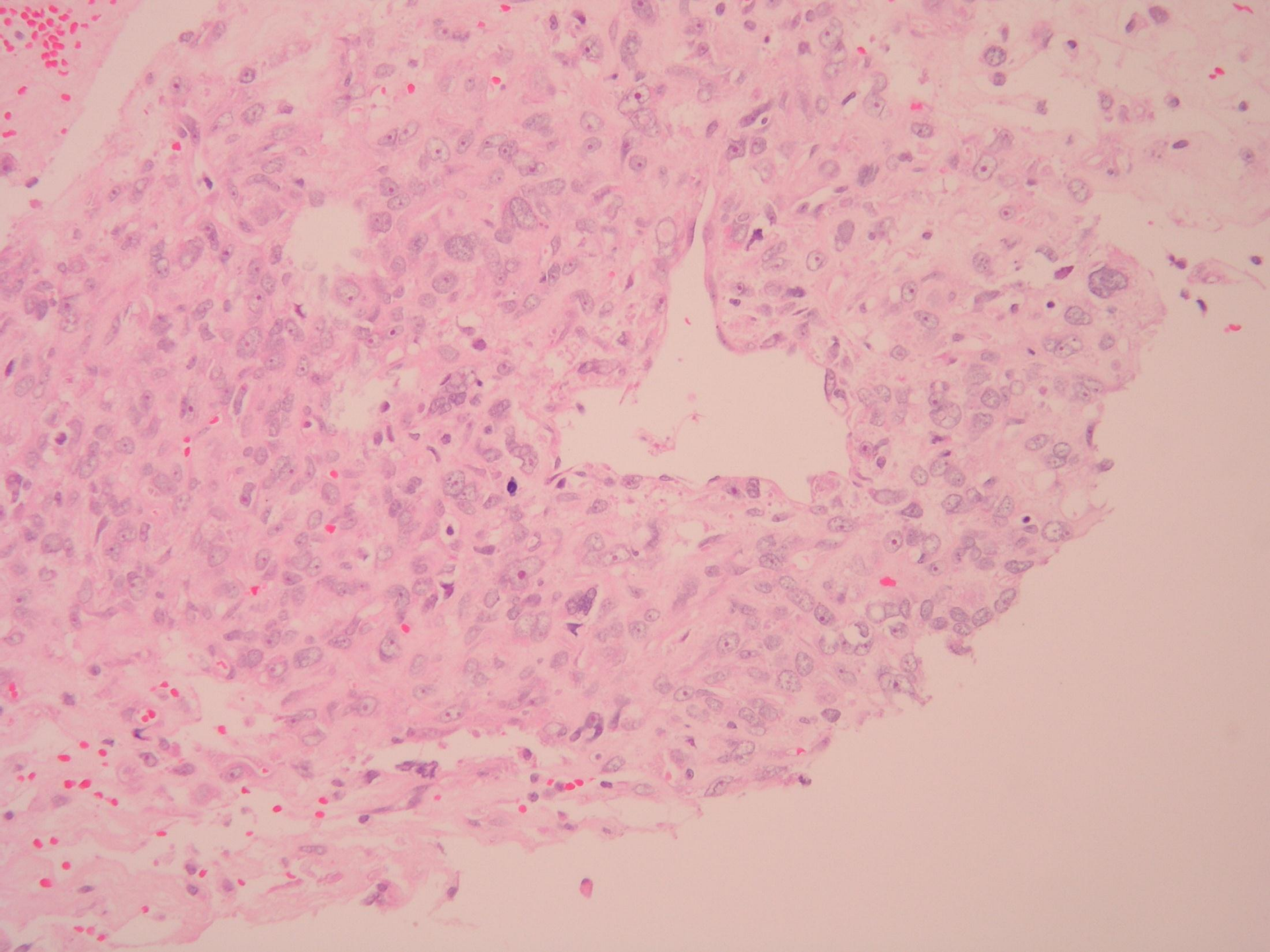

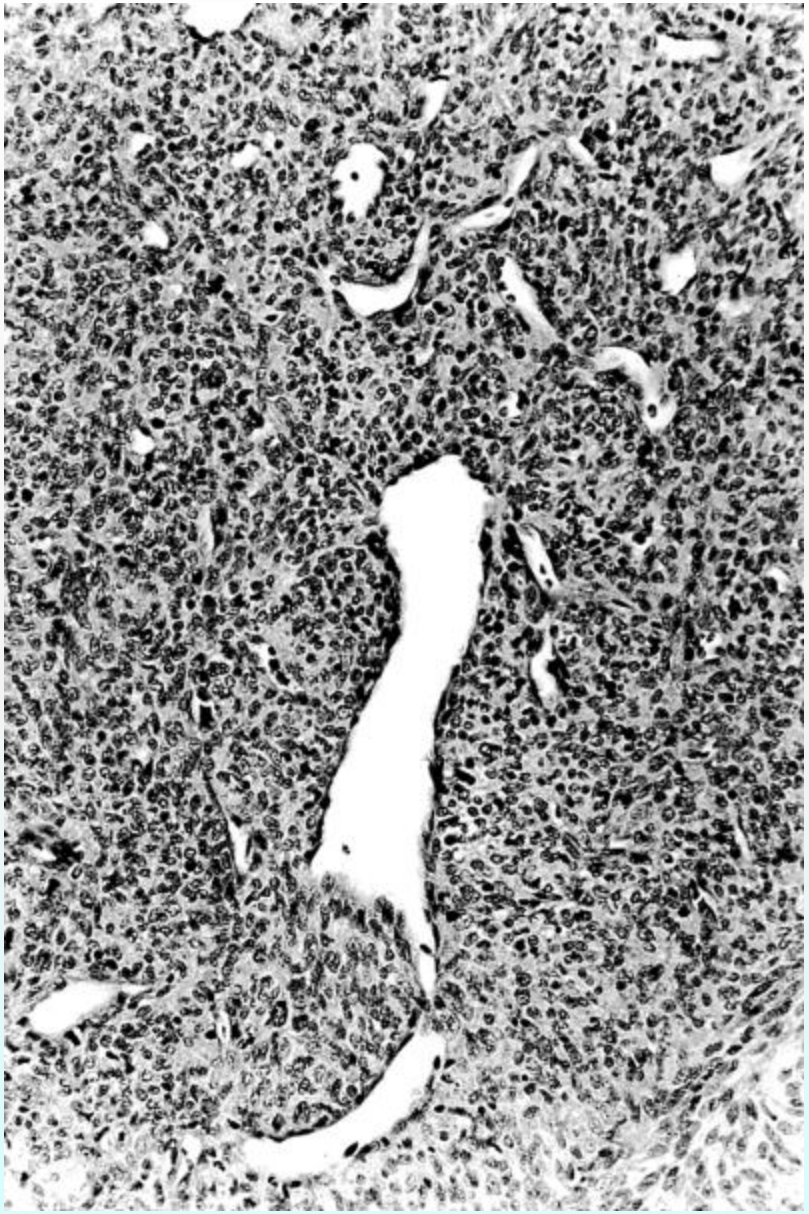

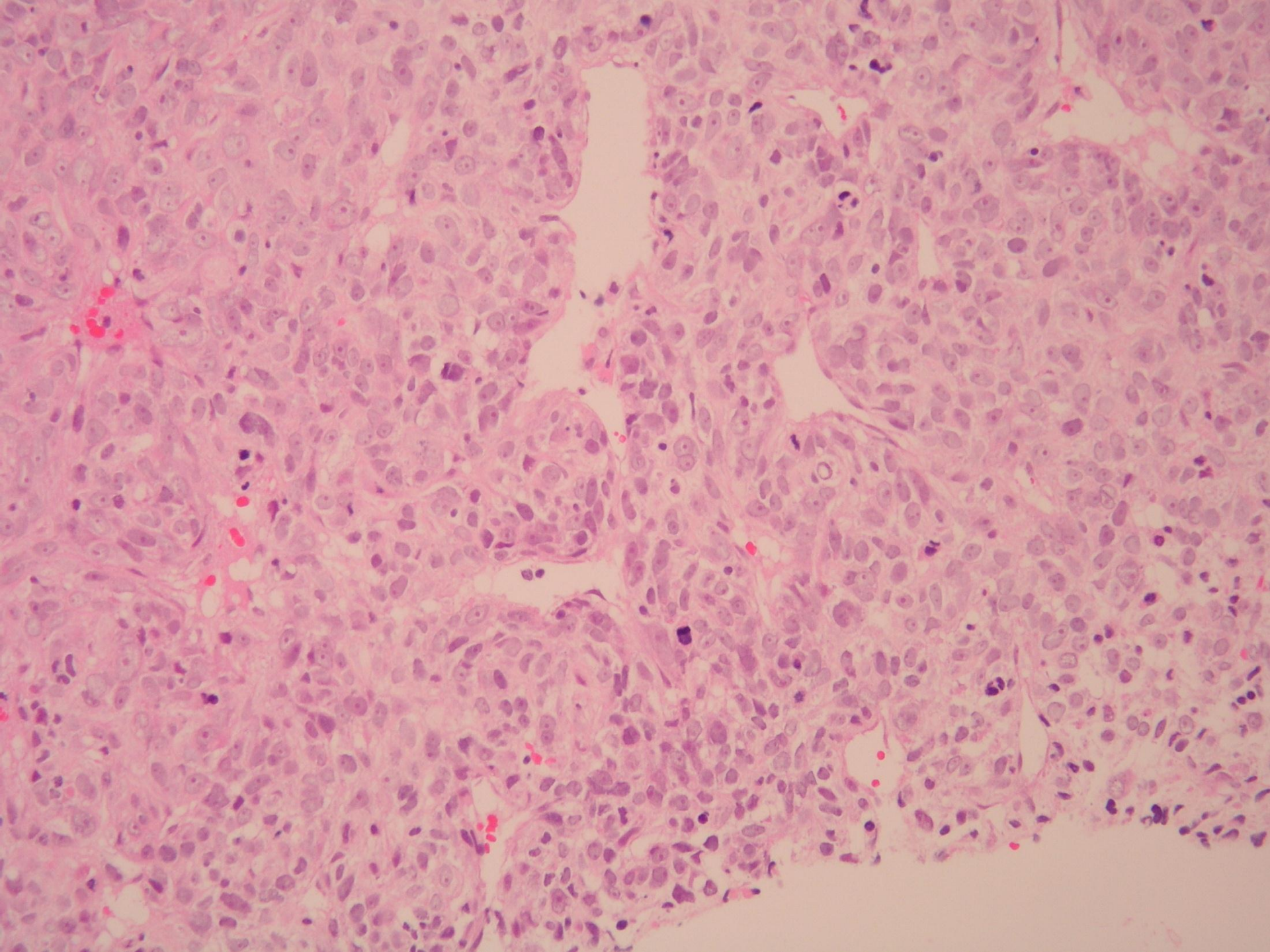

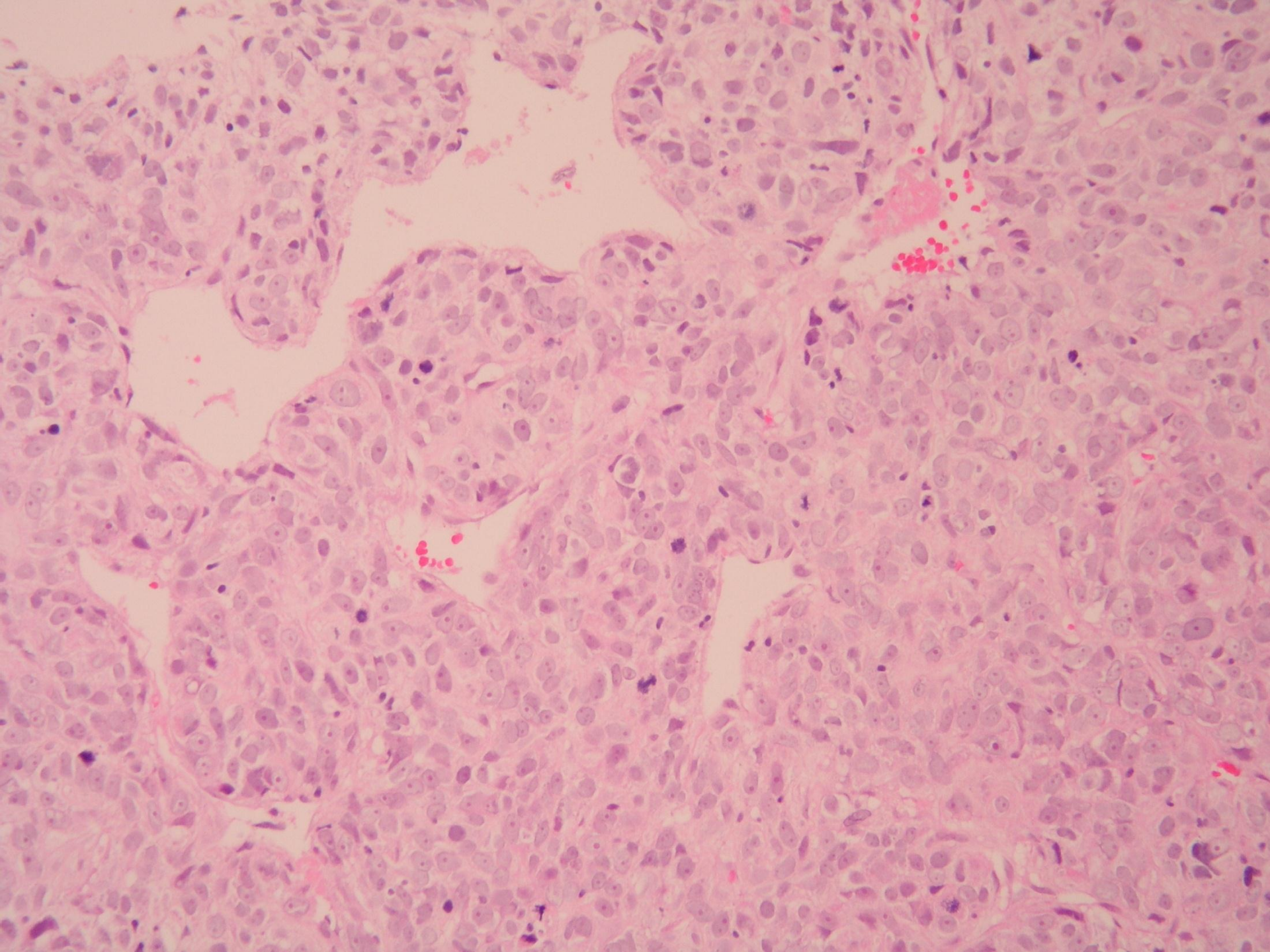

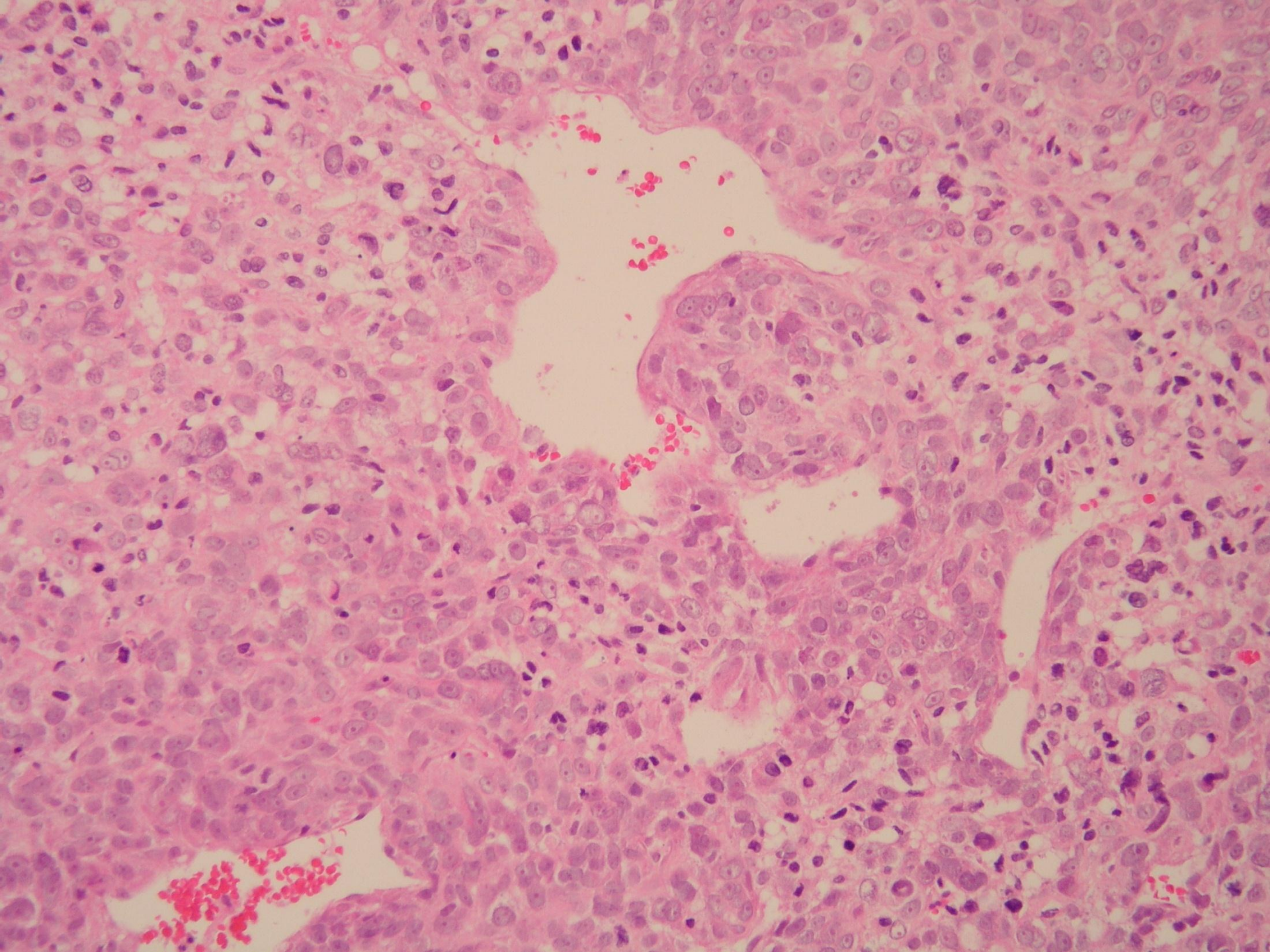

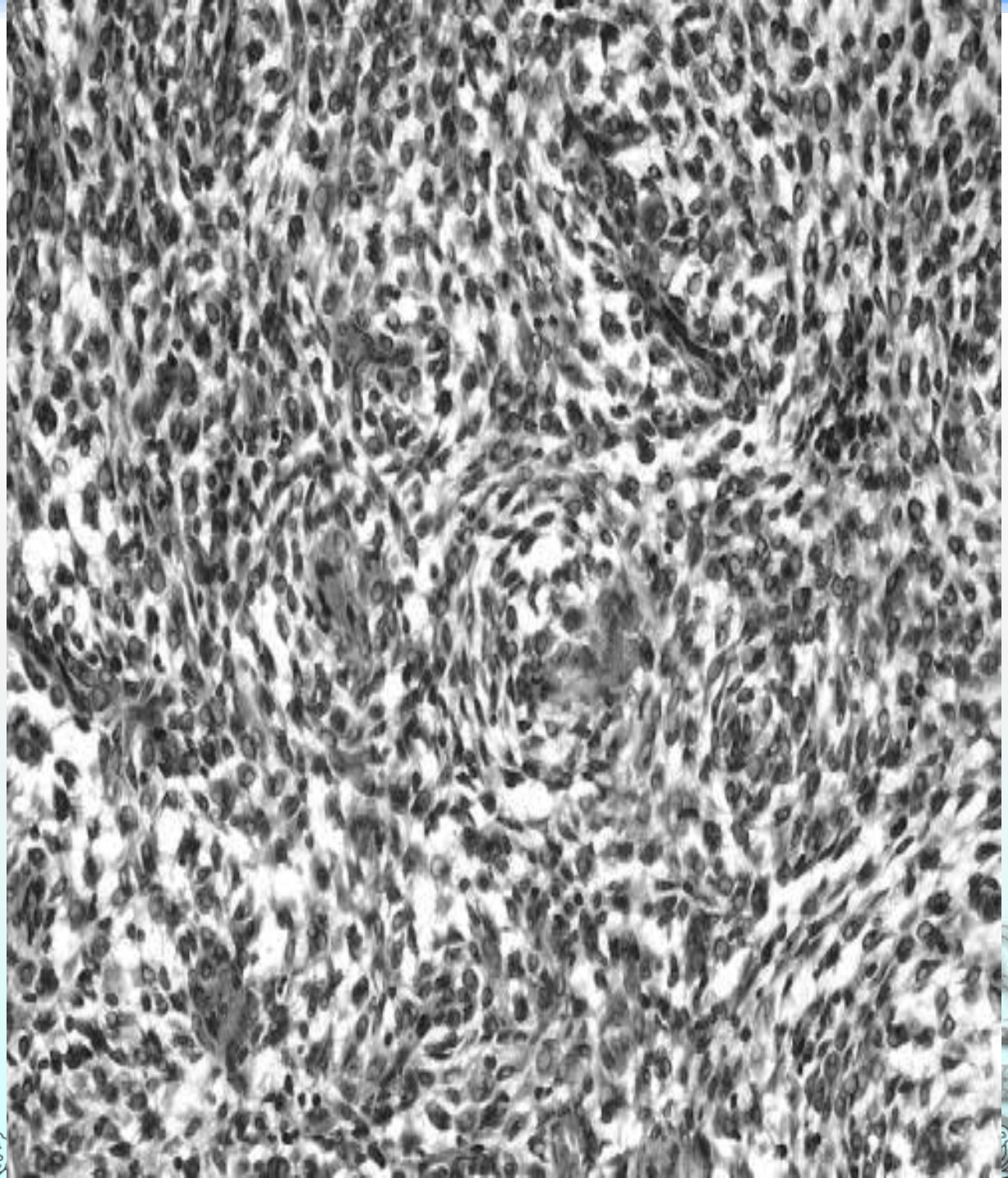

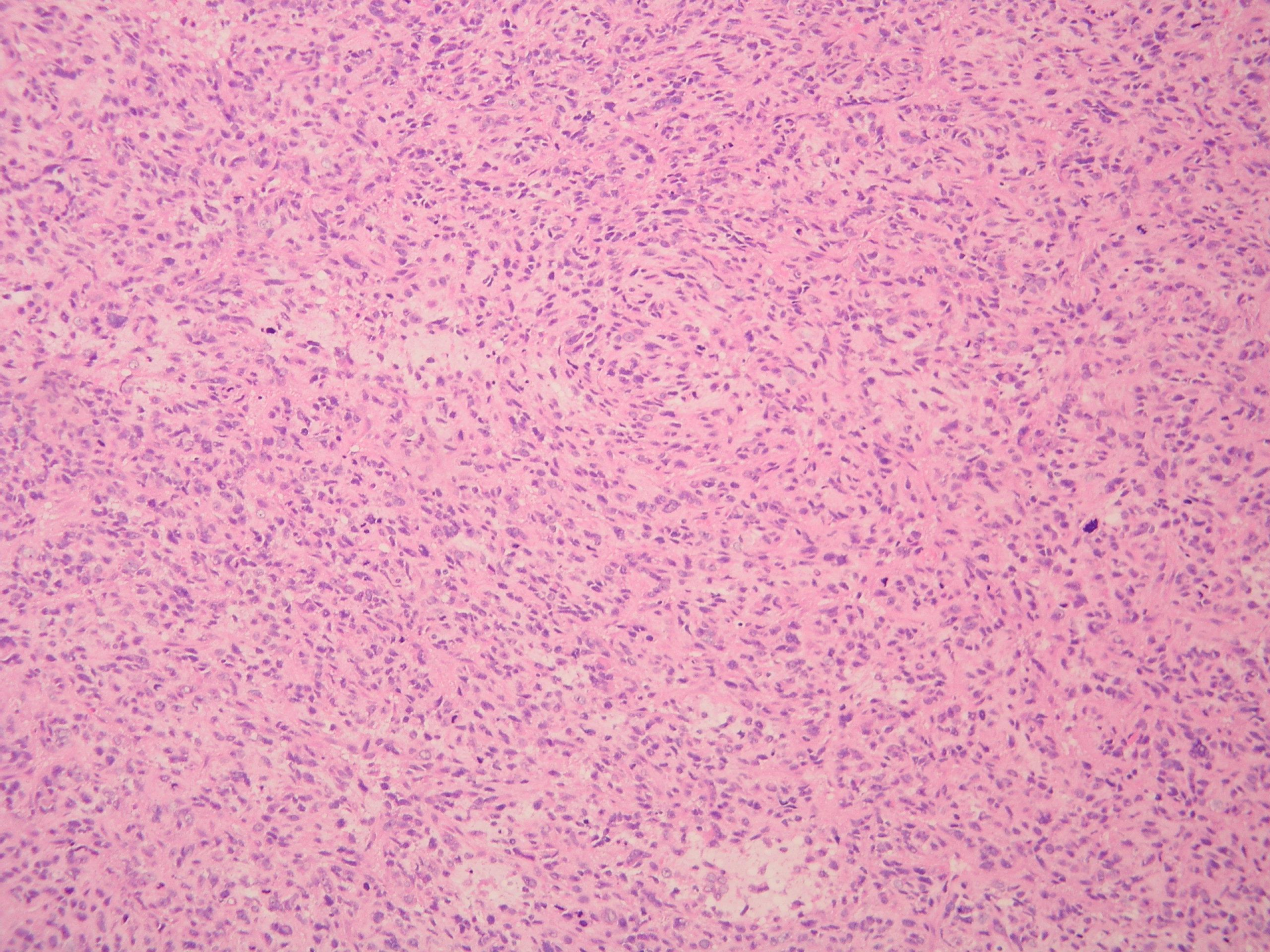

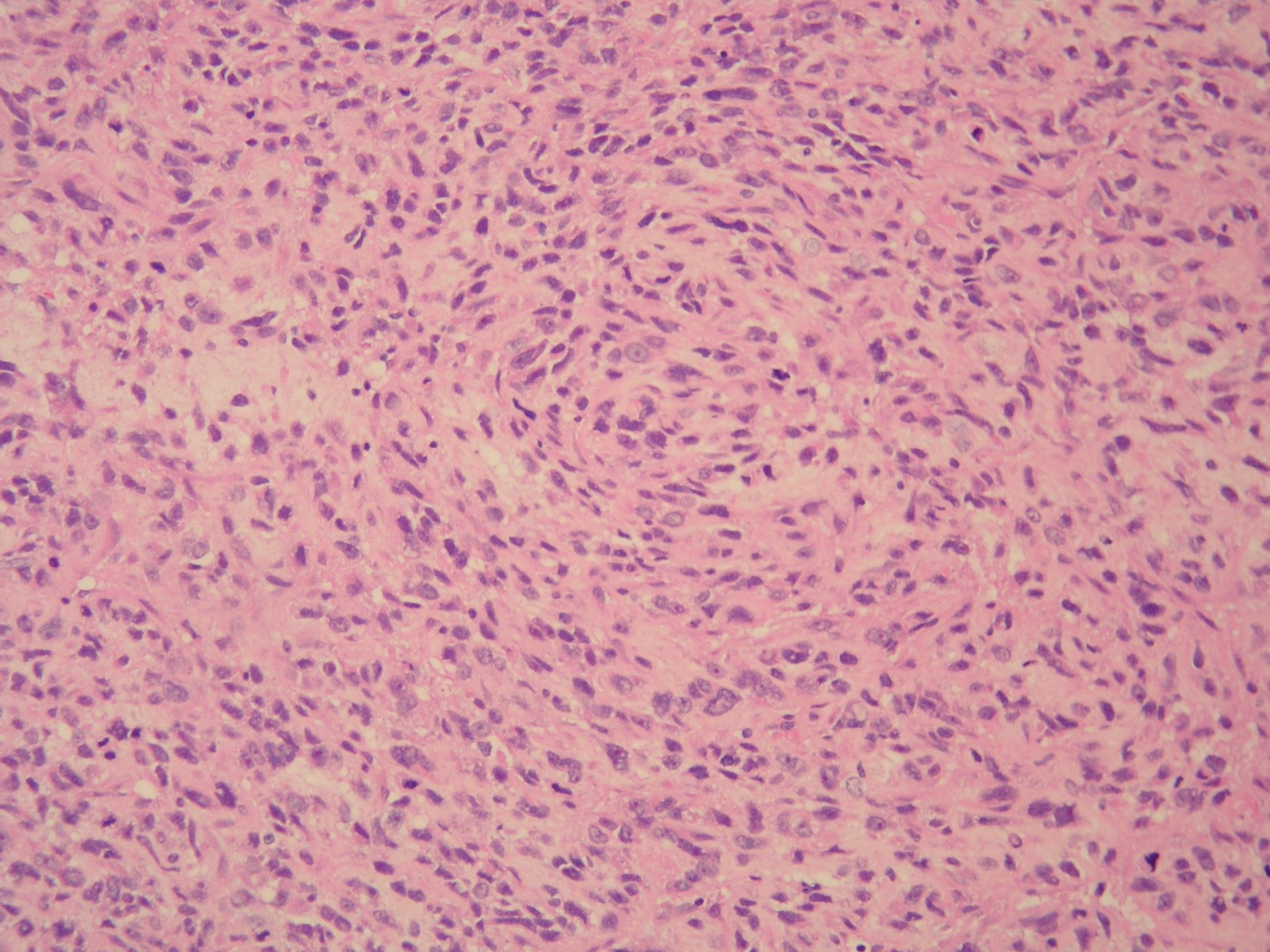

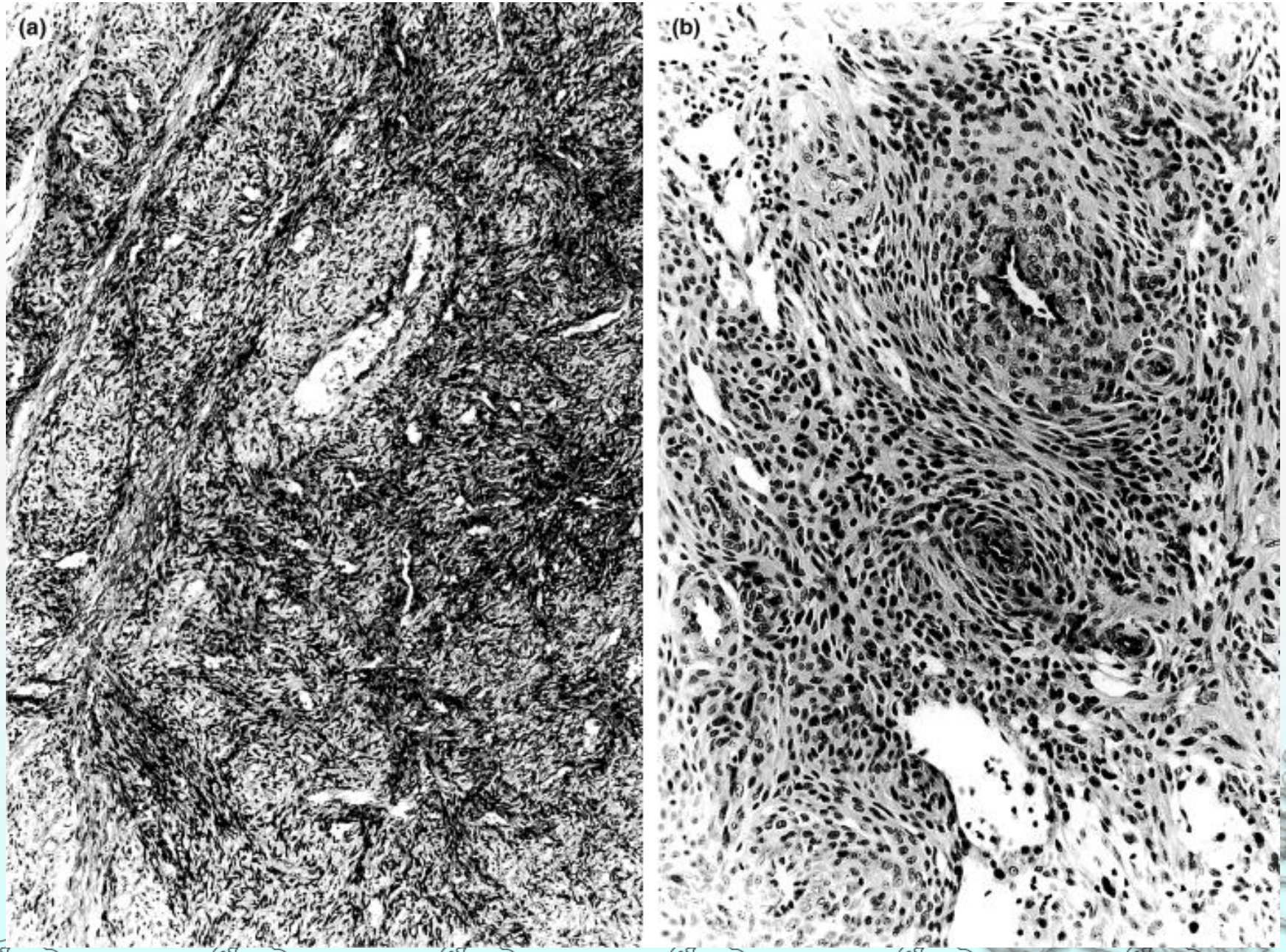
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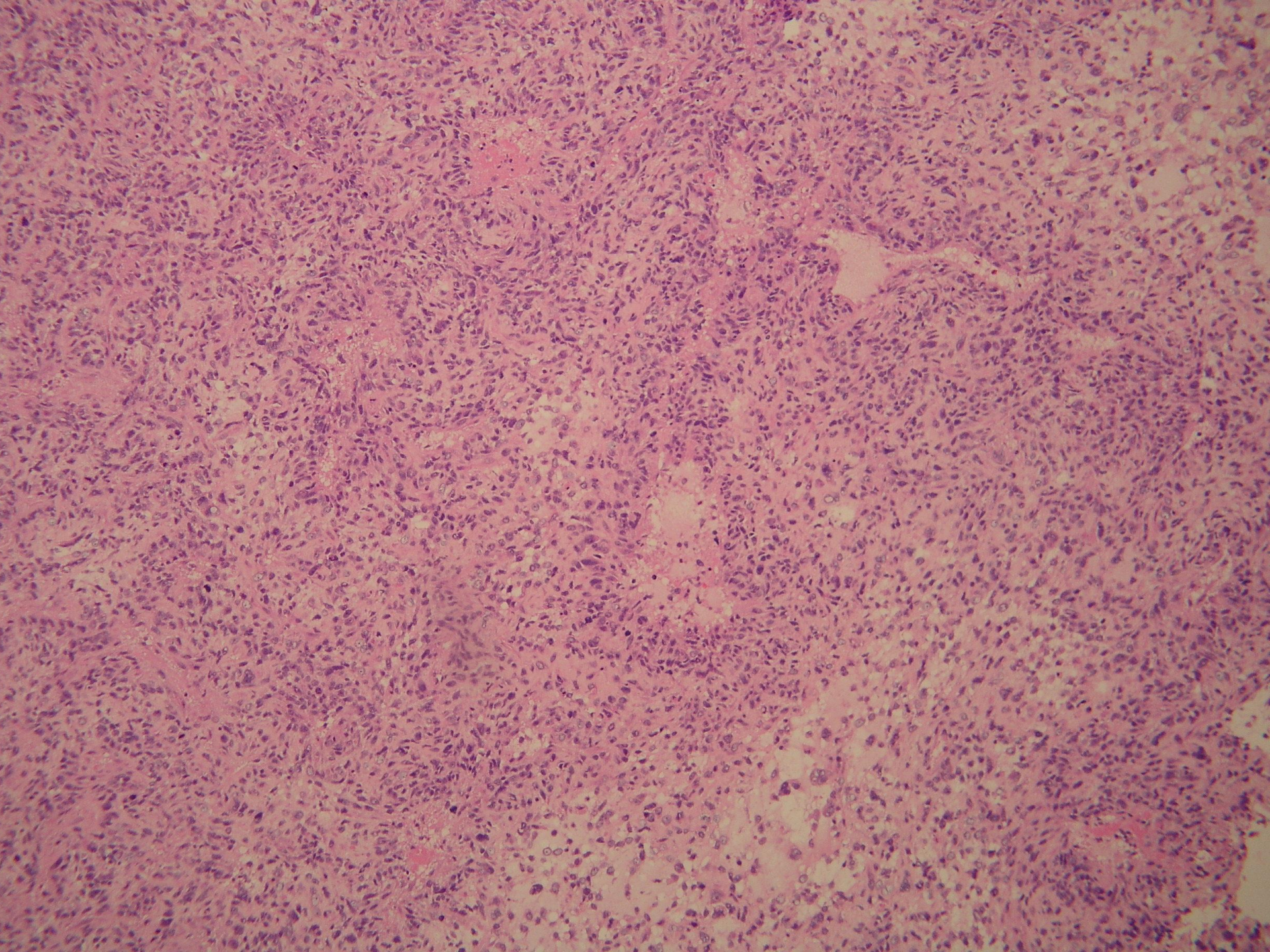
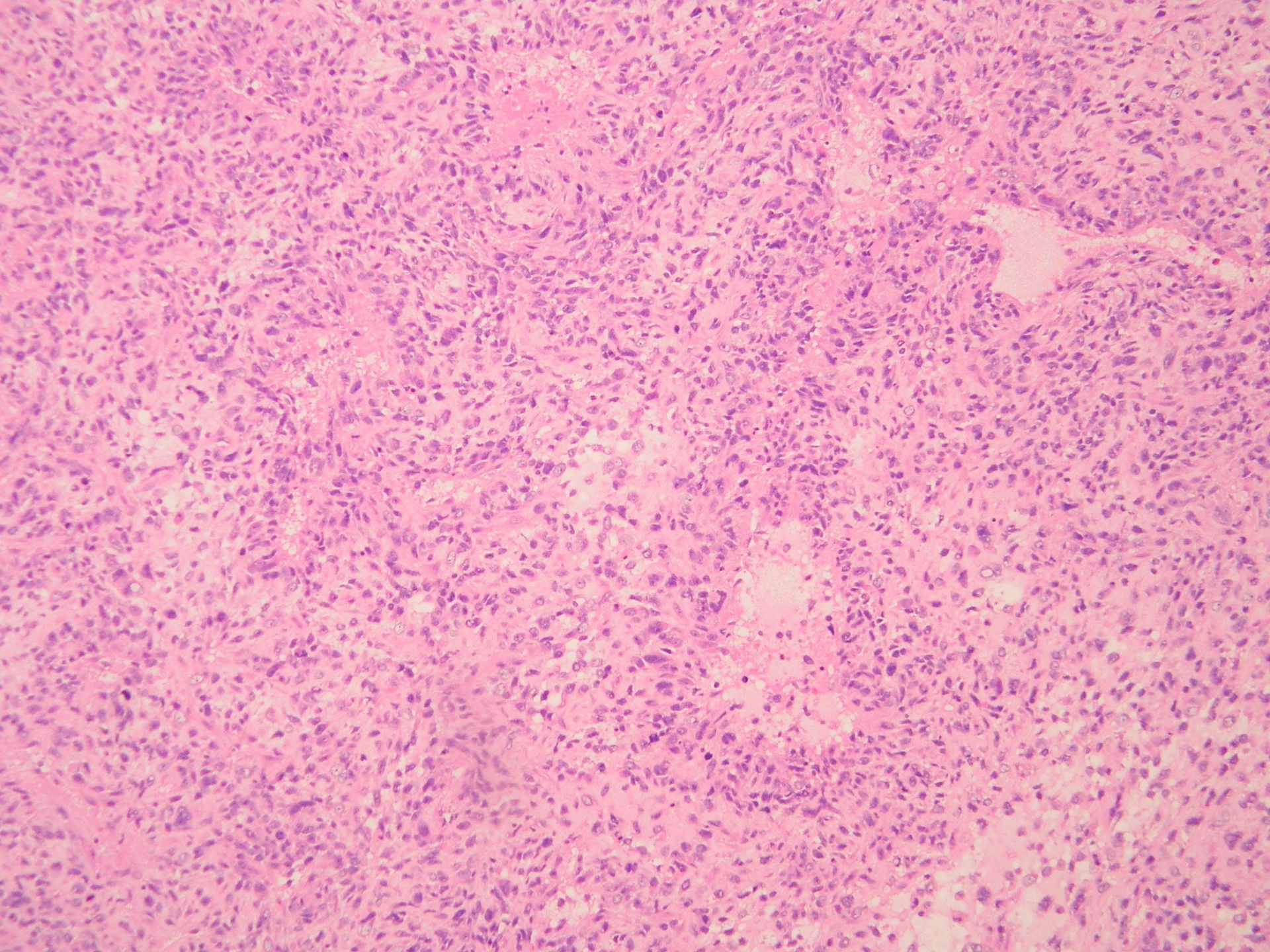
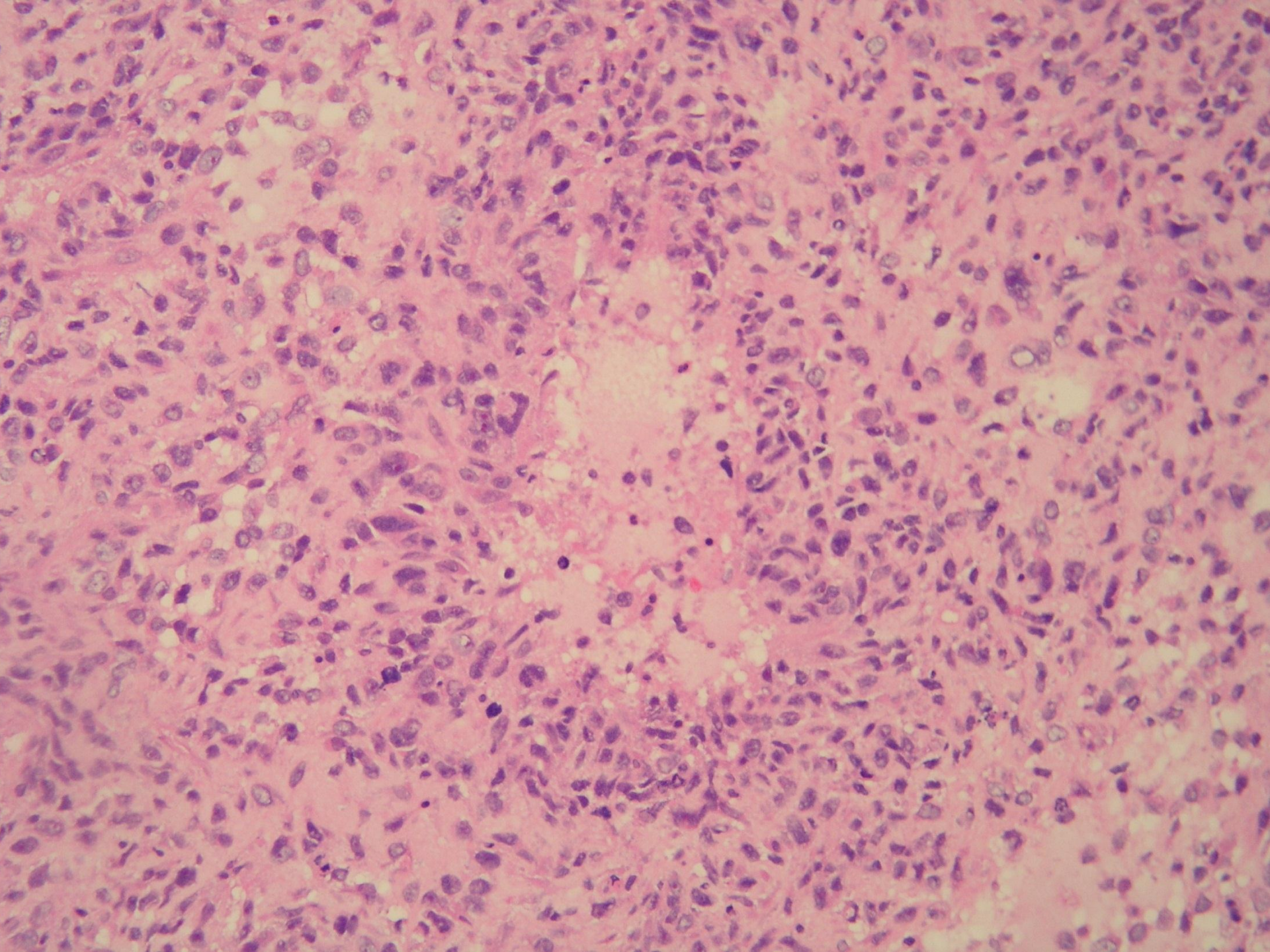
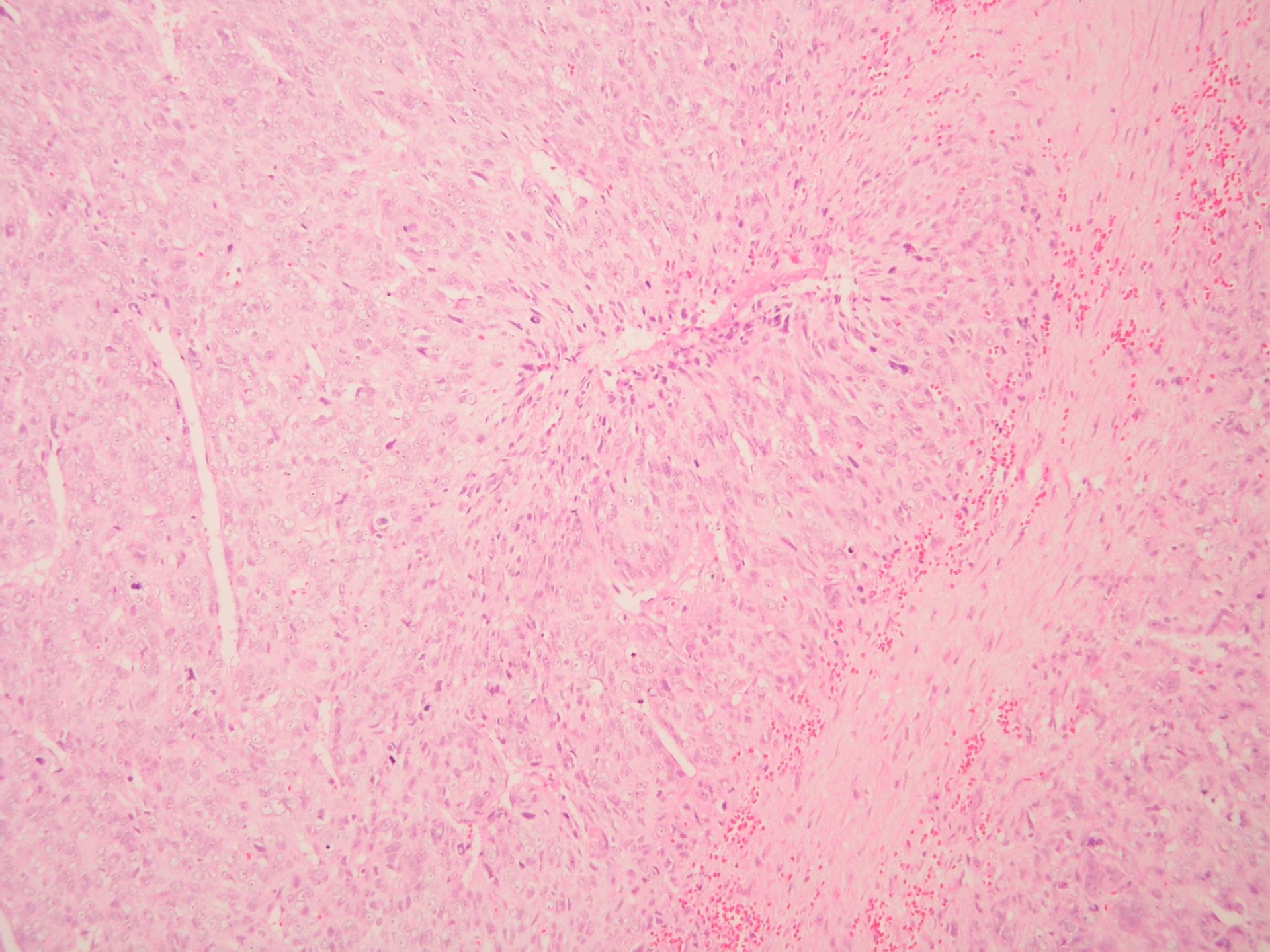
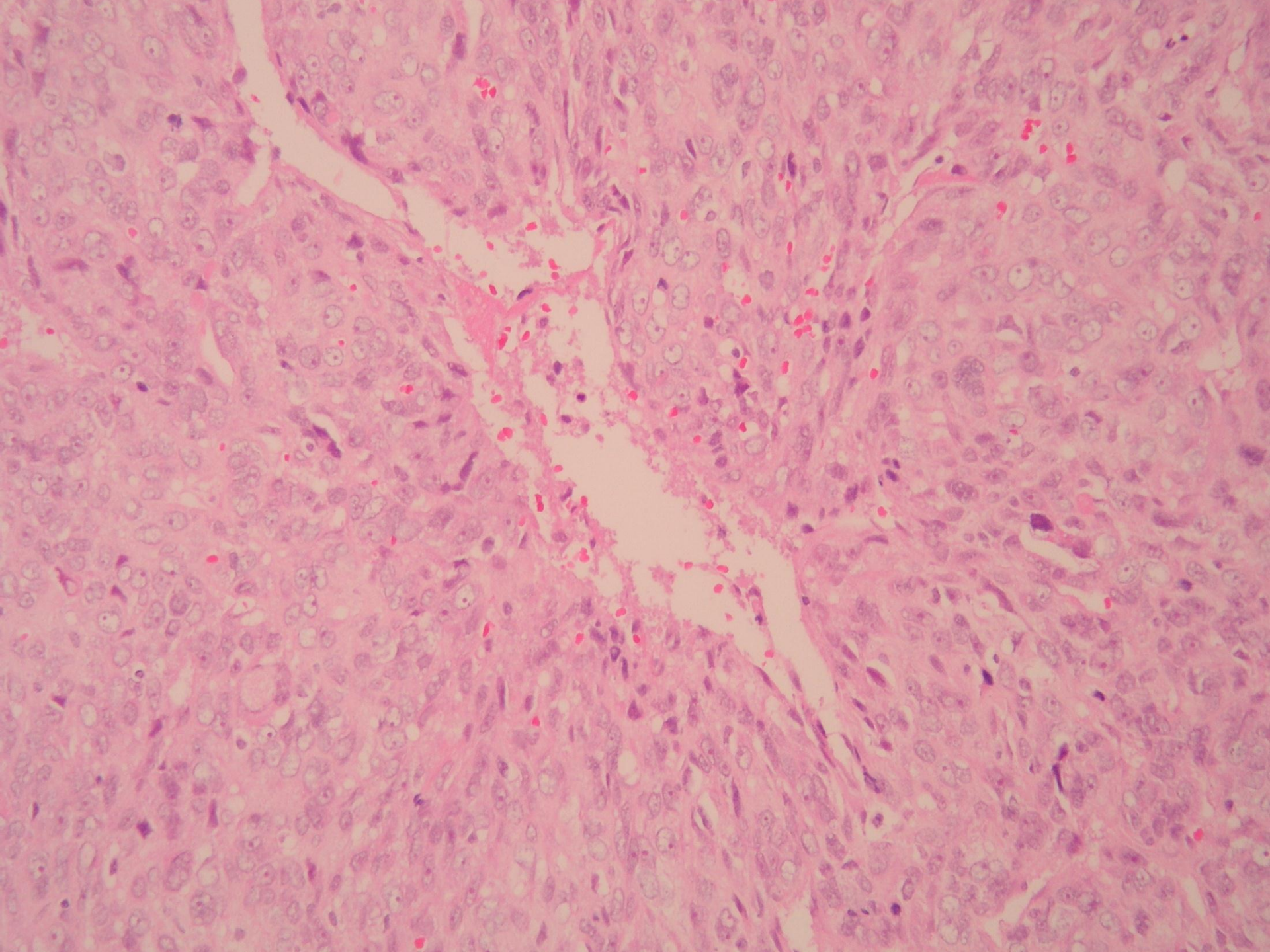
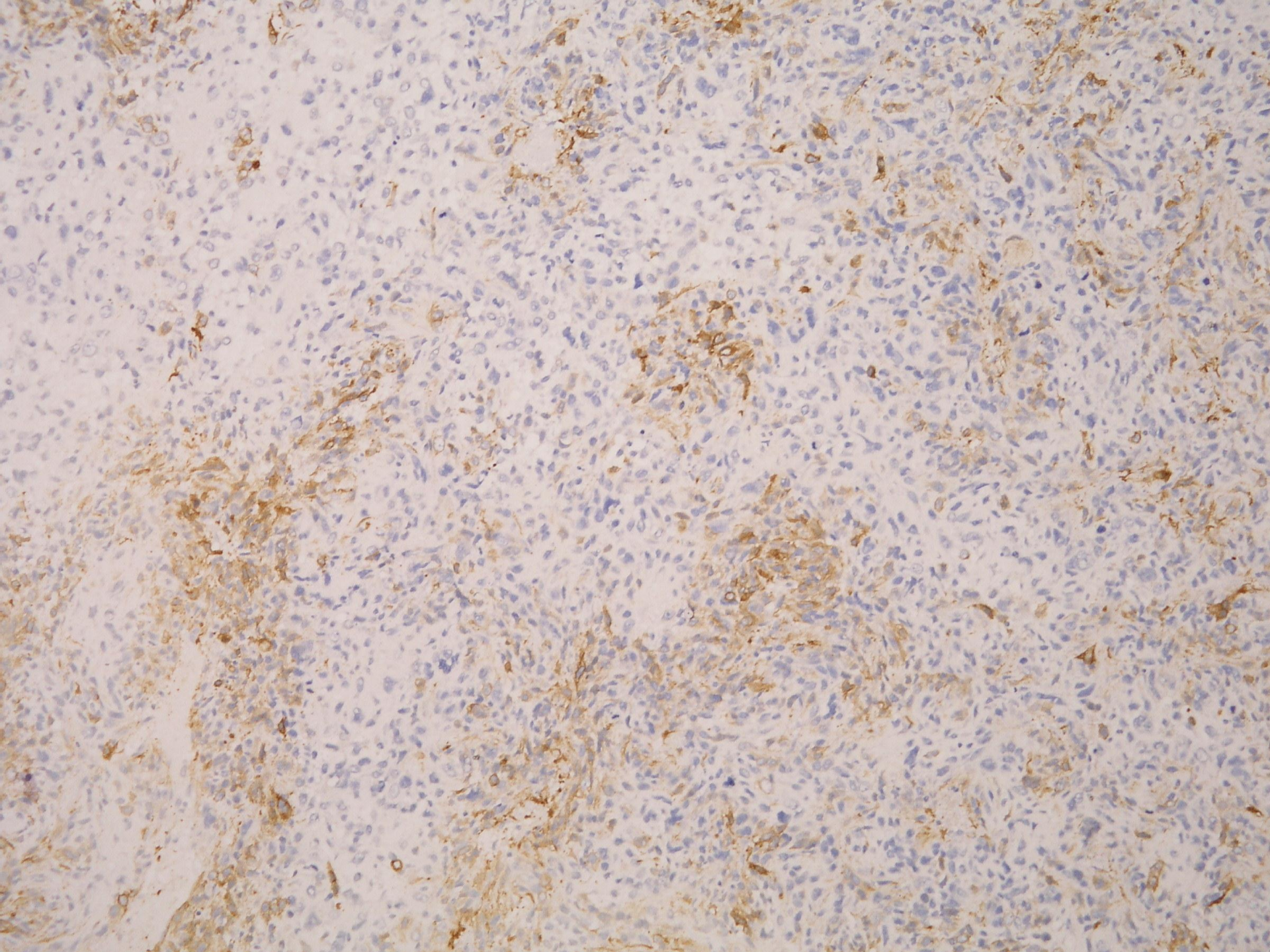


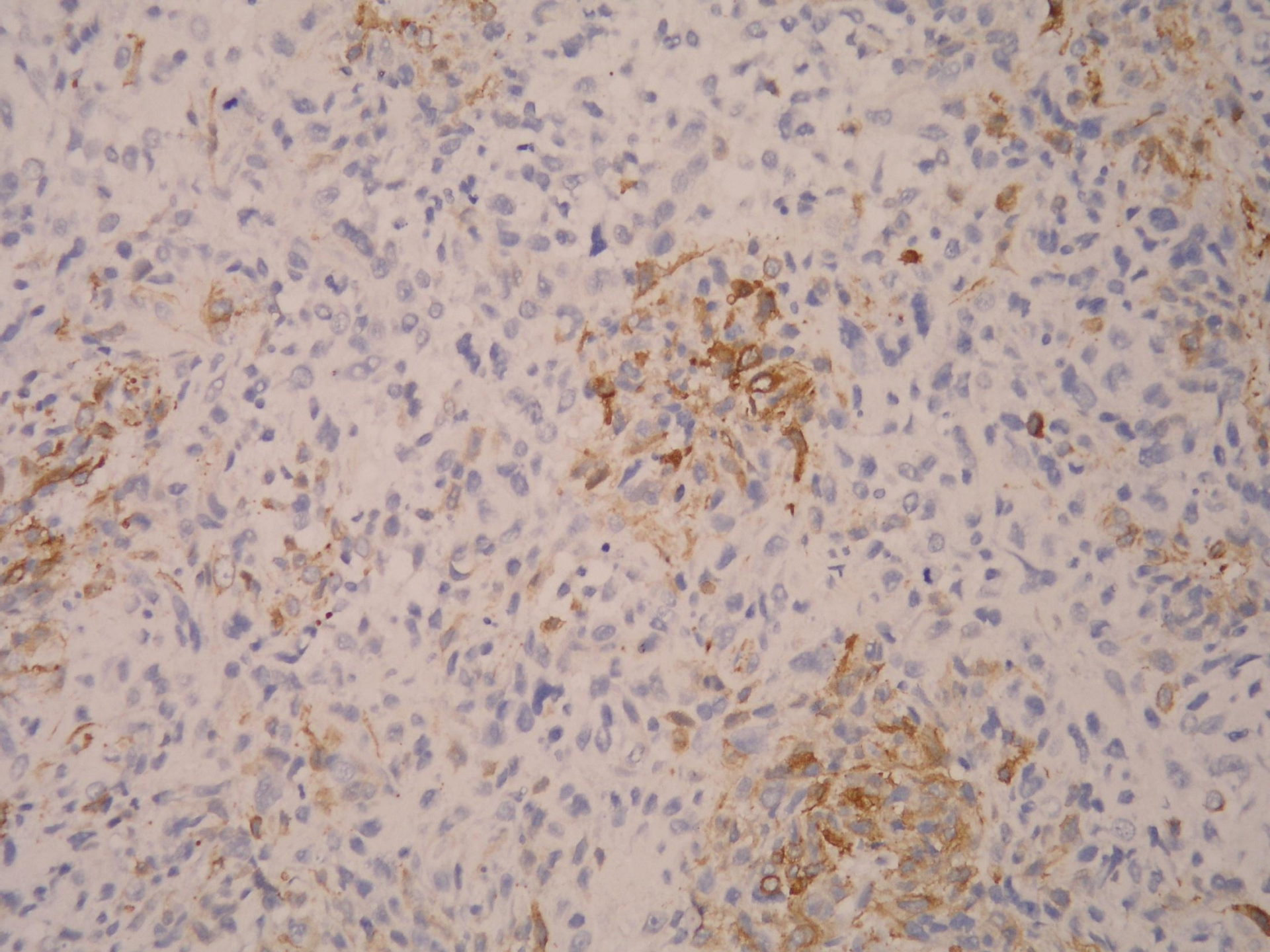


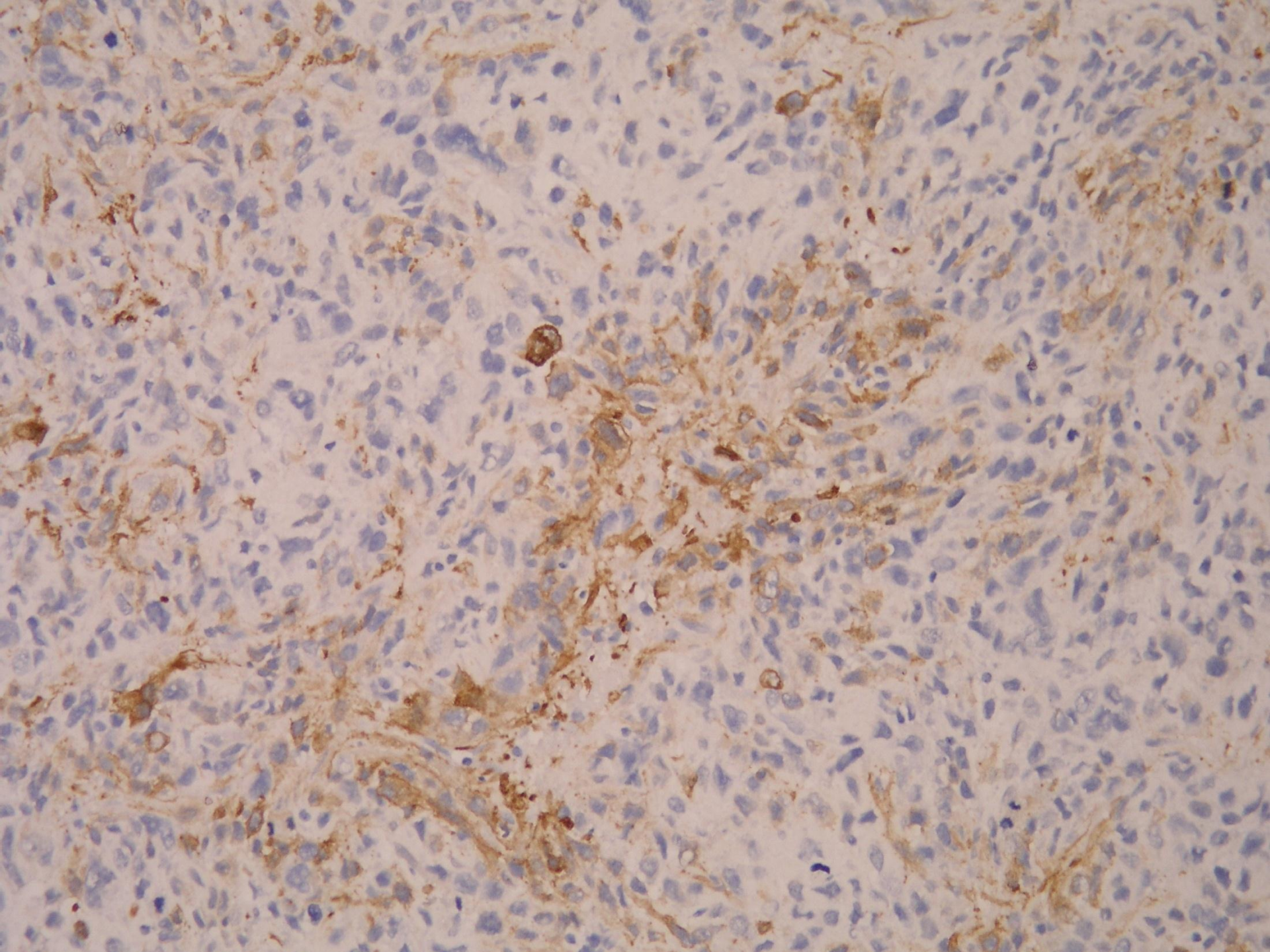


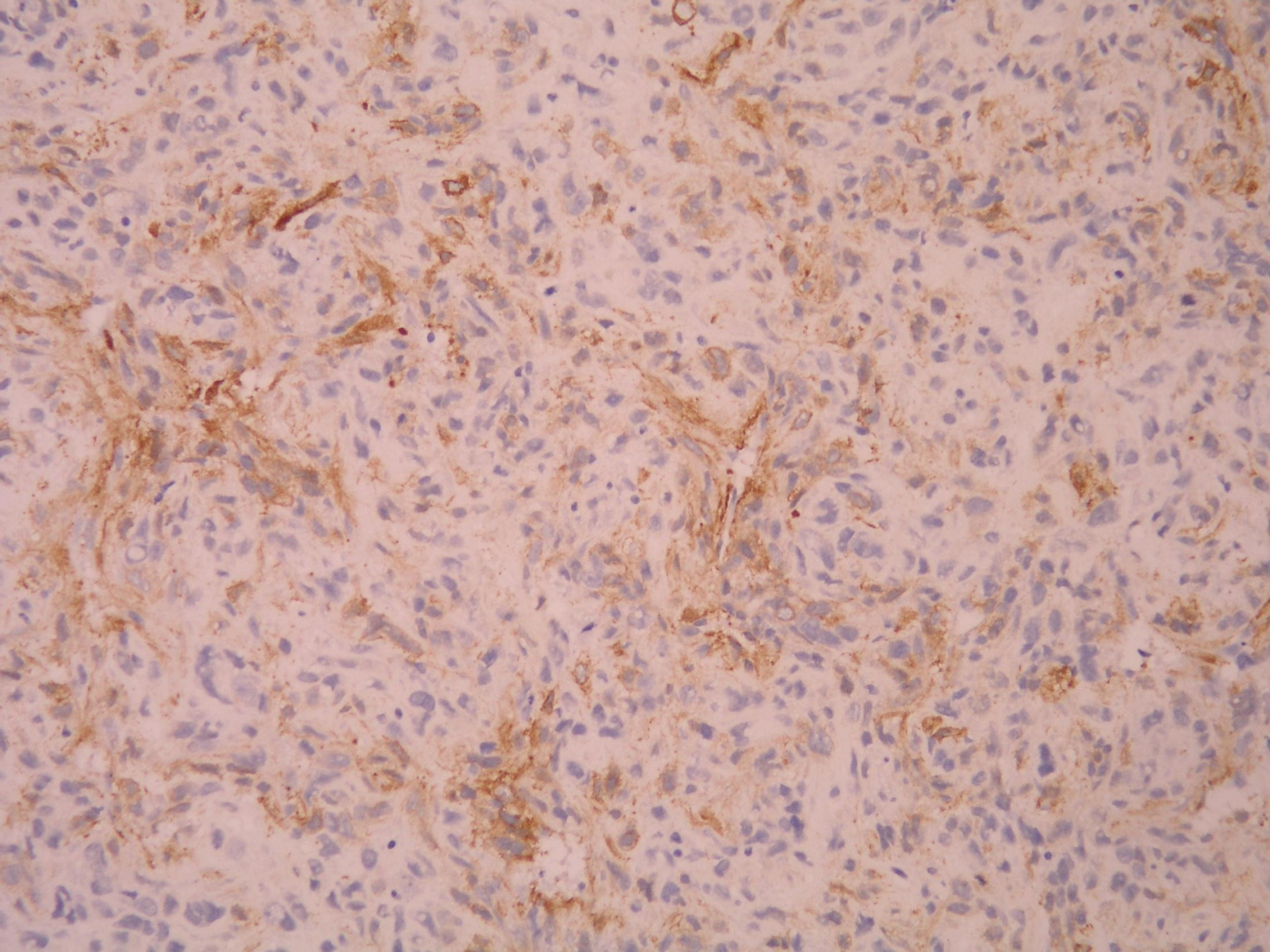


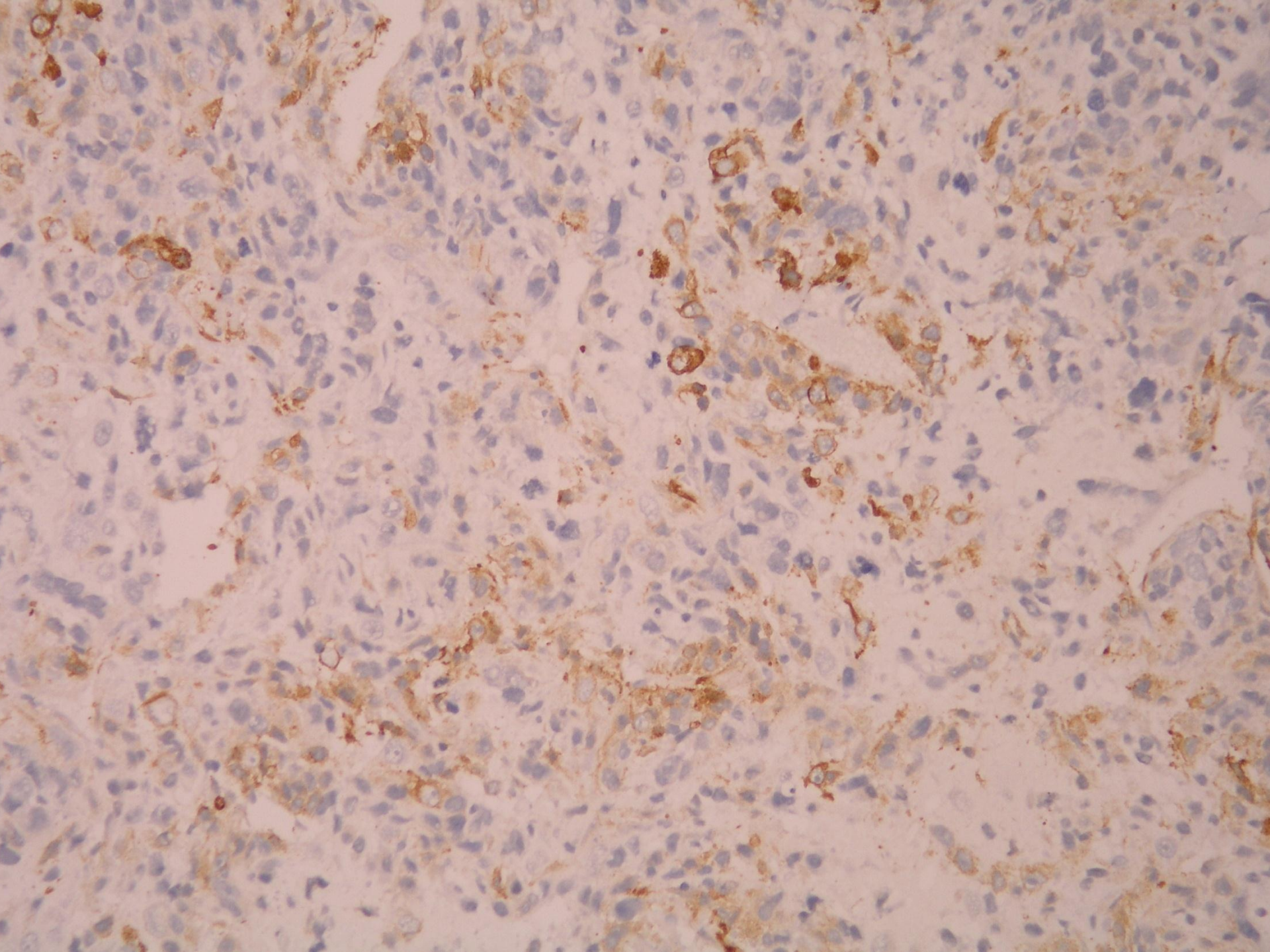


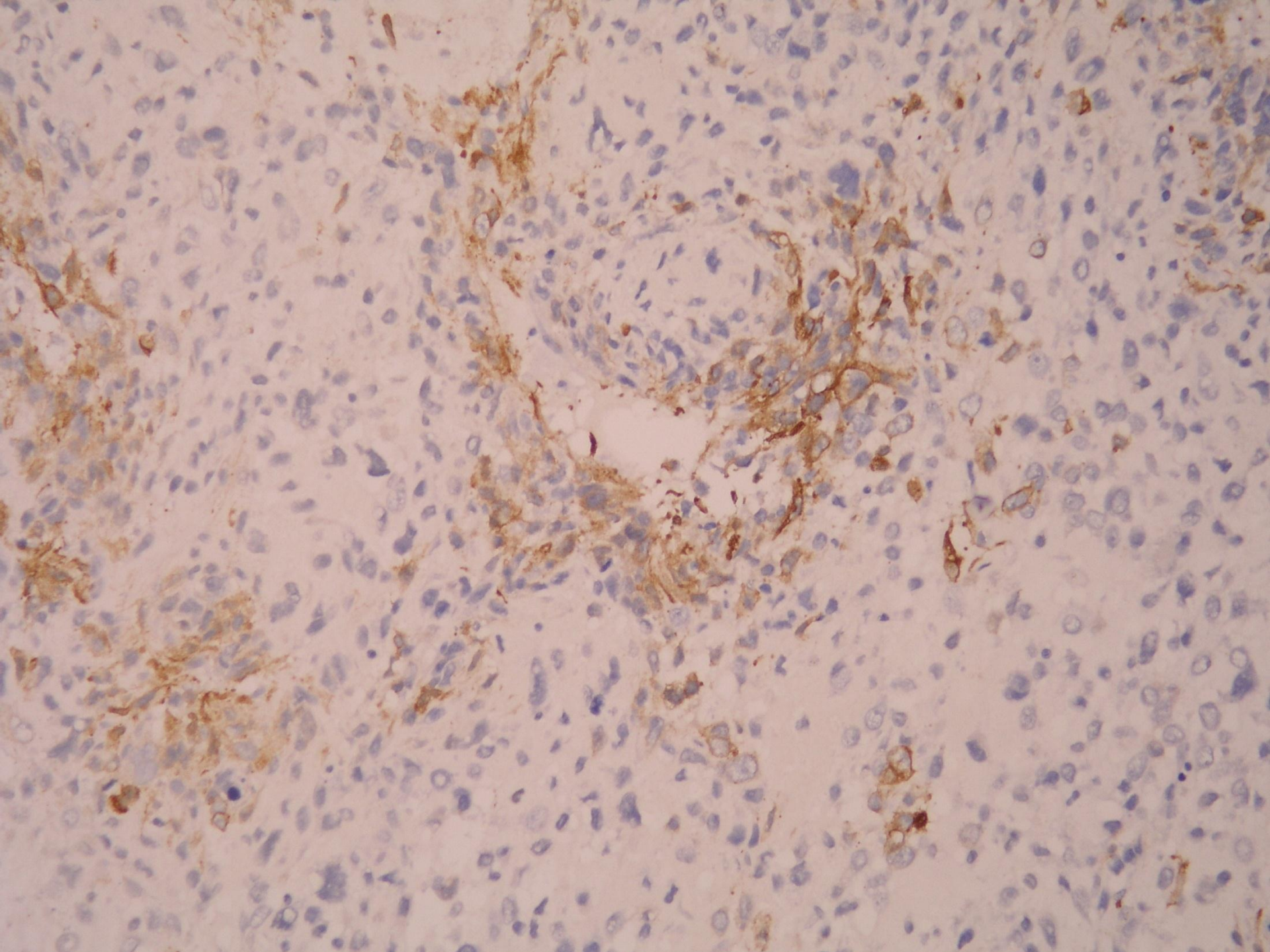


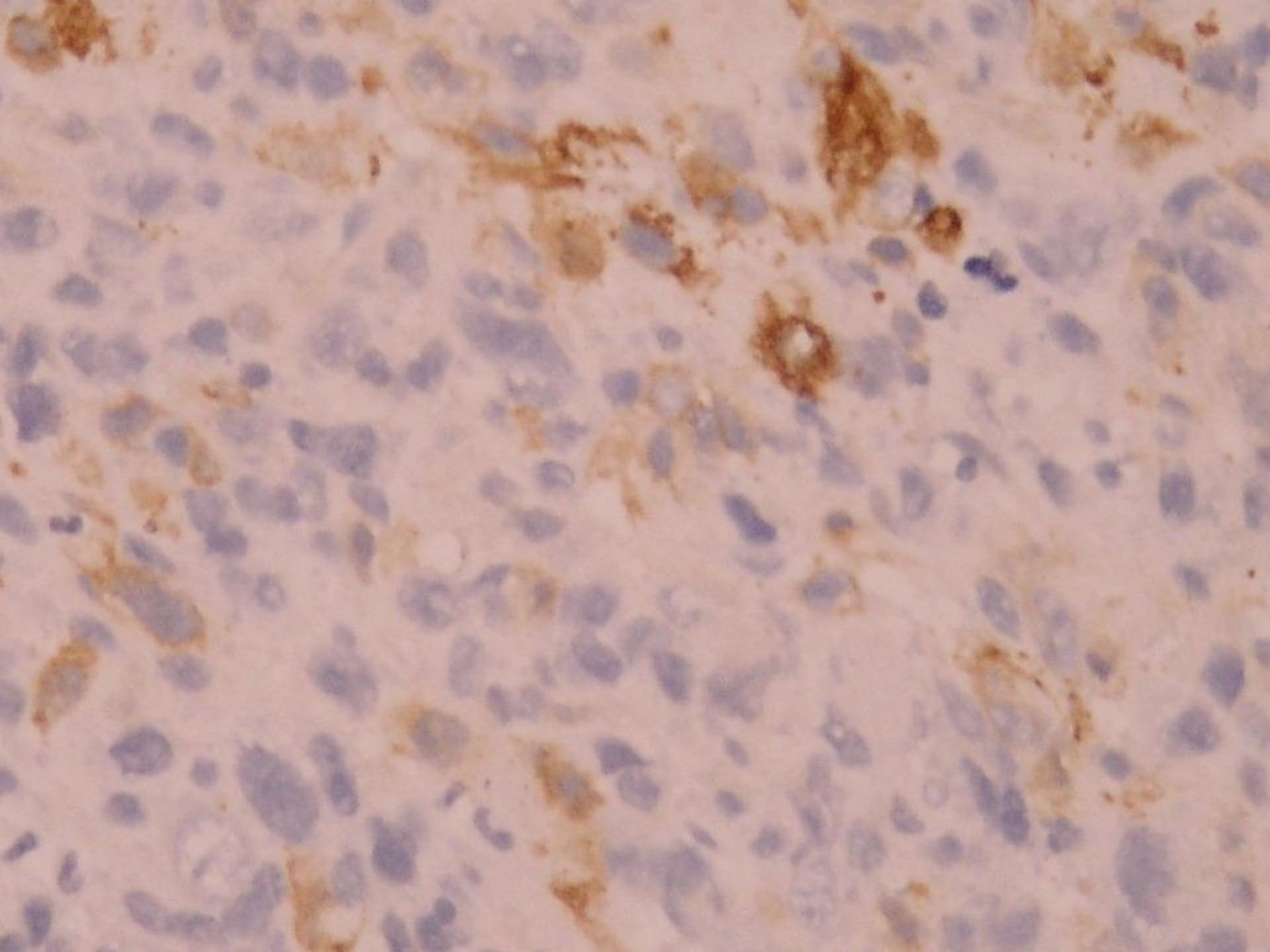












Myopericytoma

A close histological relationship exists between myopericytoma,

myofibromatosis,

solitary myofibroma

infantile haemangiopericytoma

A single morphological spectrum that shows perivascular myoid cells/pericytes.

***Am. J. Surg. Pathol.* 1998; 22; 513–525.**

Table 2 Summary of the recently described morphological categories of 'perivascular myoma'

Granter *et al.*[1]	Myofibromatosis-type	Glomangiopericytoma	Myopericytoma
	Perivascular myoma		
Kutzner *et al.*[4]	Sclerotic myofibroma	Glomangiopericytoma	Myopericytoma
	Classical biphasic-type myofibroma		Solid myopericytoma
Requena *et al.*[3]	Cutaneous adult myofibroma		
	Vascular-type		
	Nodular or cellular type		
	Multinodular or biphasic type		
	Leiomyoma-like or fascicular type		

Table 5 Nomenclature for myopericyte tumours used in various publications (modified from Mikami and colleagues[11])

Publication	Myopericytoma type	Myofibroma type	Glomangiopericytoma
WHO (2002)[5]	Myopericytoma	Myofibroma	Glomangiopericytoma
Requena *et al* (1996)[7]		Cutaneous adult myofibroma:	
		Vascular type	
		Nodular or cellular type	
		Multinodular or biphasic type	
		Leiomyoma like or fascicular type	
Granter *et al* (1998)[4]	Myopericytoma	Myofibromatosis "infantile type"	Glomangiopericytoma
Kutzner (1998)[9]	Myopericytoma	Sclerotic myofibroma	Glomangiopericytoma
	Vascular	Classic biphasic myofibroma	
	Solid		

J Clin Pathol. 2006 January; 59(1): 67 – 73.
doi: 10.1136/jcp.2005.028704.

AJCC staging system for soft tissue sarcoma

G Histologic grade of malignancy
G1 Low, well differentiated
G2 Intermediate, moderately well differentiated
G3 High, poorly differentiated
G4 Poorly differentiated or undifferentiated (4-tier systems only)
N Regional Lymph Node
N0 No histologically verified metastasis to regional lymph nodes
N1 Histologically verified regional lymph node metastasis

T Primary Tumor
T1
T1a Superficial tumor*
T1b Deep tumor*
T2 Tumor greater than 5 cm in greatest dimension
T2a Superficial tumor
T2b Deep tumor
M Distant Metastasis
M0 No distant metastasis
M1 Distant metastasis

Stage I
T1a, b N0 M0, G1-2 (G1 with a 3-tier system)
T2a, b N0 M0, G1-2 (G1 with a 3-tier system)
Stage II
T1a, 1b N0 M0,G3-4 (G2-3 with a 3-tier system)
T2a N0 M0, G3-4 (G2-3 with a 3-tier system)
Stage III
T2b N0 M0, G3-4 (G2-3 with a 3-tier system)
Stage IV
Any T N1 M0, Any G
Any T N0 M1, Any G

* Superficial tumor is located exclusively above the superficial fascia without invasion of the fascia; deep

CHEST WALL SARCOMAS

Primary soft tissue sarcomas of the chest wall are accounting for 10 percent or less of all soft tissue sarcomas.

Their clinical behavior and prognostic factors are similar to those of extremity sarcomas, and they should be treated similarly.

In a series of 55 patients treated for primary soft tissue sarcomas of the chest wall over a 32-year period, five-year overall and disease-free survival rates were 87 and 75 percent, respectively.

Surgical resection is the treatment of choice for locally recurrent chest wall sarcomas.

Chest 2005 Mar;127(3):902-8.
J Clin Oncol. 2008 Nov 1;26(31):5113-8. Epub 2008 Sep 15.

Five year actuarial local control rates according to margin status after preoperative radiation for extremity soft tissue sarcoma

Margin	Number of patients	Local control, percent
+	27	81
-	106	97
≤1 mm	27	96
>1 mm	36	97
Undefined	21	94
No tumor*	22	100

* Specimen negative for tumor.

Data from Sadoski, C, Suit, HD, Rosenberg, A, et al, J Surg Oncol 1993; 52:223.

planned radical resection

Center	Number	Percent
Memorial Hospital	70	23
University of Florida	40	15
Swedish Sarcoma Group	103	8
Brigham & Women's Hospital	24	25
Netherlands Cancer Institute	26	19
Milan	417	31
National Cancer Institute	83	0 (amputation)

Center	Number of patients	Local failure, percent	Reference
Postoperative radiation			
Massachusetts General	176	14	Suit, HD, et al, 1990
MD Anderson	300	22	Lindberg, RD, et al, 1981
IGR	89	14	Abbatucci, JS, et al, 1986
RPMI	53	14	Karakousis, CP, et al, 1986
NCI	128	10	Potter, DA, et al, 1986
Toronto	23	9	Wilson, AN, et al, 1994

Preoperative radiation			
Massachusetts General	181	10	Suit, HD, et al, 1990
MD Anderson	110	10	Barkley, HT, et al, 1988
Toronto	39	3	Wilson, AN, et al, 1994
U. Florida	58	9	Brant, TA, et al, 1990
Intraoperative brachytherapy			
Memorial	55	18	Harrison, LB, et al, 1993
Mayo Clinic	63	8*	Schray, MF, et al, 1990

IA/IV doxorubicin and radiation			
UCLA	371	about 10	Eilber, FR, et al, 1993
U. Virginia	55	2	Wanebo, HJ, et al, 1995

* Mean follow-up 20 months.

Malignant myopericytoma: expanding the spectrum of tumours with myopericytic

Methods and results: Five cases of malignant myopericytoma were identified in the authors' consultation files. Tumours arose in three females and two males (median age 67 years, range 19–81 years) on the neck, arm, thigh and foot. One patient presented with disseminated metastases. One patient had a prior history of multiple benign myopericytomas in the same location. Four patients developed metastases and three died within 1 year.

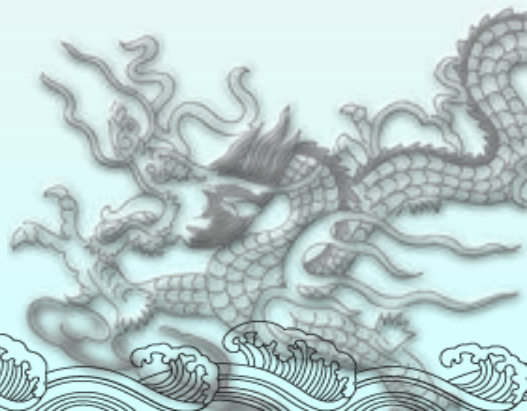
Histopathology. 41(5):450-460, November 2002.

Case	Sex/age	Site	Presenting symptoms and relevant history	Gross description	Treatment	Follow-up
1	F/80	Left side of neck	Rapidly growing painless mass Prior lentigo maligna excised at same location	20 mm mass	Marginal excision	AWD at 24 months (Liver metastases at 14 months)
2	M/46	Left thigh	Painful deep-seated intermuscular mass	130 mm myxoid mass with central firm area	Wide local excision and post-operative external beam radiation	DOD at 7 months Metastasis to liver, brain, bone and heart at 6 months
3	M/19	Heel of right foot	Painful mass Multiple benign myopericytomas on left foot × 6 years External beam radiation 5 years earlier	40 mm purple mass with extensive invasion into skeletal muscle and calcaneus bone	Below knee amputation	DOD within 1 year (exact time to death not available)

Histopathology. 41(5):450-460, November 2002

4	F/81	Left upper arm	Superficial painless mass	15 mm nodule	Local excision followed by wide excision	No recurrence after 18 months
5	F/67	Mediastinum	Short history of SVC obstruction; large mediastinal mass on imaging; rapid development of skin metastases	100 mm mediastinal mass (not biopsied). Circumscribed 8 mm skin nodule	Excision of skin nodule	DOD within 1 month Respiratory failure and metastases to skin and subcutis

AWD, Alive with disease; DOD, dead of disease.



Histopathology. 41(5):450-460, November 2002

Take home messages

We support the use of the term myopericytoma (MPC) to describe a spectrum of tumours typified by a haemangiopericytoma-like vascular architectural pattern with features of perivascular myoid (myopericytic) differentiation

MPC shows a spectrum of growth patterns that overlap with myofibroma (MF)

Tumours can be categorised as MPC or MF depending on the growth pattern: the presence of a concentric perivascular arrangement of plump spindle shaped cells in MPC and a zonation/biphasic appearance in MF

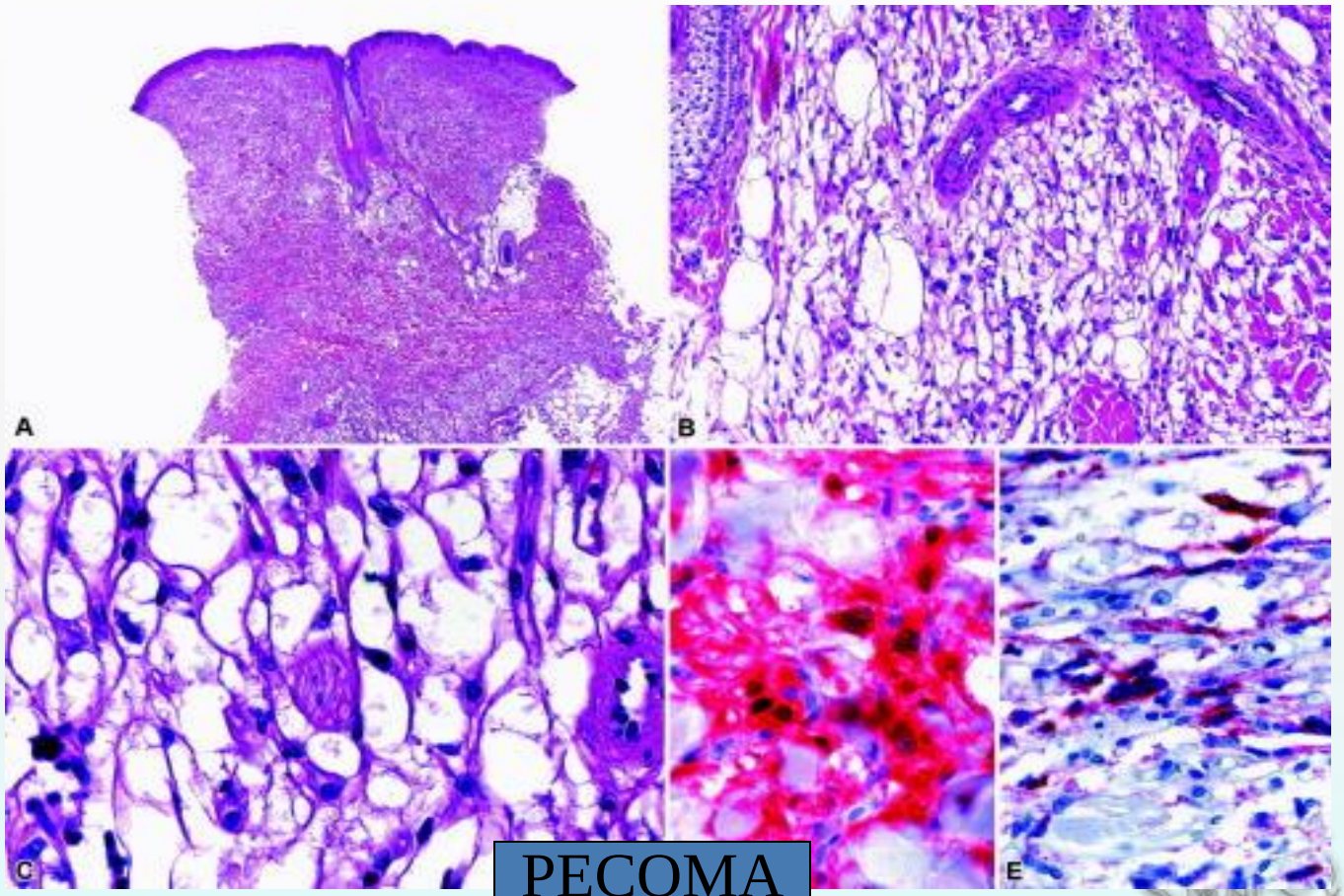
We suggest that the glomangiopericytoma-like pattern should be included within the MPC group

J Clin Pathol. 2006 January; 59(1): 67–73.

PECOMAS

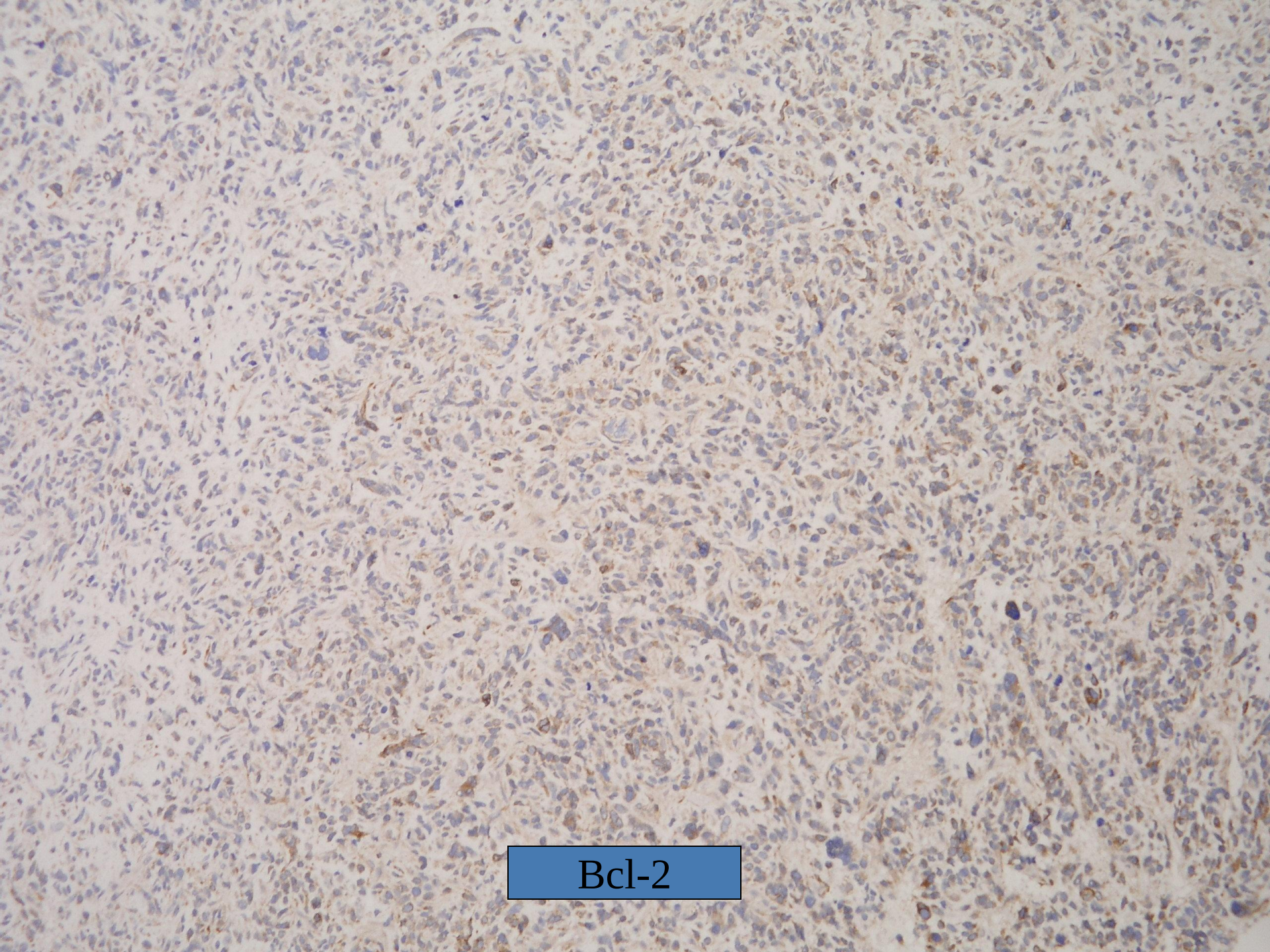
PECs are typically round to polygonal with distinct cell borders and ample clear to eosinophilic granular cytoplasm. They have a centrally placed round to oval nucleus with an inconspicuous nucleolus. Spindle cell forms also occur. The pathognomonic feature of the PEC is its unusual immunohistochemical staining pattern. These cells coexpress melanocytic (HMB-45, Melan-A, MITF, NKI-C3, tryrosinase) and myogenic (actin, desmin, myosin, calponin) markers.

Am J Surg Pathol. 1992;16:307–308.



PECOMA

Pathology Case Reviews. 13(6):247-257, November/December 2008.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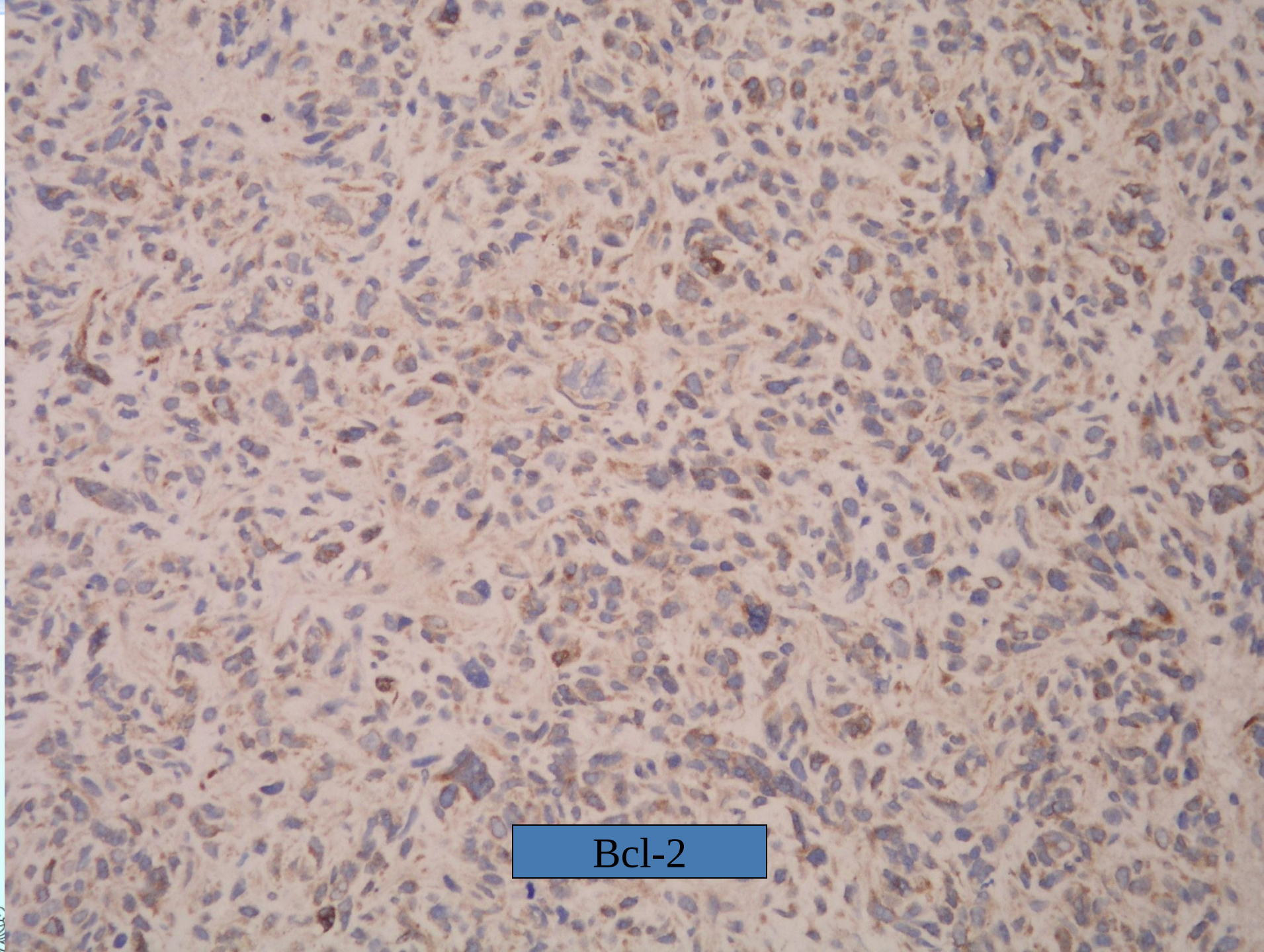
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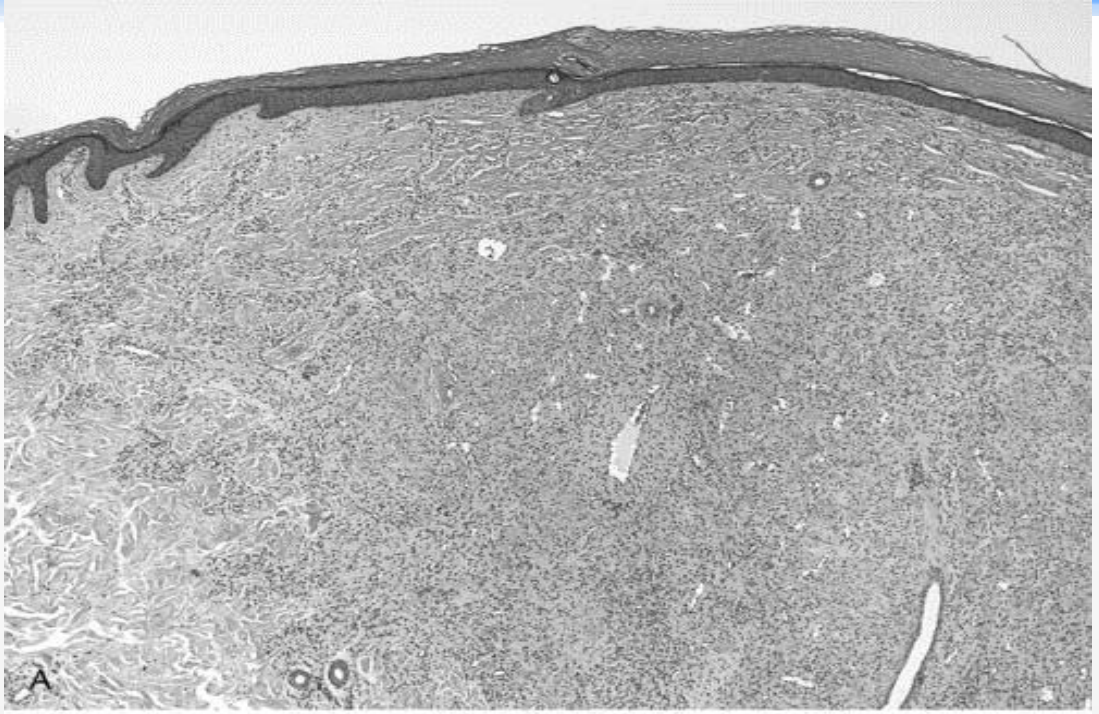
Antibody	Clone Designation	Vendor	Working Dilution	Method of Epitope Retrieval
Smooth muscle myosin heavy chain	SMMS-1	Dako (Carpinteria, CA)	1:200	20 min pH 6 citrate buffer followed by 10 min 0.1% pronase
Calponin	CALP	Dako (Carpinteria, CA)	1:100	20 min pH 6 citrate buffer followed by 10 min 0.1% pronase
p63	4A4	LabVision (Fremont, CA)	1:250	8 min pH6 citrate buffer
CD10	56C6	Vector Laboratories (Burlingame, CA)	1:100	8 min pH6 citrate buffer
CK5/6	D5/16 B4	Dako (Carpinteria, CA)	1:50	20 min pH 6 citrate buffer followed by 10 min 0.1% pronase

Am J Surg Pathol, Volume 30(8).August 2006.1002-1007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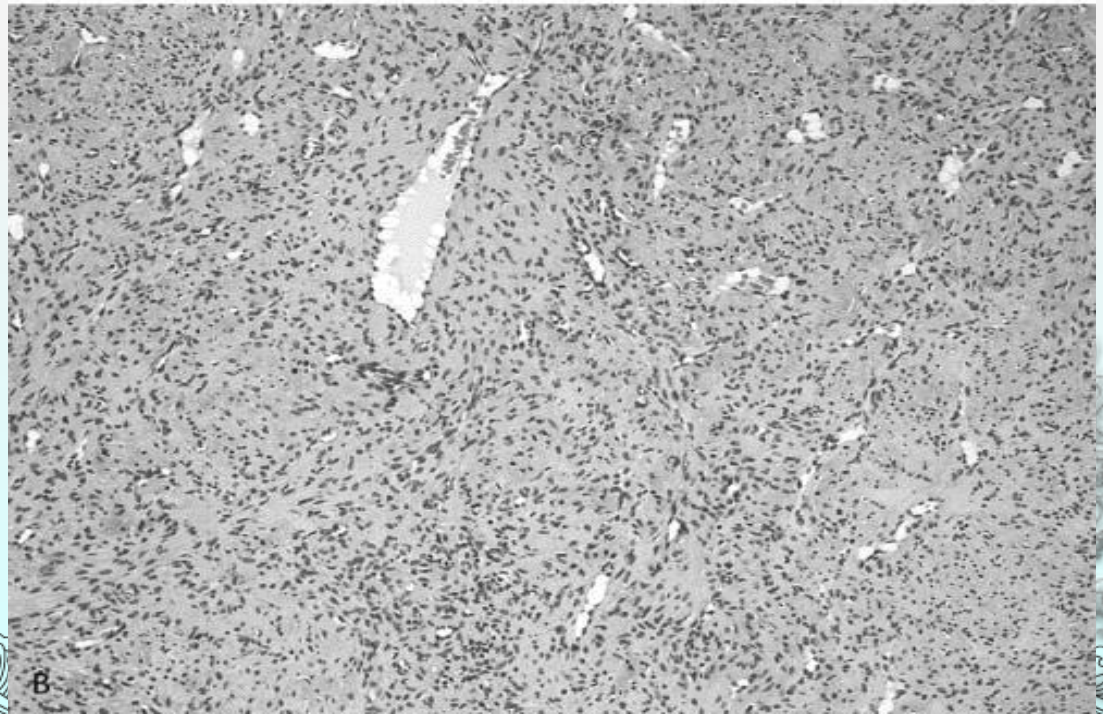
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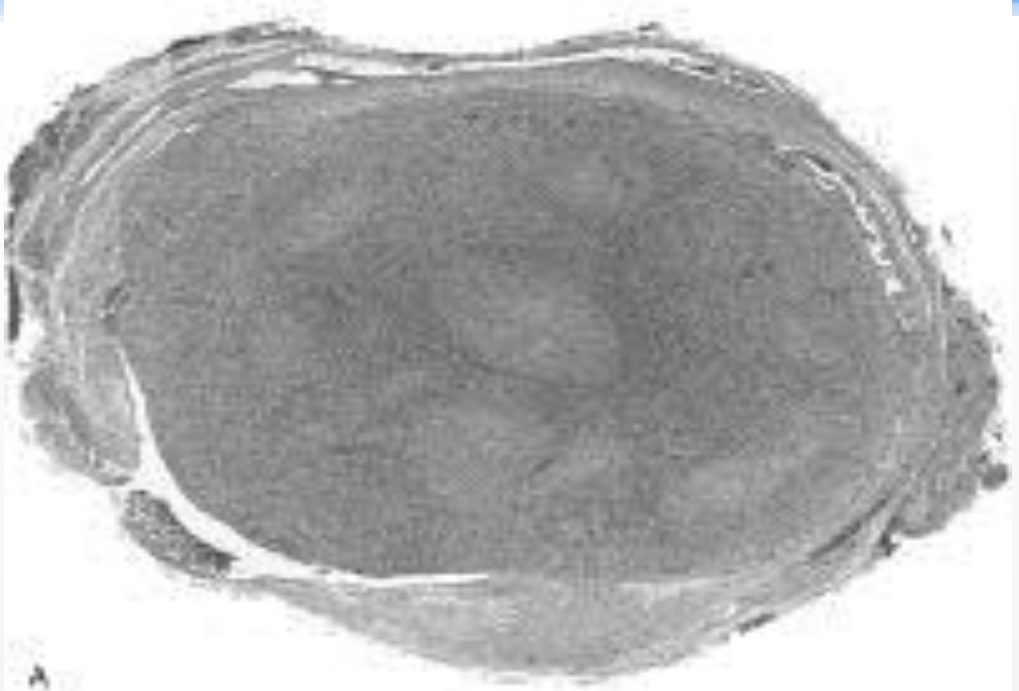
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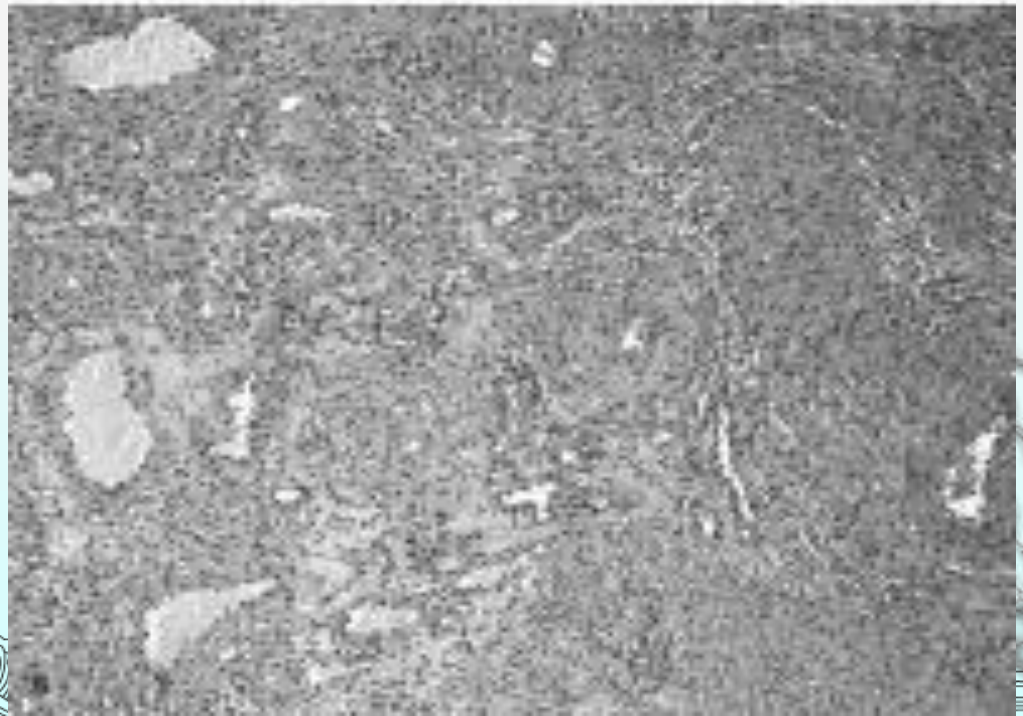

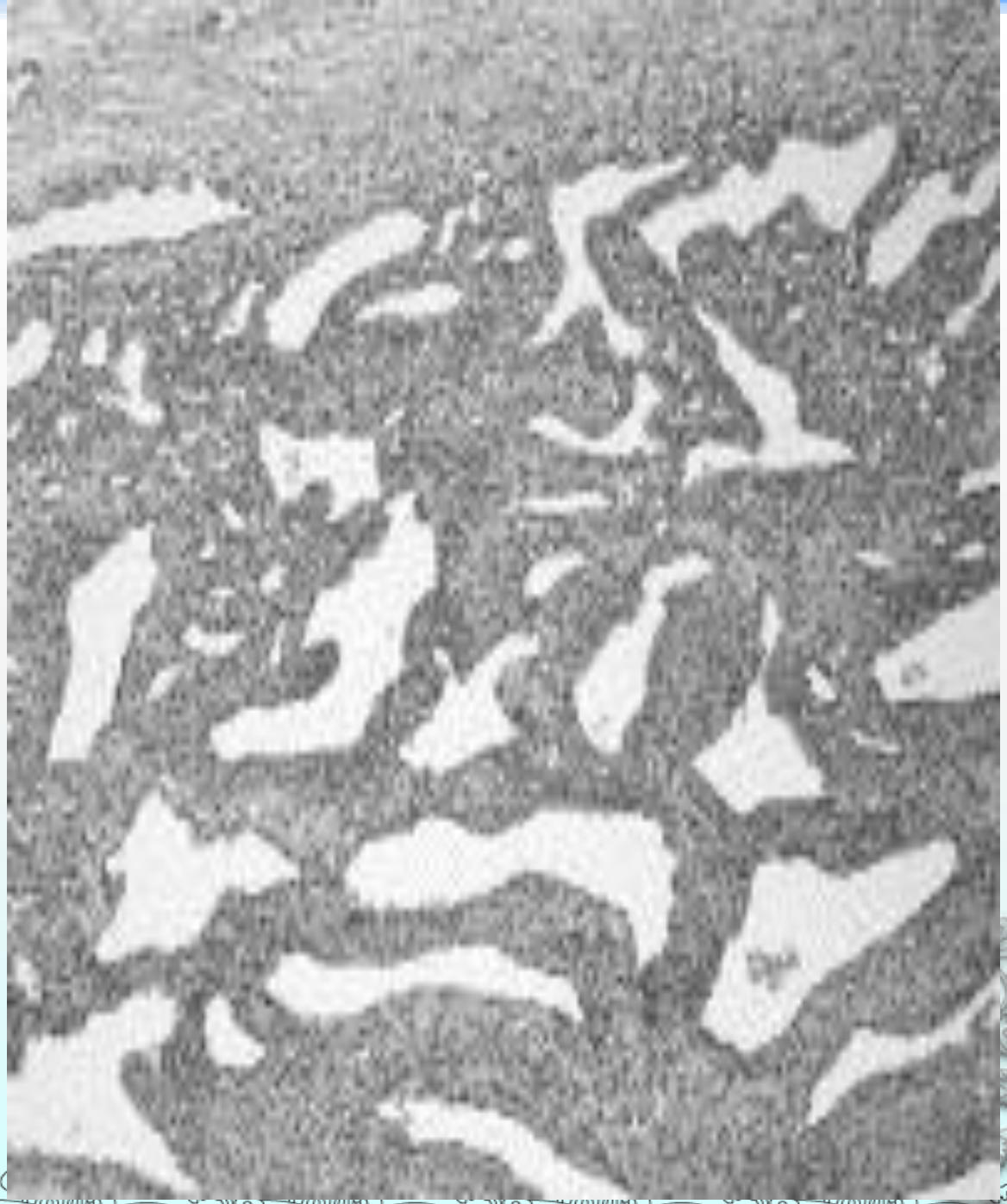

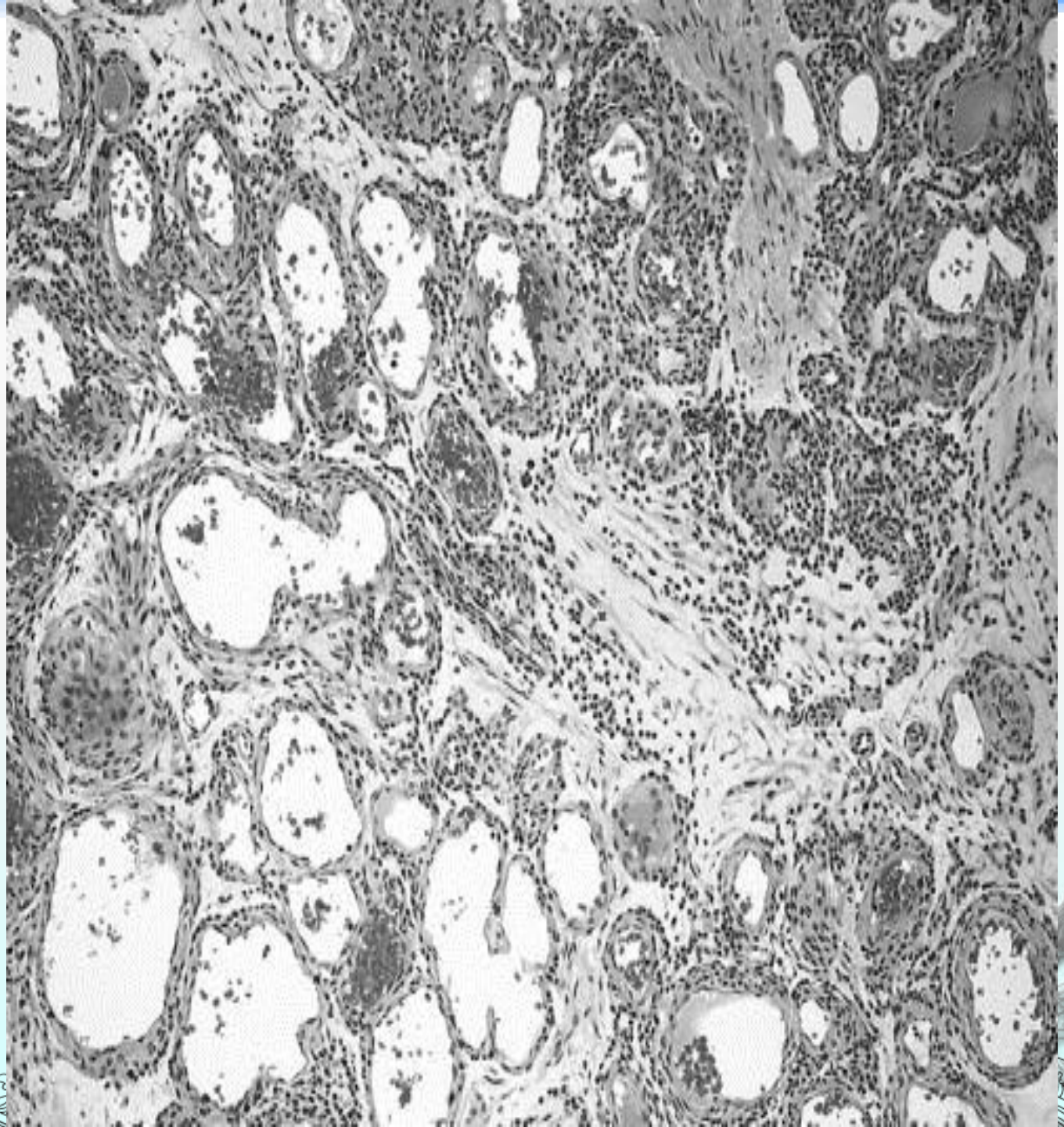

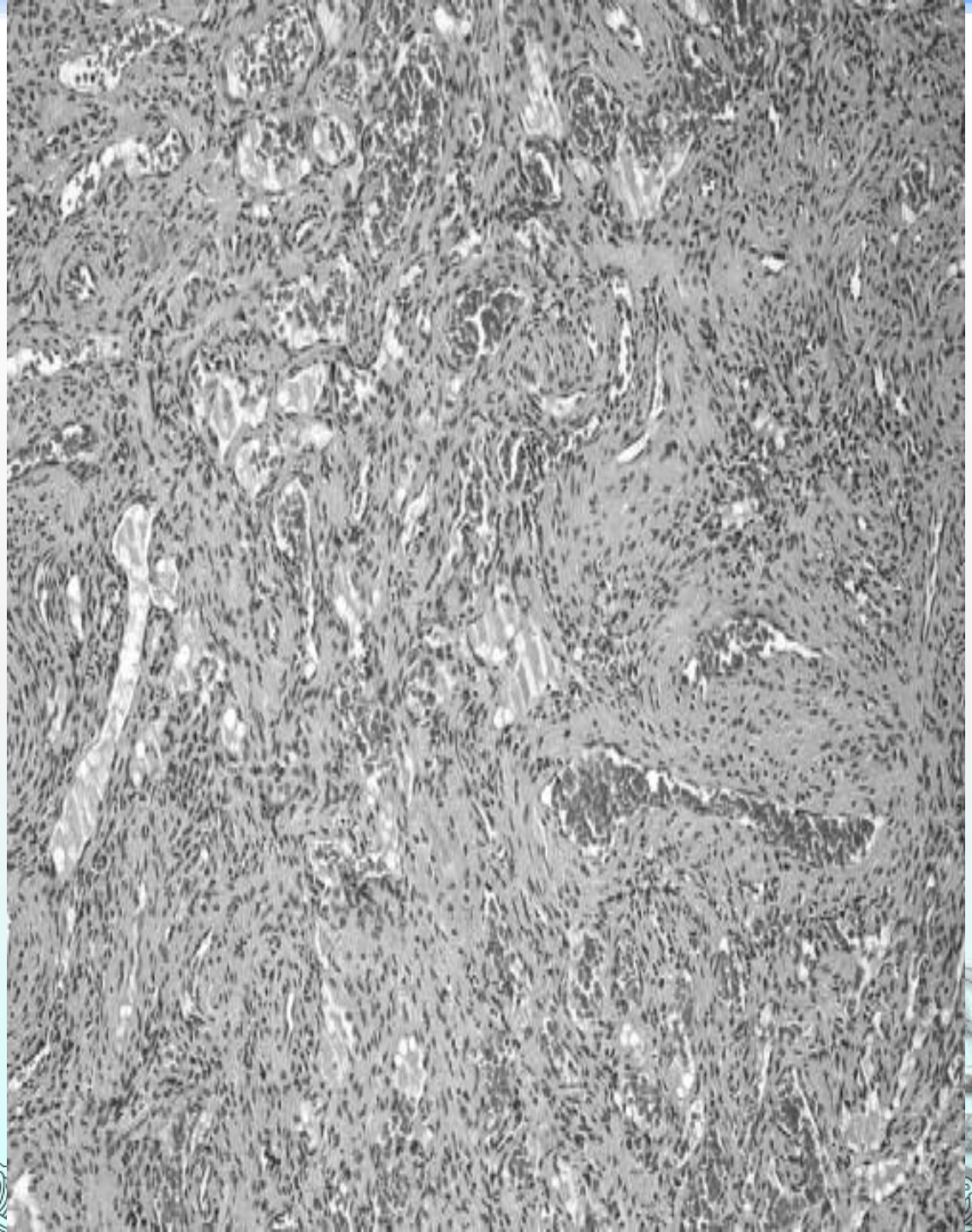

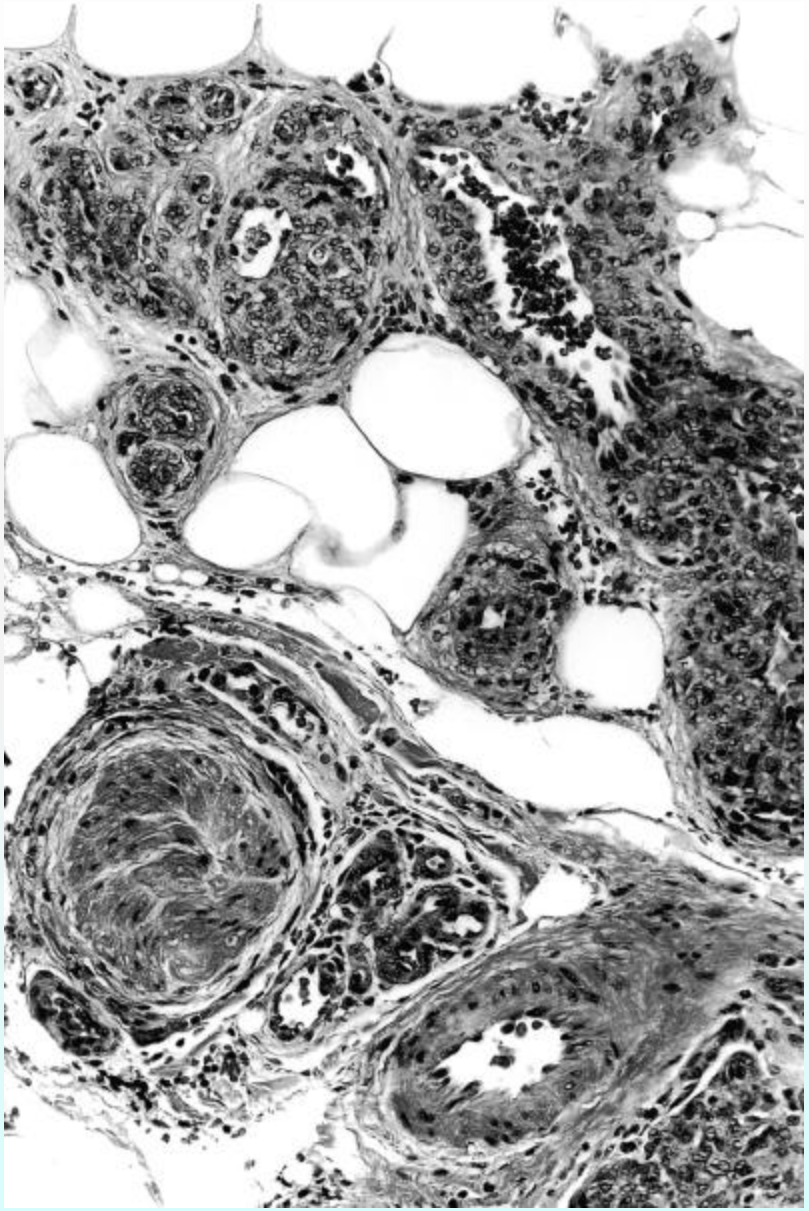

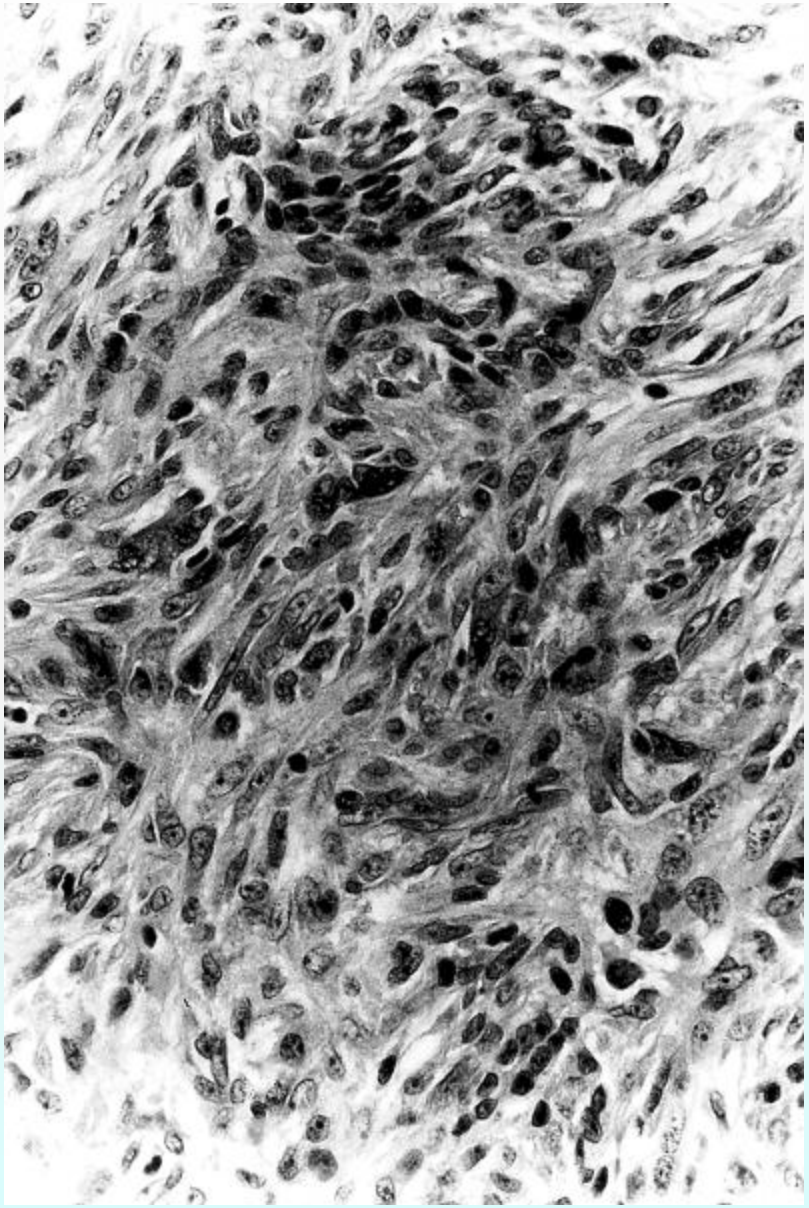

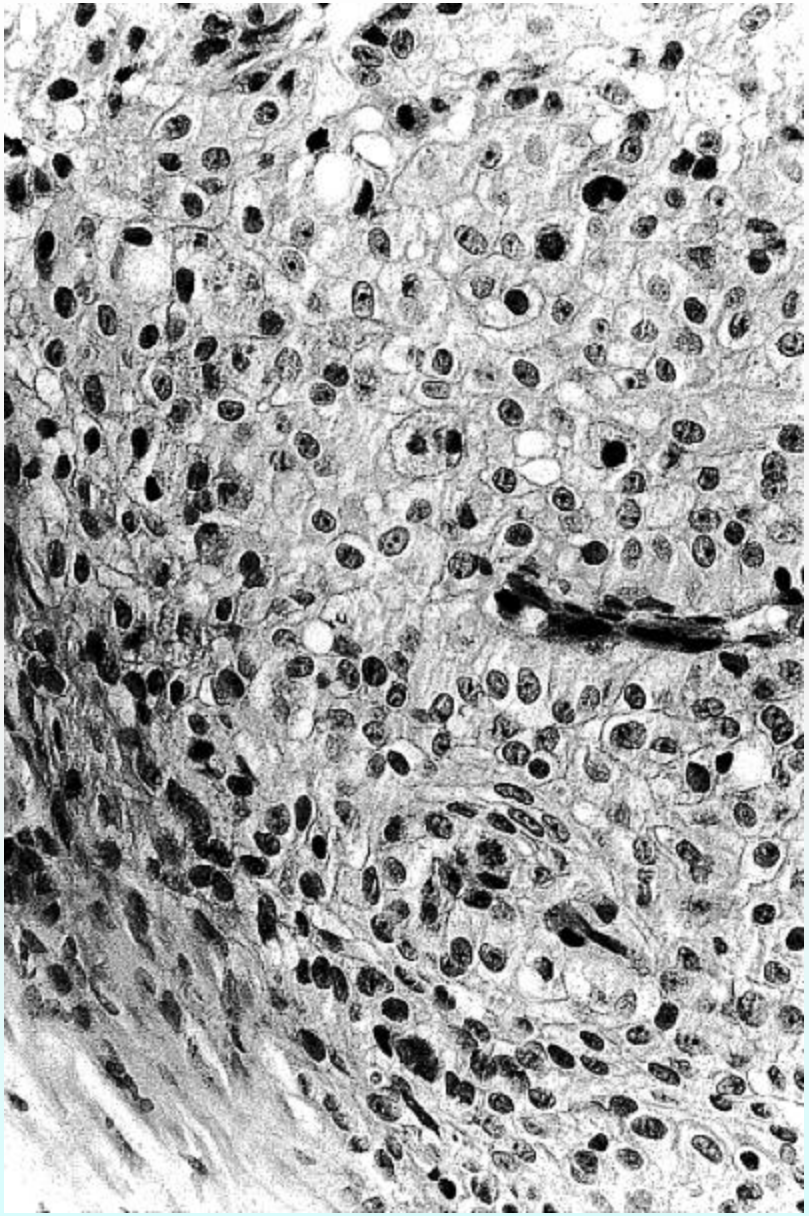

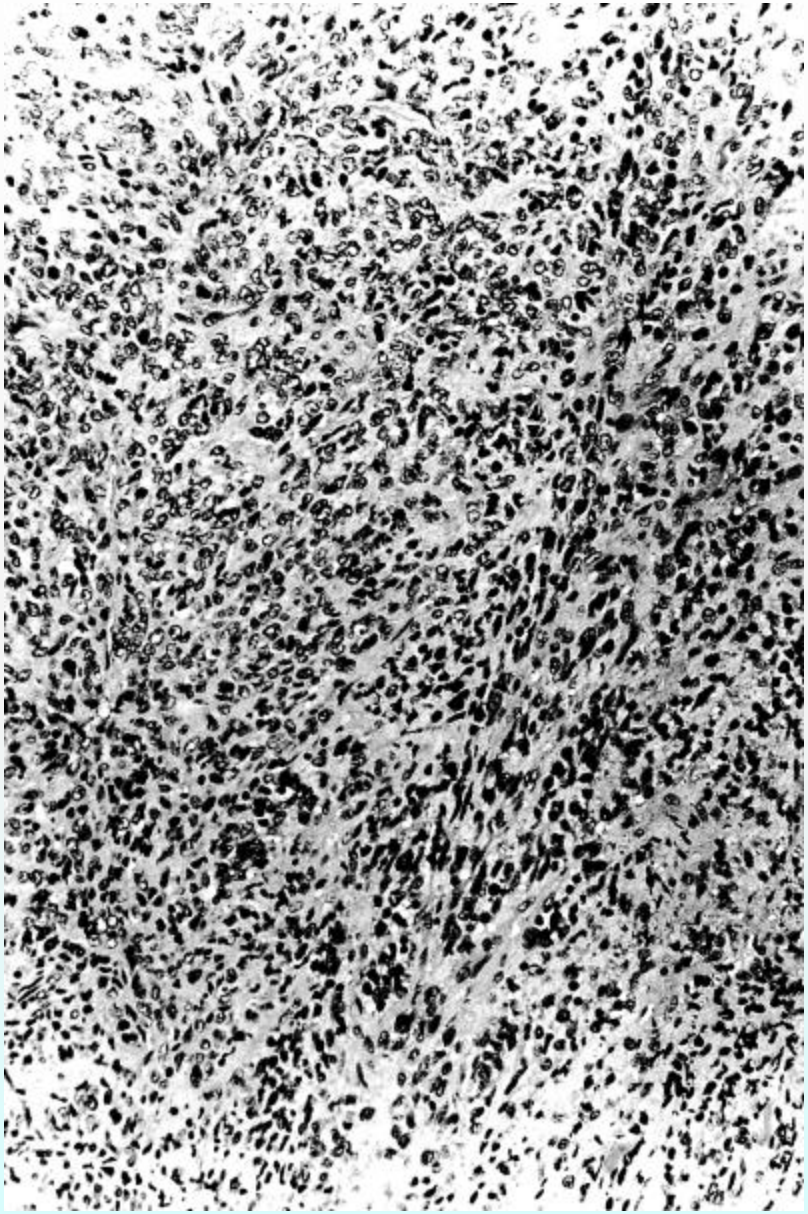

Five year actuarial local control results, margin status, and size of sarcoma after preoperative radiation therapy for extremity soft tissue sarcoma

Size, mm	Margin positive		Margin negative	
	N	LC	N	LC
<25	-	-	7	100
26-50	2	100	15	93
51-100	14	78	34	100
101-150	5	100*	22	100
151-200	2	100•	21	87
>200	4	100	7	100

LC: local control, values in percent.
* Only 2 year follow-up.
• Only 1 year follow-up.

Haemangiopericytoma

Immunohistochemistry

vimentin	positive (intensity variable)
factor XIIIa	50% of cases
muscle-specific actin	a few cases
CD57	a few cases
desmin	negative
CD31	negative
cytokeratin	negative
S-100	negative