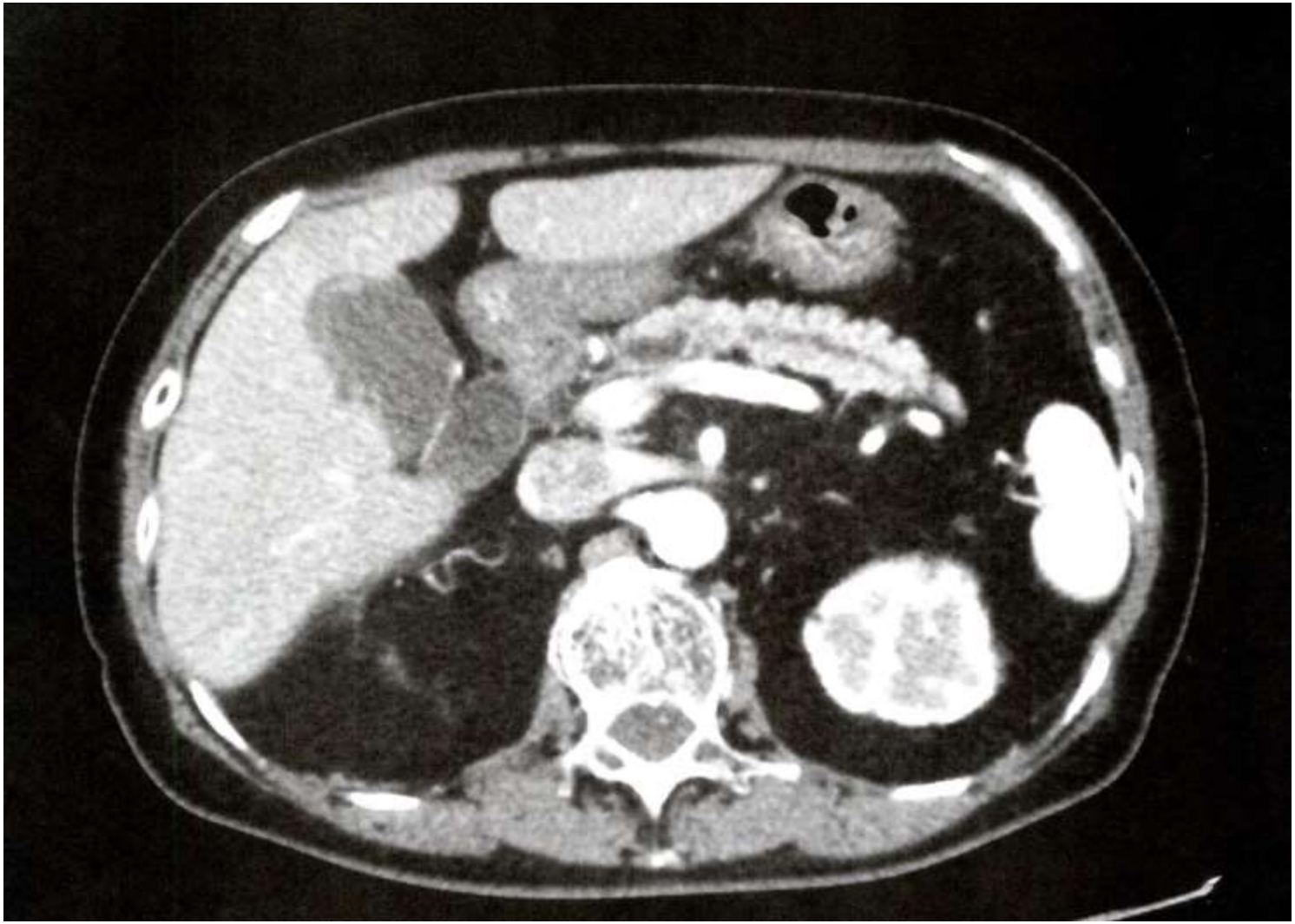
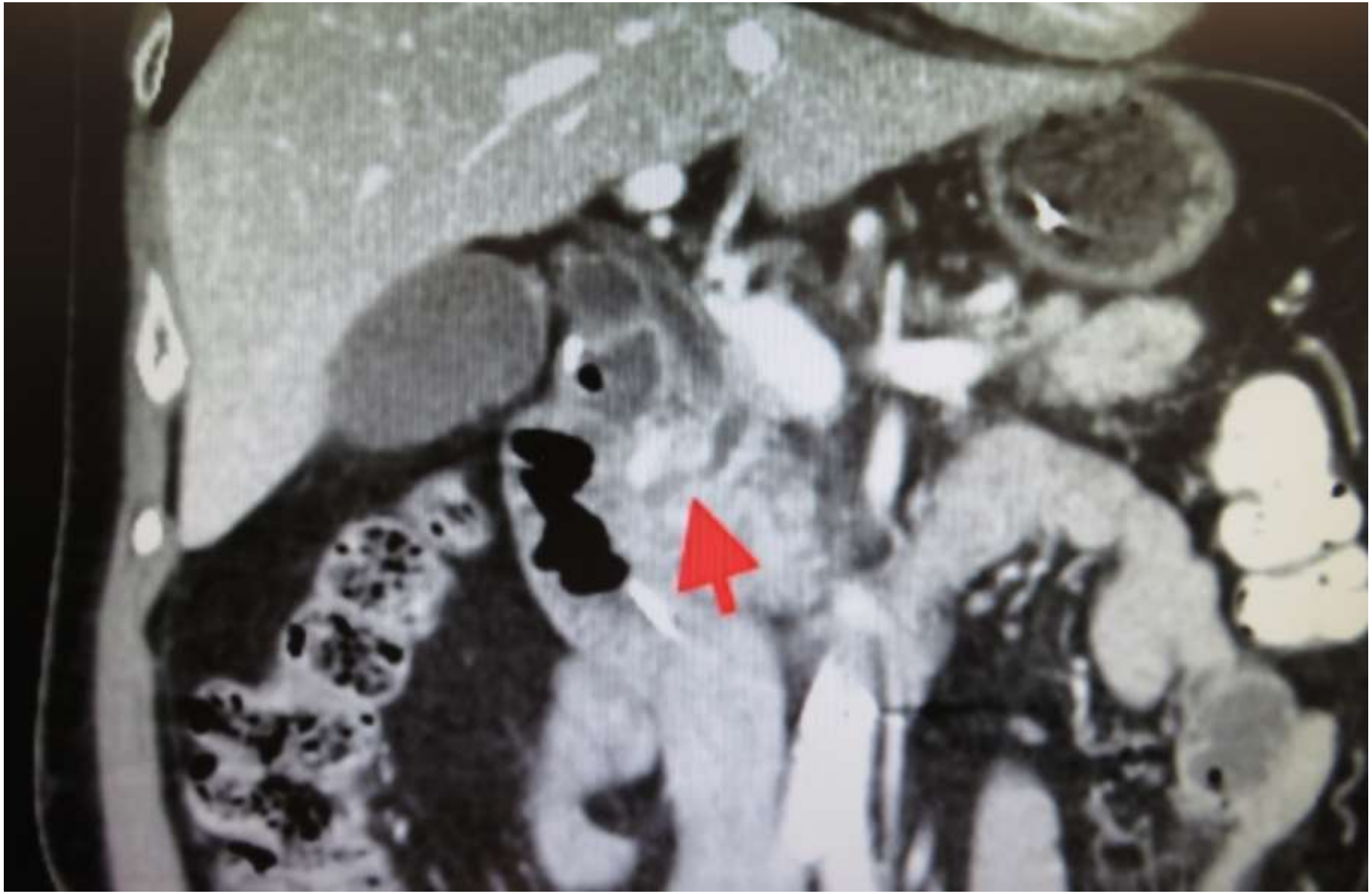
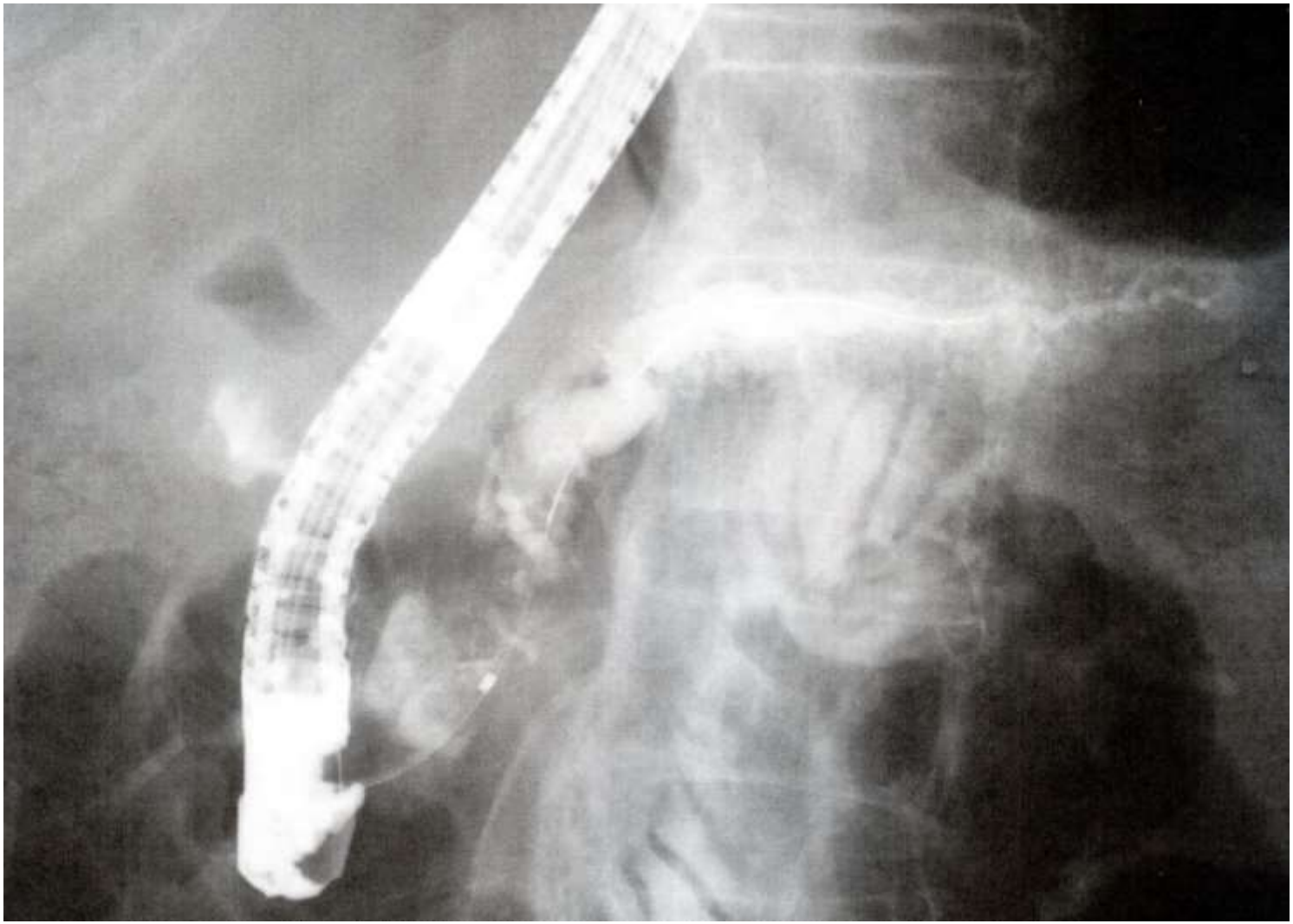
造影CT



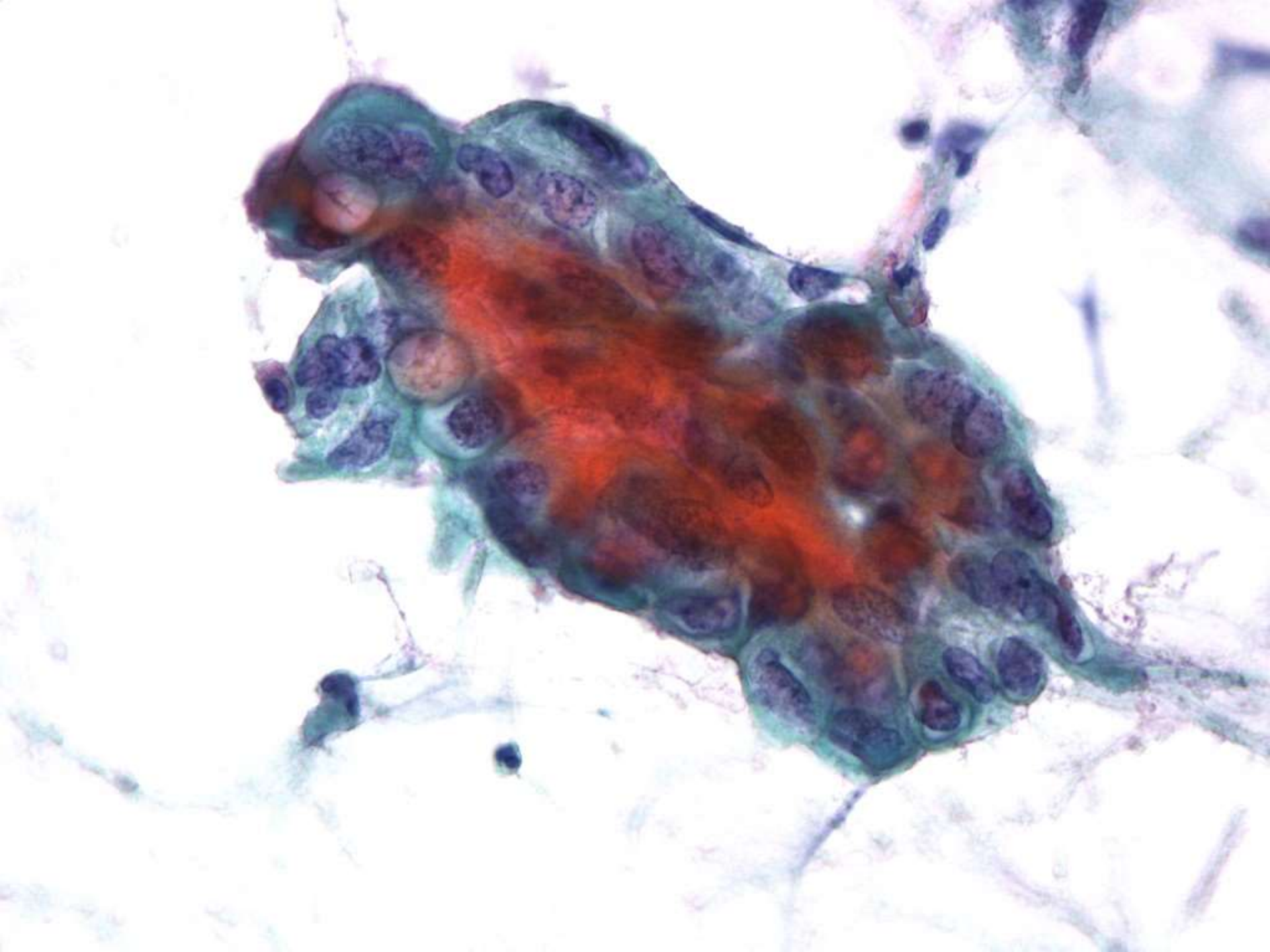
造影CT

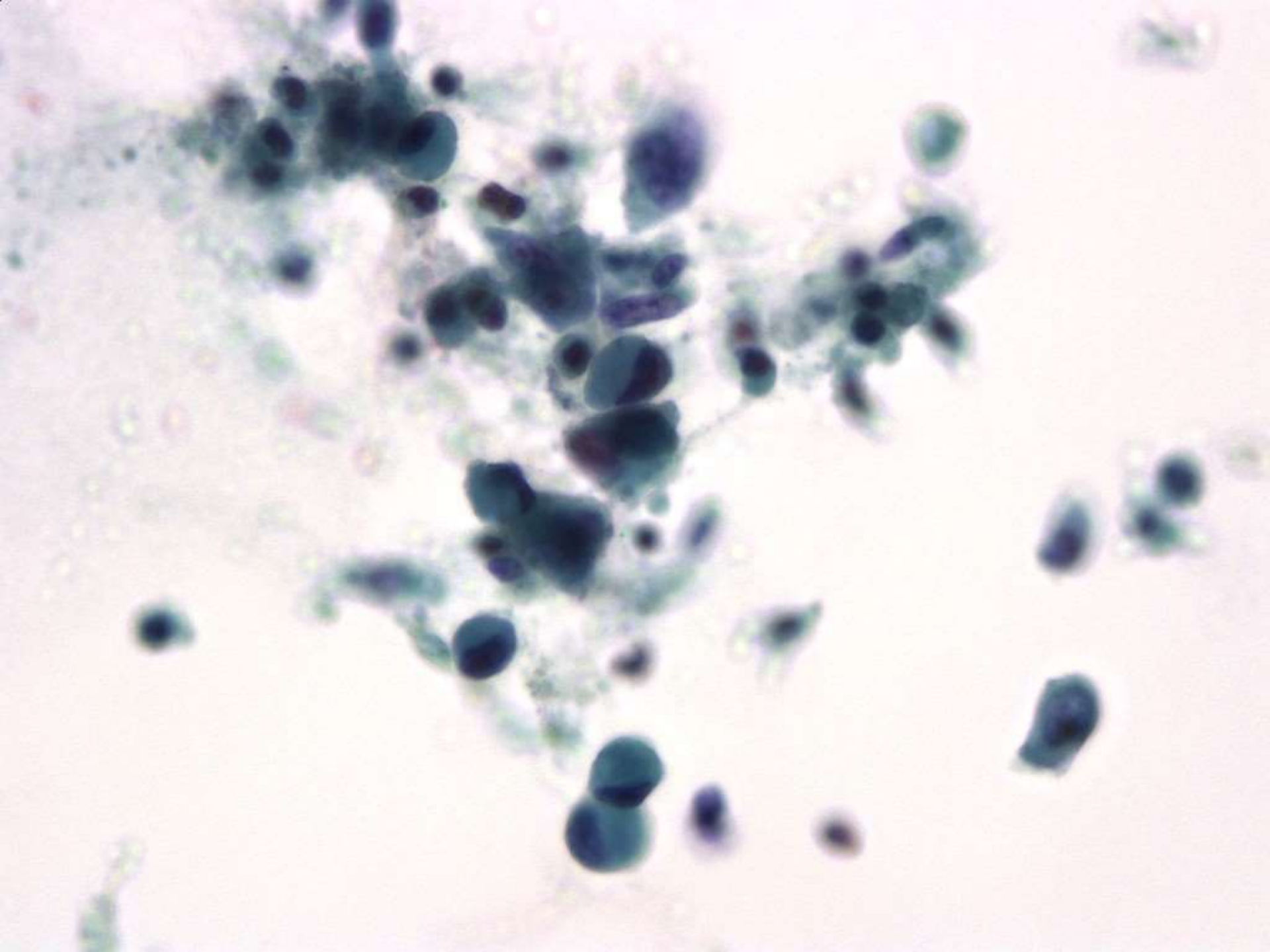


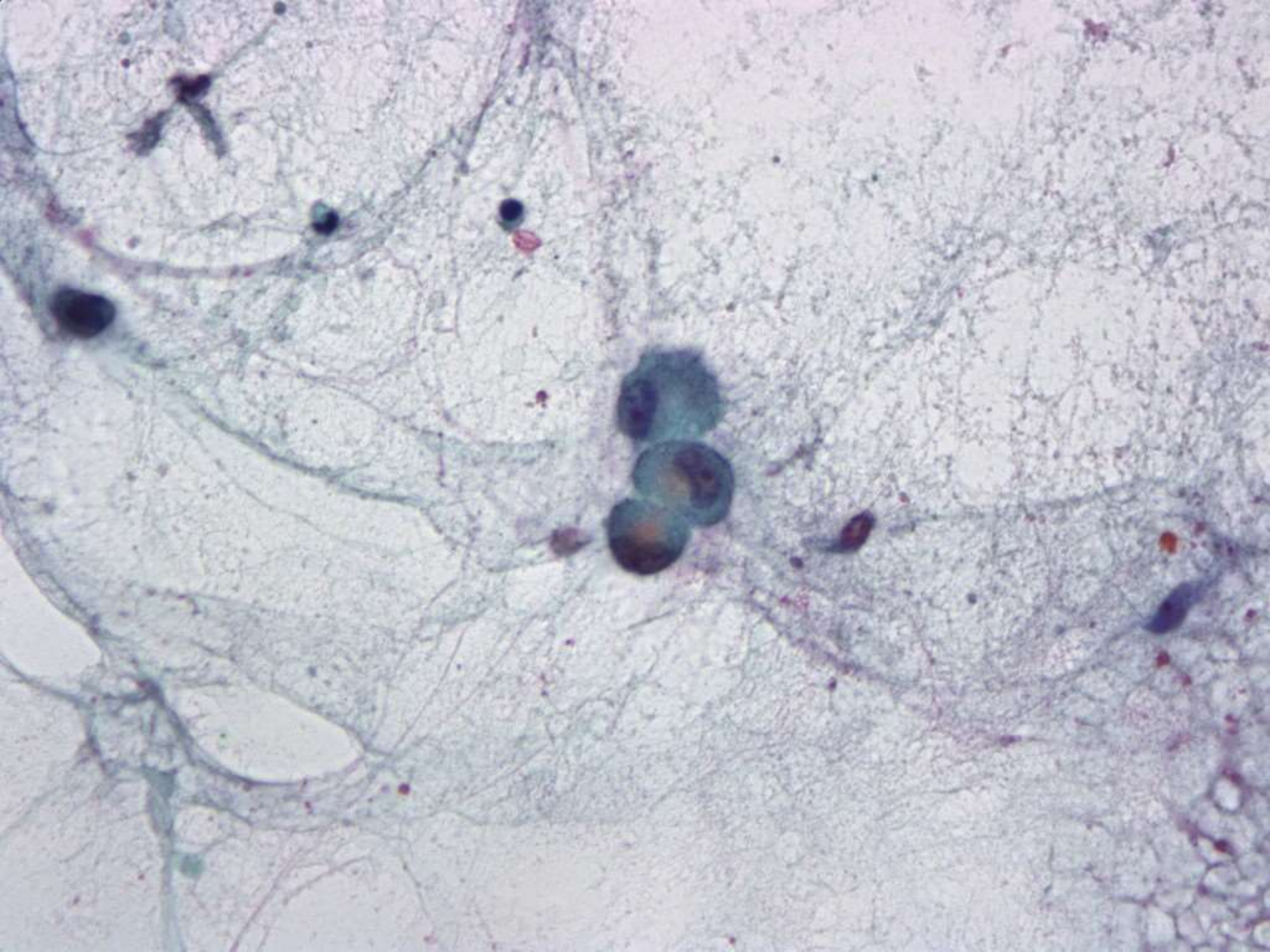
ERCP



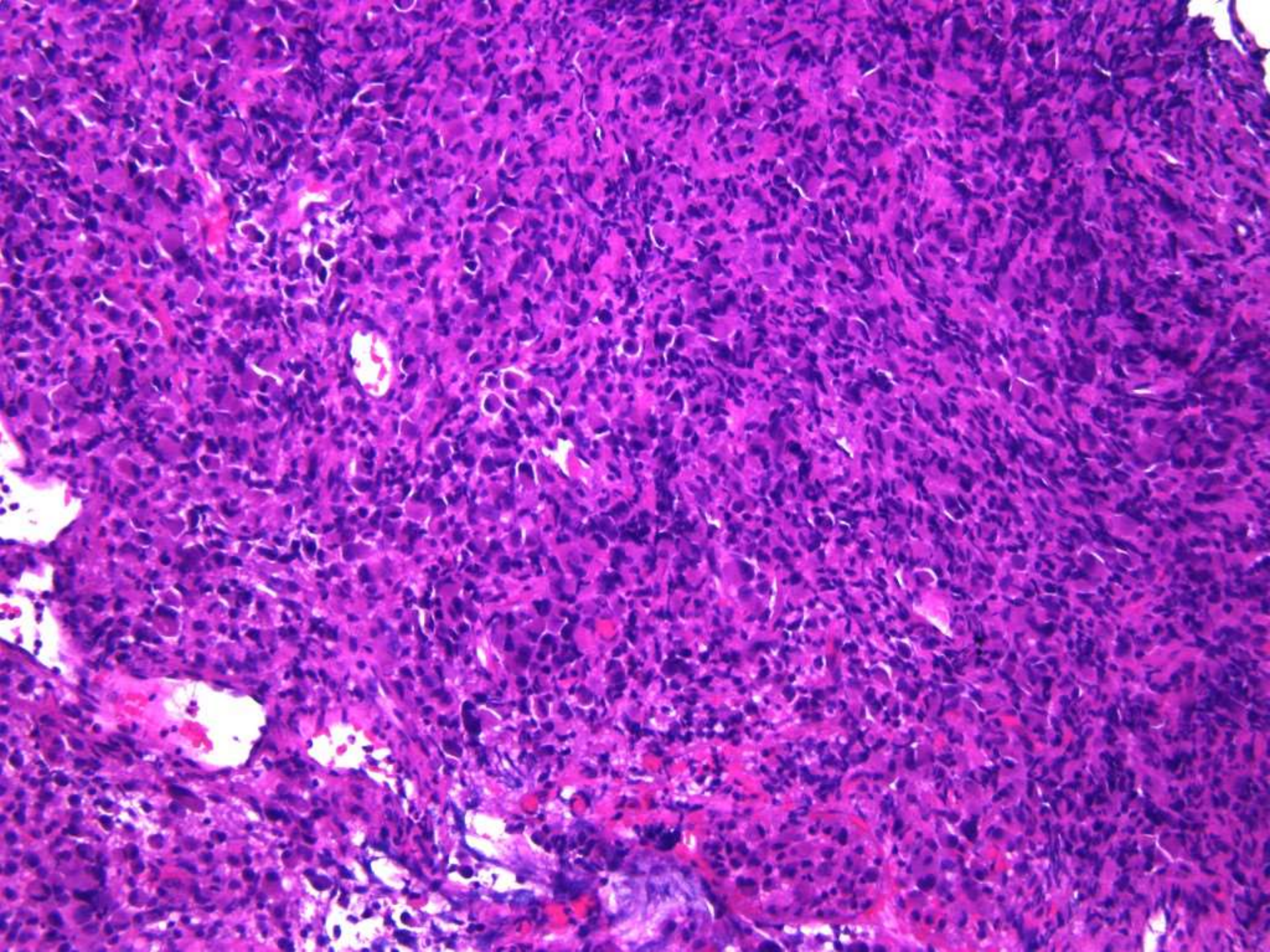
膵液細胞診標本

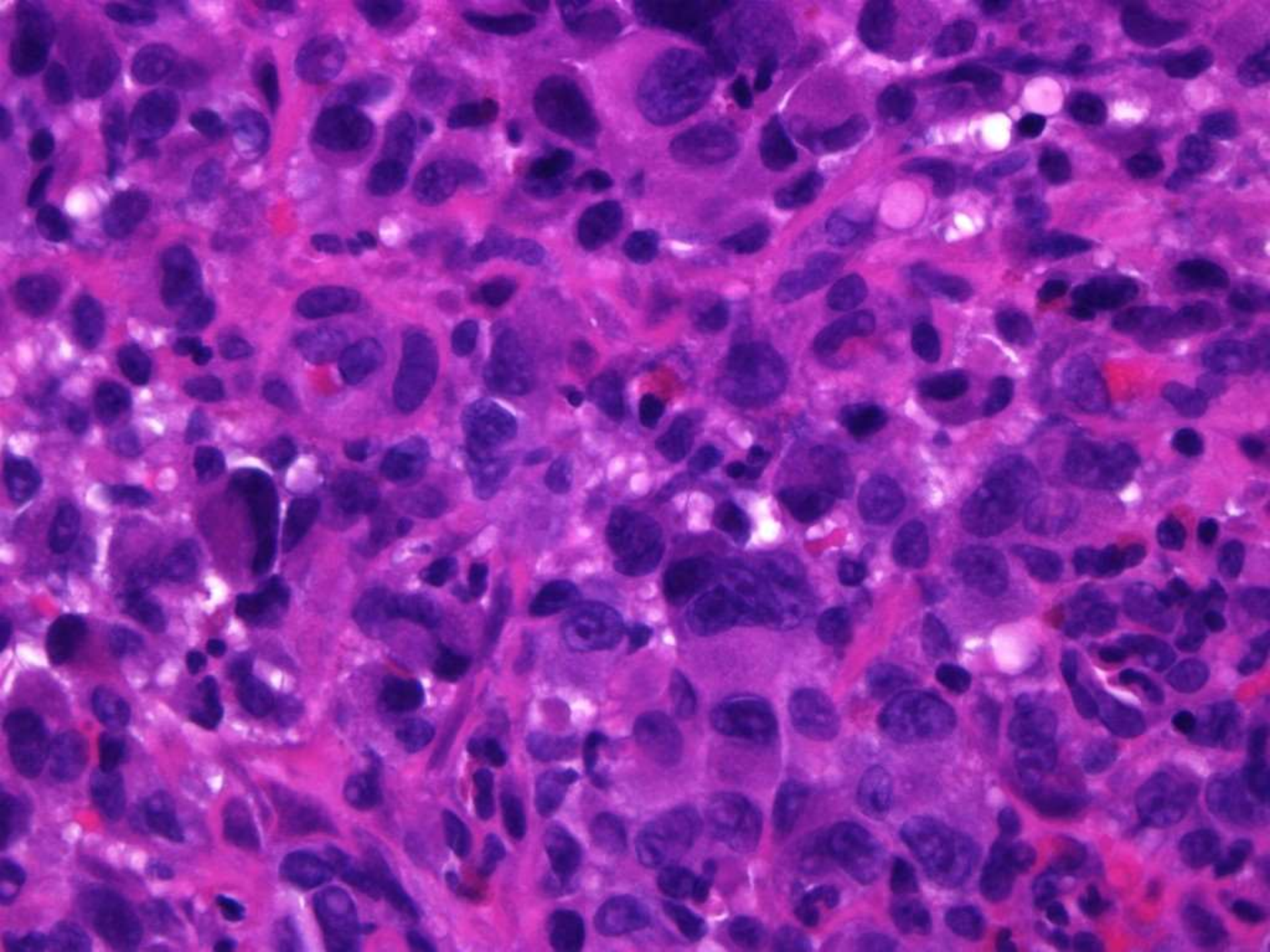


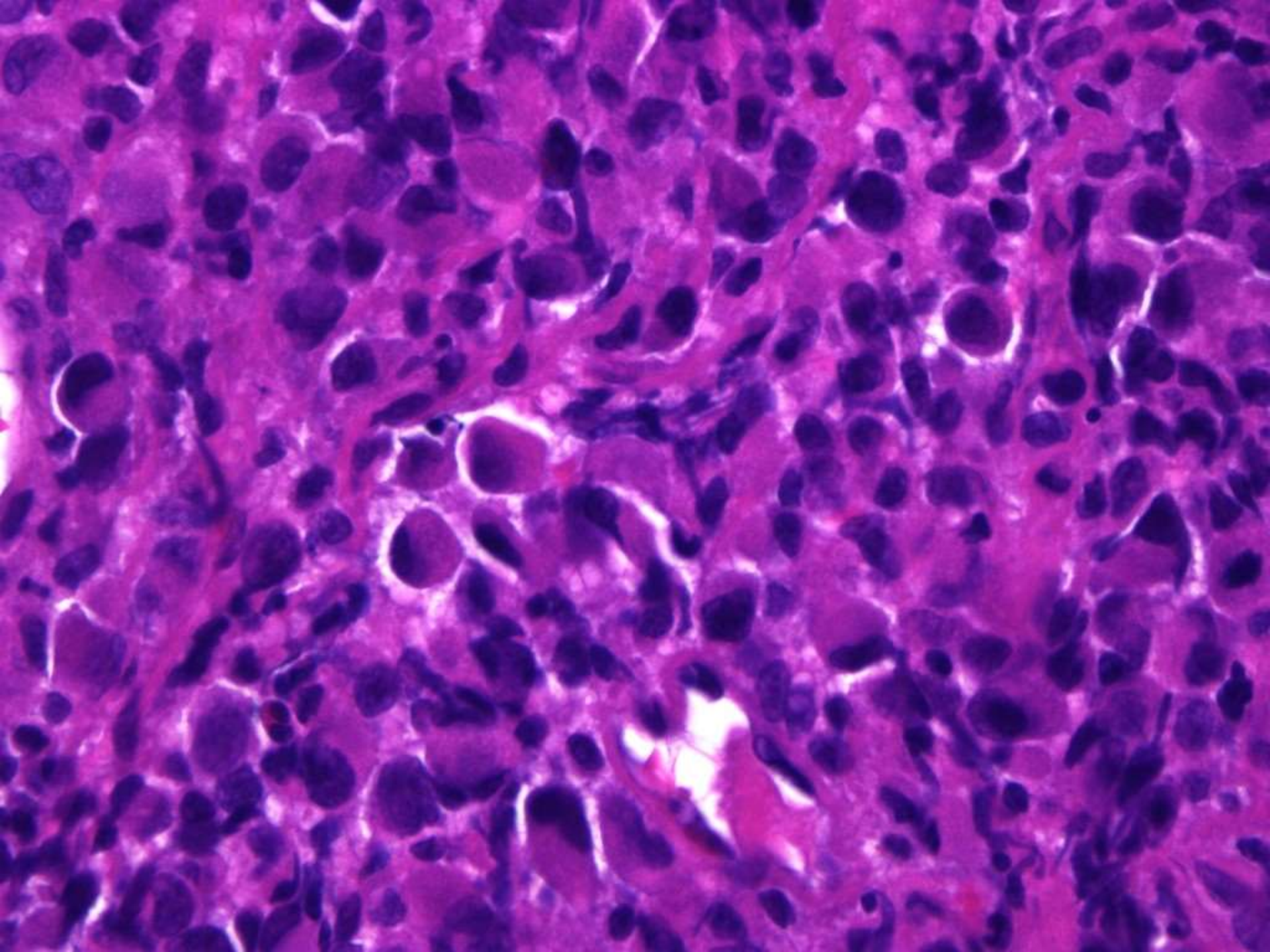


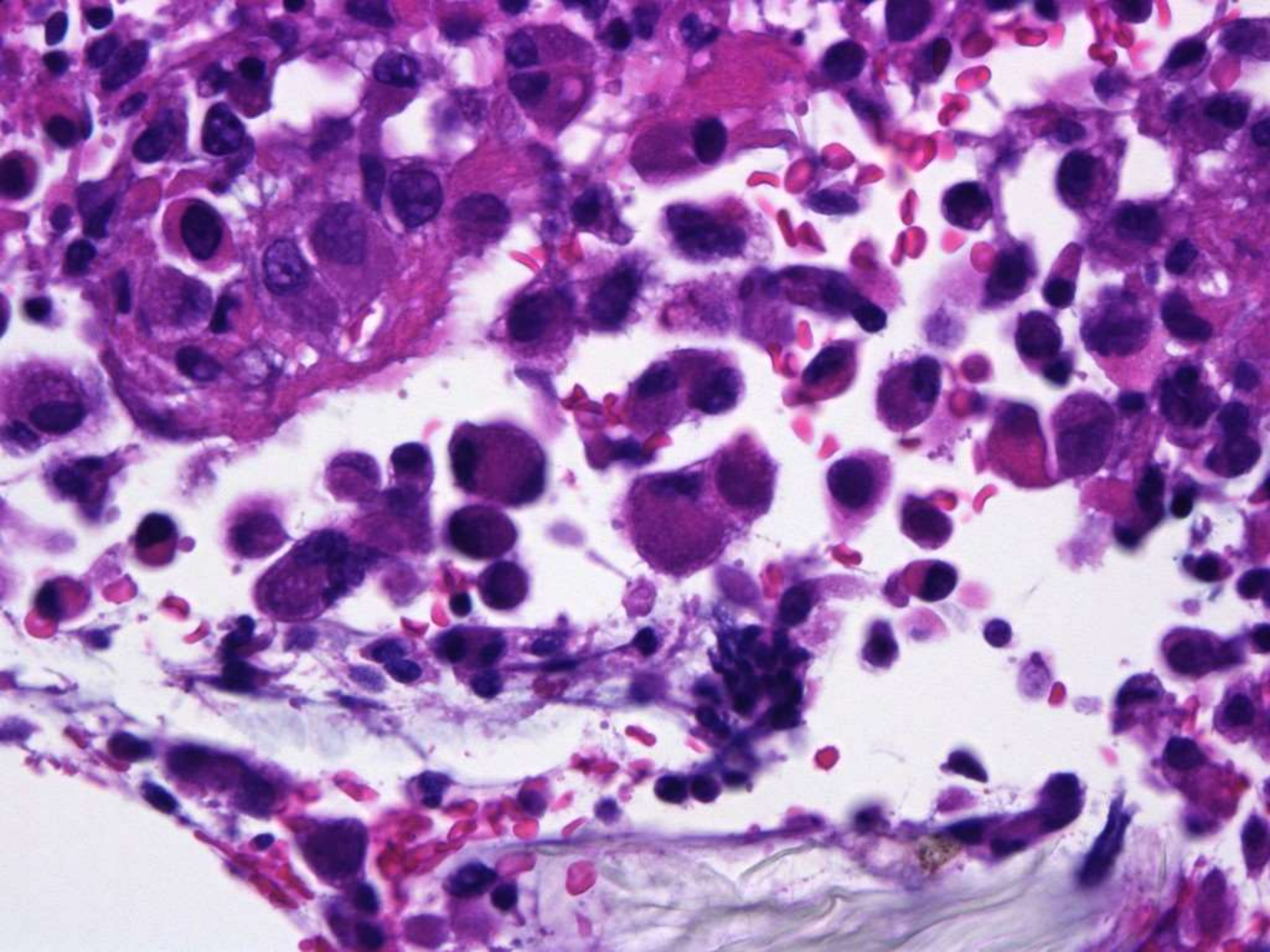


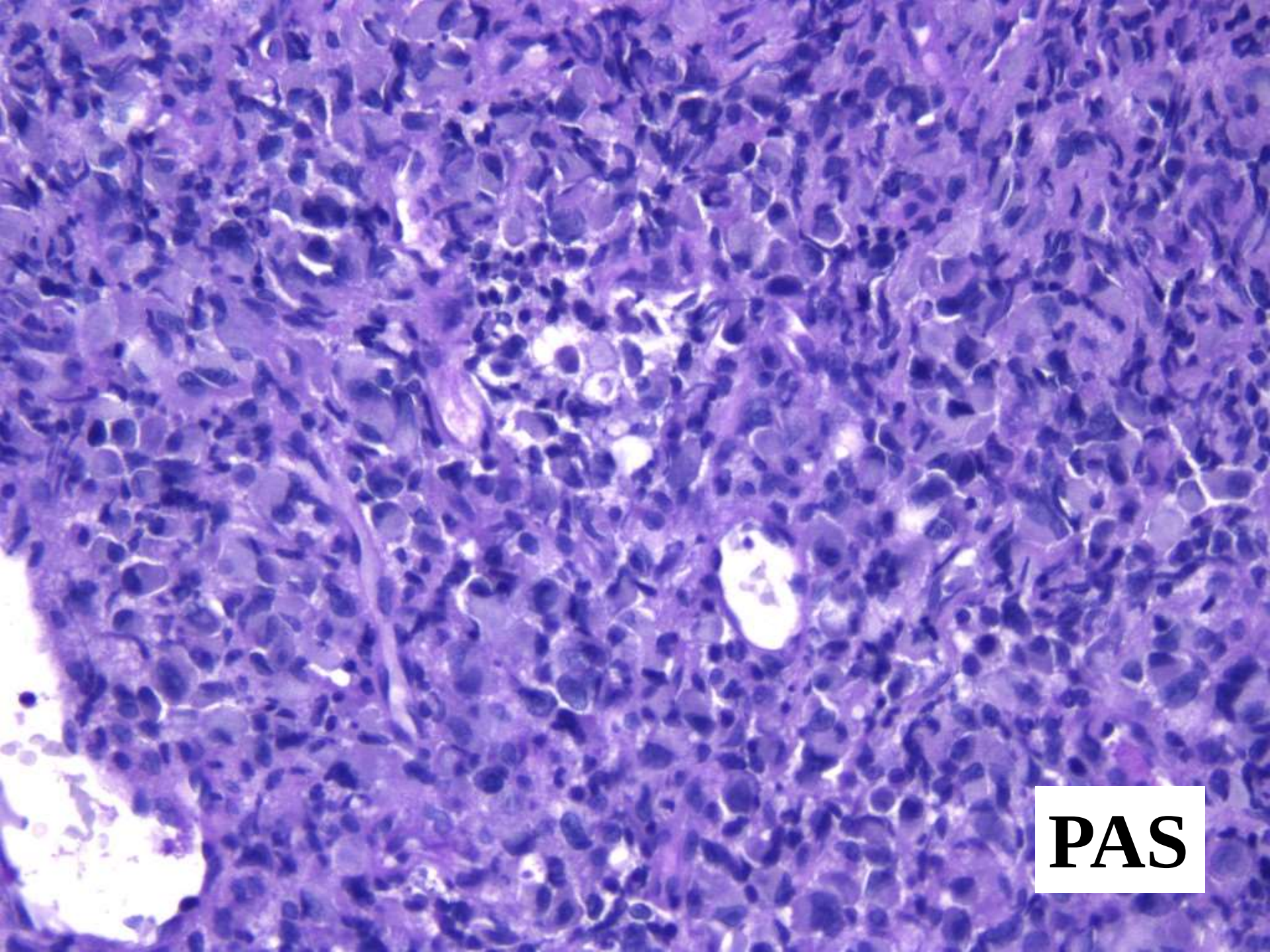
膵管生検
標本

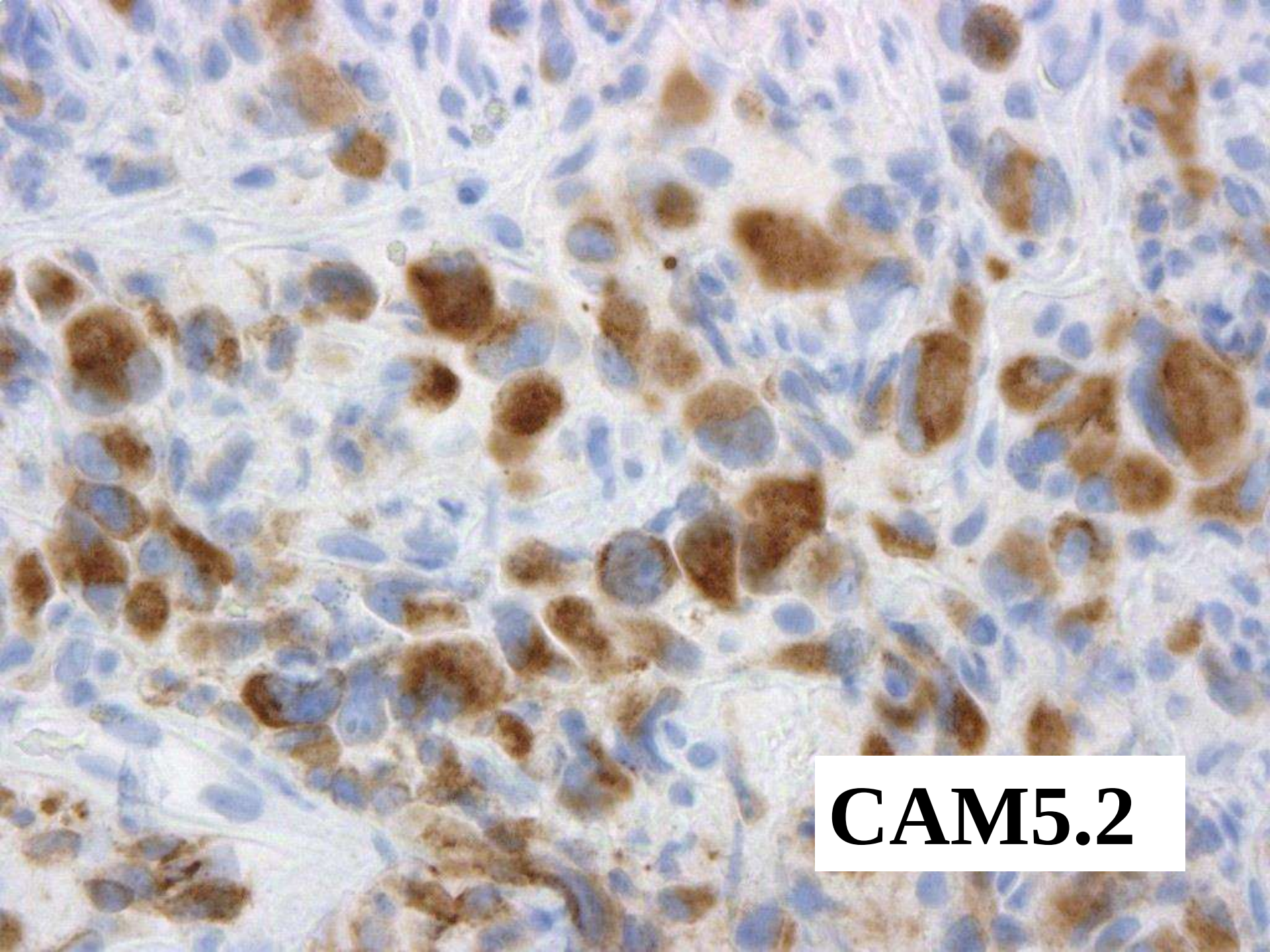


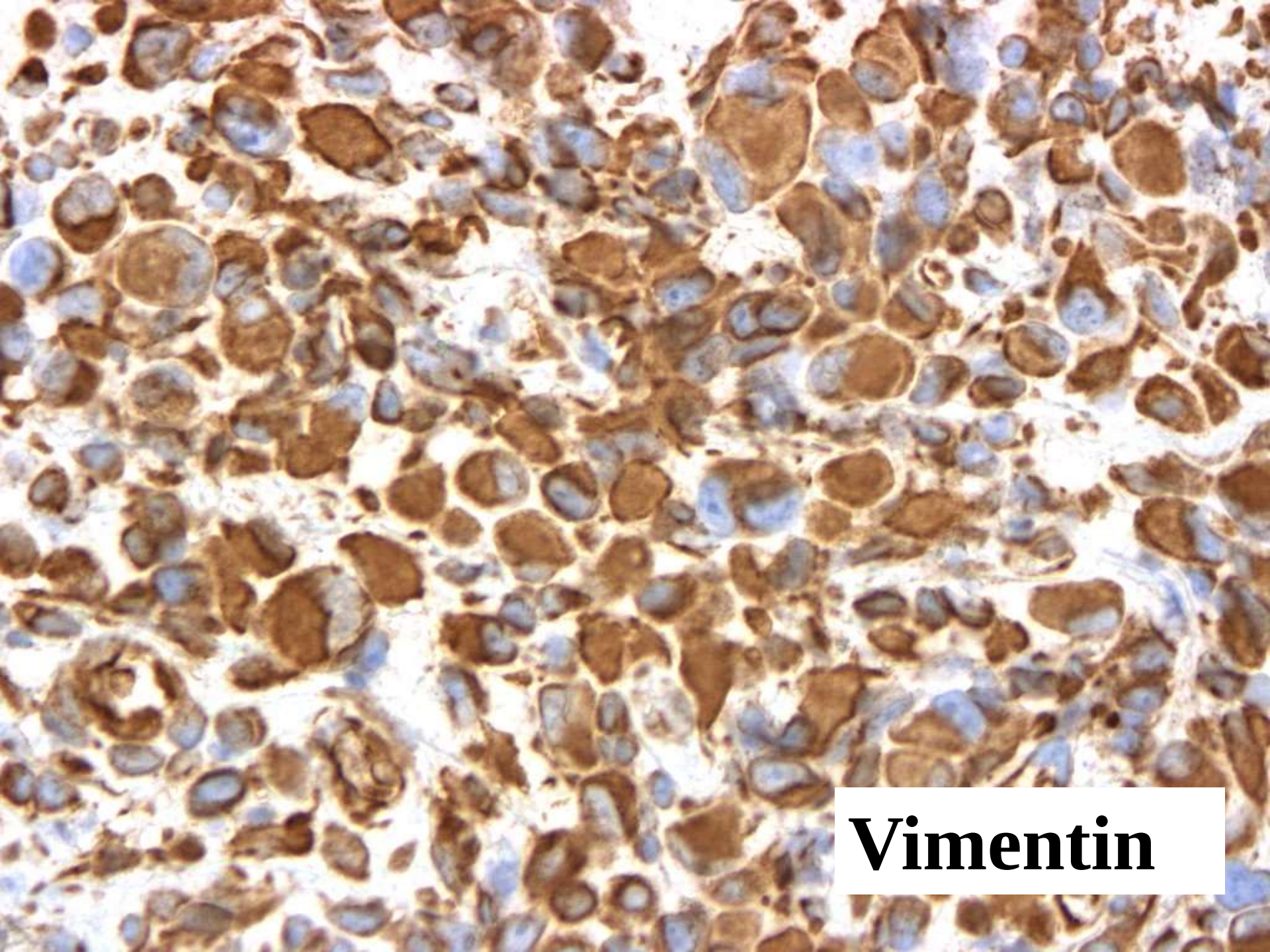


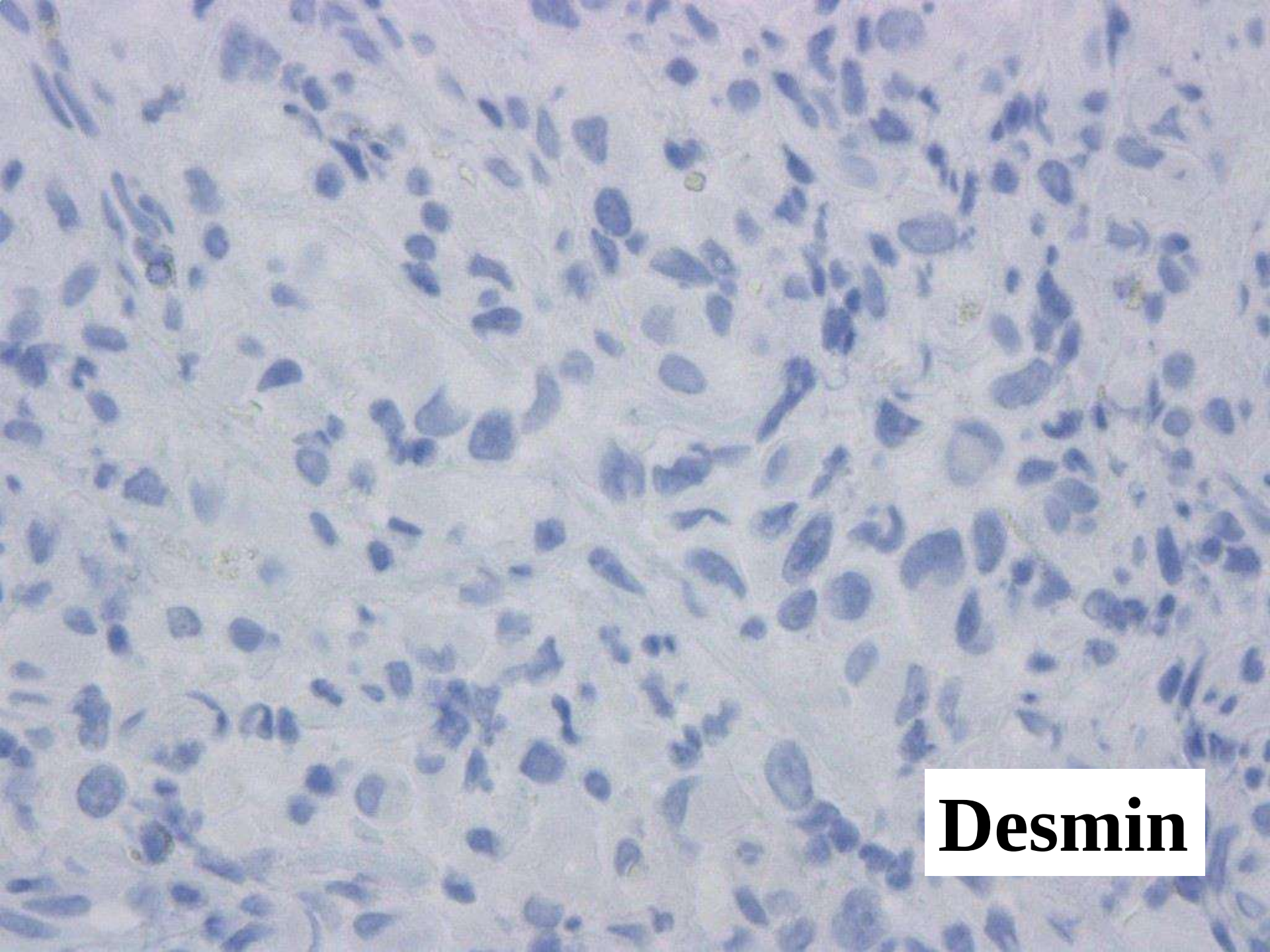


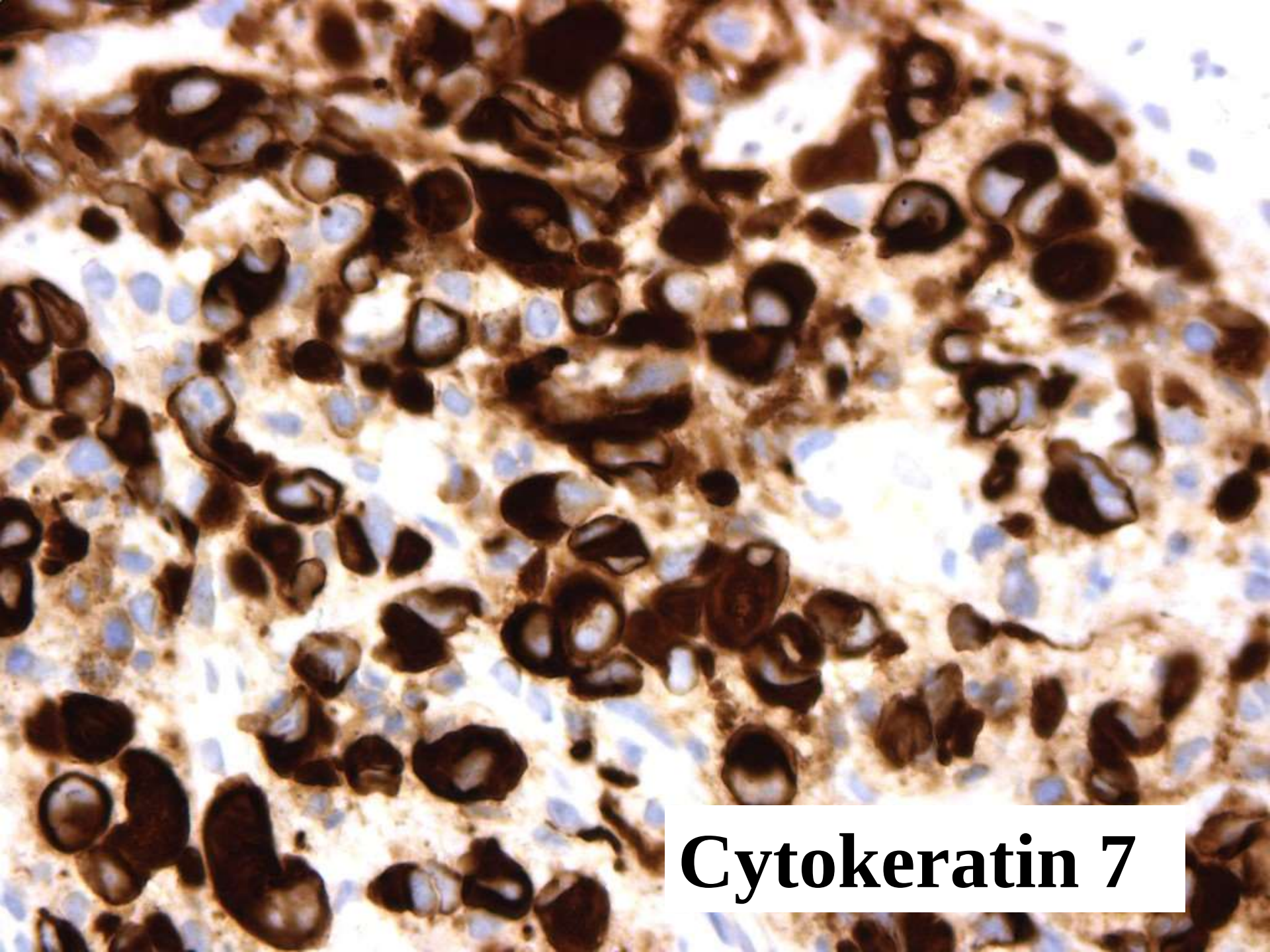


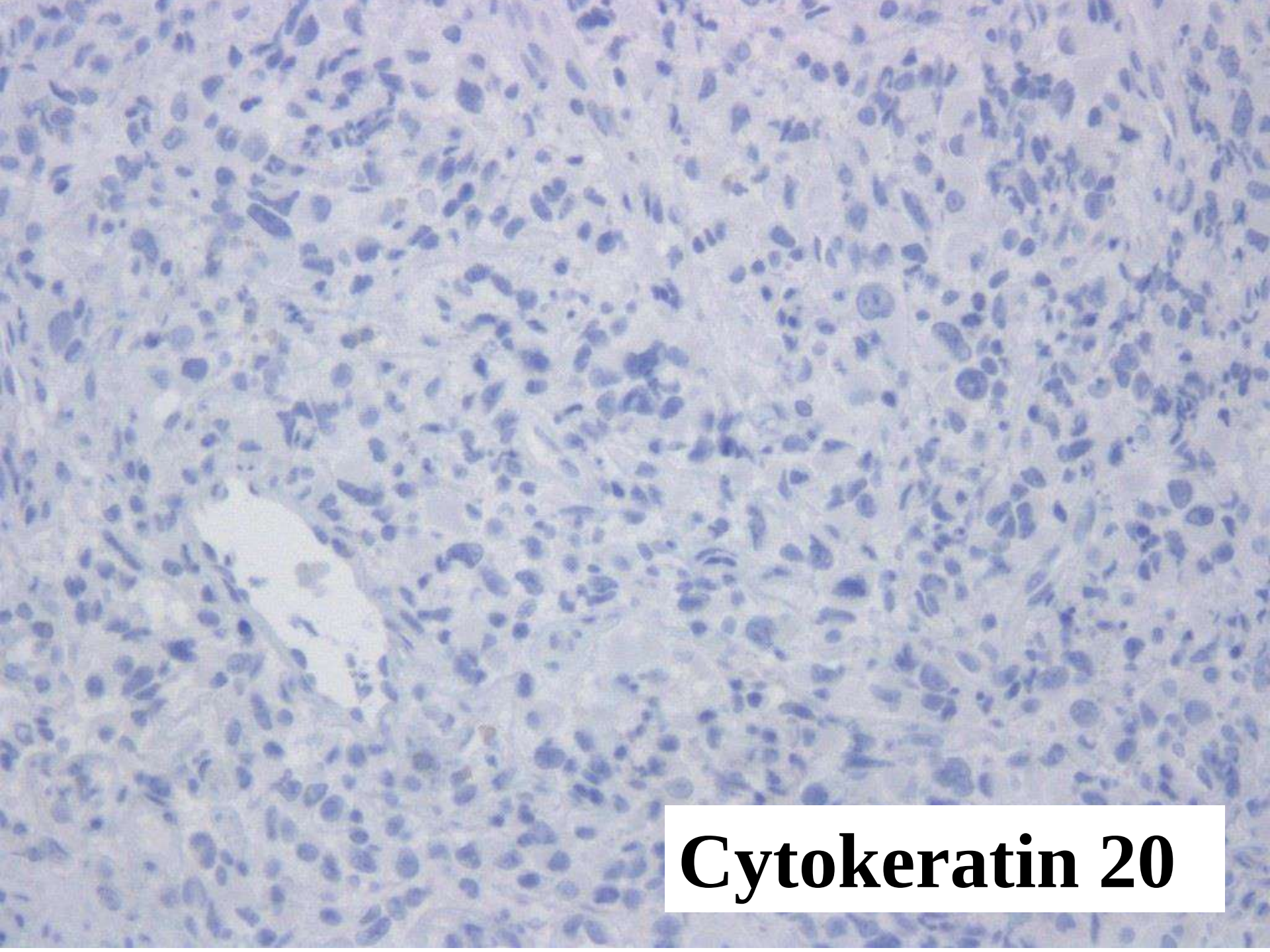

PAS


CAM5.2


Vimentin


Desmin


Cytokeratin 7


Cytokeratin 20

免疫染色結果

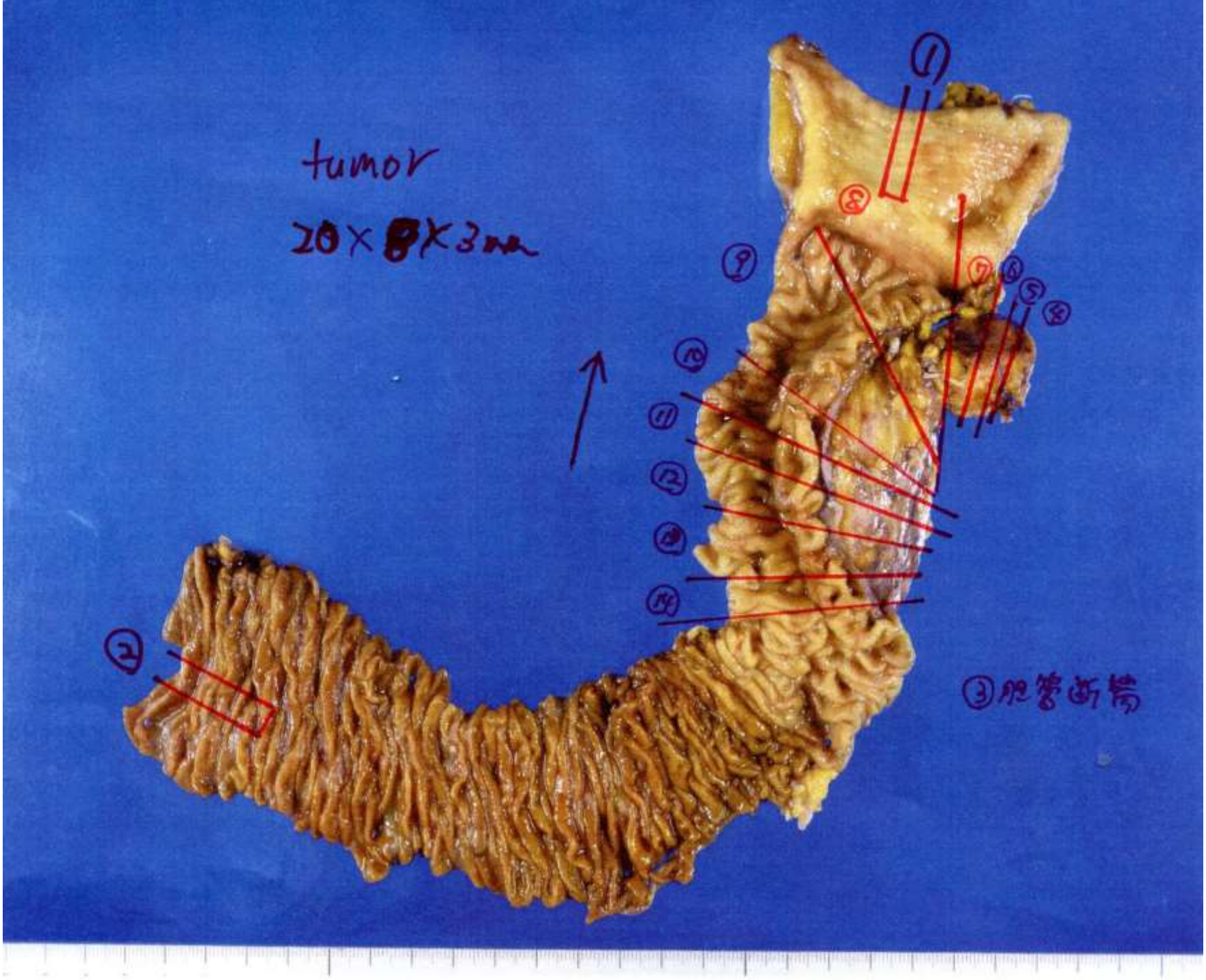
陽性

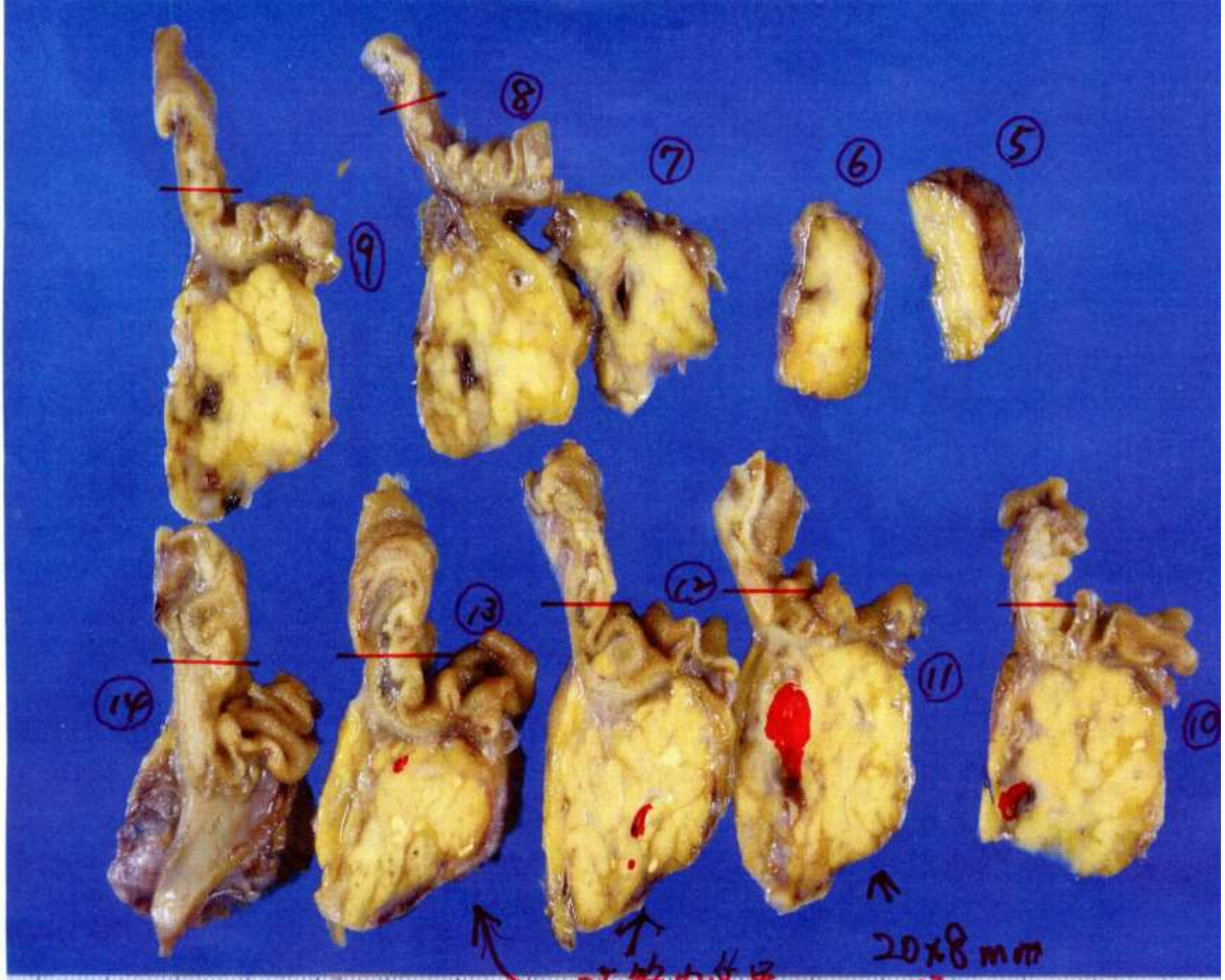
CK7, cytokeratin CAM5.2, AE1/AE3, vimentin

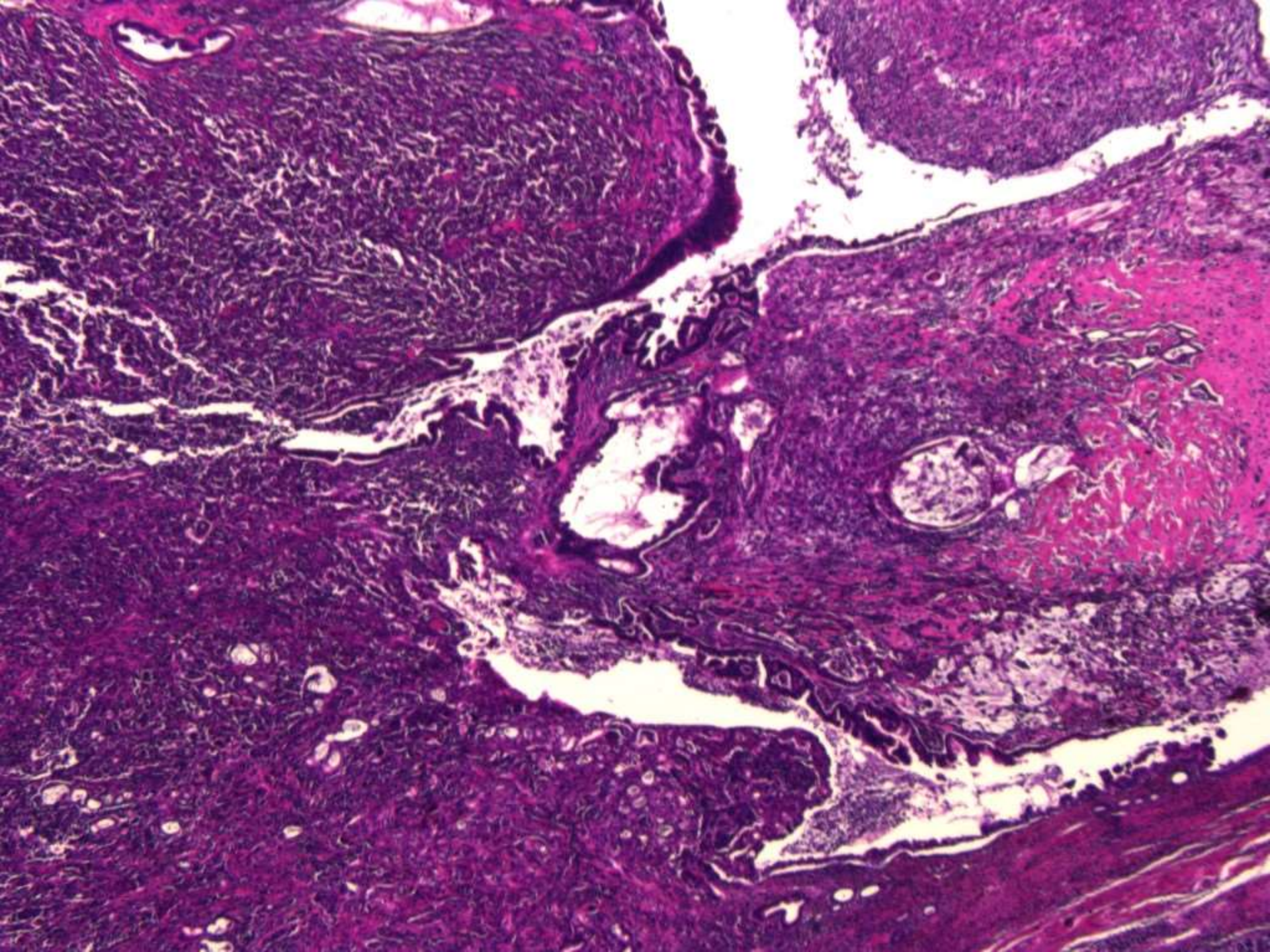
陰性

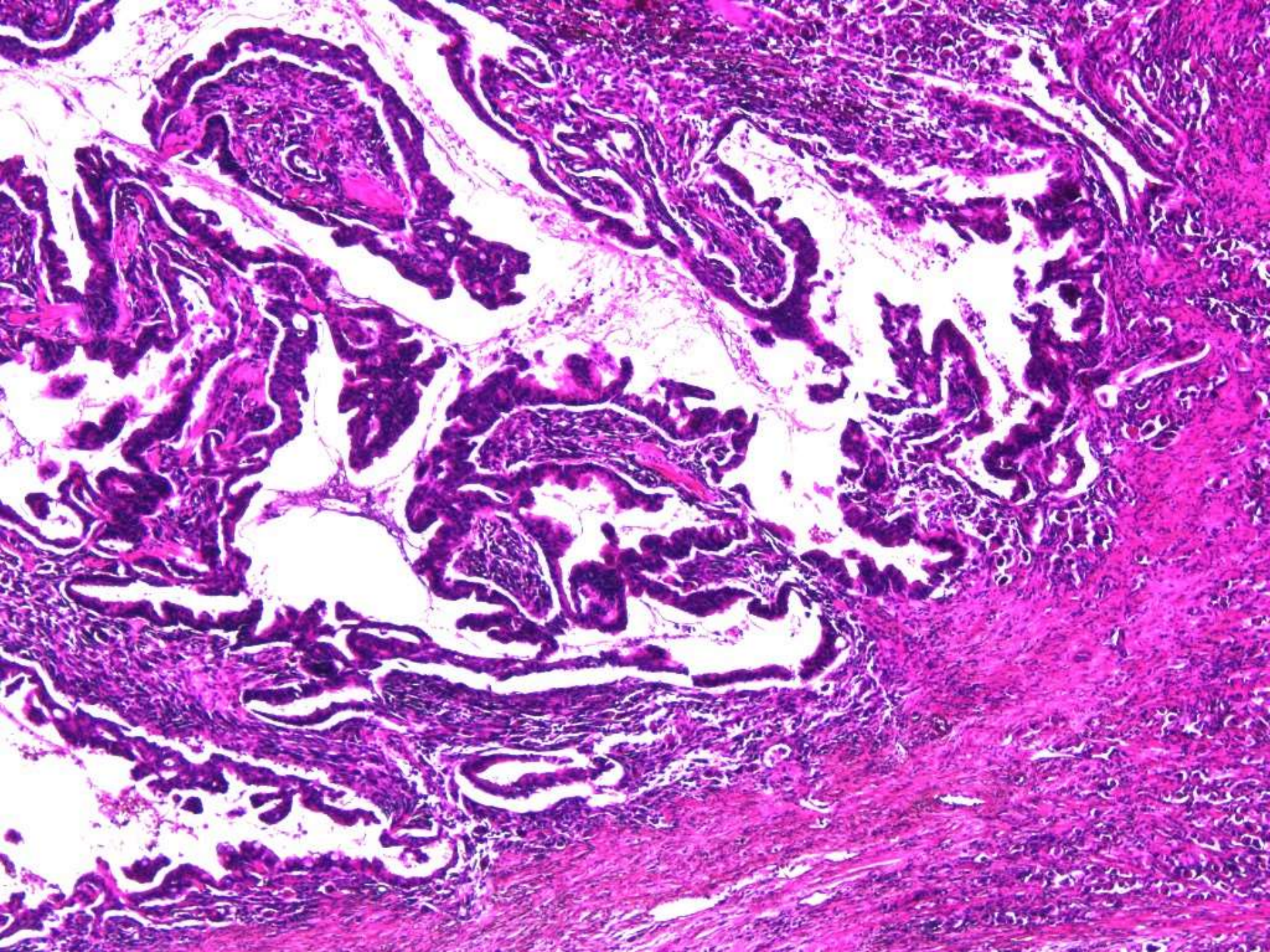
Desmin, Trypsin, Bcl-10, chromogranin A, synaptophysin, CD56. S-100 protein

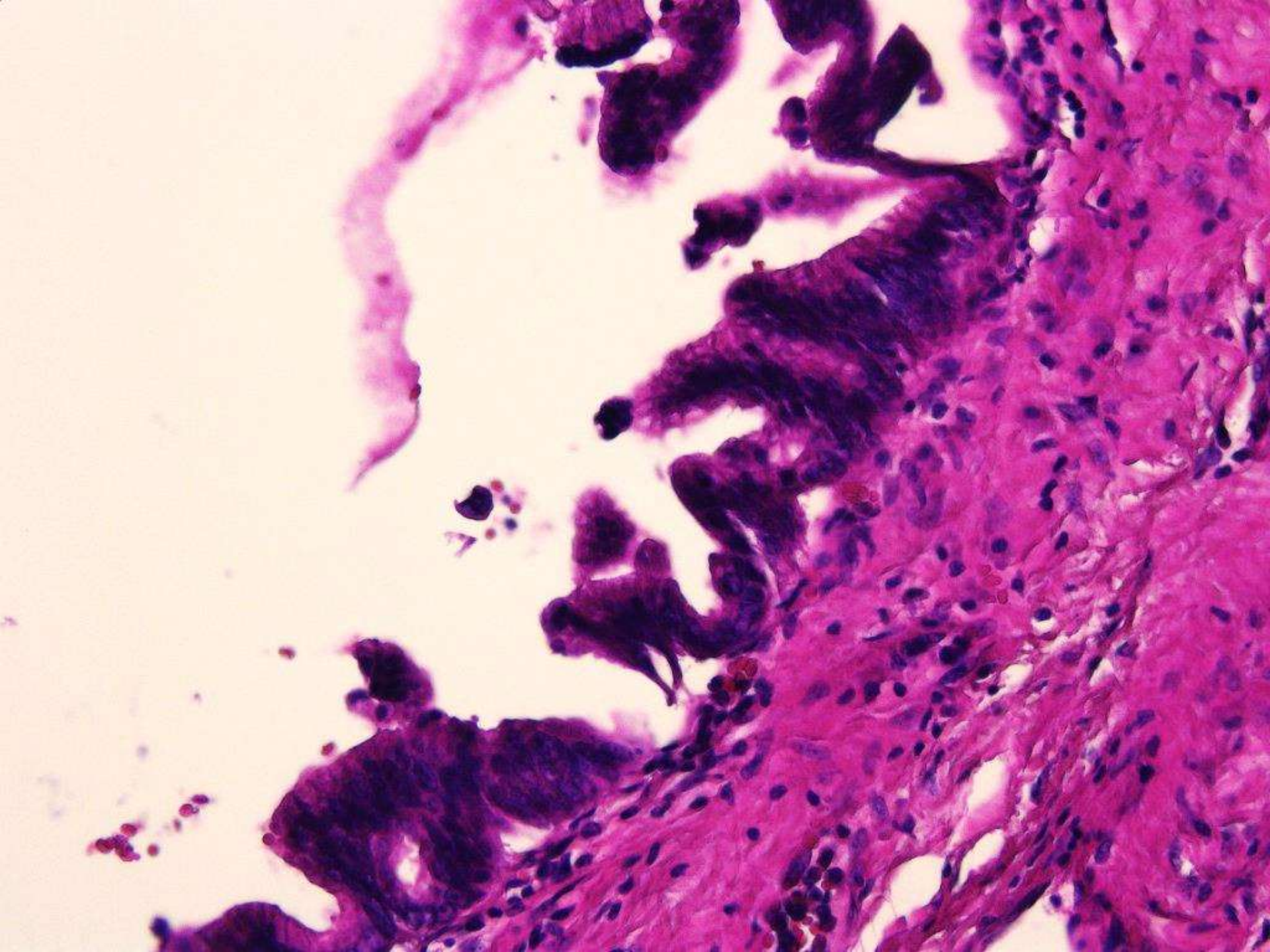
膵頭十二指腸切除手術標本

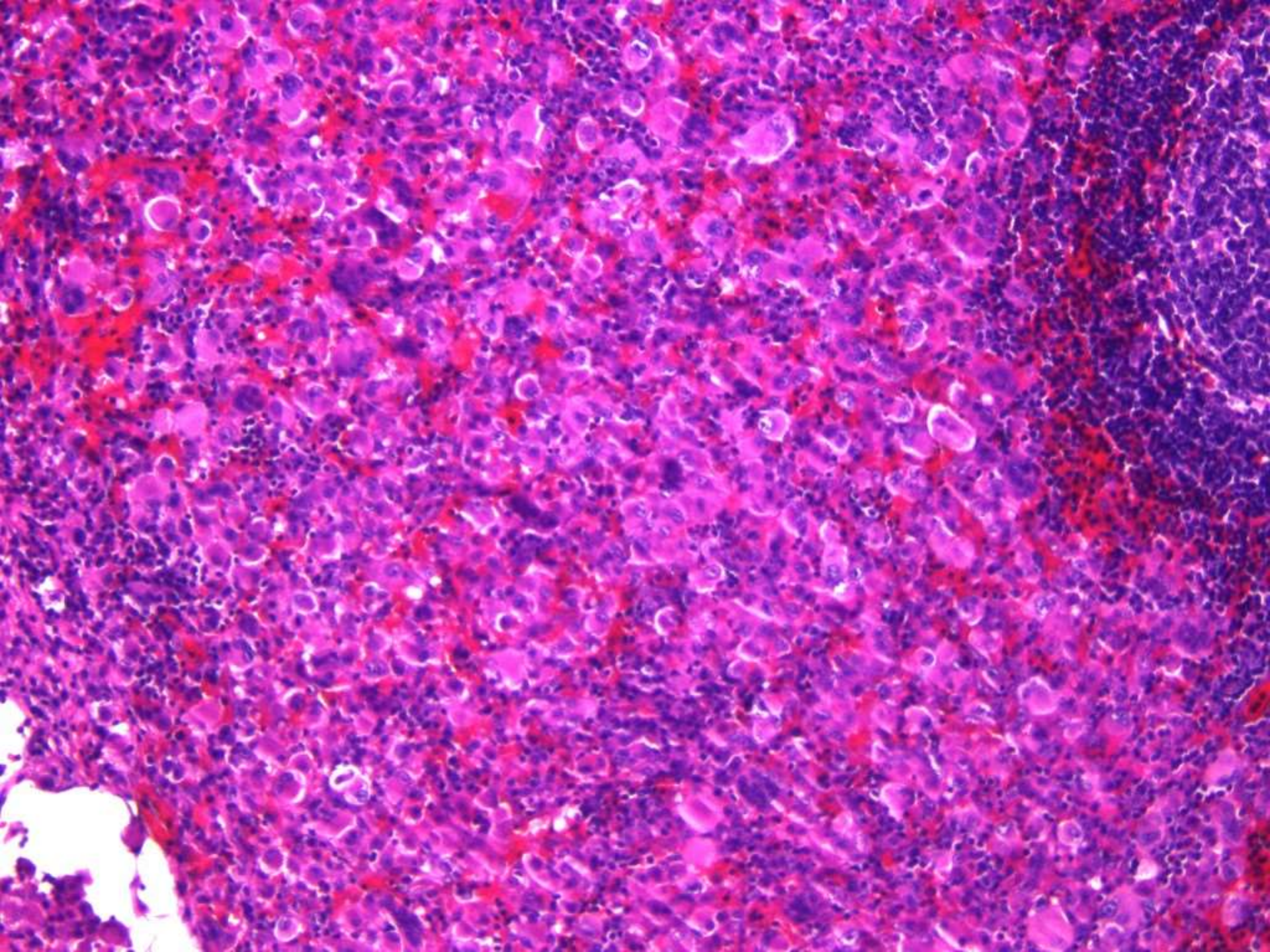

tumor
20×8×3mm
①
②
③
④
⑤
⑥
⑦
⑧
⑨
⑩
⑪
⑫
⑬
⑭
⑮
⑯
③肿瘤断端

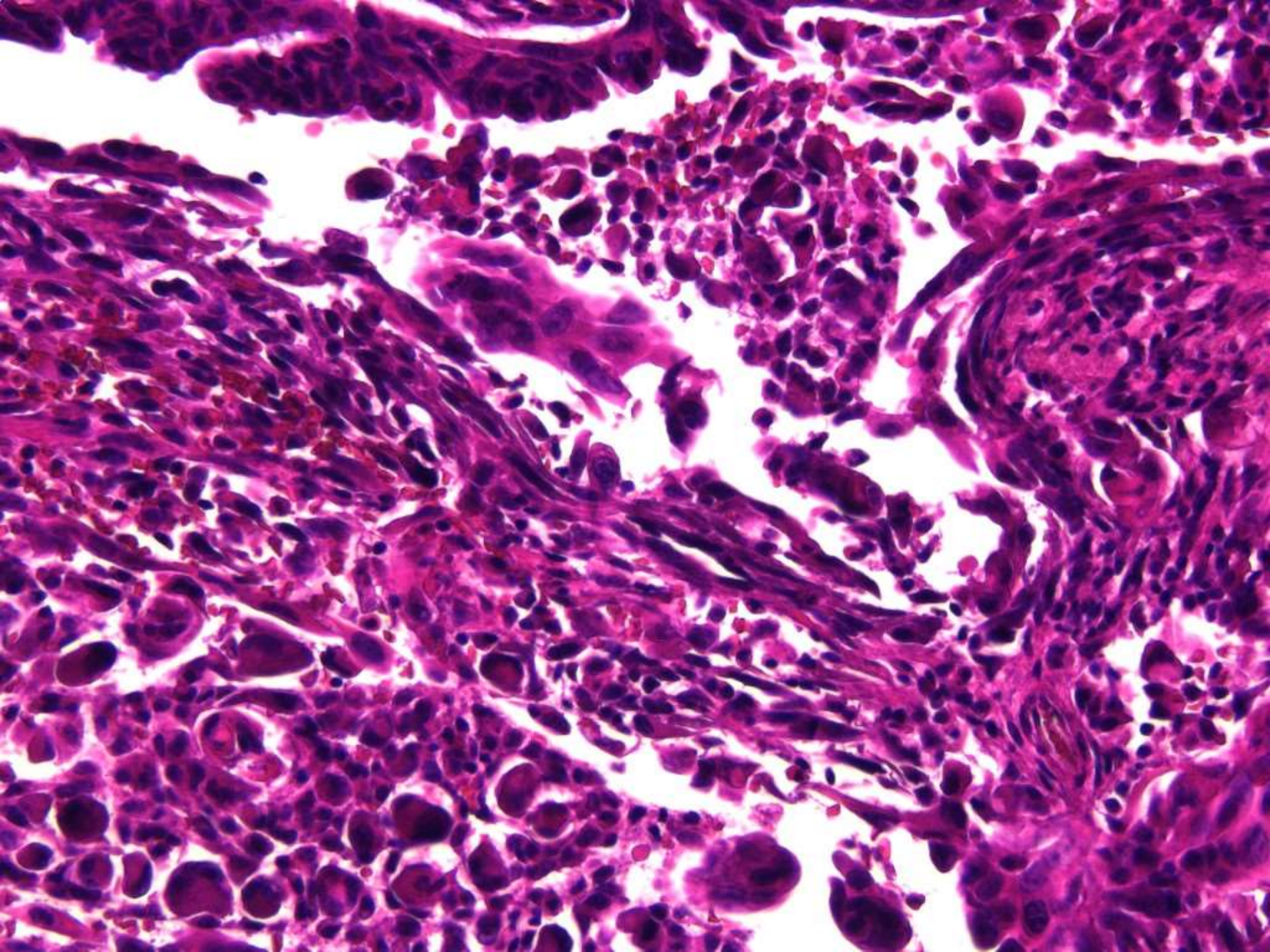

⑧
⑦
⑥
⑤
⑨
⑫
⑬
⑭
⑪
⑩
20×8mm
×3

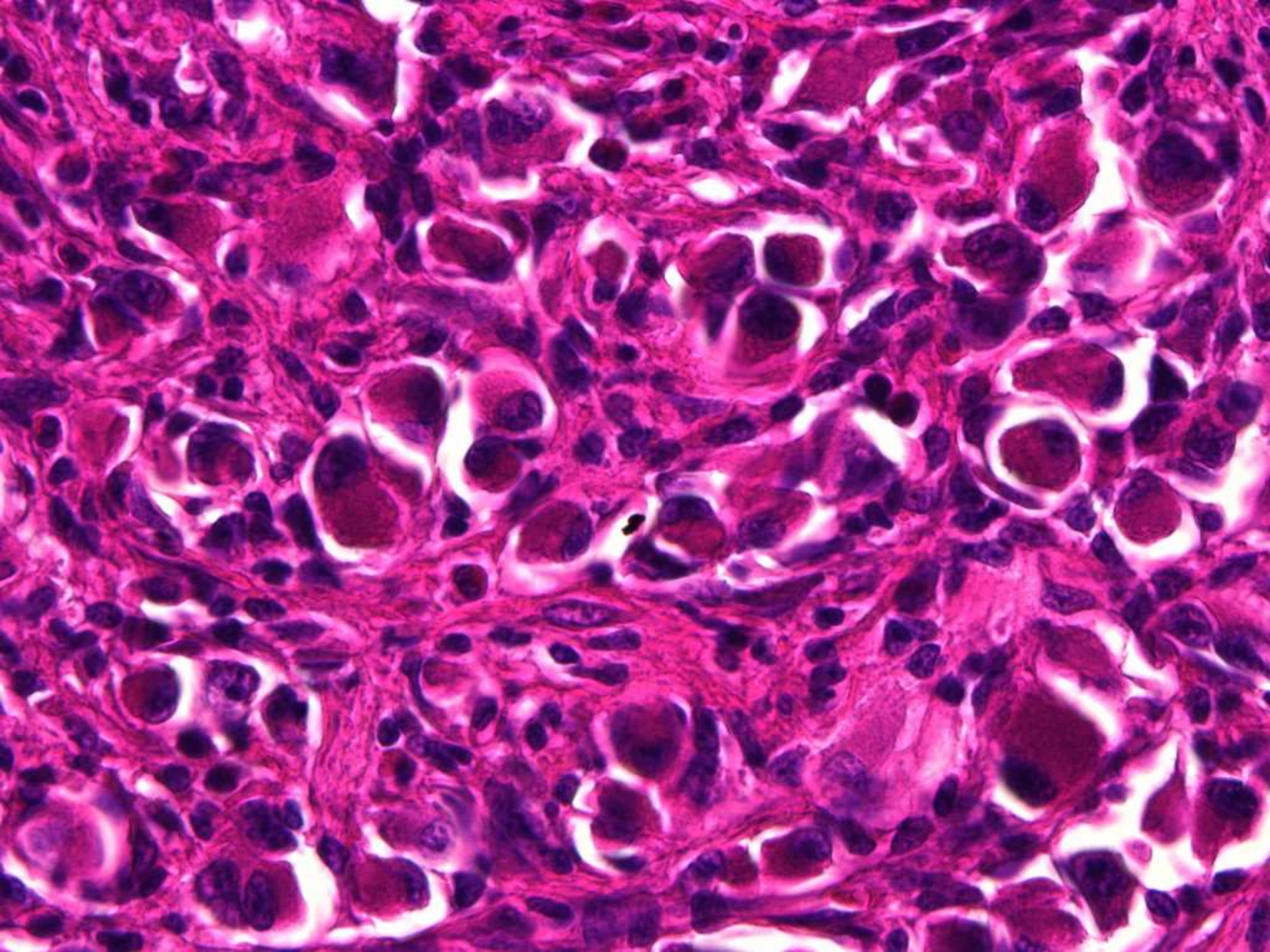


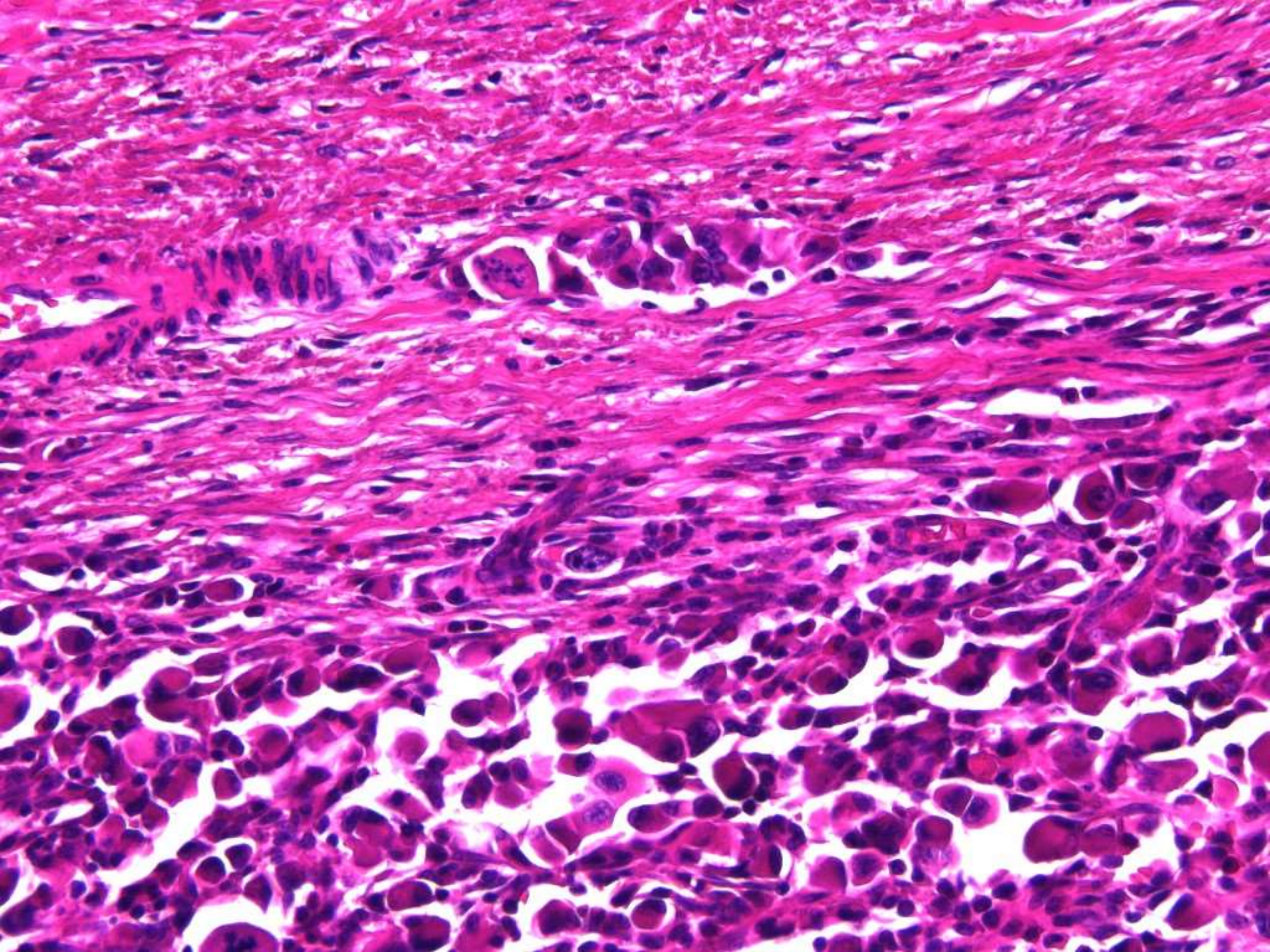


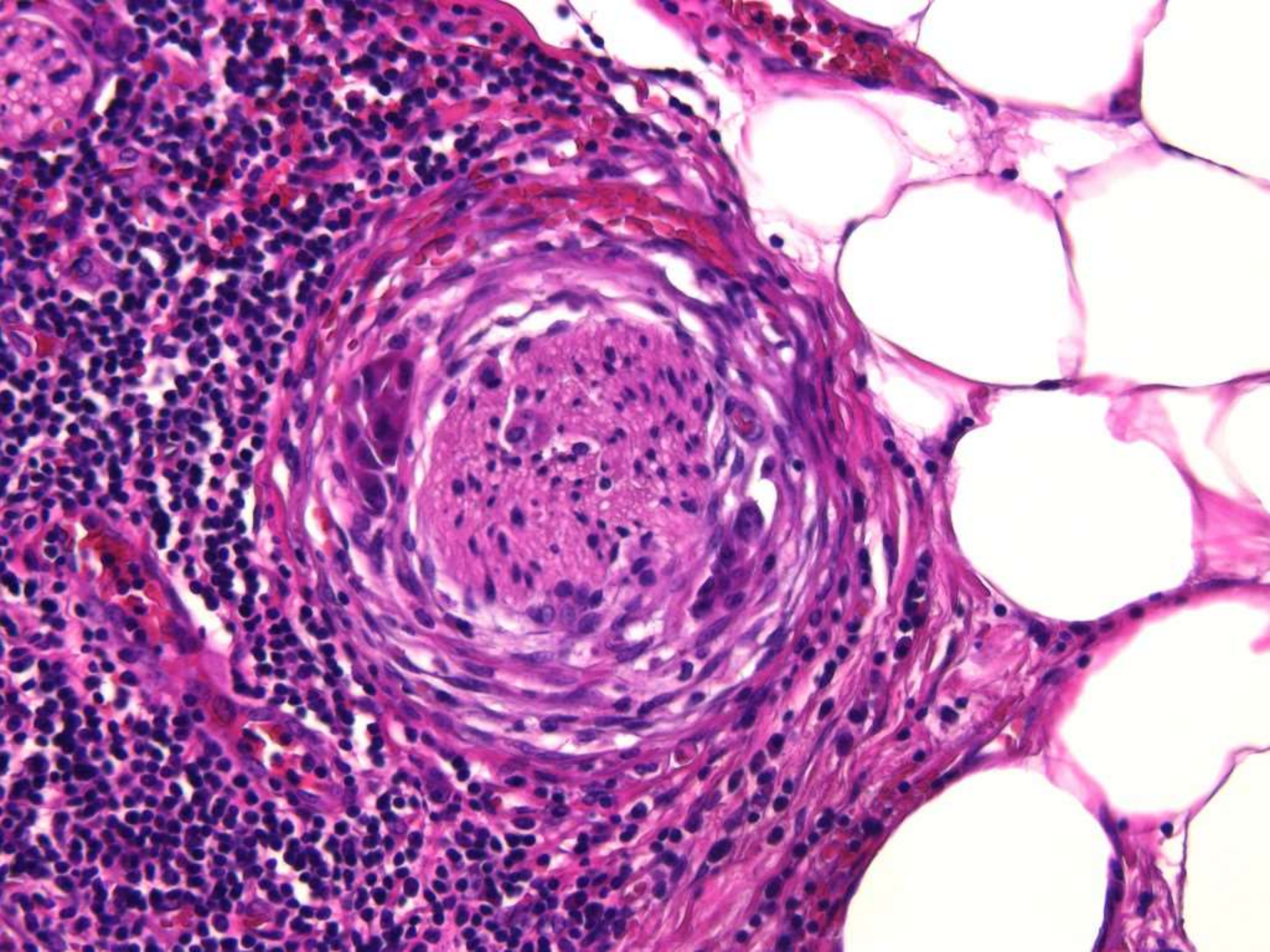












Pathological Findings

Pancreatic cancer, Ph, TS1 (20x8x3mm), Nodular type, anaplastic carcinoma with rhabdoid features, pT3, med, INFa, ly0, v0, ne1, mpd1, pCH0, pDu0, pS0, pRP1, pPV0, pA0, pPL0. pPCM0, pBCM0, pDPM0, pN0, Stage IIB, subtotal stomach-conserving pancreaticoduodenectomy

膵癌取扱い規約

General Rules for the Study of Pancreatic Cancer

2016年7月

第7版

日本膵臓学会●編

July 2016（The 7th Edition）
Japan Pancreas Society

金原出版株式会社

d）退形成癌 Anaplastic carcinoma（同義語*：未分化癌 undifferentiated carcinoma）（図 94～97）

細胞分化が不明瞭な癌腫。多くの場合，一部に膵管癌成分がみられるので，膵管癌の一型と考えられる。腫瘍細胞の形態により，多形細胞型 pleomorphic type，紡錘細胞型 spindle cell type，および非腫瘍性の破骨型多核巨細胞 osteoclast-like giant cells を伴う退形成癌に分けるが，これらが混在することも稀ではない。破骨型多核巨細胞は CD68 陽性，サイトケラチン陰性を示す。

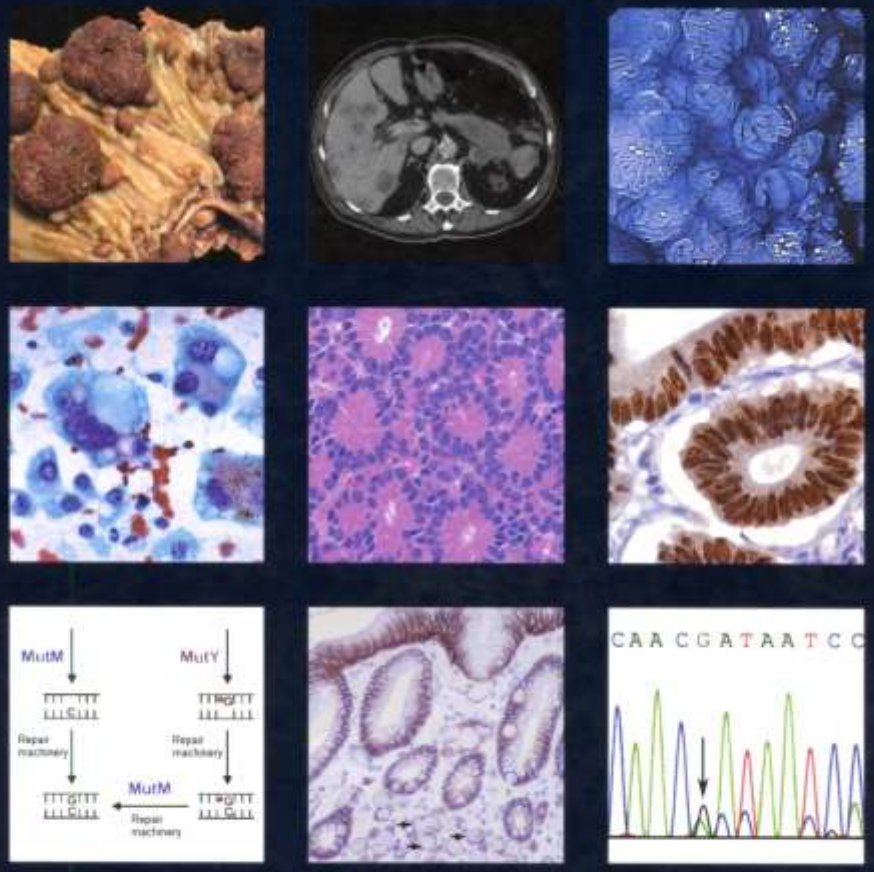
i）多形細胞型退形成癌 Anaplastic carcinoma，pleomorphic type

ii）紡錘細胞型退形成癌 Anaplastic carcinoma，spindle cell type

iii）破骨型多核巨細胞を伴う退形成癌 Anaplastic carcinoma with osteoclast-like giant cells

WHO Classification of Tumours of the Digestive System

Edited by Fred T. Bosman, Fátima Carneiro, Ralph H. Hruban, Neil D. Theise



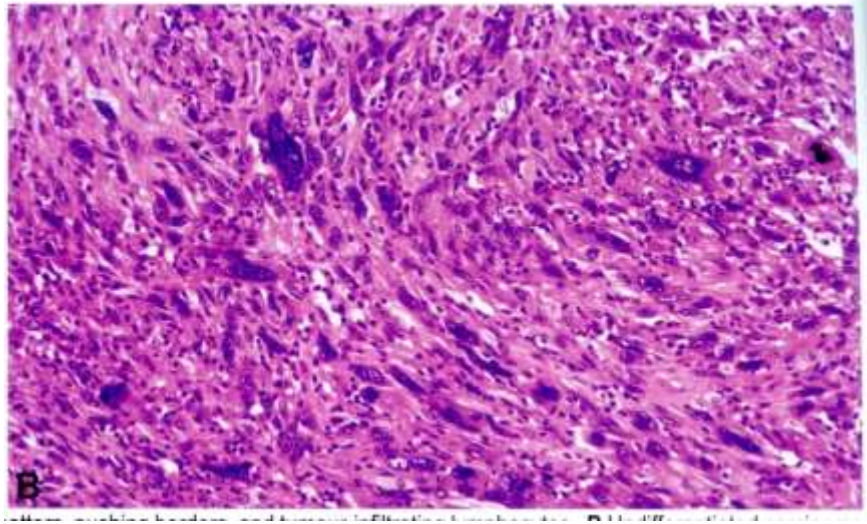
WHO

Undifferentiated (anaplastic) carcinoma

Synonyms

Anaplastic carcinoma, pleomorphic carcinoma, pleomorphic large cell carcinoma, pleomorphic giant cell carcinoma, spindle cell carcinoma, sarcomatoid carcinoma and carcinosarcoma.

Undifferentiated carcinoma is a malignant epithelial neoplasm in which a significant component of the neoplasm does not show a definitive direction of differentiation. These neoplasms usually occur in elderly people, and males and females are equally affected.



Three histological variants have been described. Anaplastic giant cell carcinomas are composed of pleomorphic mononuclear cells admixed with bizarre-appearing giant cells with eosinophilic cytoplasm. Cells with spindle-cell morphology predominate in the sarcomatoid carcinoma, and carcinosarcomas have cells recognizable as adenocarcinoma as well as a high-grade spindle-cell component. In contrast to ductal adenocarcinoma, undifferentiated carcinomas are poorly cohesive, cellular and often have only scant stroma {628, 1604, 3299}. Nuclear pleomorphism, mitoses as well as perineural, lymphatic, and vascular invasion are easily identified. Immunohistochemically, most of these carcinomas express keratin and vimentin but not E-cadherin {1236, 2428, 3527}. Electron microscopy reveals epithelial differentiation, microvilli and occasional mucin in some cases {628}. The prognosis is extremely poor, and average survival is just 5 months {1236, 2428, 3299}.

Final Diagnosis

Anaplastic carcinoma of the pancreas with rhabdoid features

Anaplastic Carcinoma Showing Rhabdoid Features Combined With Mucinous Cystadenocarcinoma of the Pancreas

Kazuyoshi Nishihara, MD; Fujio Katsumoto, MD; Yoshikatsu Kurokawa, MD; Satoshi Toyoshima, MD; Shigeaki Takeda, MD; Ryuji Abe, MD

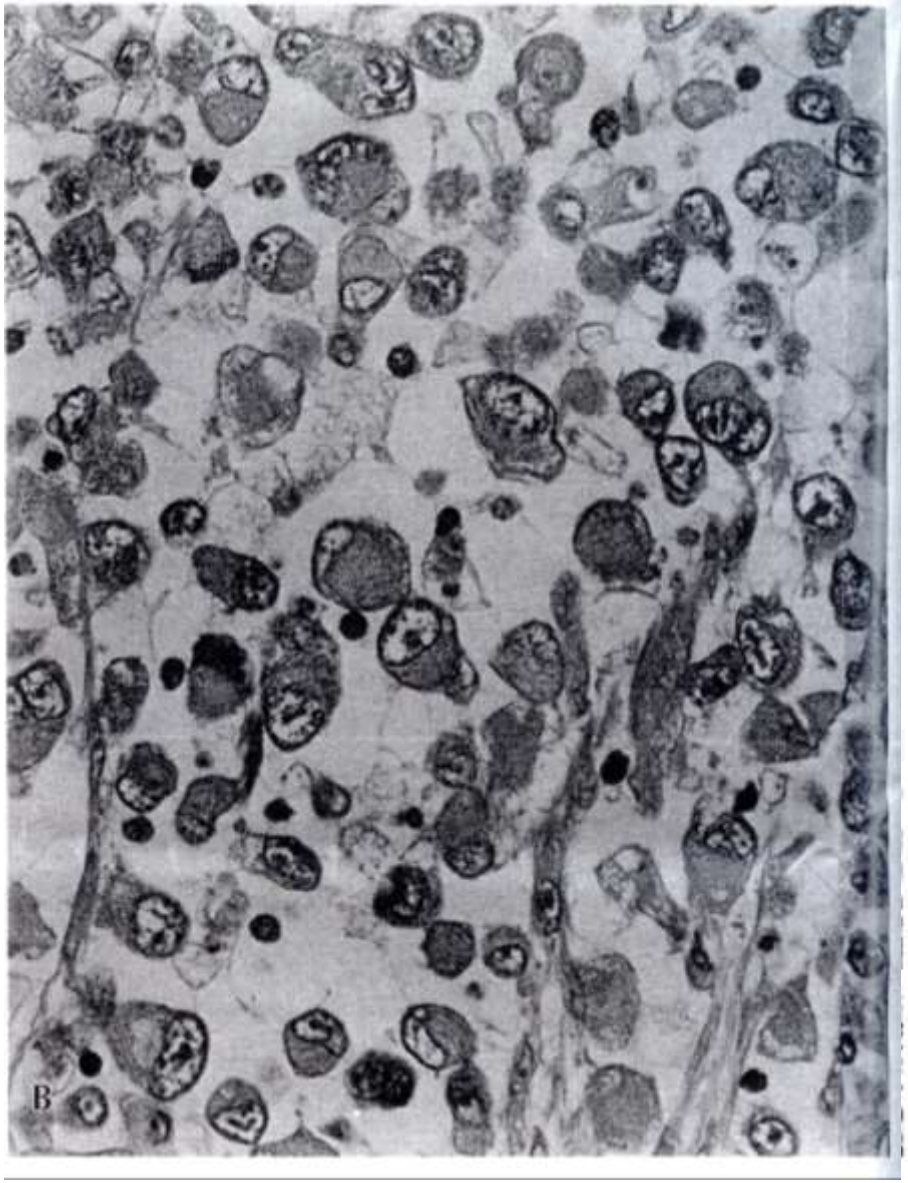
• A 52-year-old Japanese woman presented with complaints of back pain, loss of appetite, and weight loss; her diagnosis was a mass in the pancreatic tail. A distal pancreatectomy combined with lymph node dissection was performed. The tumor measured 10.0 × 9.5 × 8.0 cm and consisted of a cystic mass and a solid area. Histologically, the cystic mass represented a typical mucinous cystadenocarcinoma, whereas the solid portion was composed of anaplastic tumor cells with round eosinophilic intracytoplasmic inclusions, as seen in malignant rhabdoid tumor of the kidney. The hyaline-like intracytoplasmic inclusions were weakly positive for periodic acid–Schiff and were resistant to diastase. There was a gradual transition between the mucinous cystadenocarcinoma and the anaplastic carcinoma showing rhabdoid features. Additionally, the tumor cells of the anaplastic carcinoma were immunoreactive to both epithelial membrane antigen and vimentin. These findings suggest that the anaplastic carcinoma with rhabdoid features developed from the mucinous cystadenocarcinoma of the pancreas.

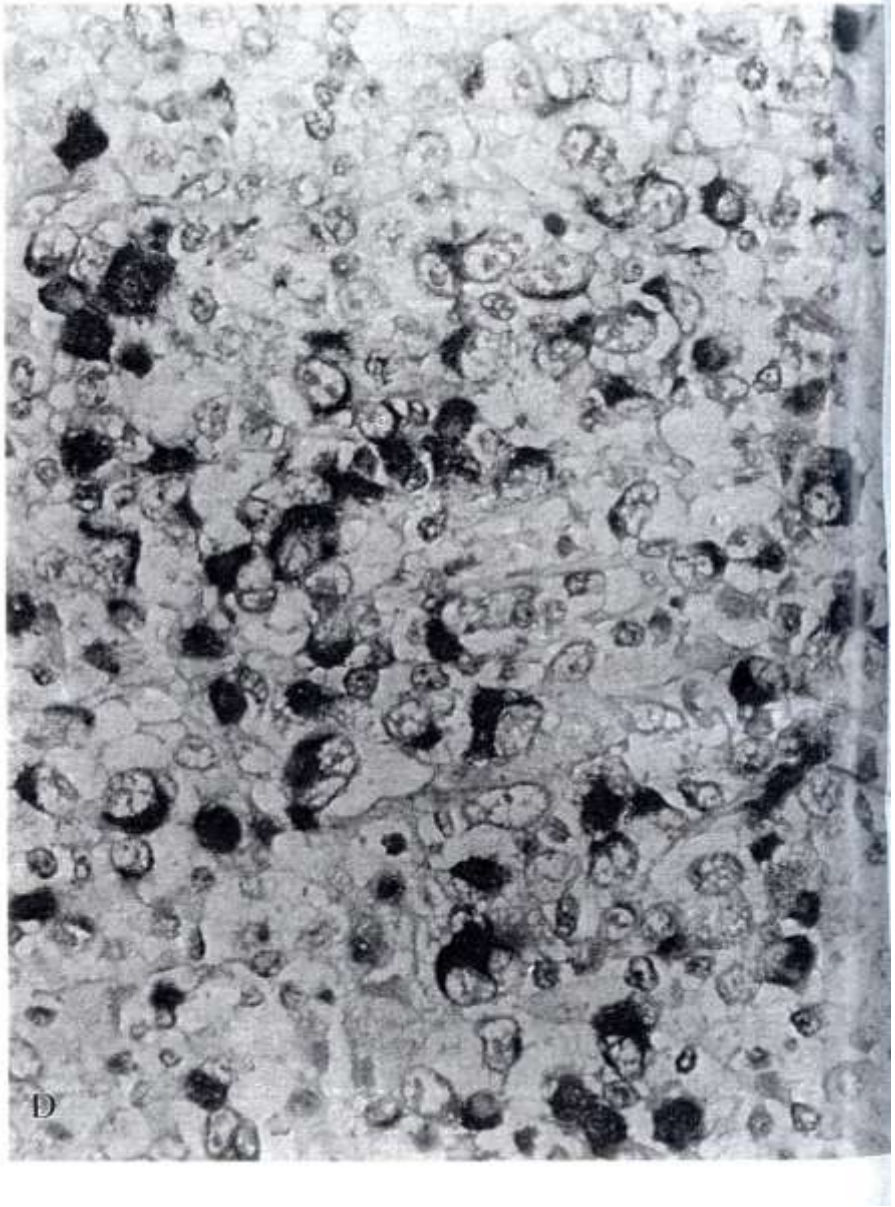
(*Arch Pathol Lab Med.* 1997;121:1104–1107)

(−7.5 kg); she was admitted to our hospital with dull back pain in February 1994. Physical examination showed mild anemia and a round mass, measuring 6.5 cm in diameter, in her left upper abdomen. Routine abdominal ultrasonography demonstrated a cystic mass with a solid area in the tail of the pancreas. By computed tomography and magnetic resonance imaging, the anterior wall of the cystic mass appeared to be thickened (Fig 1). The serum carcinoembryonic antigen (9.9 ng/mL, normal <5.0 ng/mL), CA 19-9 (655.0 U/mL, normal <60 U/mL), elastase 1 (467 ng/dL, normal 80–400 ng/dL), and Span-1 (148.0 U/mL, normal <30 U/mL) levels were elevated, whereas the DUPAN-II (50 U/mL) and α-fetoprotein (0.6 ng/mL) levels were all within normal limits. A distal pancreatectomy/splenectomy, along with lymph node dissection, was performed in March 1994 under the preoperative diagnosis of mucinous cystadenocarcinoma of the pancreas. The postoperative course was uneventful, and the patient was discharged from the hospital on the 40th postoperative day. Ten months later, she was diagnosed as having metastatic nodules in the liver, and she eventually died of the disease in October 1995 (19 months after the initial operation). Consent for autopsy was denied.

GROSS FINDINGS

The resected tumor measured 10.0 × 9.5 × 8.0 cm, and


B


D

Pathology International 2000; **50**: 57–62

Case Report

Anaplastic carcinoma of the pancreas with rhabdoid features

Naoto Kuroda,[1] Taeko Sawada,[2] Eriko Miyazaki,[1] Yoshihiro Hayashi,[1] Makoto Toi,[1] Keishi Naruse,[1] Takayuki Fukui,[1] Hirofumi Nakayama,[3] Makoto Hiroi,[1] Hirokuni Taguchi[2] and Hideaki Enzan[1]

[1]First Department of Pathology, [2]Third Department of Internal Medicine, Kochi Medical School, Kochi and [3]Department of Pathology, Kure National Hospital, Hiroshima, Japan

The malignant rhabdoid tumor (MRT) is histologically characterized by the invasive proliferation of polygonal to ovoid cells with abundant eosinophilic cytoplasm and eccentric vesicular nuclei with a prominent nucleolus. MRT frequently occurs in the kidney, but may also arise in other organs. However, MRT should be strictly distinguished from carcinomas with rhabdoid features. A post-mortem examination of a 68-year-old woman found an anaplastic carcinoma of the pancreas with rhabdoid features displaying extensive invasion into the neighboring tissues. To the best of our knowledge, this is the first case of a pancreatic tumor with rhabdoid features. Pathologists should consider that carcinomas showing rhabdoid features may also appear in the pancreas. As pancreatic tumors with rhabdoid features have characteristic histopathological features and poor prognosis compared to other pancreatic tumors, careful histopathological differential diagnosis is important.

and received rehabilitation from March 1998. Although she could maintain a standing position on her own, she gradually began to develop lumbago. She had a history of diabetes mellitus and ischemic heart disease and received insulin therapy, for which she received medication 38 years and 18 years, respectively. She had a family history of diabetes mellitus (father) and gastric cancer (mother), there was no history of smoking. Ultrasonographic examination revealed a marked swelling of the lymph nodes in her abdominal cavity. She was transferred to the Kochi Medical School Hospital, Kochi, Japan. Her condition gradually deteriorated before biopsy examination was possible for final diagnosis, and she received chemotherapy on suspicion of malignant lymphoma. She subsequently developed aspiration pneumonia. The serum CA 19-9, carcinoembryonic antigen (CEA) and alpha-fetoprotein (AFP) levels were constantly within normal

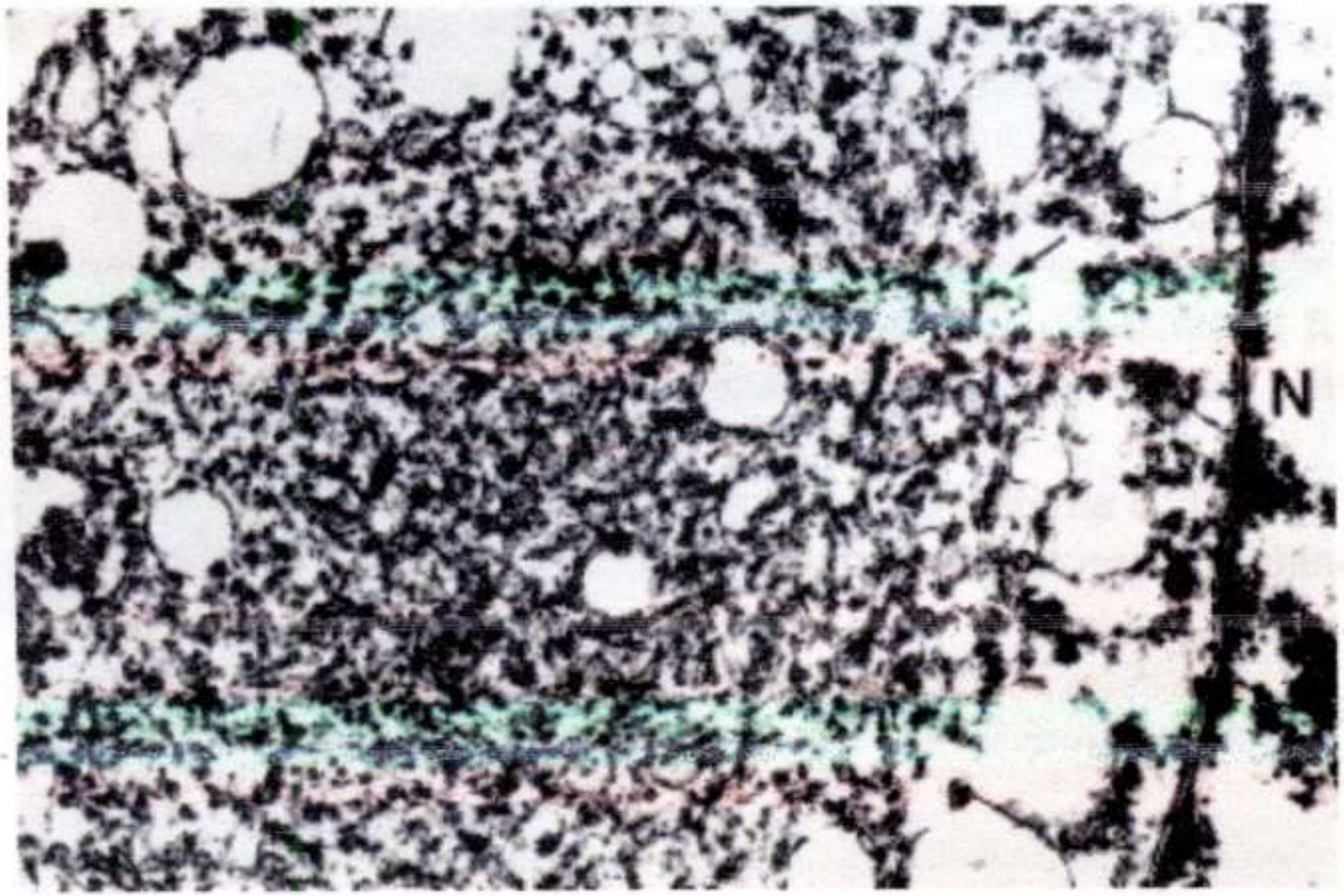


Figure 8 Electron micrograph of paranuclear aggregates of intermediate filaments. N, nucleus of a neoplastic cell.

Med Mol Morphol (2007) 40:168–171
DOI 10.1007/s00795-006-0349-0

ORIGINAL PAPER

Naoto Kuroda · Shin-ichi Iwamura · Nokiaki Fujishima
Masahiko Ohara · Takashi Hirouchi · Keiko Mizuno
Yoshihiro Hayashi · Gang-Hong Lee

Anaplastic carcinoma of the pancreas with rhabdoid features and hyaline globule-like structures

Received: September 5, 2006 / Accepted: October 10, 2006

Abstract A 59-year-old Japanese man presented with a giant submucosal tumor with ulceration during follow-up of duodenal ulcer. Pancreaticoduodenectomy was undertaken, and subsequent histological examination of the tumor disclosed anaplastic carcinoma of the pancreas head. The carcinoma components contained adenocarcinoma and squamous cell carcinoma. Additionally, undifferentiated spindle or pleomorphic cells were seen in continuity with the carcinoma component. Undifferentiated neoplastic cells with rhabdoid features and with hyaline globule-like structures positive for PAS stain with diastase pretreatment were also observed. Immunohistochemically, cytoplasmic inclusions corresponding to rhabdoid features showed aggregates of vimentin. Ultrastructurally, hyaline globule-like structures corresponded to lysosomes. Finally, we report here the first case of anaplastic carcinoma of the pancreas with hyaline globule-like structures.

the pancreas with rhabdoid features.[2] Hyaline globule-like structures in the cytoplasm of neoplastic cells have been reported in malignant neoplasms of several anatomic sites. In this article, we report the second case possessing rhabdoid features and the first case possessing hyaline globule-like structures in the cytoplasm of a pancreatic anaplastic carcinoma.

Case report

When a 59-year-old Japanese man underwent endoscopic examination of the upper digestive tract during follow-up of duodenal ulcer, a giant submucosal tumor with ulceration that measured 10 cm in maximum diameter was discovered in the duodenum. On the suspicion of malignancy, pancre-

Pathology International 2015; **65**: 264–270 doi:10.1111/pin.12283

Case Report

CD44-expressing undifferentiated carcinoma with rhabdoid features of the pancreas: Molecular analysis of aggressive invasion and metastasis

Takuji Ohmoto,[1,2*] Nobuyuki Yoshitani,[1,2*] Kazuchika Nishitsuji,[2] Tetsuji Takayama,[3] Yuto Yanagisawa,[4] Motohiro Takeya[5] and Naomi Sakashita[2]

[1]*Undergraduate student, The University of Tokushima School of Medicine,* [2]*Department of Human Pathology and* [3]*Department of Gastroenterology and Oncology, Institute of Health Biosciences, The University of Tokushima Graduate School,* [4]*Undergraduate student, Faculty of Pharmaceutical Sciences, The University of Tokushima, Tokushima and* [5]*Department of Cell Pathology, Graduate School of Medical Sciences, Kumamoto University, Kumamoto, Japan*

Carcinoma with rhabdoid features is a rare malignant tumor with a poor prognosis whose molecular mechanism for aggressive behavior is unclear. We describe an undifferentiated pancreatic carcinoma with rhabdoid features that demonstrated extensive invasion and metastasis. Examination of a 63-year-old man with back pain disclosed a retroperitoneal tumor with multiple metastases. Lymph node biopsy revealed an undifferentiated carcinoma of unknown origin. Intensive chemotherapy was ineffective; the patient died 3 months after initial symptoms. Autopsy showed that the tumor displaced the retroperitoneal space: it diffusely invaded and destroyed the pancreas and duodenum. Histology demonstrated tumor cells with eccentric vesicular nuclei, large nucleoli, juxtanuclear eosinophilic inclusions, and poor cell adhesion. Immunohistochemistry showed that tumor cells expressed cytokeratin and vimentin, and electron microscopy confirmed a perinuclear mass of intermediate fibrils and lipid droplets, which indicated an undifferentiated carcinoma with rhabdoid features. Tumor tissue contained hyaluronan; tumor cells strongly expressed CD44, matrix metalloproteinase-9, hypoxia-inducible factor-1α, hyaluronan synthase 2, and acyl-CoA:cholesterol acyltransferase 1 and had a high Ki-67$^+$ ratio. Since hyaluronan is a ligand for CD44, formation of CD44-hyaluronan complex on the cell surface activates CD44 and this activation may explain why the tumor manifested aggressive invasion and metastasis throughout the clinical course.

Key words: CD44, hyaluronan, hypoxia-inducible factor-1α (HIF-1α), matrix metalloproteinase-9 (MMP-9), pancreas, undifferentiated carcinoma with rhabdoid features

The rhabdoid tumor was originally described as an extremely aggressive renal tumor arising mainly in childhood,[1] and cumulative data indicated that similar malignant tumors may appear in various organs, even in elderly patients. Its cytological characteristics include a large eccentric nucleus with a prominent nucleolus and abundant cytoplasm with large eosinophilic inclusions.[2] Ultrastructural and immunocytochemical analyses provided evidence that the intracytoplasmic inclusions consisted of intermediate fibrils, i.e., vimentin and cytokeratin.[3] Rhabdoid tumors frequently demonstrate extensive invasion and metastasis at the time of initial diagnosis, so complete surgical resection and chemoradiotherapy are usually ineffective. Occasionally, this tumor develops as a carcinoma with rhabdoid differentiation in various organs, such as the lung, esophagus, gastrointestinal tract, pancreas, thyroid gland, urinary tract, and thymus.[4–11] Previous case studies reporting pancreatic

Virchows Arch (2014) 465:531–538
DOI 10.1007/s00428-014-1631-5

ORIGINAL ARTICLE

Clinicopathological characteristics of anaplastic carcinoma of the pancreas with rhabdoid features

Makoto Sano · Taku Homma · Emiko Hayashi · Hiroko Noda · Yusuke Amano · Ryusuke Tsujimura · Tsutomu Yamada · Brian Quattrochi · Norimichi Nemoto

Received: 5 March 2014 / Revised: 18 June 2014 / Accepted: 7 July 2014 / Published online: 17 July 2014

Abstract Undifferentiated (anaplastic) carcinoma with rhabdoid features is a rare and aggressive subtype of pancreatic carcinoma. Here, we report the clinical, histological, and immunohistochemical phenotypes in six autopsy cases of anaplastic carcinoma with rhabdoid features. The patients ranged between 44 and 76 years of age (median, 61 years) and consisted of four males and two females. All patients except one case died within 3 months of diagnosis, as these tumors were found at an advanced stage and were chemoresistant. At autopsy, tumor masses measuring 4–22 cm in maximum diameter were mainly located in the pancreatic body and tail. Microscopically, all cases showed anaplastic carcinoma with rhabdoid features that were discohesive with round to polygonal eosinophilic cytoplasm with occasional inclusions, and that had vesicular nuclei, and prominent nucleoli. Immunohistochemistry showed that the rhabdoid cells, particularly the inclusions, were strongly positive for pan-cytokeratin (AE1/AE3) and vimentin. Meanwhile, downregulation or aberrant cytoplasmic localization with focal aggregation of E-cadherin, β-catenin, and EMA were frequently observed in the rhabdoid cells. Moreover, the intracytoplasmic inclusions were labeled with selective autophagy-related molecules including p62/SQSTM1, ubiquitin, and kelch-like ECH-associated protein 1 (KEAP1). In addition, nuclear factor erythroid 2-related factor 2 (NRF2) and overexpression of its target molecule multidrug resistance-associated protein 1 (MRP1) were commonly observed in the rhabdoid cells. Therefore, these results suggest that p62-mediated aggregation of ubiquitinated intermediate filaments and membranous proteins is an important phenomenon in the rhabdoid phenotype. Indeed, the ubiquitinated aggregates of p62 and KEAP1 would induce activation of NRF2 and upregulation of MRP1, leading to potential chemoresistance of anaplastic carcinoma with rhabdoid features.

Keywords Undifferentiated carcinoma · Pleomorphic carcinoma · Rhabdoid feature · Autophagy · Chemoresistance

Table 1 Clinicopathological characteristics of anaplastic carcinoma with rhabdoid features

Case	Age[a]	Sex	Tumor size (cm)	Location	Gross appearance	Direct invasion/metastasis	Rhabdoid cells (%)	Premortal therapy	Period
1	51	F	10×10×5	Body/tail	Whitish and solid mass with necrosis	Stomach, transverse colon, liver, spleen, left kidney, retroperitoneal adipose tissue (serous ascites; 2,300 ml), peripancreatic, and pulmonary hilar lymph nodes (LNs)	30	TS-1 (80 mg/m^2/day, 3 weeks; PD), 5-FU (333 mg/m^2×5)+DTX (60 mg/m^2; PD), CDDP (60 mg/m^2)+VP-16 (60 mg/m^2; PD), LV (32 mg/m^2×5; PD)	3 m
2	44	M	21×17×13	Body/tail	Whitish and solid mass with necrosis	Stomach, duodenum, liver, transverse colon, inferior vena cava, and omentum (serosanguinous ascites; 6,100 ml)	15	[CDDP (80 mg/m^2)+VP-16 (100 mg/m^2; PD), Radiation (PD)][b], *5-FU (540 mg/m^2× 2; PR)*, TS-1 (65 mg/m^2/day; 9 m; PD)[b]	2 years
3	68	F	10×9×9	Body	Ocher and solid mass with necrosis and hemorrhage	Liver, kidneys, lungs, right adrenal gland, omentum, and peritoneum (hemorrhagic ascites; 2,800 ml)	50	Palliative care	2 weeks
4	59	M	22×14×9	Body	Ocher and solid mass with necrosis and hemorrhage	Transverse and descending colon, spleen, peritoneum (hemorrhagic ascites; 3,000 ml), hepatic hilar, and mesenteric LNs	75	Palliative care	3 m
5	68	M	4 in diameter	Head	Whitish-yellow and solid mass with necrosis	Bile duct, duodenum, mesenterium, peritoneum (hemorrhagic ascites; 4,100 ml), liver, heart, stomach, small and large intestine, right ureter, thyroid gland, adrenal glands, bone marrow, pulmonary hilar and para-aortic LNs, and bone marrow	40	Palliative care	1 m
6	76	M	9×5×3.5	Body/tail	Grayish and solid mass with hemorrhage	Liver, lungs, diaphragm, and peritoneum (serosanguinous ascites 3,750 ml); peripancreatic, splenic hilar, and para-aortic LNs	80	Palliative care	2 m

PD progressive disease, *PR* partial response

[a] Age (years) at death

[b] Performed in other hospital

Table 2 Immunohistochemical phenotype of rhabdoid cells in anaplastic pancreatic carcinoma

Case	1	2	3	4	5	6
AE1/AE3	+++ (c)	+++ (c)	+++ (c)	+++ (c)	+++ (c)	+++ (c)
CK7	+++ (c)	−	+ (c)	+++ (c)	++ (c)	+++ (c)
CK20	−	−	−	−	−	−
Vimentin	++ (c)	+++ (c)	++ (c)	++ (c)	+++ (c)	+++ (c)
EMA	+++ (c)	+++ (c)	+++ (c)	++ (c>m)	+ (c)	+++ (c)
INI1	+++ (n)	+++ (n)	−	+++ (n)	+++ (n)	+++ (n)
E-cadherin	+ (m^a>c^a)	−	−	−	−	+ (c^a>m^a)
β-catenin	+ (m^a>c^a)	+ (m^a>c^a)	+ (c)	++ (c>m^a)	+ (c>m^a)	+ (c>m^a≫n)
p62/SQSTM1	++ (c)	++ (c>n)	++ (c)	++ (c>n)	++ (c>n)	+++ (c>n)
Ubiquitin	+++ (c>n)	++ (c>n)	++ (c)	+++ (c>n)	+ (c>n)	+++ (c>n)
KEAP1	++ (c)	++ (c>n)	++ (c)	++ (c>n)	+ (c>n)	++ (c>n)
NRF2	++ (n/c)	+++ (n/c)	++ (n/c)	++ (n/c)	+++ (n/c)	+++ (n/c)
MRP1	++ (c)	+++ (c)	++ (c)	++ (c)	+++ (c)	+++ (c)

−, absent; +, less than 25 %; ++, 25-50 %; +++, more than 50 %

m membranous, *c* cytoplasmic, *n* nuclear immunoreactivity

[a] Faint expression

A spindle cell anaplastic pancreatic carcinoma with rhabdoid features following curative resection

TOMOYUKI ABE[1], HIRONOBU AMANO[1], KEIJI HANADA[2], AKIHISA OKAZAKI[2], SHUJI YONEHARA[3], FUMITO KURANISHI[4], MASAHIRO NAKAHARA[1], YOSHINORI KURODA[1] and TOSHIO NORIYUKI[1,5]

Departments of [1]Surgery, [2]Gastroenterology and [3]Pathology, Onomichi General Hospital, Onomichi, Hiroshima 722-8508; [4]Department of Surgery, Innoshima-Ishikai Hospital, Innoshima, Hiroshima 1722-2323; [5]Department of Gastroenterological and Transplant Surgery, Applied Life Sciences, Institute of Biomedical and Health Sciences, Hiroshima University, Onomichi, Hiroshima 1734-8551, Japan

Received February 1, 2016; Accepted May 9, 2016

DOI: 10.3892/mco.2016.914

Abstract. Anaplastic pancreatic carcinoma (ANPC) accounts for ~5% of all pancreatic ductal adenocarcinoma cases. Due to its rarity, its clinical features and surgical outcomes remain to be clearly understood. A 74-year-old woman was admitted to Onomichi General Hospital (Onomichi, Japan) in April 2015 without any significant past medical history. Contrast-enhanced computed tomography (CT) revealed a 9.5x8.0 cm tumor in the body and tail of the pancreas. The patient developed acute abdominal pain 3 weeks later and the CT revealed massive abdominal bleeding caused by tumor rupture. The tumor increased in size and reached 12.0x10.0 cm in maximal diameter. The tumor doubling time was estimated to be 13 days. ^{18}F-fluorodeoxyglucose (FDG) positron emission tomography/CT confirmed the absence of distant metastasis since FDG accumulation was detected only in the tumor lesion. Emergency distal pancreatectomy and splenectomy were performed. Histologically, the tumor was classified as a spindle cell ANPC with rhabdoid features. The patient succumbed to mortality 8 months following the surgery while undergoing systemic adjuvant chemotherapy for multiple liver metastases. ANPC is difficult to detect in the early stages due to its progressive nature and atypical radiological findings. Long-term survival can be achieved only by curative resection; therefore, surgical resection must be performed whenever possible, even if the chance of long-term survival following surgery is considered dismal. As the present case suggested, spindle cell ANPC with rhabdoid features is highly aggressive and curative-intent resection must not be delayed.

Introduction

Since anaplastic pancreatic carcinoma (ANPC) is rare and accounts for only 2-7% of all pancreatic carcinoma cases, its clinical features and surgical outcomes remain to be elucidated (1,2). A total of three pathological subtypes of ANPC exist: Spindle cell carcinoma, giant cell carcinoma, and pleomorphic carcinoma. Surgical resection is the only curative therapy for patients with ANPC since no effective systemic chemotherapy or other interventions are available; however, long-term survival remains to be achieved in patients with ANPC even following curative surgery (1,3). ANPC is often associated with a delay in clinical presentation as a result of its asymptomatic nature until the tumor has progressed to an advanced stage. In several reports that have been published since the early 1900s, ANPC was referred to as 'giant-cell tumor', 'undifferentiated carcinoma' with or without osteoclast-like giant cells and 'pleomorphic carcinoma' of the pancreas (3-6).

Establishing the precise pre-operative diagnosis is difficult due to the radiological findings being atypical and similar to those of gastrointestinal stromal tumors, mucinous cyst adenocarcinomas and pancreatic carcinomas. Upon admission, the present patient's tumor was already huge and revealed enhanced rims with hypodense lesions on computed tomography (CT) scans, which appears to be a common radiological feature of ANPCs (1).

Of the three subtypes, spindle cell carcinoma is the most aggressive subtype of sarcomatoid carcinoma and currently no

Summaries of previously reported cases

Case	Sex	Age	Site	Rhabdoid	Duration	Outcome
1	F	52	T	?	19 months	DOD
2	F	68	H	?	2 months	DOD
3	M	59	H	60%	2 months	DOD
4	M	63	BT	?	3 months	DOD
5	F	51	BT	30%	3 months	?
6	M	44	BT	15%	24 months	?
7	F	68	B	50%	2 weeks	Palliative care
8	M	59	B	75%	3 months	Palliative care
9	M	68	H	40%	1 month	Palliative care
10	M	76	BT	80%	2 months	Palliative care
11	F	74	BT	?	8 months	DOD
12	F	81	H	60%	5 months	AWOD

12例のまとめ

男女比: 6:6

年齢: 44~81歳、平均 63.6歳

発生部位 :

頭部 4:体部 2:尾部 1:体尾部 5

予後:不良