OS-OM Combined Case Conference
Pleomorphic Rhabdomyosarcoma with Telangiatasia

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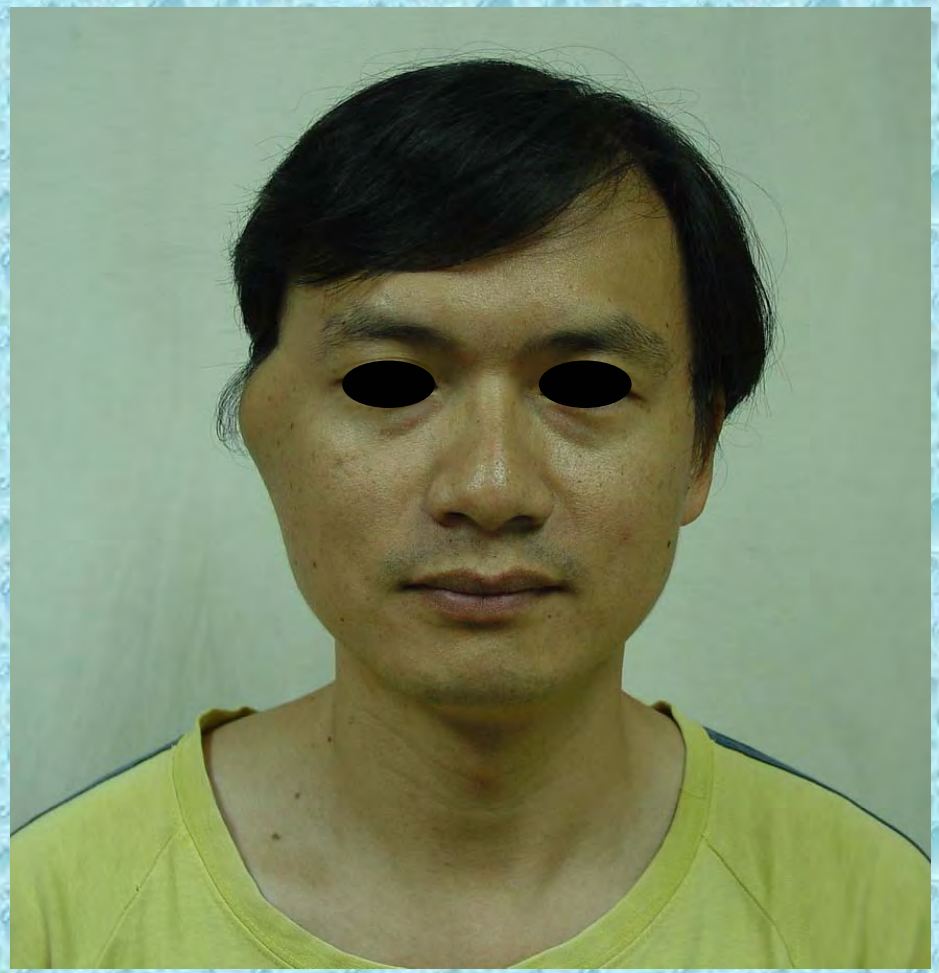
指導醫師 ： 黃逸岳 、陳玉昆

94-06-23

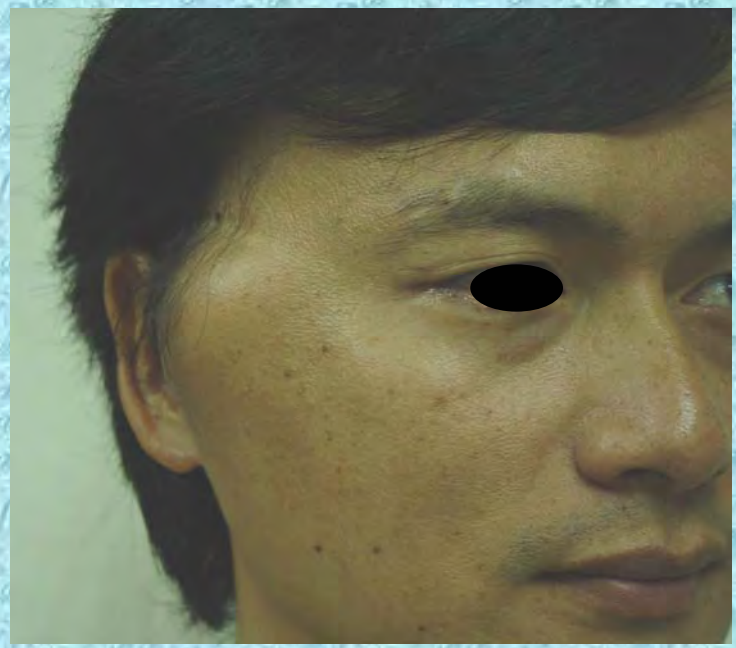
Case Data

- Name : 鍾X X
- Gender : Male
- Age : 39
- Marital status : Yes
- Native : 屏東縣
- Occupation : 商
- Attending V.S. :
  黃逸岳醫師
- First visit : 900614



Chief Complaint

- Swelling over right cheek and preauricular to temporal area for half a year.



Present Illness

- This 39 y/o male patient found swelling over right cheek and preauricular to temporal area for four years.
- 900614 He noted the swelling and mouth open limitation when he took food and felt discomfort over bilateral faces, so he came to 屏基 H for help.
- Referred to KMU OM department for further exam.
  - R/O sialolithiasis or TMD
  - referred to OMS department --- op suggestion – patient think
  - ~901119 OPD F/U
- 9307 patient found the lesion got larger.
- 940226 came back to our OPD

Past History

- Past medical history
  - Denied systemic disease
  - Denied drug allergy
- Past dental history
  - OD , prosthesis : acceptable

Personal History

- Oral habit : A(-)B(-)C(-)
- Occupation : 從商
- Education : 高職
- Marital status : 已婚
- Economic state : 小康
- Psychic status : 樂觀

Family History

- 家族無系統性疾病史
- No contribution to this problem
- Family support： well--住院期間將由父母及妻子照顧

Review of System (nothing particular)

- Cardiovascular system
- Central nervous system
- Respiratory system
- Digestive system
- Endocrine system
- Hematopoietic system
- Genitourinary system
- Bone and joint

Physical Examination

(Essentially normal)

- Conscious : clear , vital sign : stable
- Head & Face : asymmetry
- Eye : conjunctiva : not pale sclera : not icteric, EOM : normal
- Ear : no hearing impairment
- Nose : patent
- Throat : no discharge, tonsil : not injected
- Neck : LAP (-), JVE (-), supple, free extension
- Chest : symmetric expansion H.S. : no murmur, RHB B.S. : clear
- Abd. : soft, no palpable mass, normative bowel sound
- L/L : free extension, no pitting edema
- CN12 : N.P.

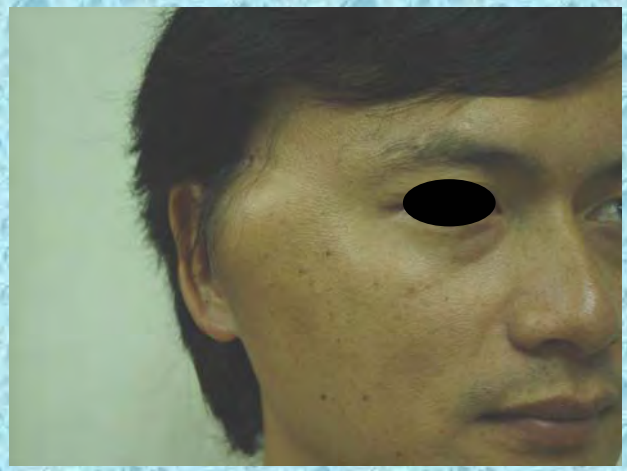
Lab. Data

(Essentially normal)

- RBC : 4.17 x10^6 /ul PLT : 26.2 x10^4 /ul
- WBC : 4.31 x10^3 /ul HGB : 12.4 g/dl
- HCT : 38.8%
- PT: 11.1/ 11.1 PTT :34.2/ 27.5
- GOT / GPT : 23/23
- BUN / Cr. : 20/1.0
- NA / K / Cl : 142 / 3.6 / 108

OMF Examination

- Swelling mass over right temporal region
- Soft to rubbery in consistency
- Size : 4.5 x 4.0 x 1.5 cm
- Pain (+)
- Tenderness (-)
- LAP (-)
- MMO : 43mm from 11 to 41
- TMJ function : OK



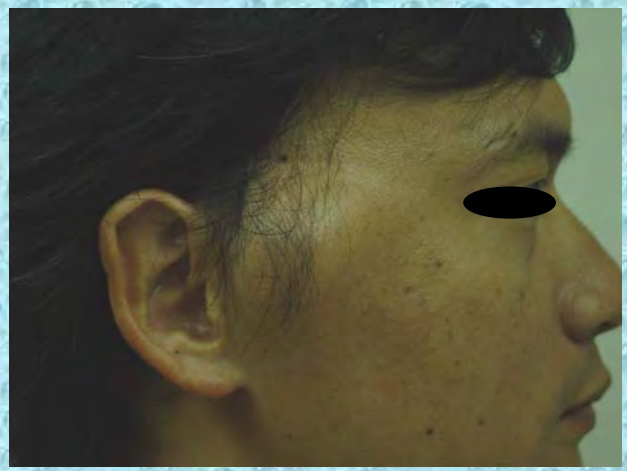


Image 940503

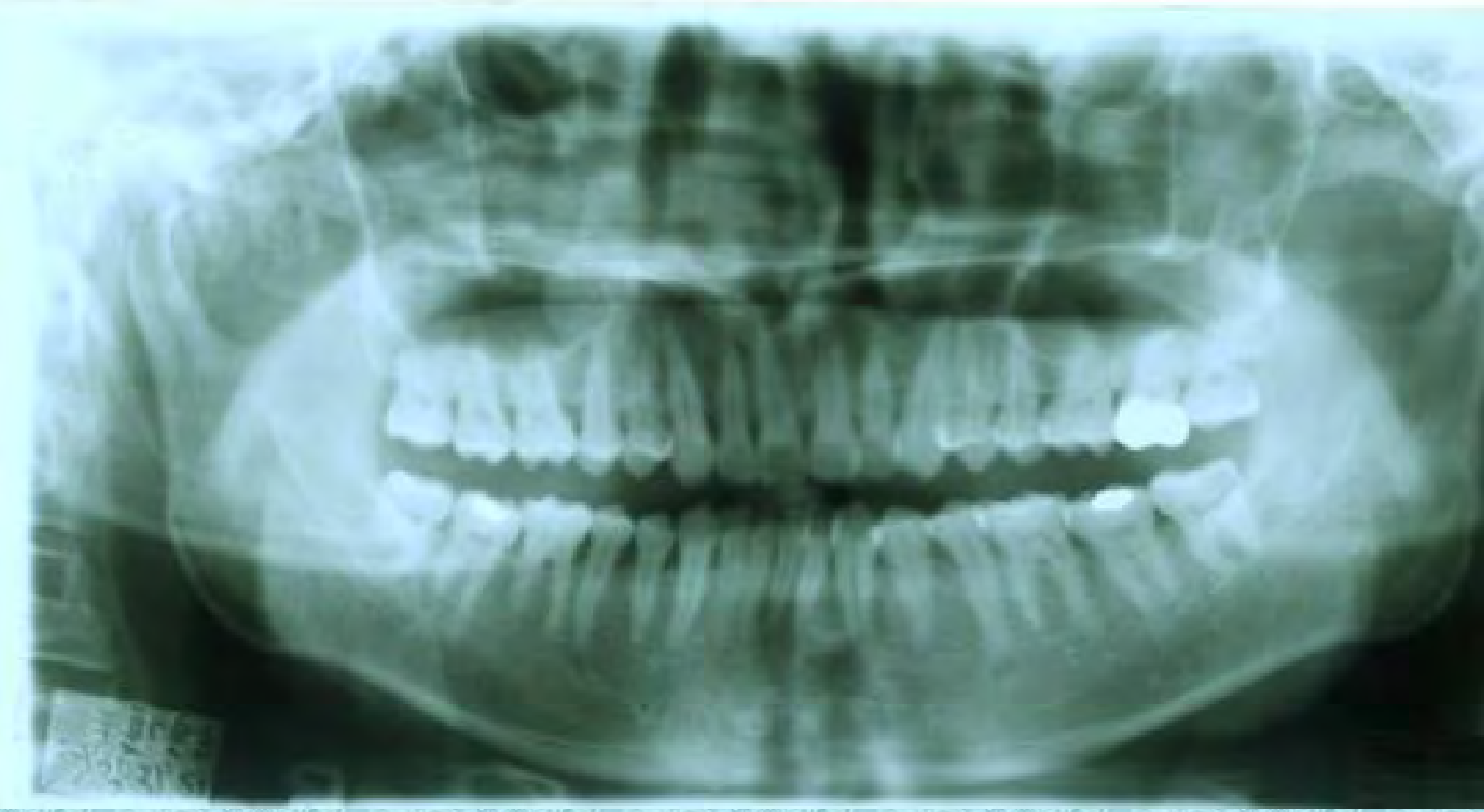
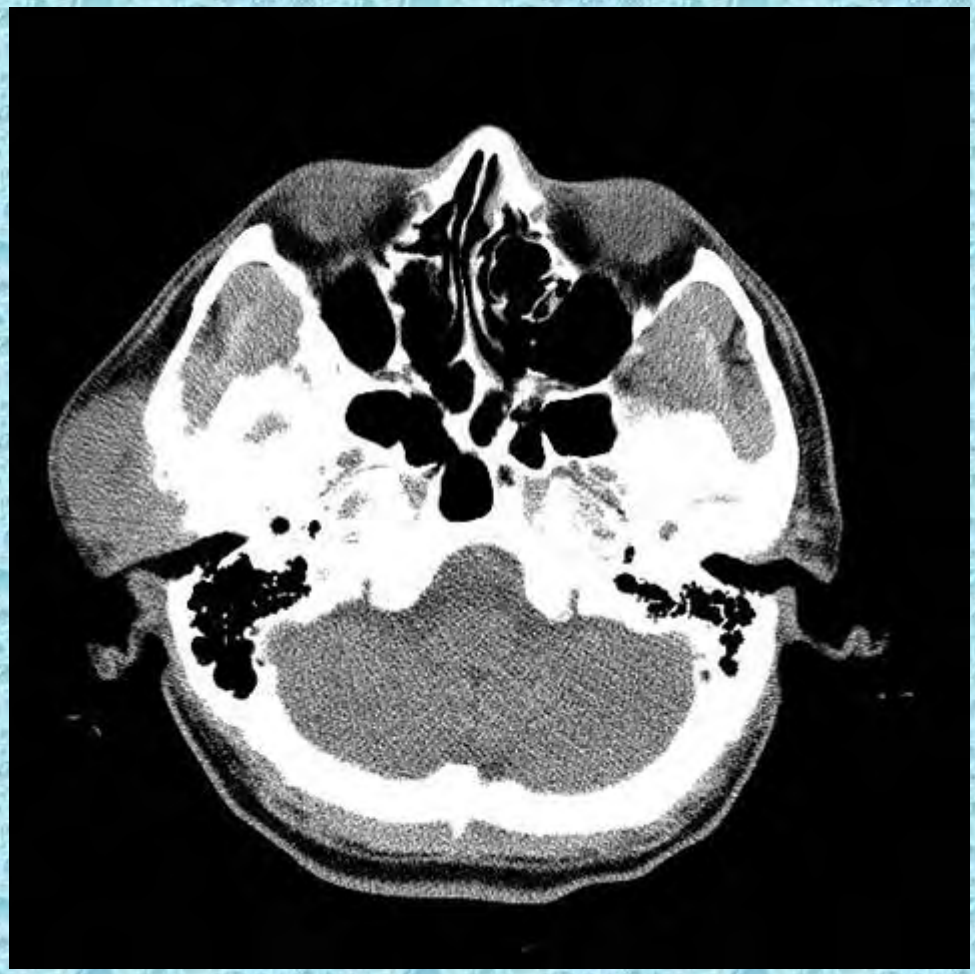


Image Findings – CT scan940304



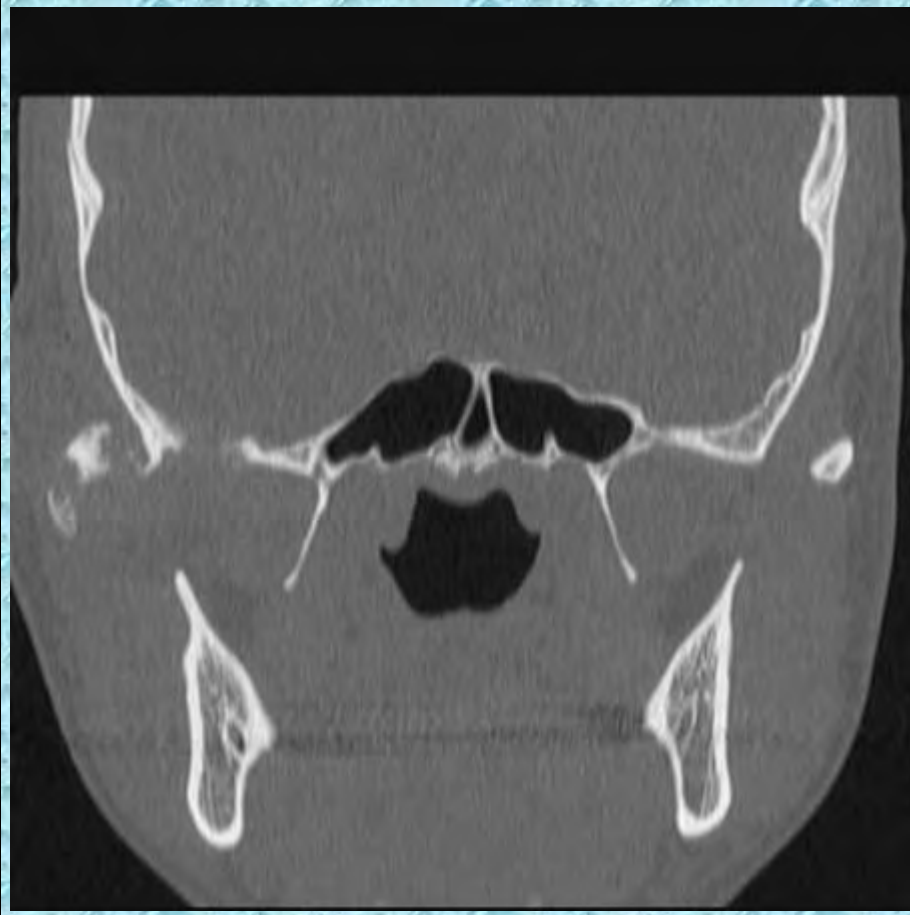
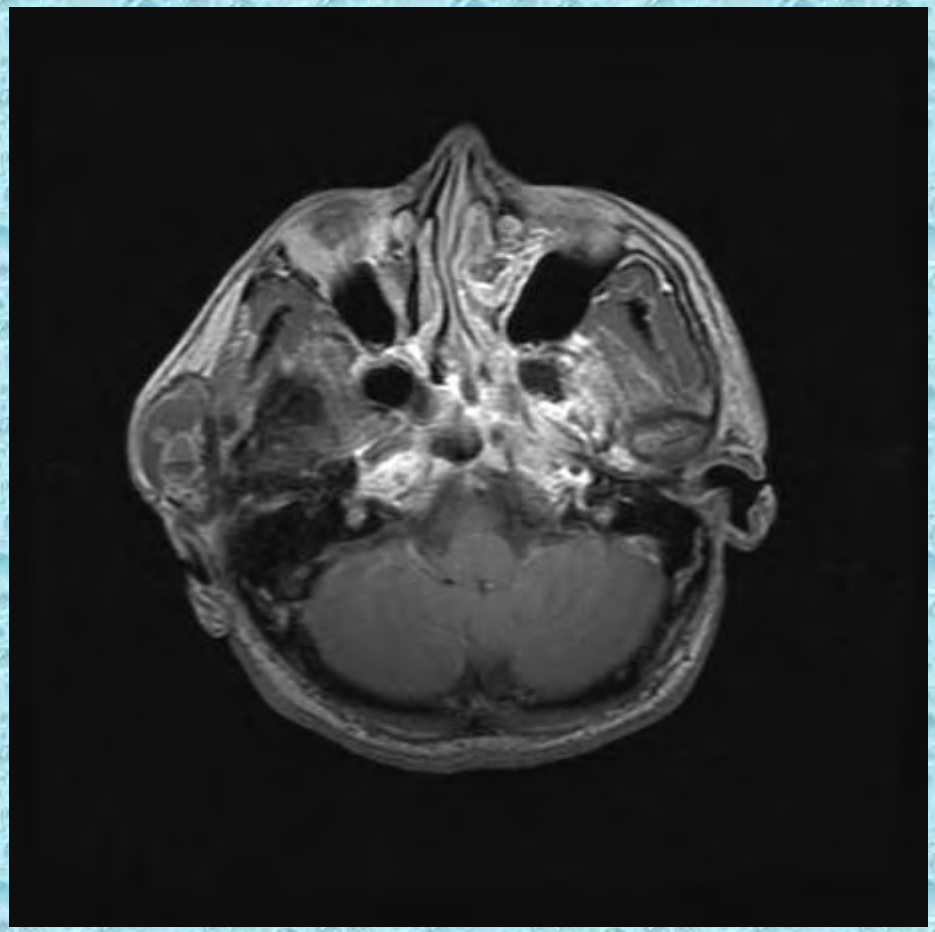


Image Findings – CT scan

- 1) Prominent soft tissue involving right infratemporal fossa and right periauricular corresponding destruction and involving of right temporal joint.R/O metastasis, recommend biopsy.
- 2) Retention cysts in bilateral maxillary sinuses.
- 3) Bilateral ethmoid sinusitis.

Image Findings – MRI 940314



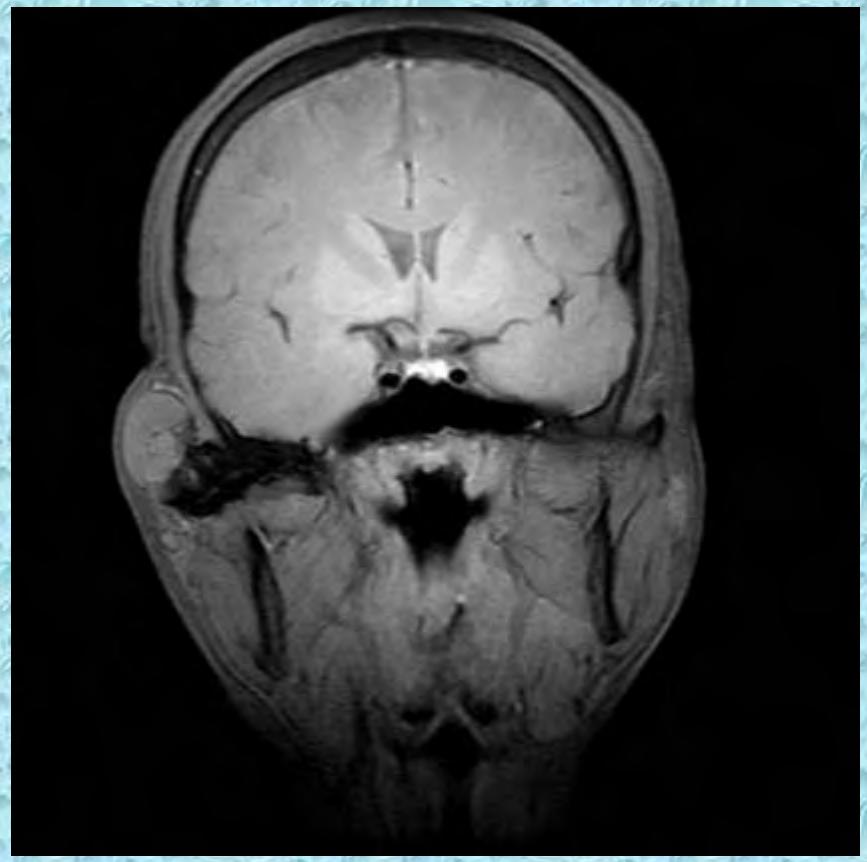


Image Findings – MRI

- 1) Interval enlargement of the soft tissue mass lesion involving right infratemporal fossa and right periauricular region with cystic component and adjacent bony remodeling of temporal bone. Interval development of hemorrhage in the cystic component of lesion.
  - D/D: hemangioma and metastasis.
  - Recommend biopsy for further evaluation.
- 2) Retention cysts in the bilateral maxillary sinuses.

Other Survey

- CXR : no active cardiopulmonary disease
- Bone scan : low probability of local bone invasion from parotid gland tumor
- SCC marker : pending

Impression and D.D.

- Pleomorphic adenoma near right parotid gland
- Warthin’s tumor
- Hemagioma
- R/O Malignant tumor

Treatment Plan

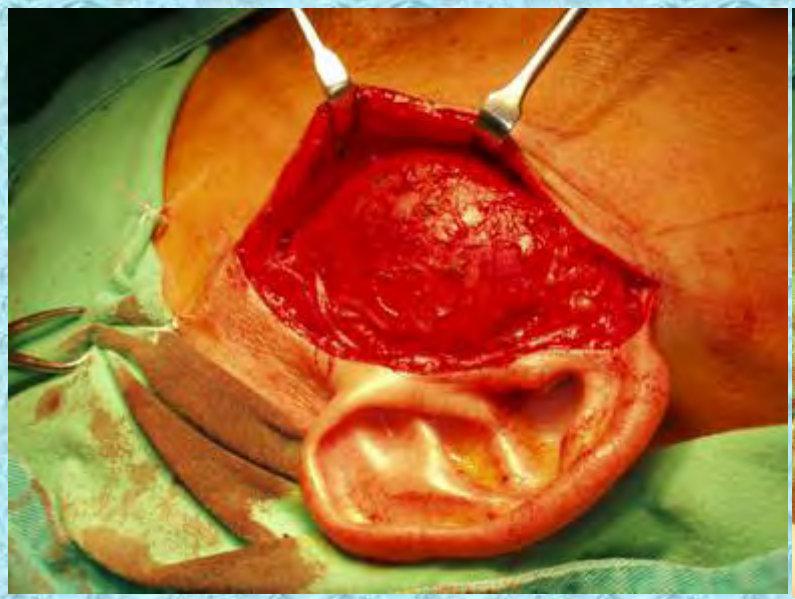
OP :

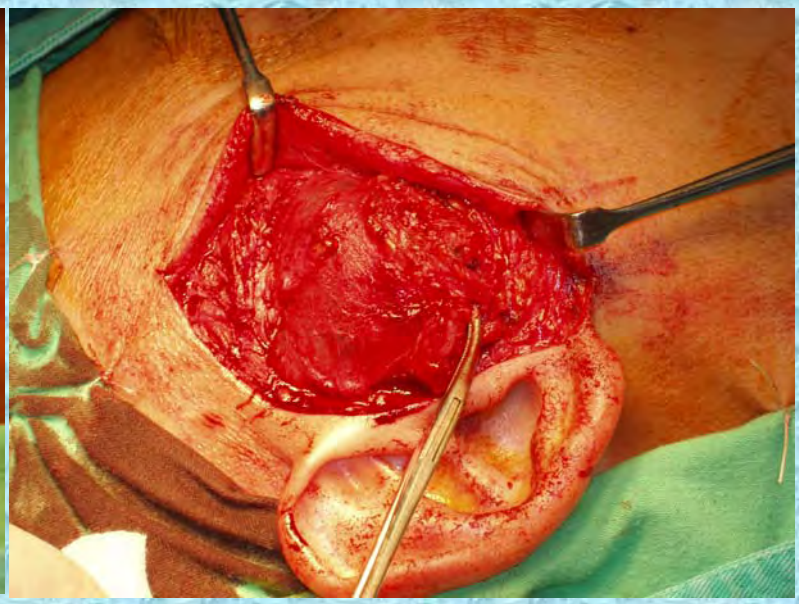
Wide excision + bone trimming

Treatment Course

- **940510 Admission**
- **GA and ENT dept. -- consultation**
- OP on 940512

OP Procedure

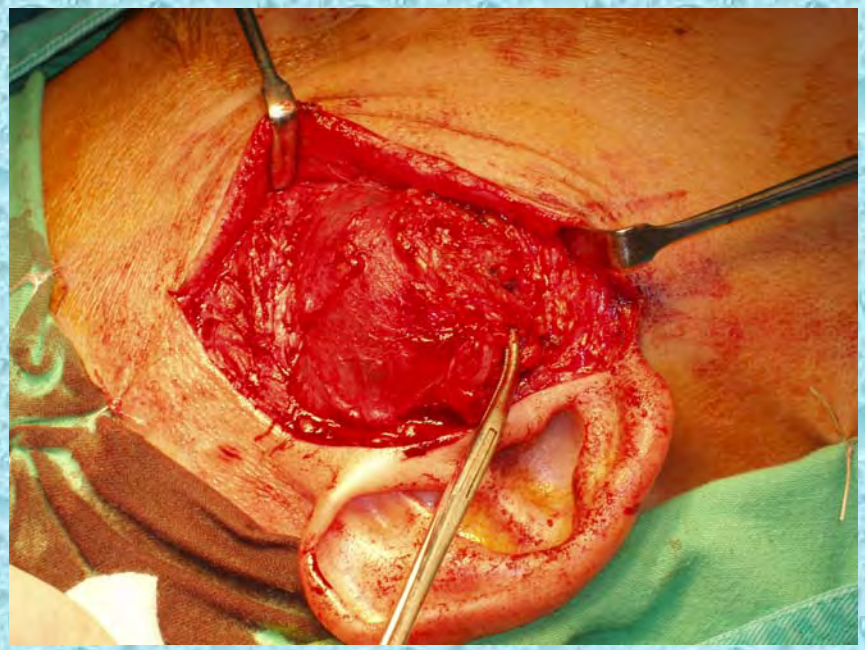


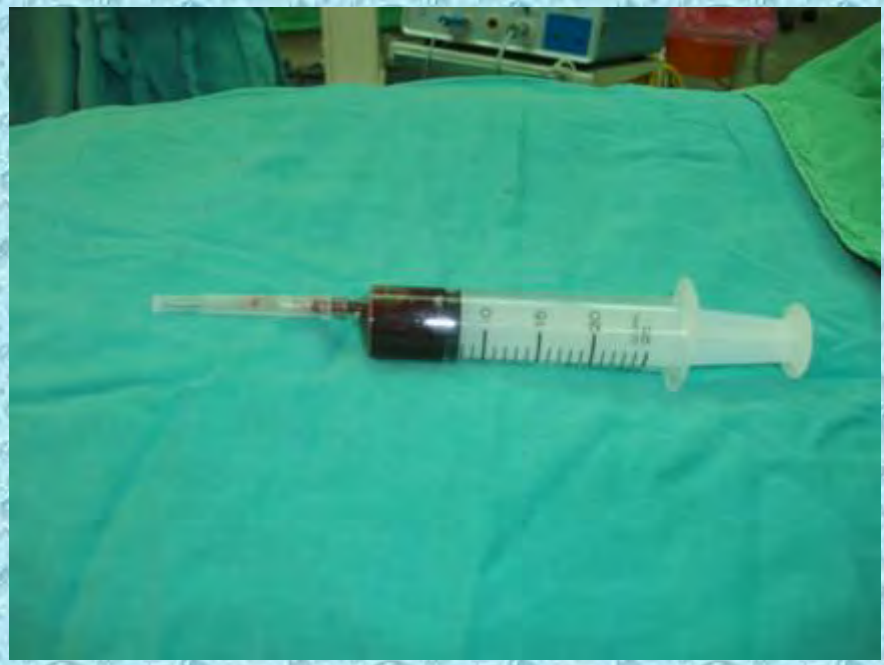


- Preauricular incision was made then dissected to expose the lesion
- Well encapsulated at superficial portion

OP Procedure(cont.)

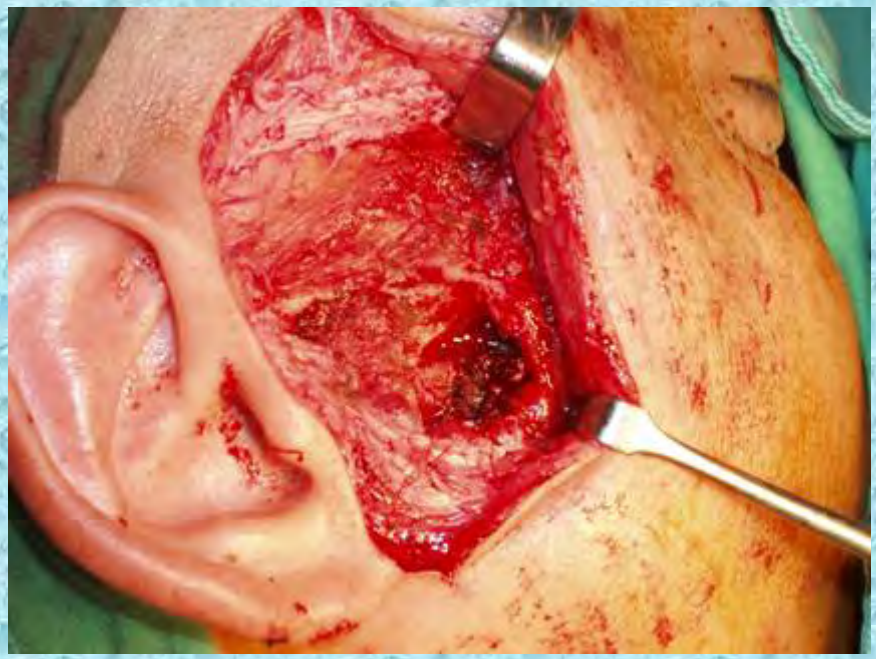
- Aspiration : yellowish fluid followed by dark reddish fluid
- Ruptured during dissection : dark reddish jelly-like mass

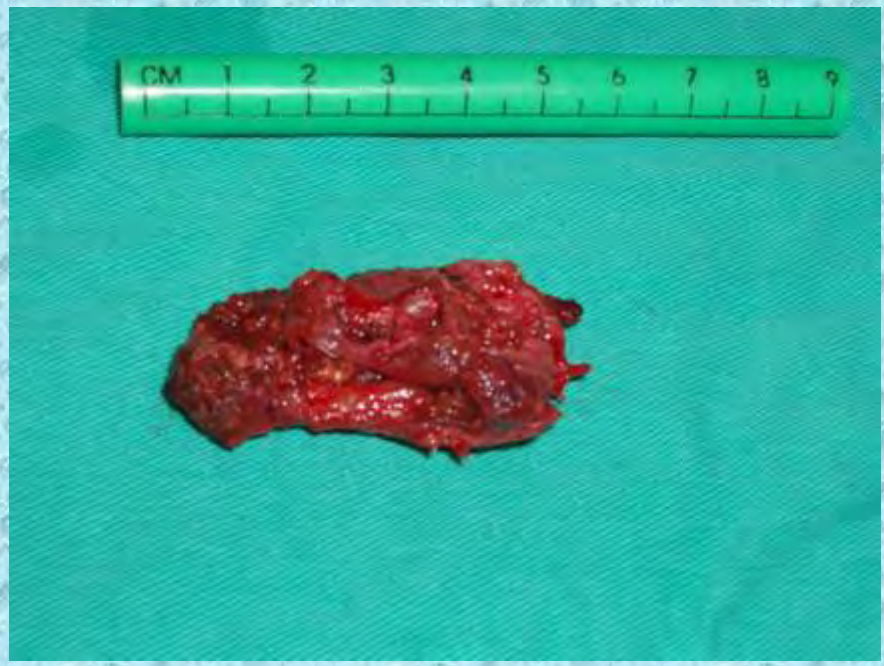




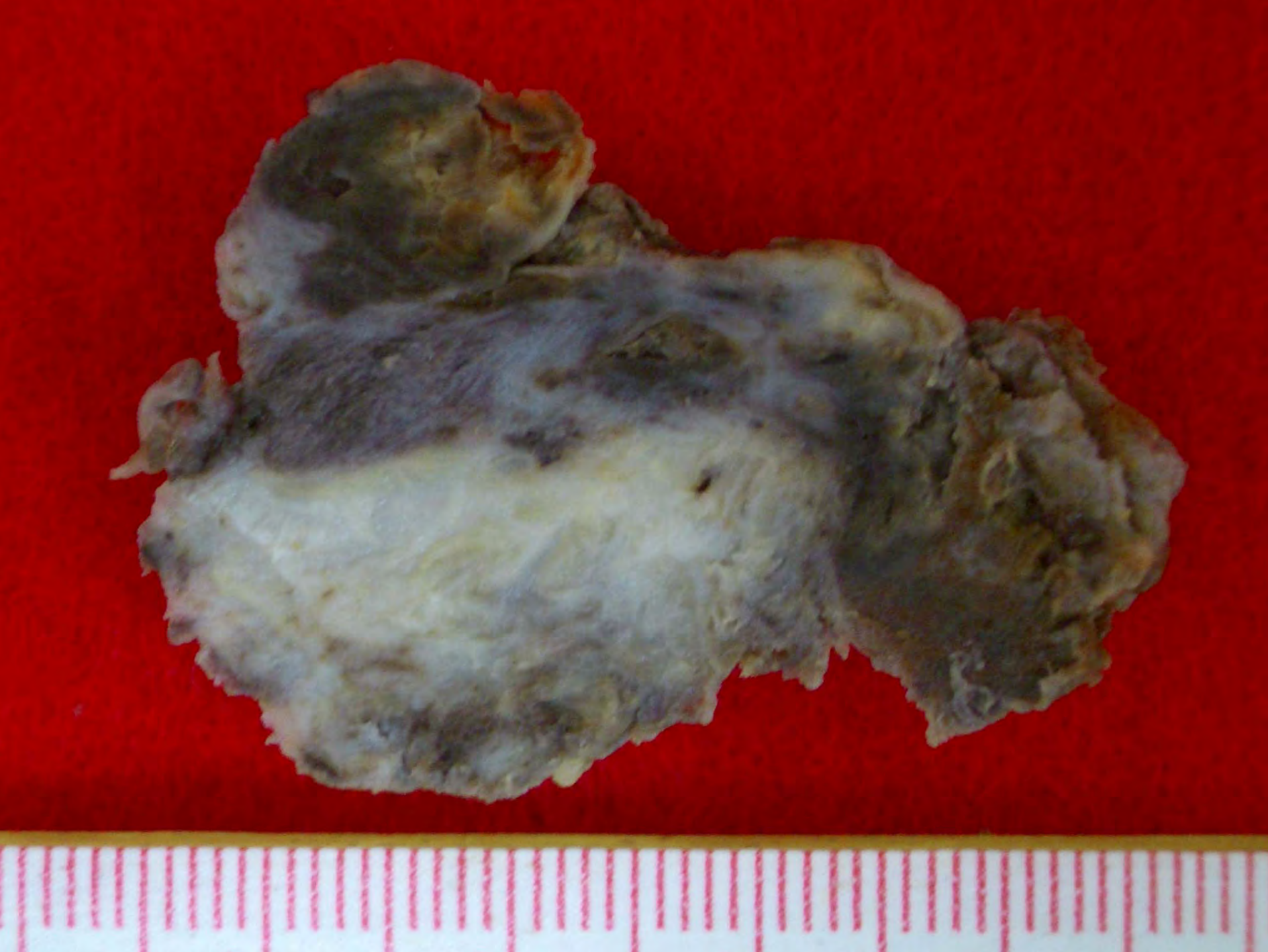
OP Procedure (cont.)

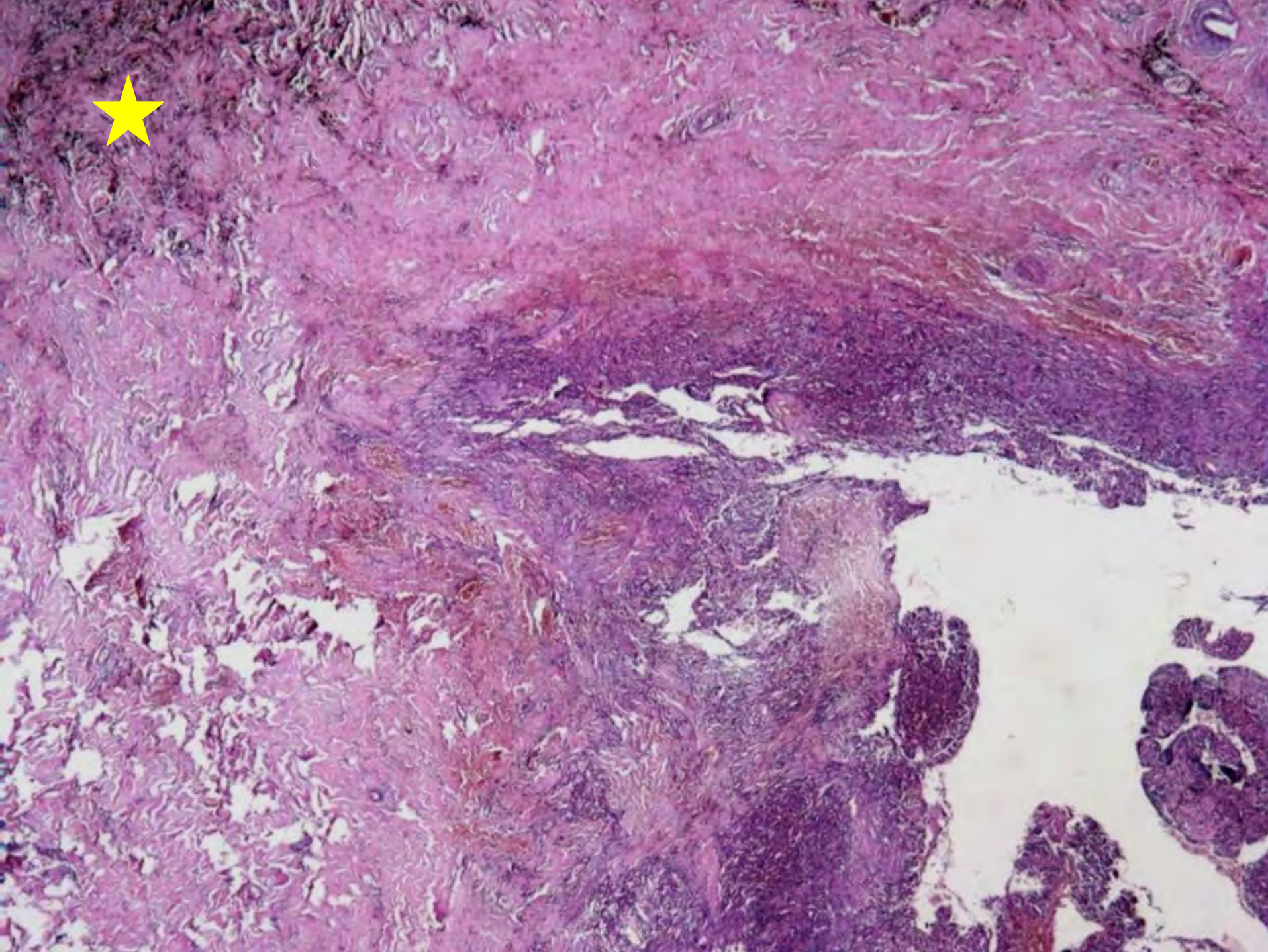
- Bony destruction of zygoma, pterygoid plate, zygomatic arch
- Bone trimming was done
- Closed the wound layer by layer after placed of penrose

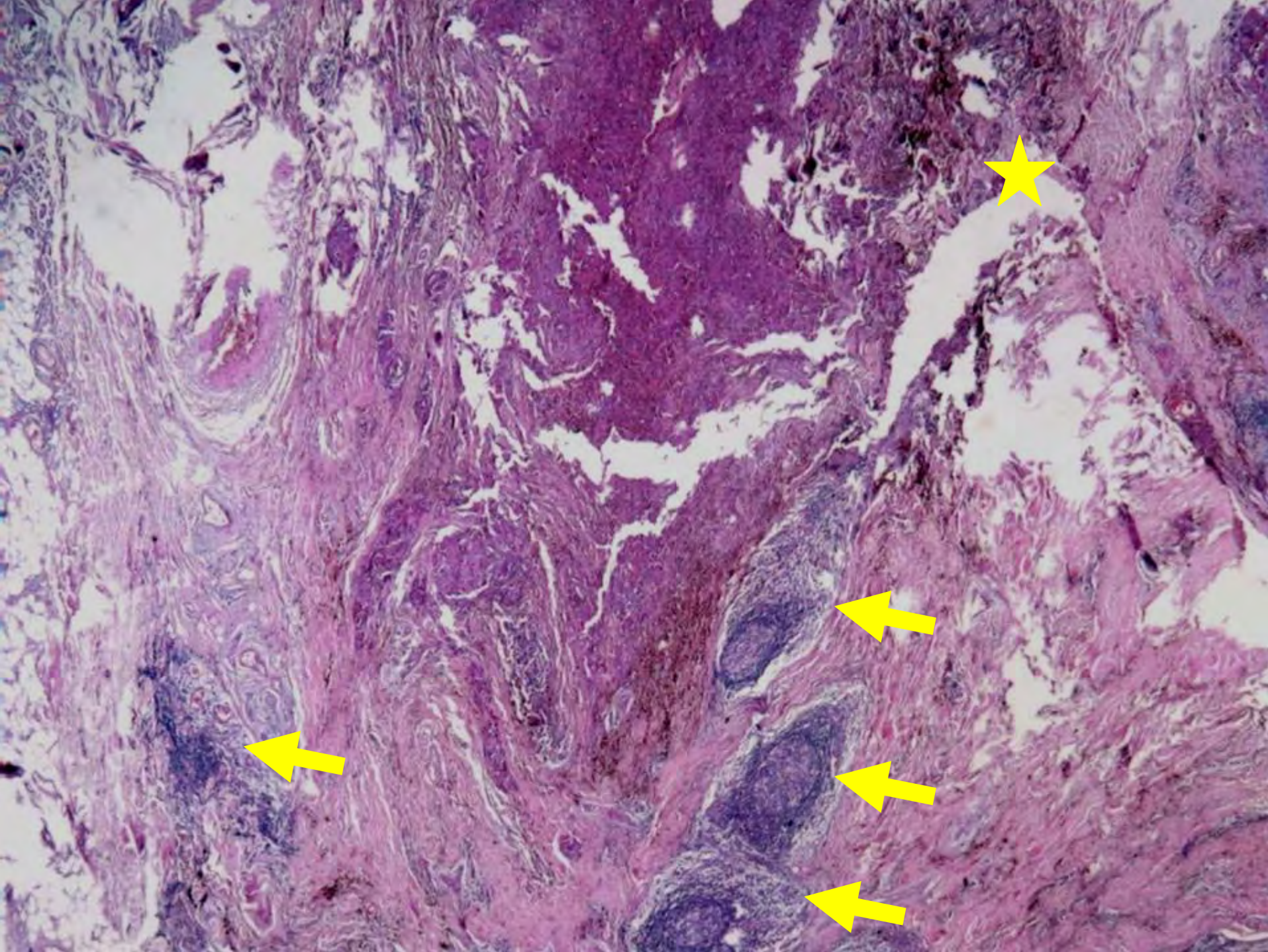


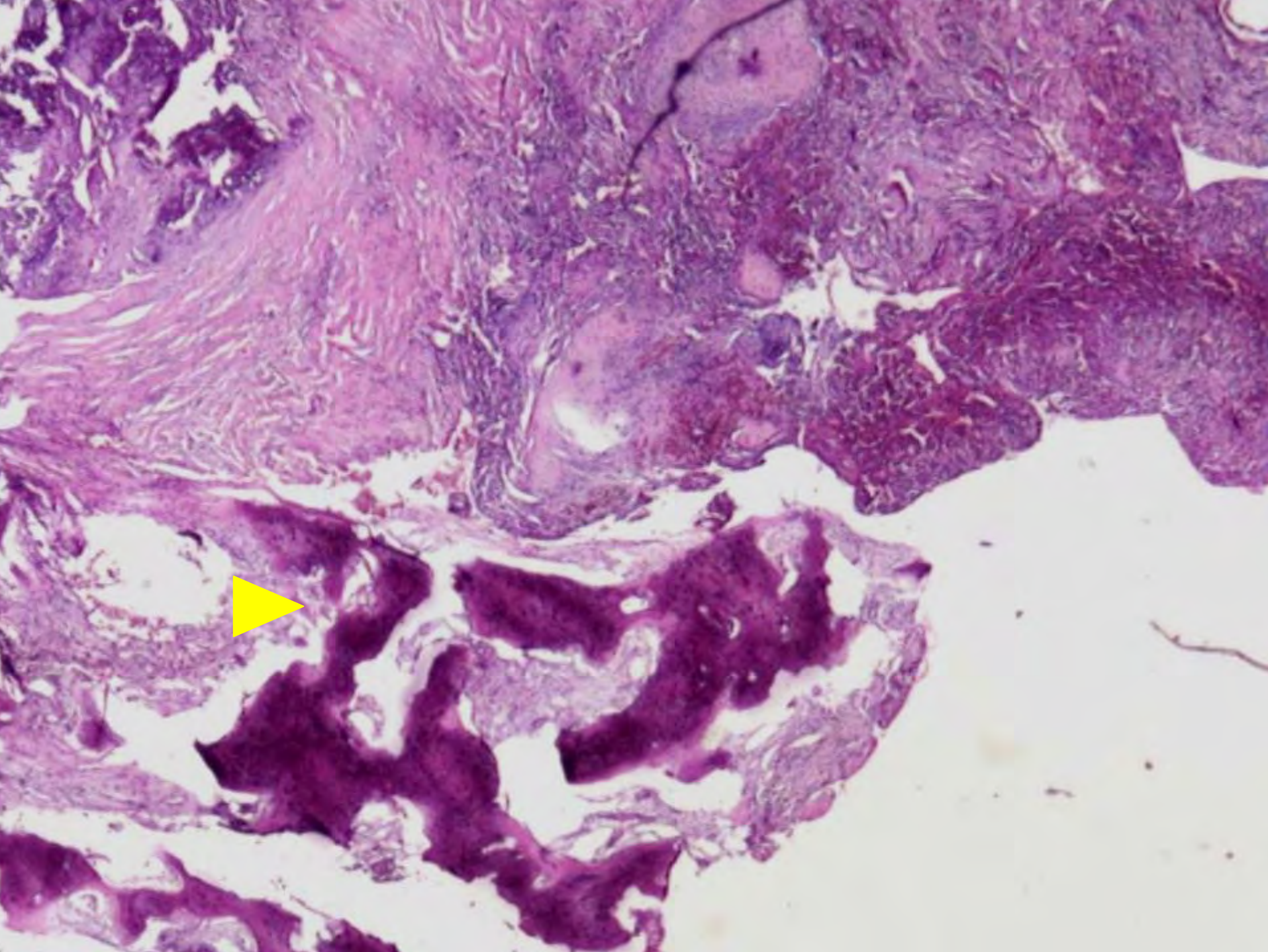


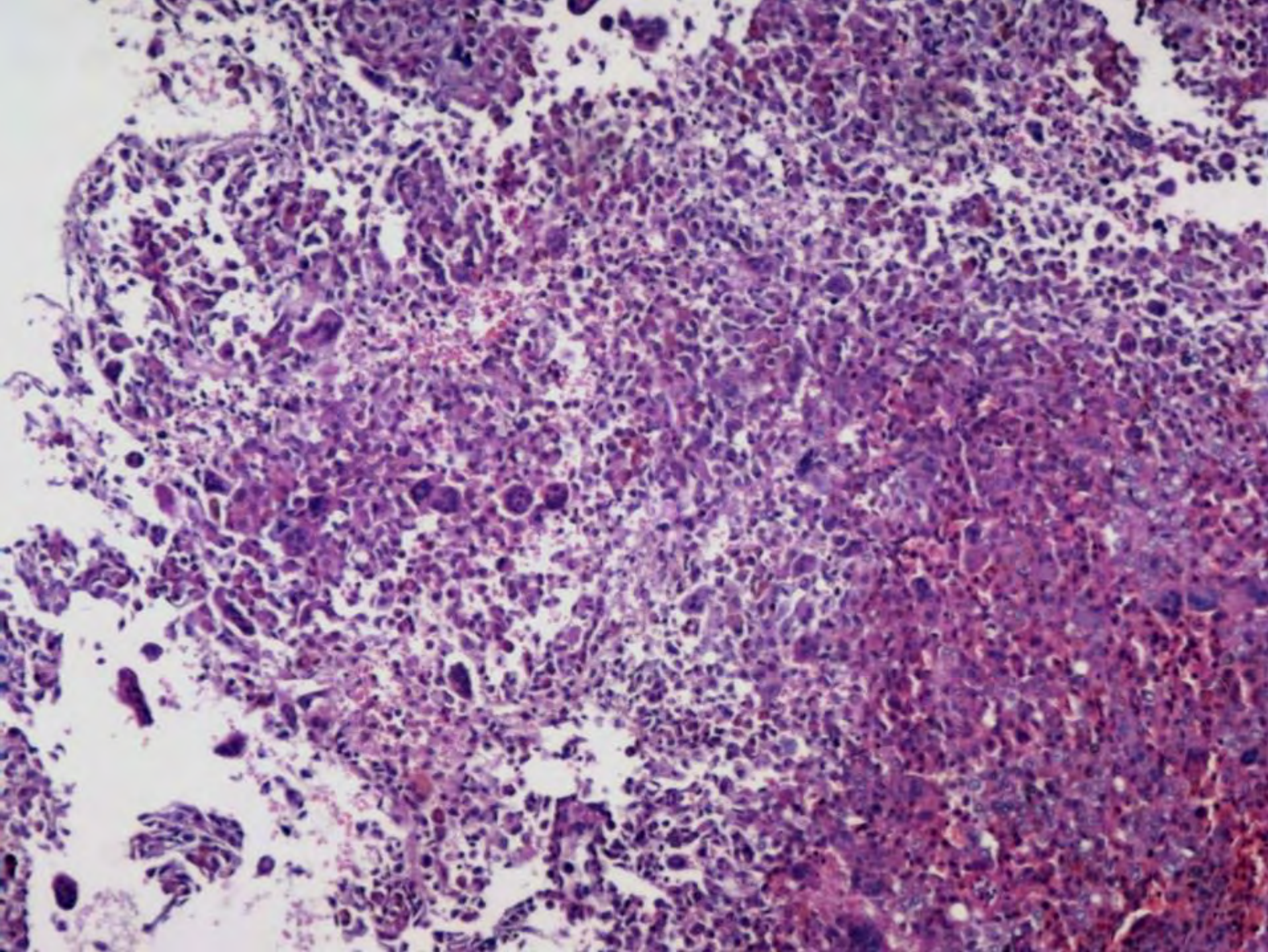
Histopathological findings

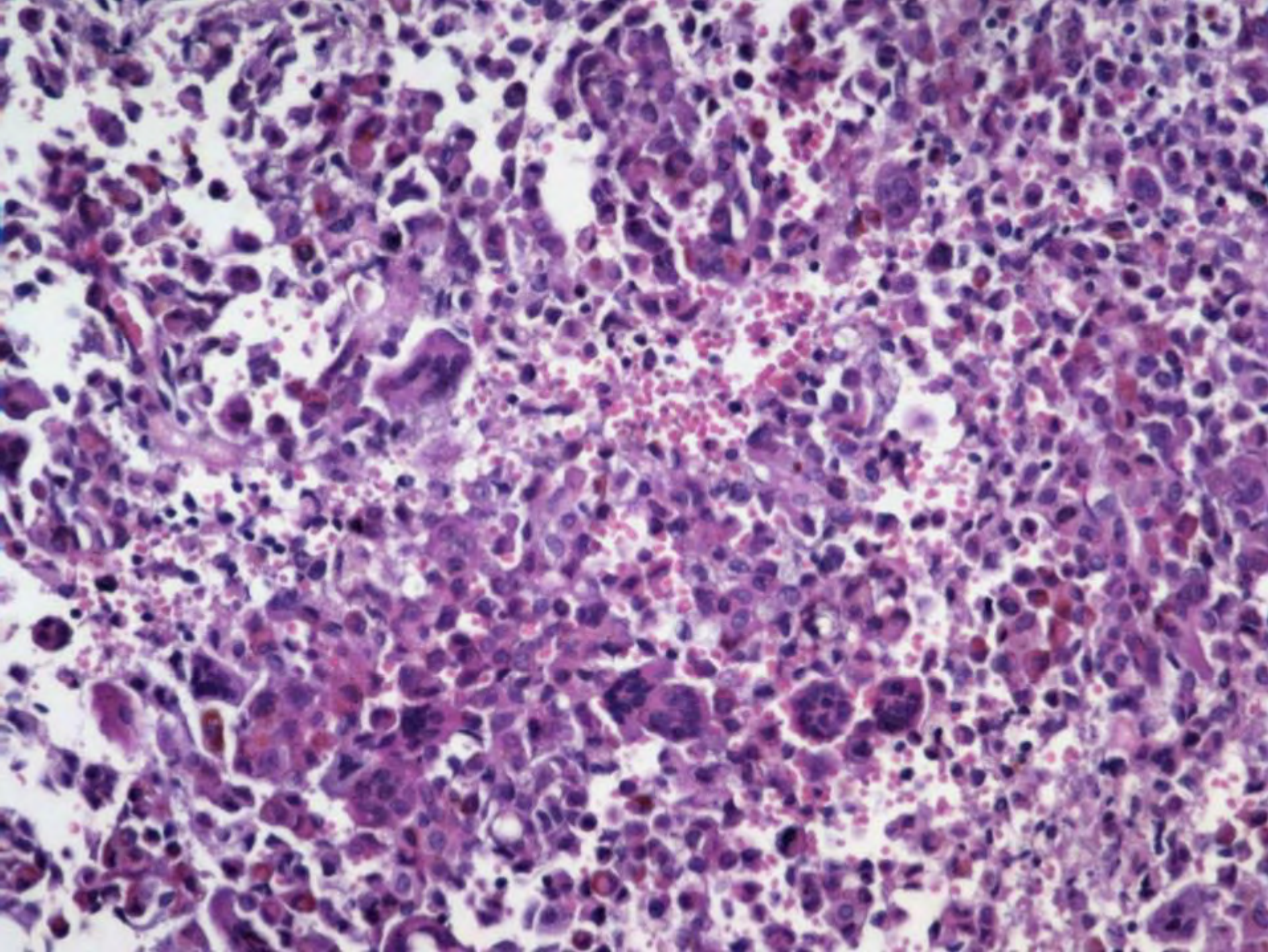


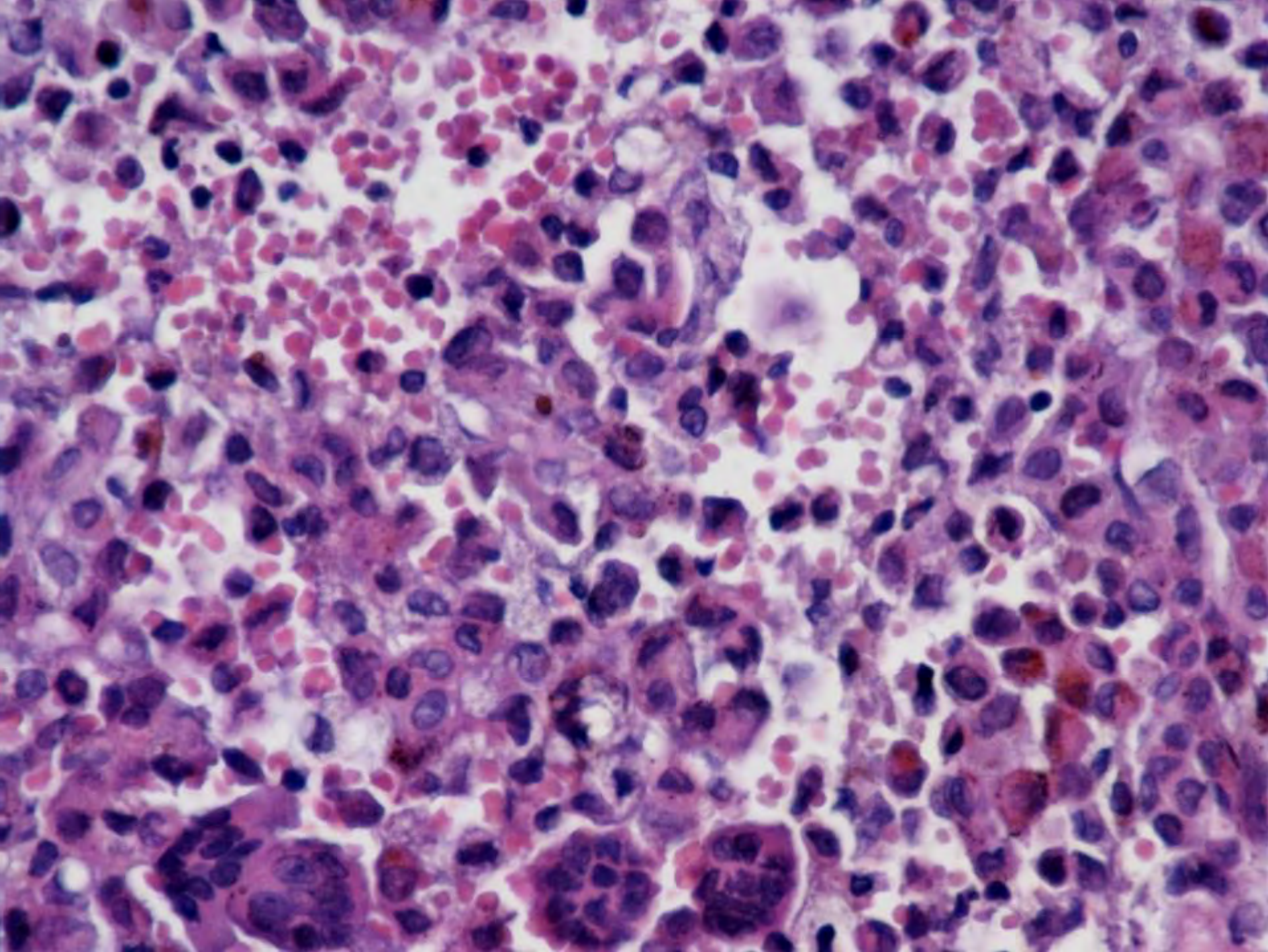


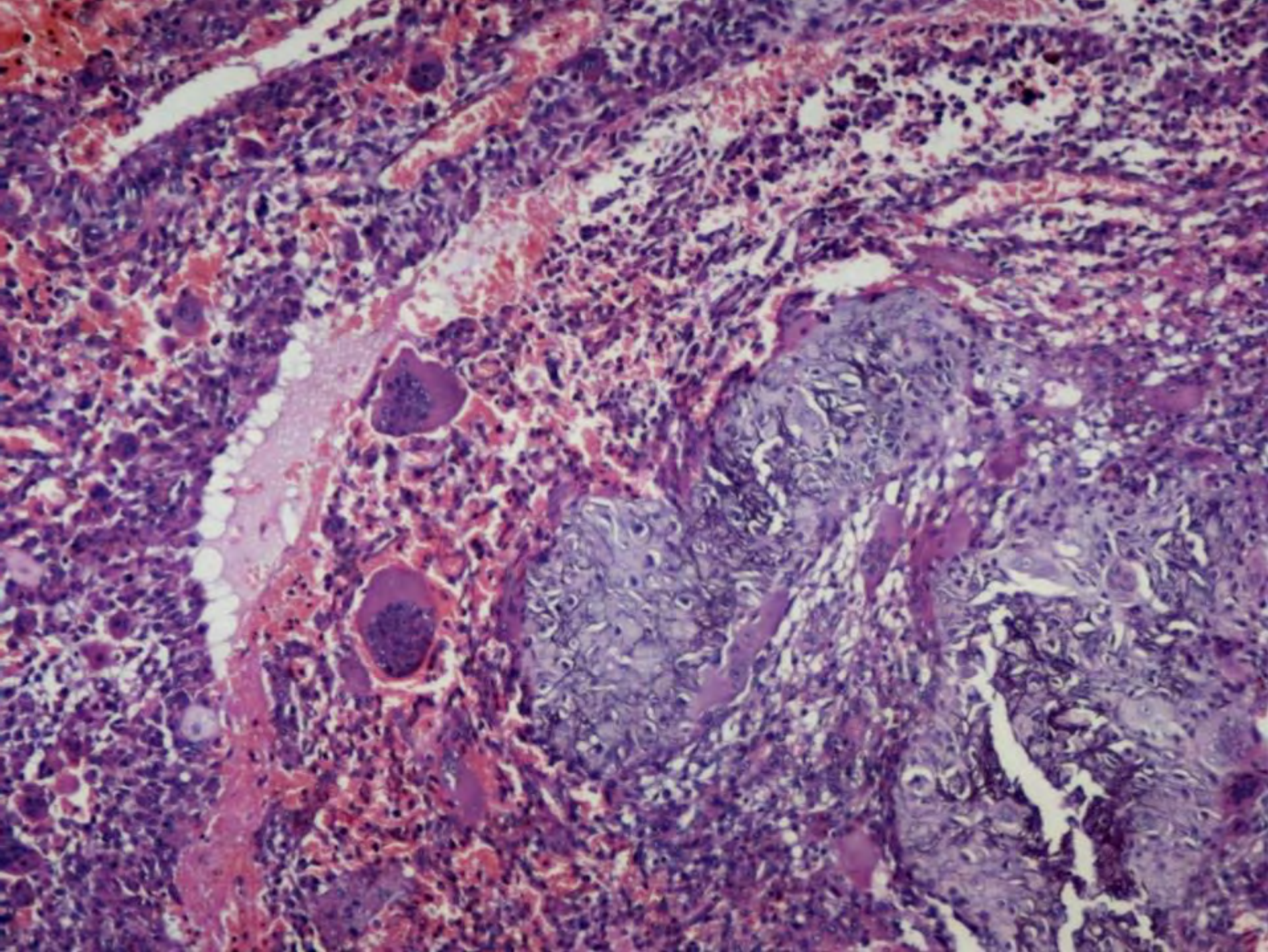


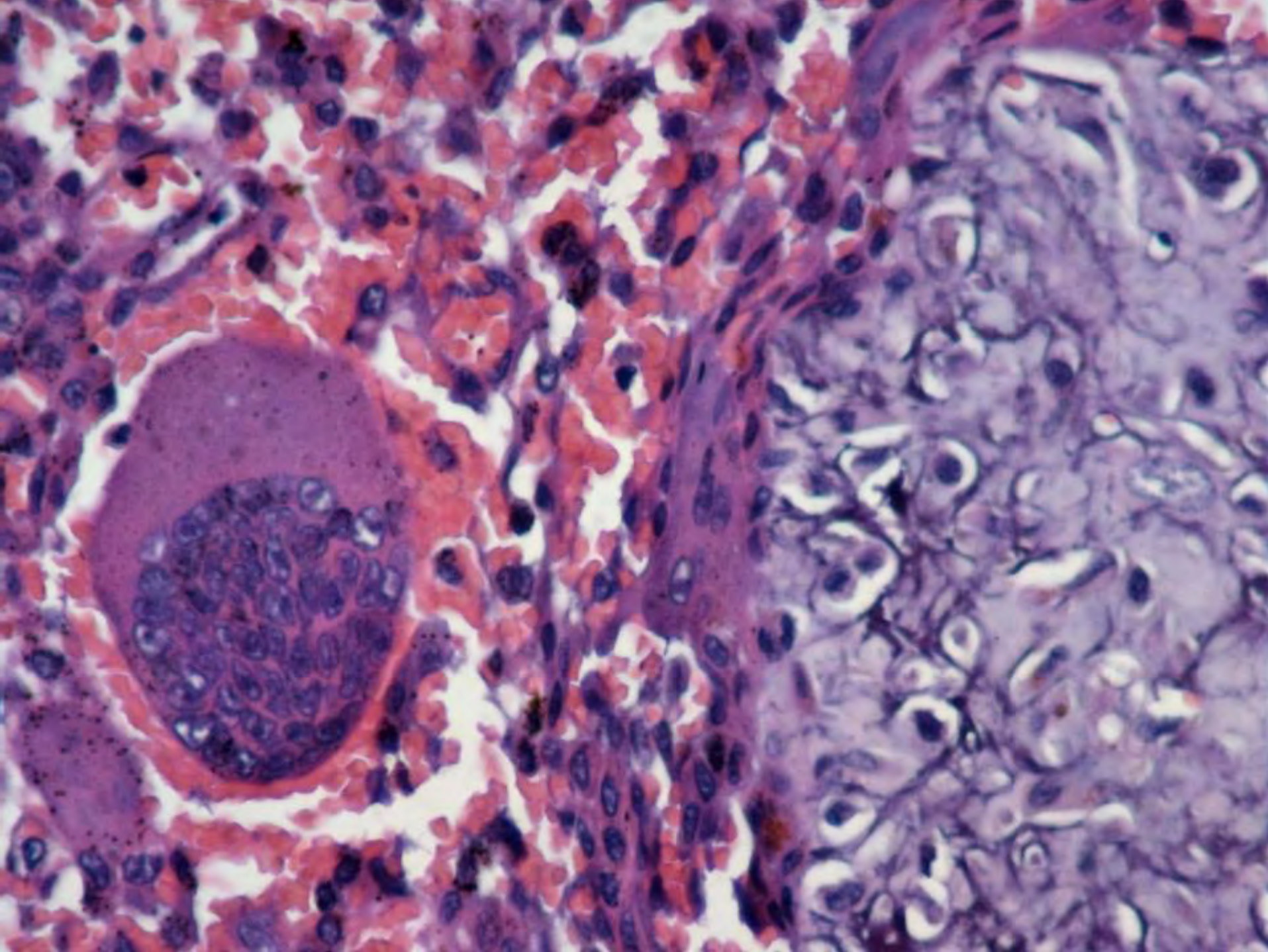


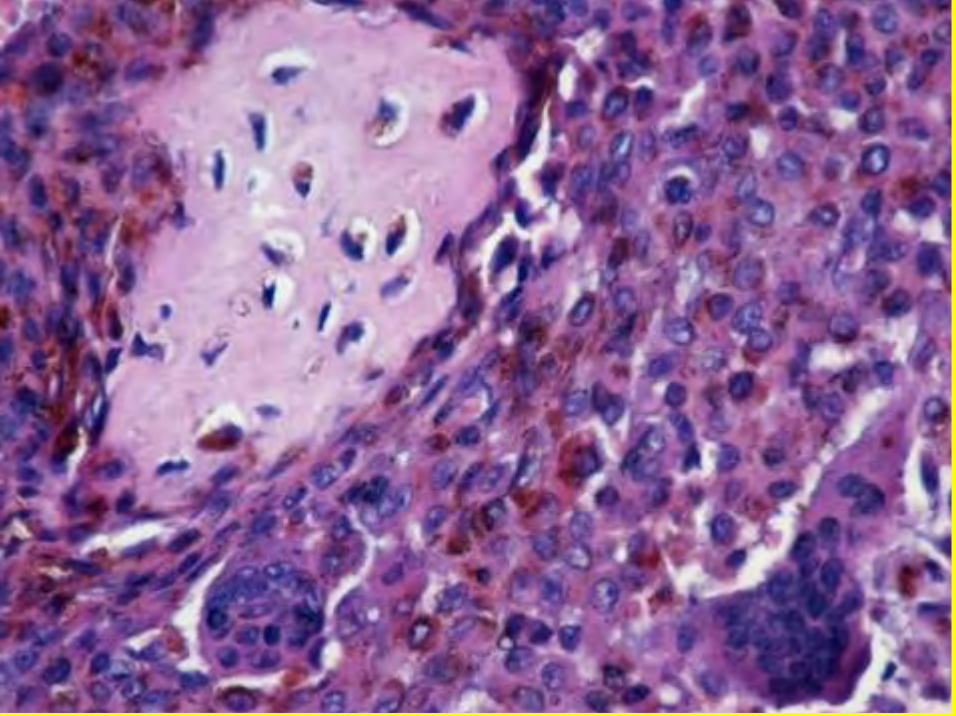
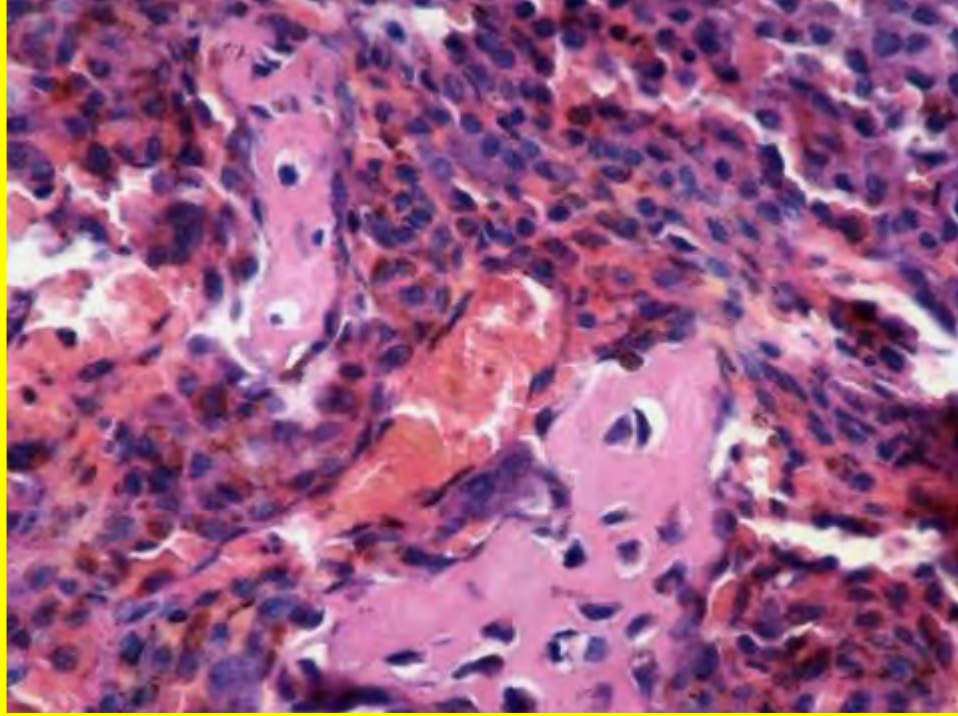
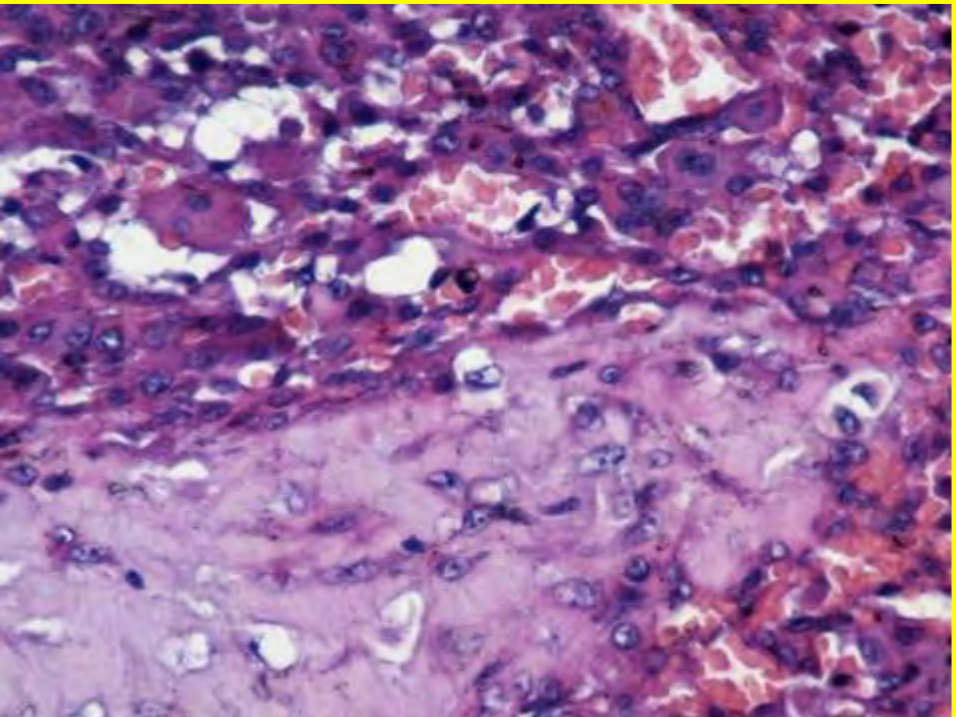
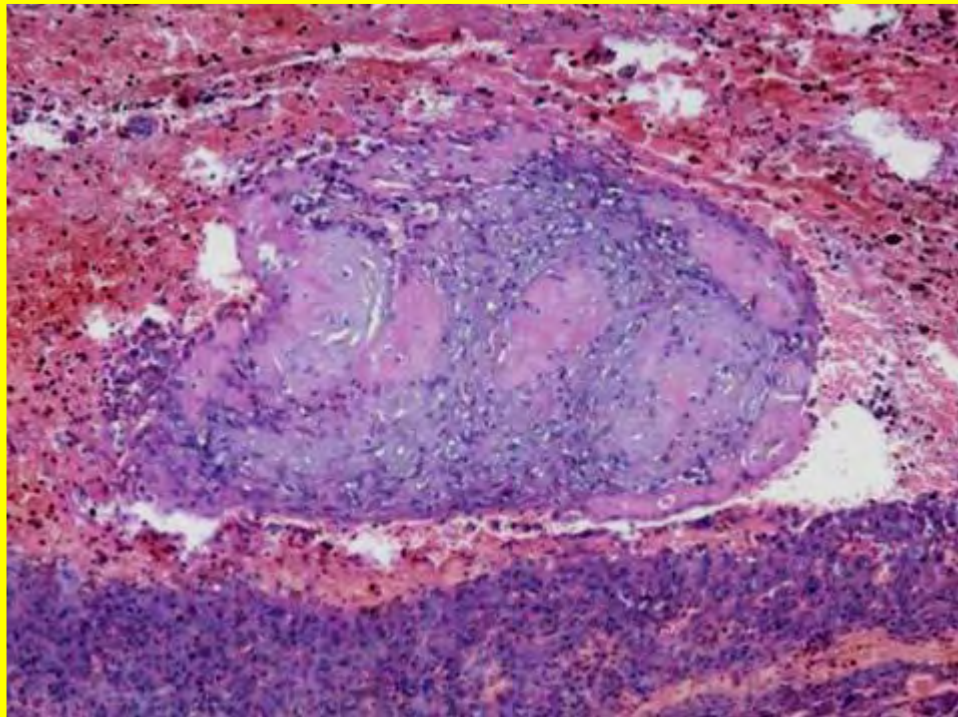


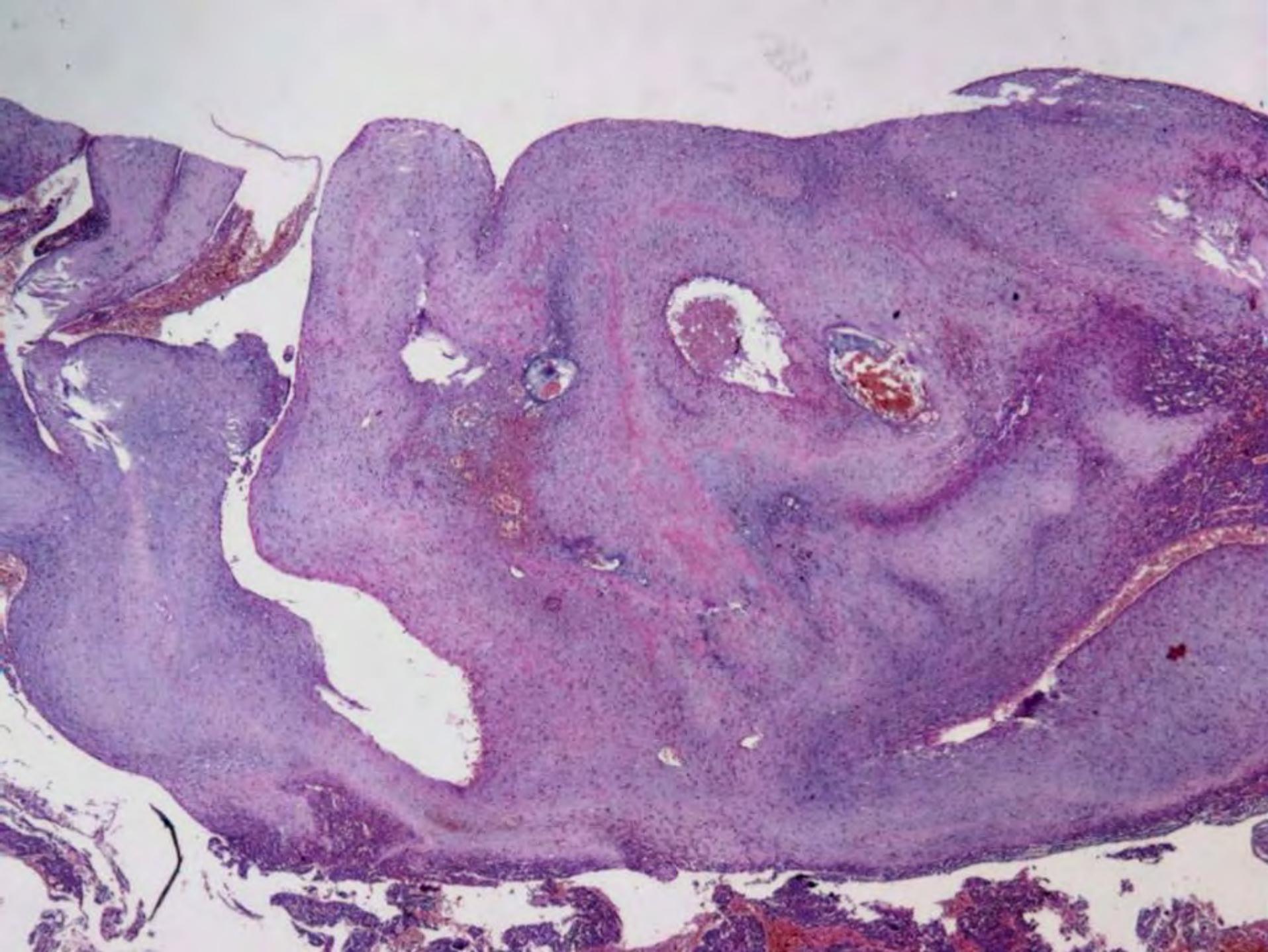


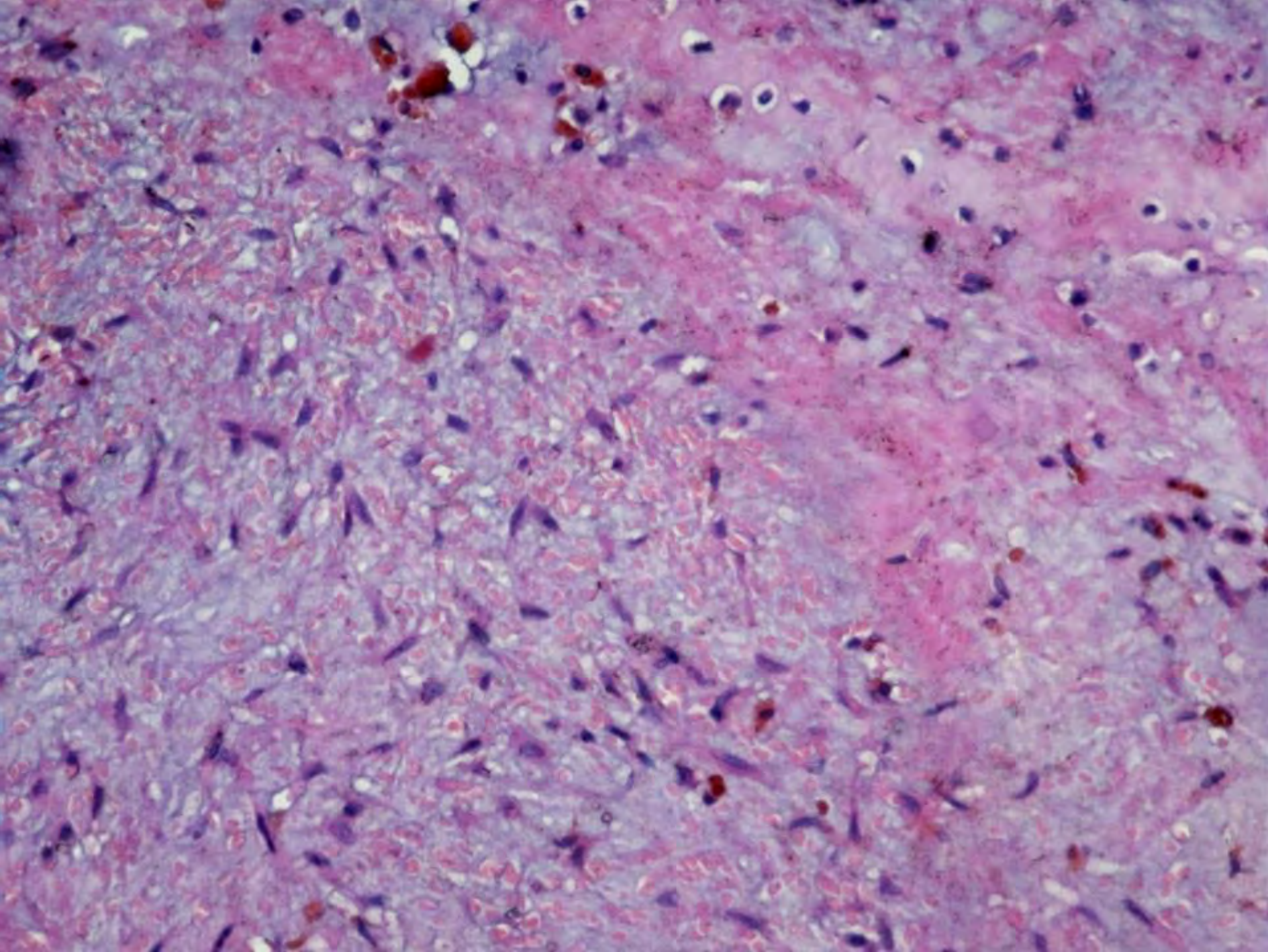


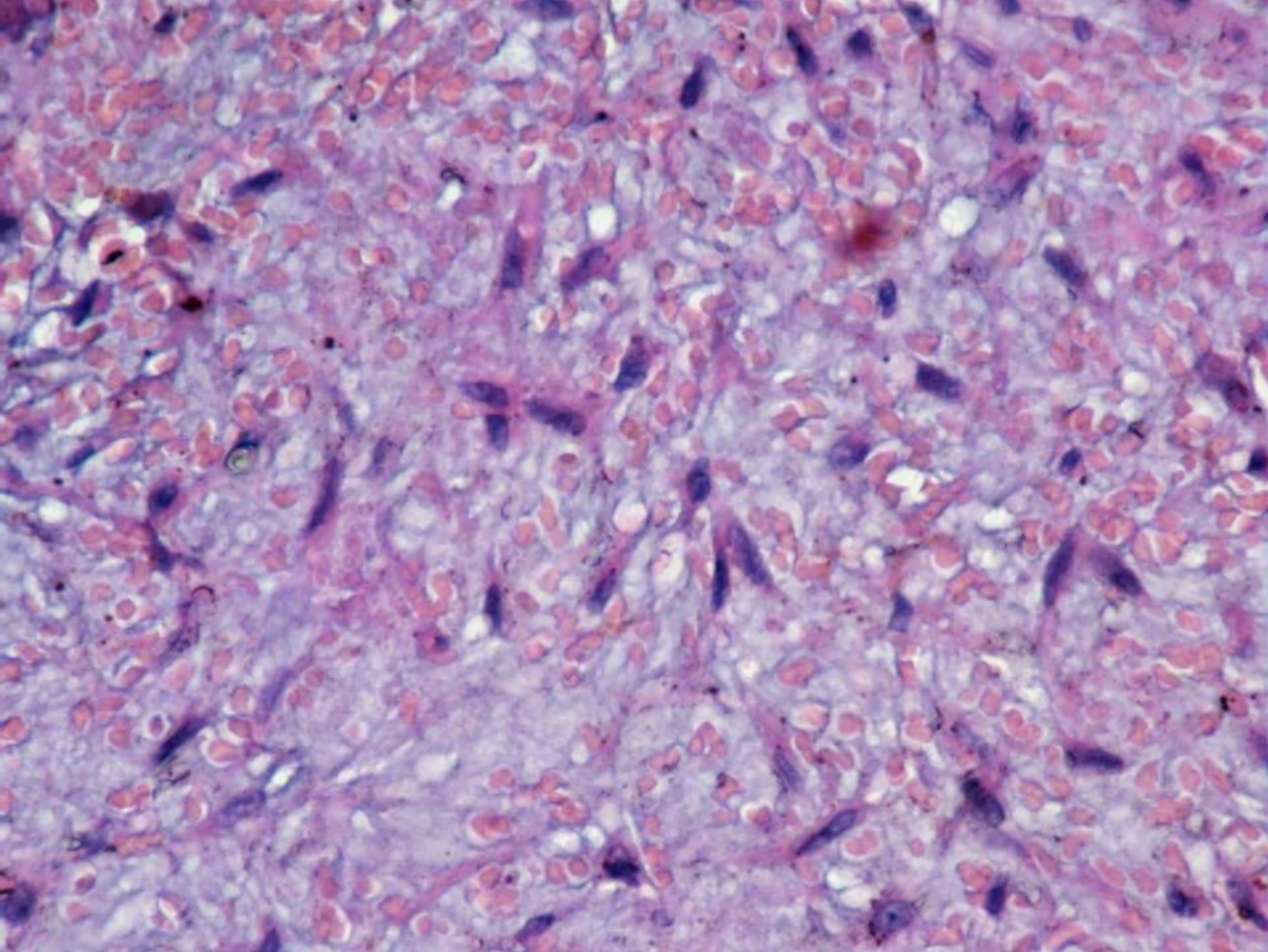




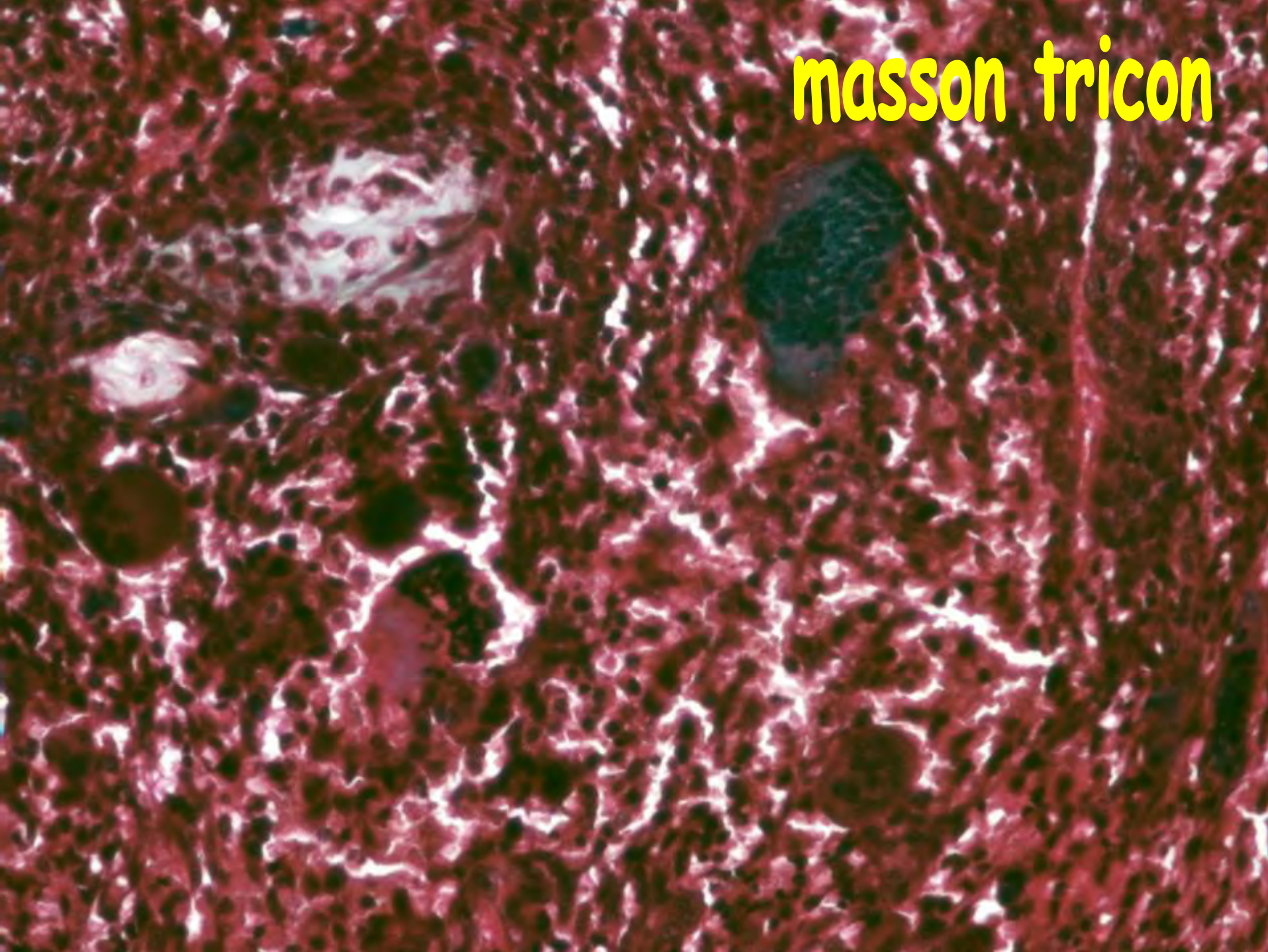





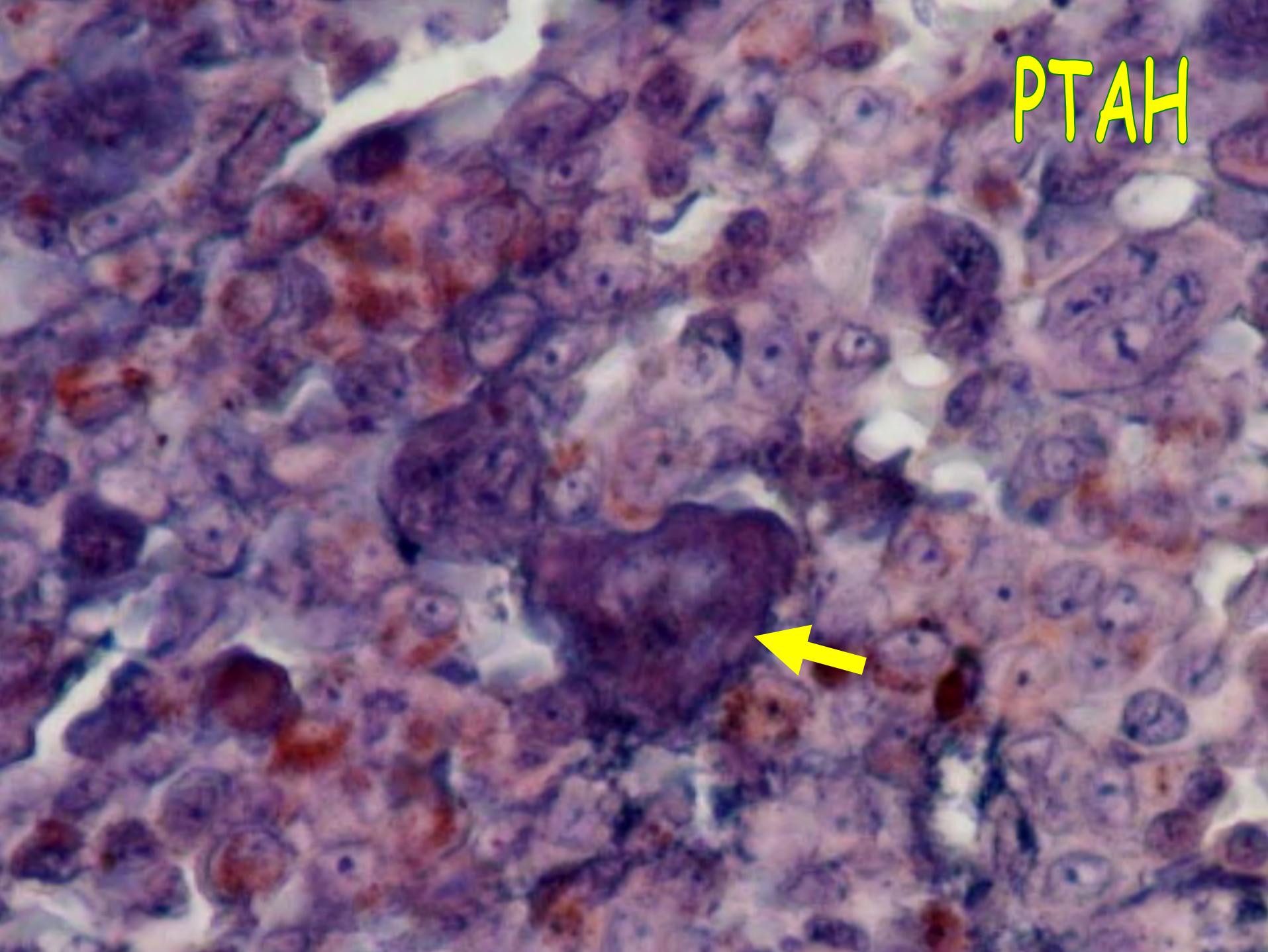






- Infiltrative margin
- Small blue polygonal tumor cells with eosinophilic cytoplasm
- Prominent pleomorphism, hyperchromatism and abnormal mitosis
- Numerous multinucleated giant cells
- Chondroid, myxoid substance and metaplastic bone formation
- Numerous small blood vessles


masson tricon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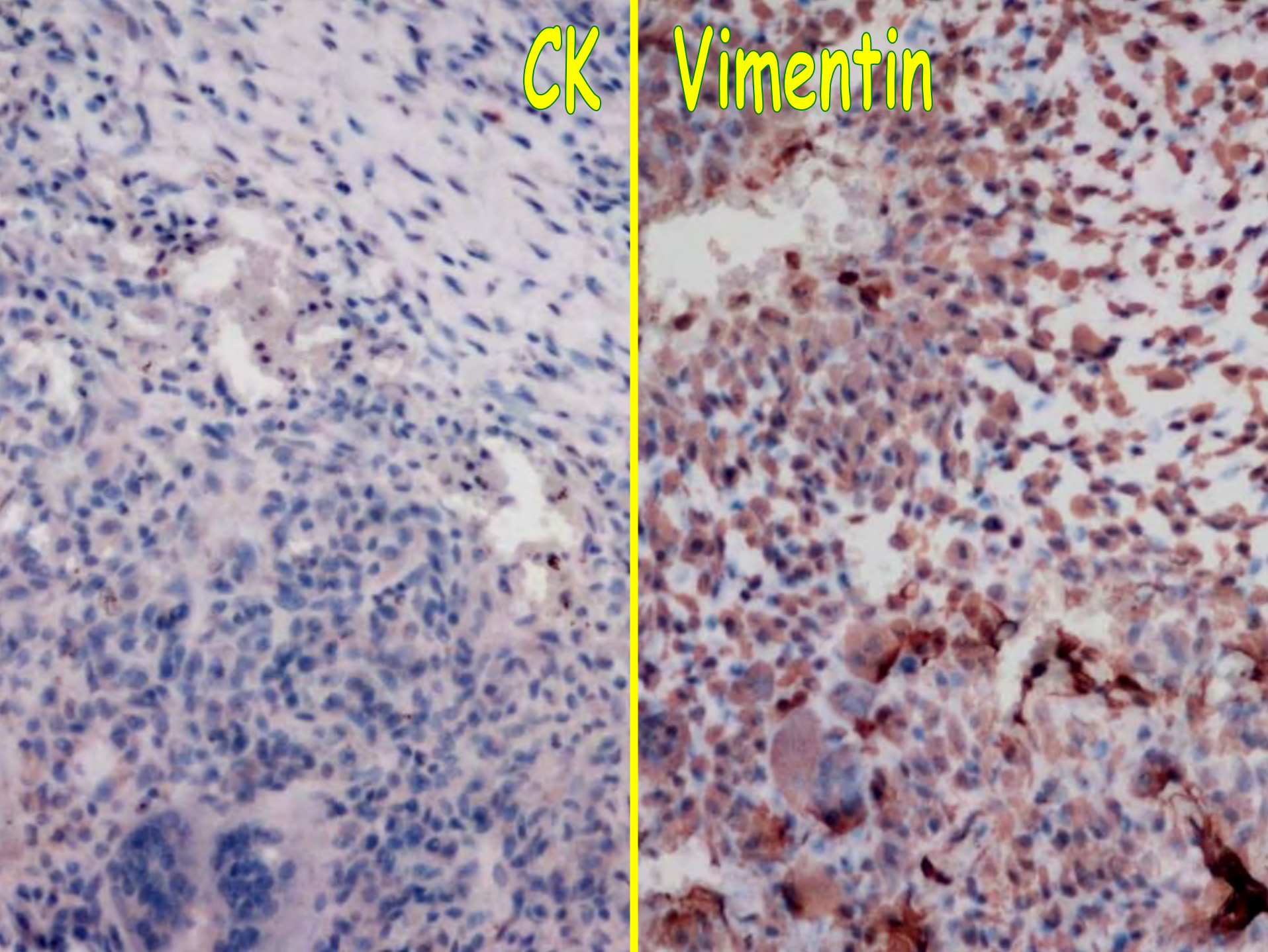
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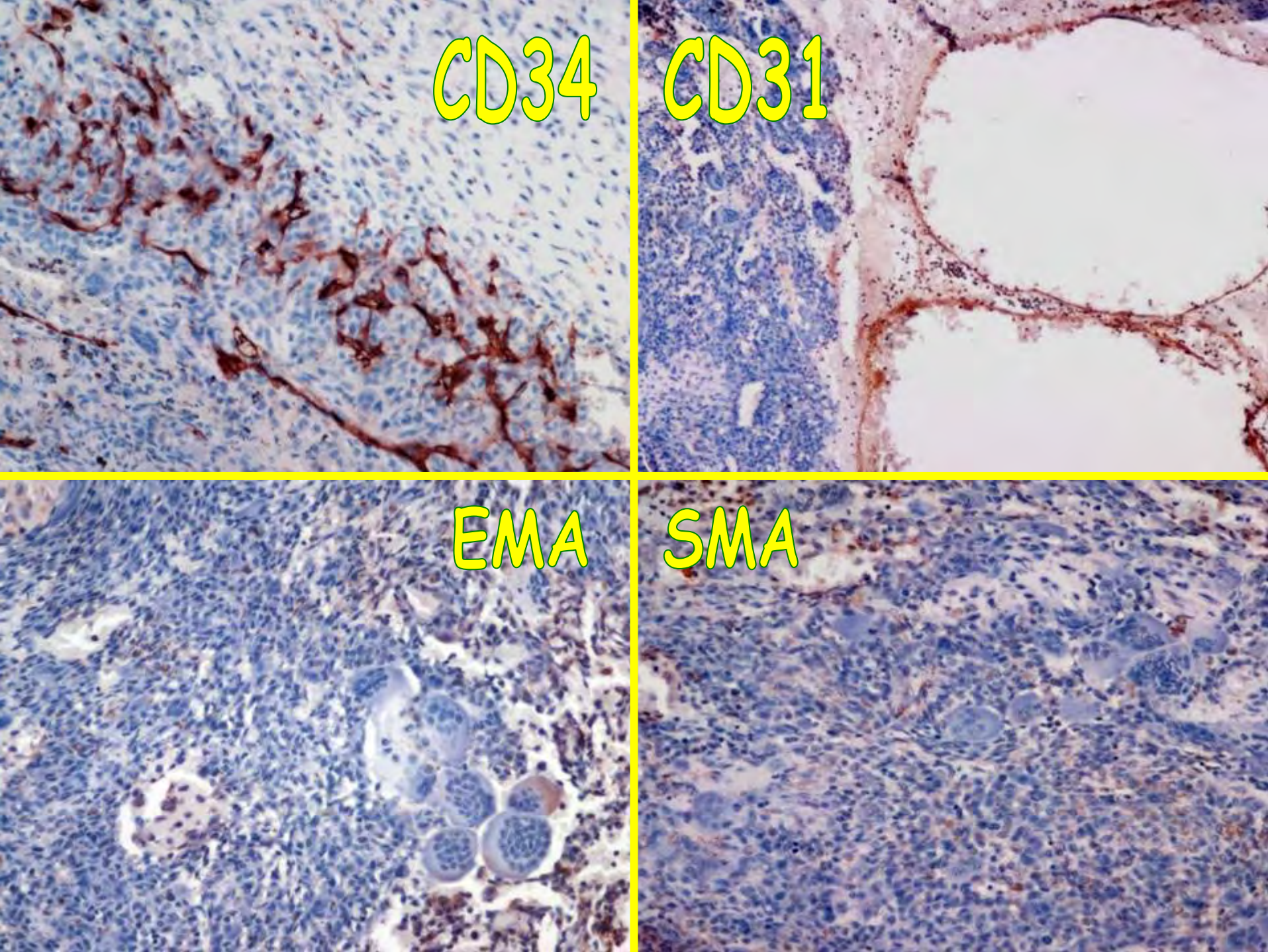
Vimentin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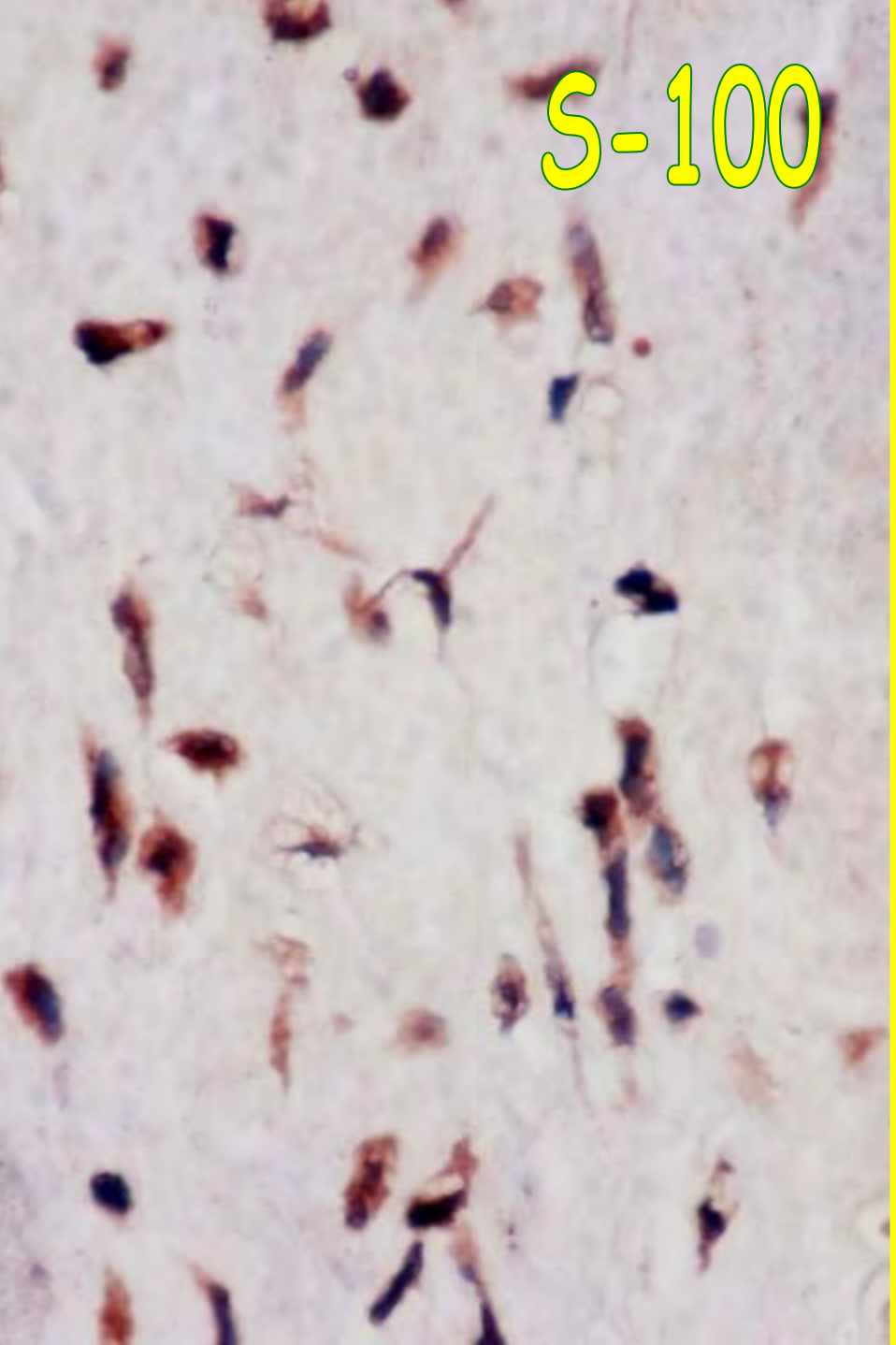
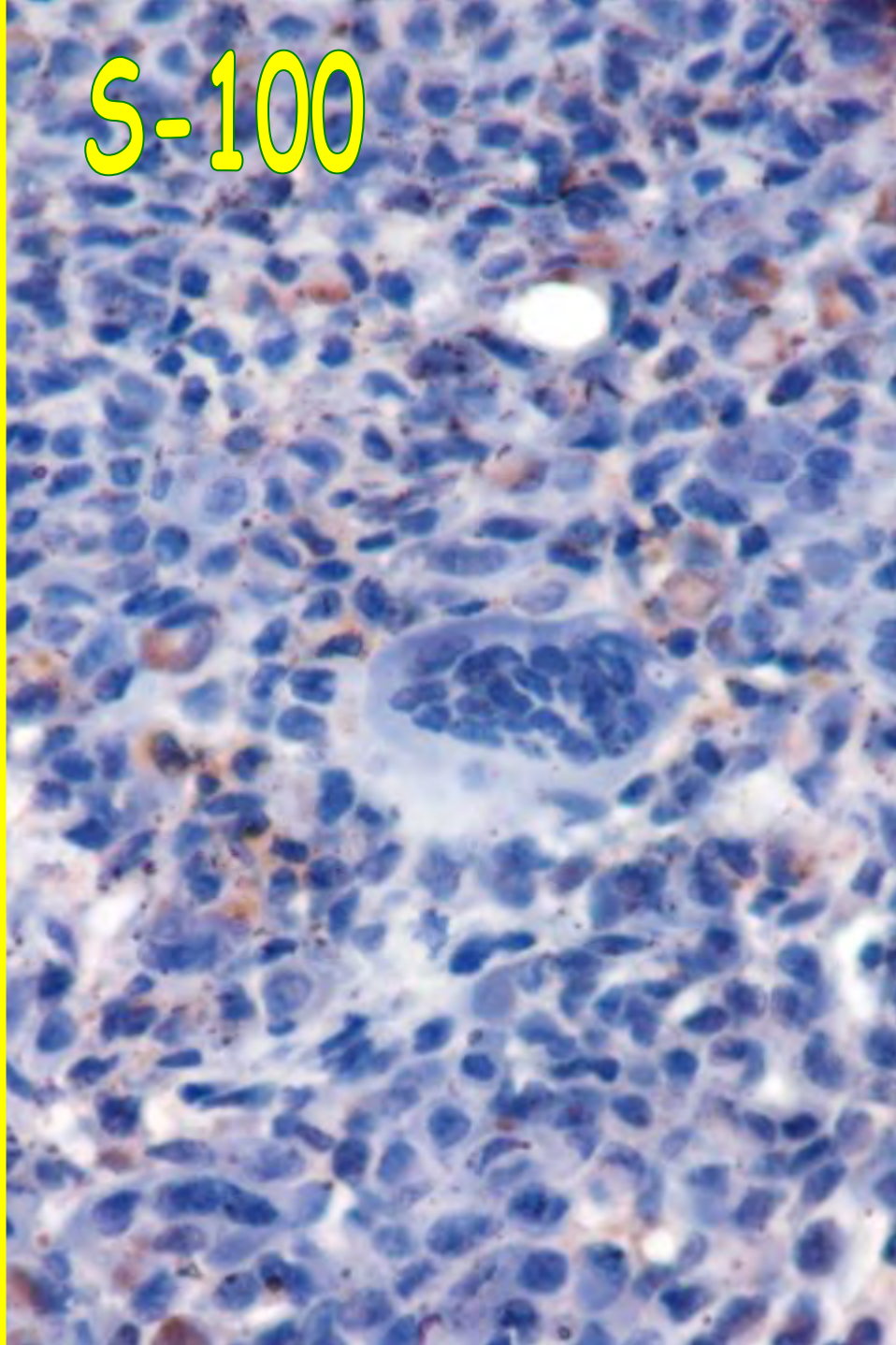
CD31
EMA
SMA


S-100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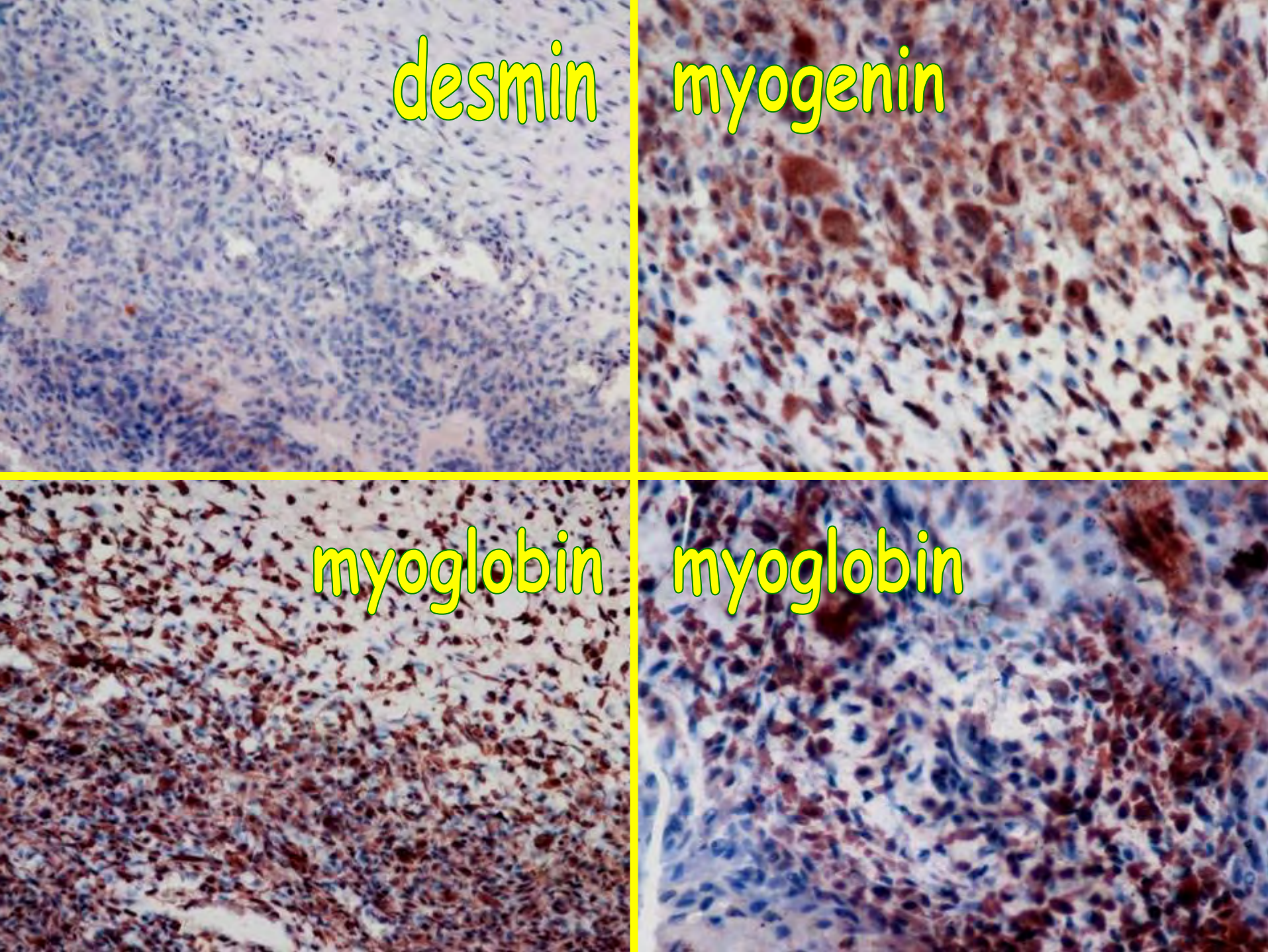
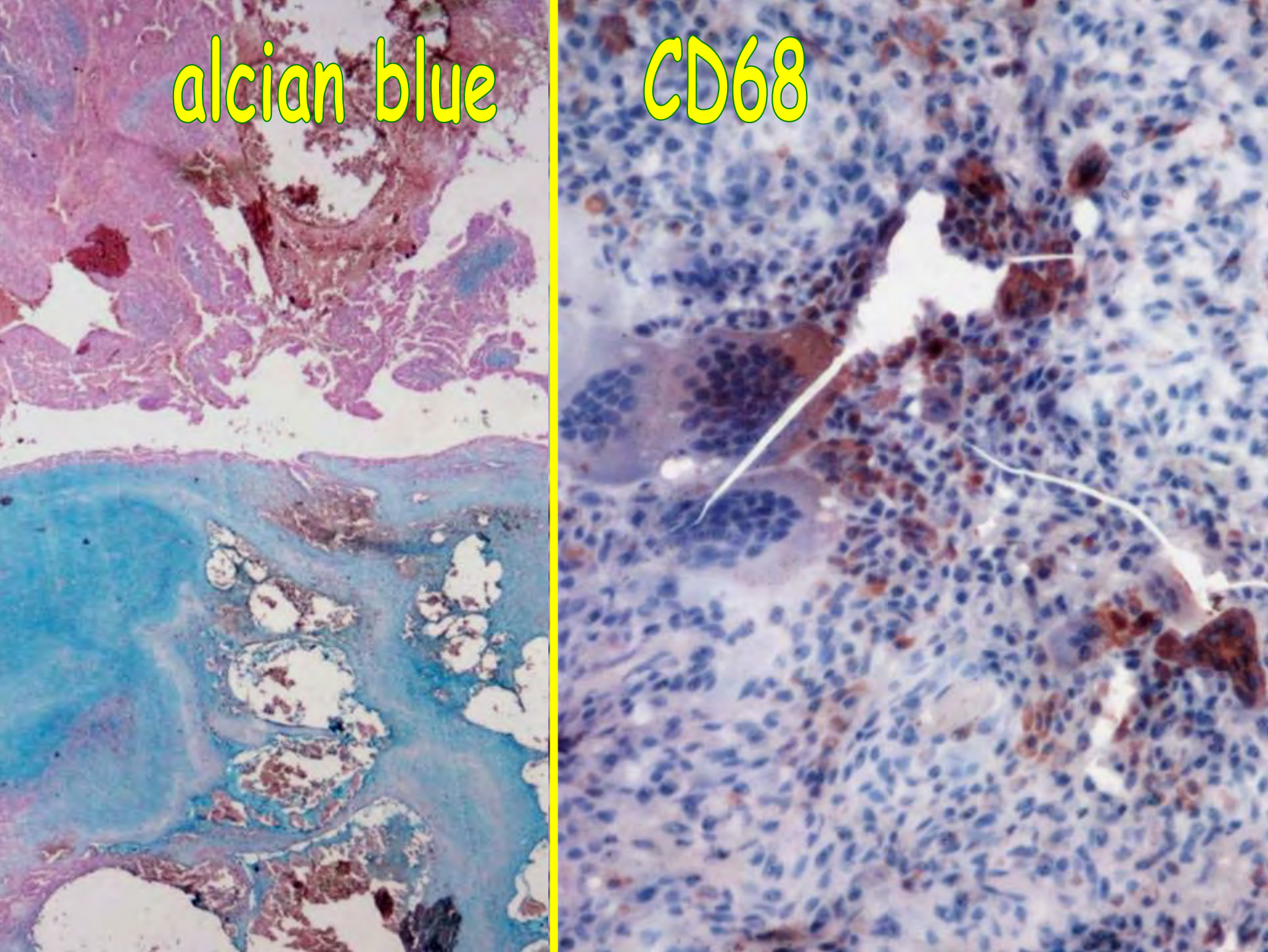
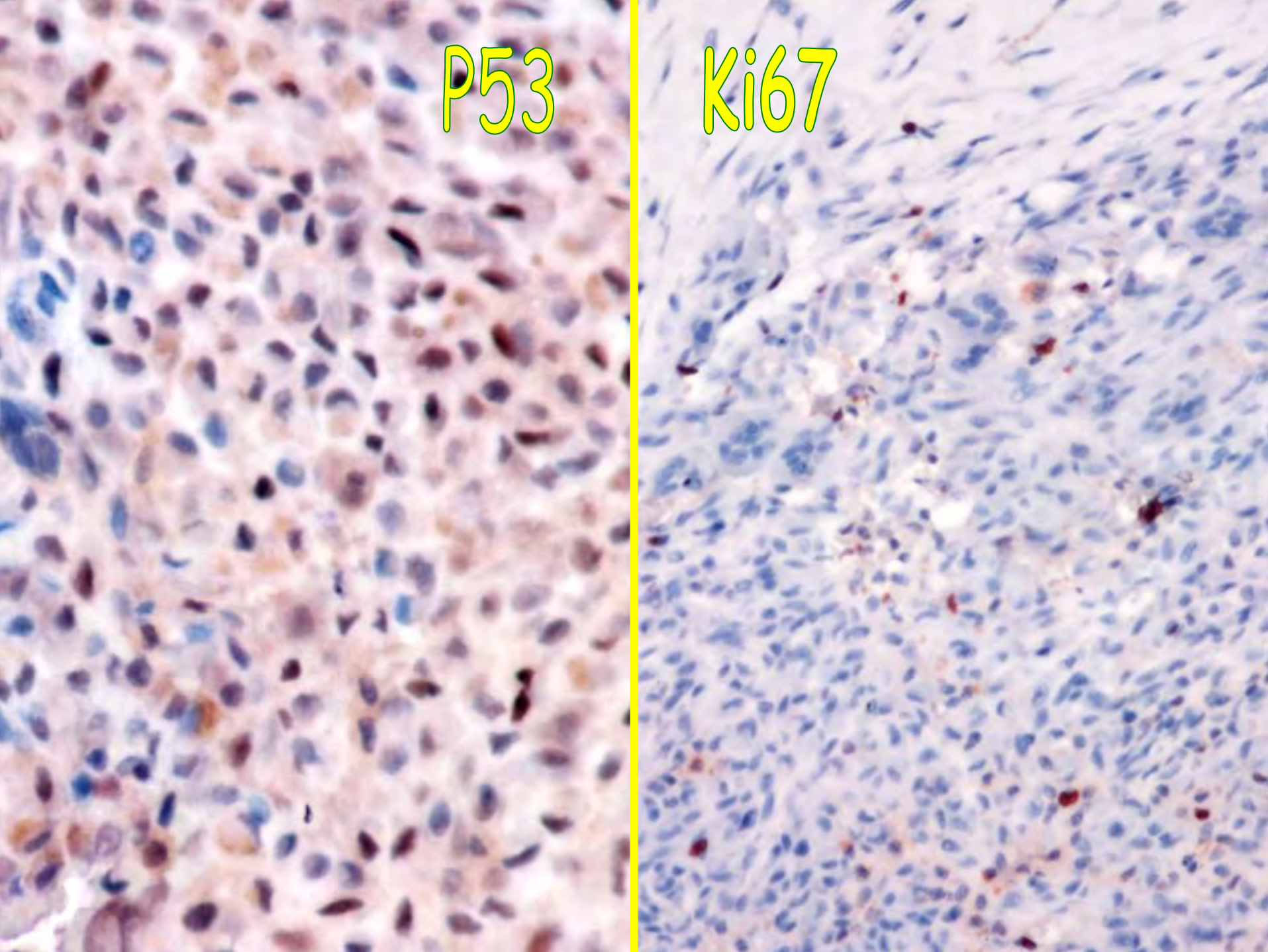

desmin
myogenin
myoglobin
myoglobin


alcian blue
CD68


P53
Ki67

Masson tricon	Brown	S-100	-
PTAH	+	Desmin	-
CK	-	Myogenin	+
Vimentin	+	Myoglobin	+
CD31	-	Alcian blue	+
CD34	-	CD68	-
EMA	-	P53	Focal +
SMA	-	Ki67	Focal +

Classification of rhabdomyosarcoma

- Embryonal (ERMS)
- Alveolar (ARMS)
- Pleomorphic (PRMS)

Morphologic variants of PRMS

- Classic
- Round-cell
- Spindle-cell

Histopathological diagnosis

Pleomorphic rhabdomyosarcoma with telangiatasia

Right pre-auricular area

Treatment Course (cont.)

- Post-OP :
  - No significant complaint
  - No evidence of facial nerve injury
  - Reinforce change dressing
- 940519 - Discharged

Diagnosis of Surgical Specimen

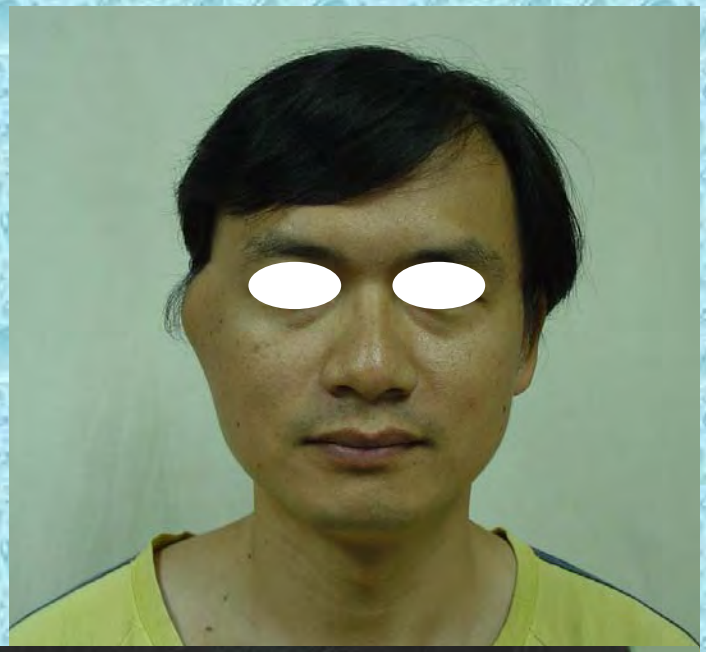
- Pleomorphic Rhabdomyosarcoma with Telangiatasia

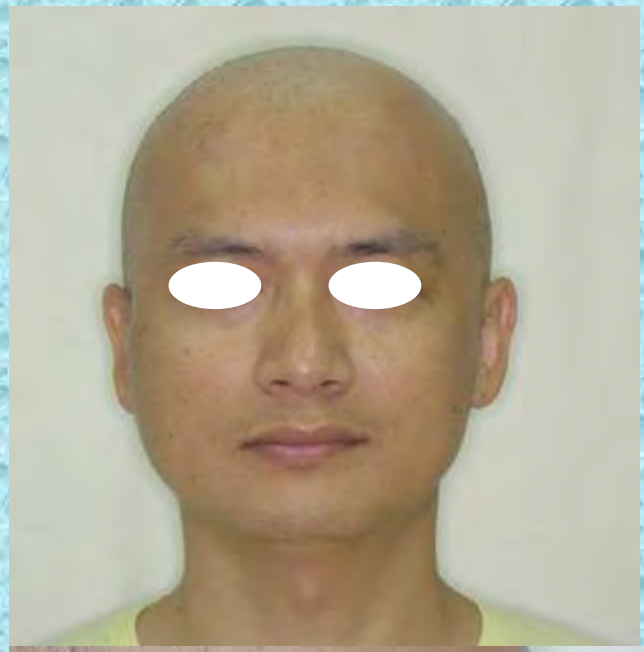
Treatment Course (cont.)

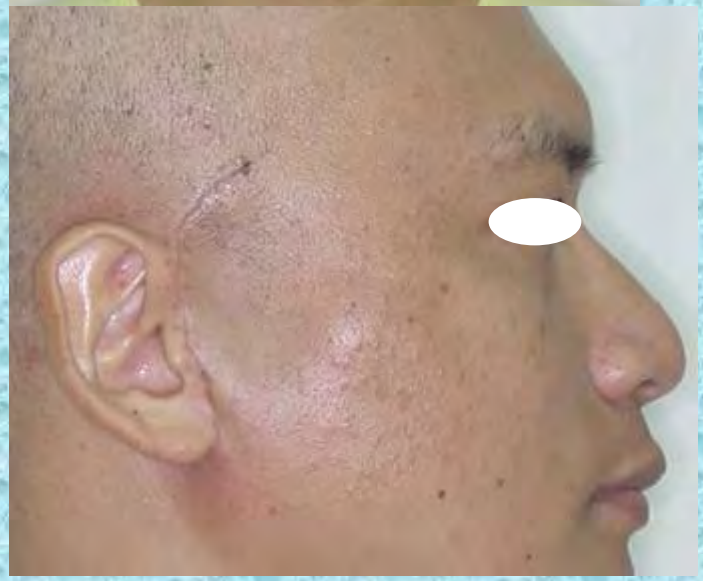
- OMS OPD F/U
- 940525~0605 chemotherapy 血液腫瘤內科
  - Dacarbazine + Epirubicin + Ifosfamind +Mesna

術前

術後一個月







Discussion

Introduction

- A malignant neoplasm of skeletal muscle origin
- 8~19% of all soft tissue sarcoma
- 35~45% occur in head and neck
- Primary in children and young adults
- Most cases arising in patient under 12 y/o and 43% seen in patient less than 5 y/o
- The most common sarcoma in children

Clinical features

- A painless, infiltrative mass that may grow rapidly
- In the head and neck region, the orbit is the most frequent location, followed by the nasal cavity and nasopharynx.
- The palate is the most frequent intraoral site.
- 60% occur in male

	ERMS	ARMS	PRMS
Incidence of RMS	60%	20~30%	<5%
Age	< 12 y/o	15~25 y/o	40~60 y/o
Occur in head and neck	70~90%	18%	7%
Regional nodal meta	10~38%	33% Distant: 75~85%	9%

Image features

- A densly cellular infiltrating neoplasm
- Skeletal muscle is always seen as the site of origin
- On both CT and MRI, masses are typically homogeneous, with destruction of adjacent bone.
- Necrosis can be seen, but intratumoral hemorrhage is uncommon and calcification is typically absent.

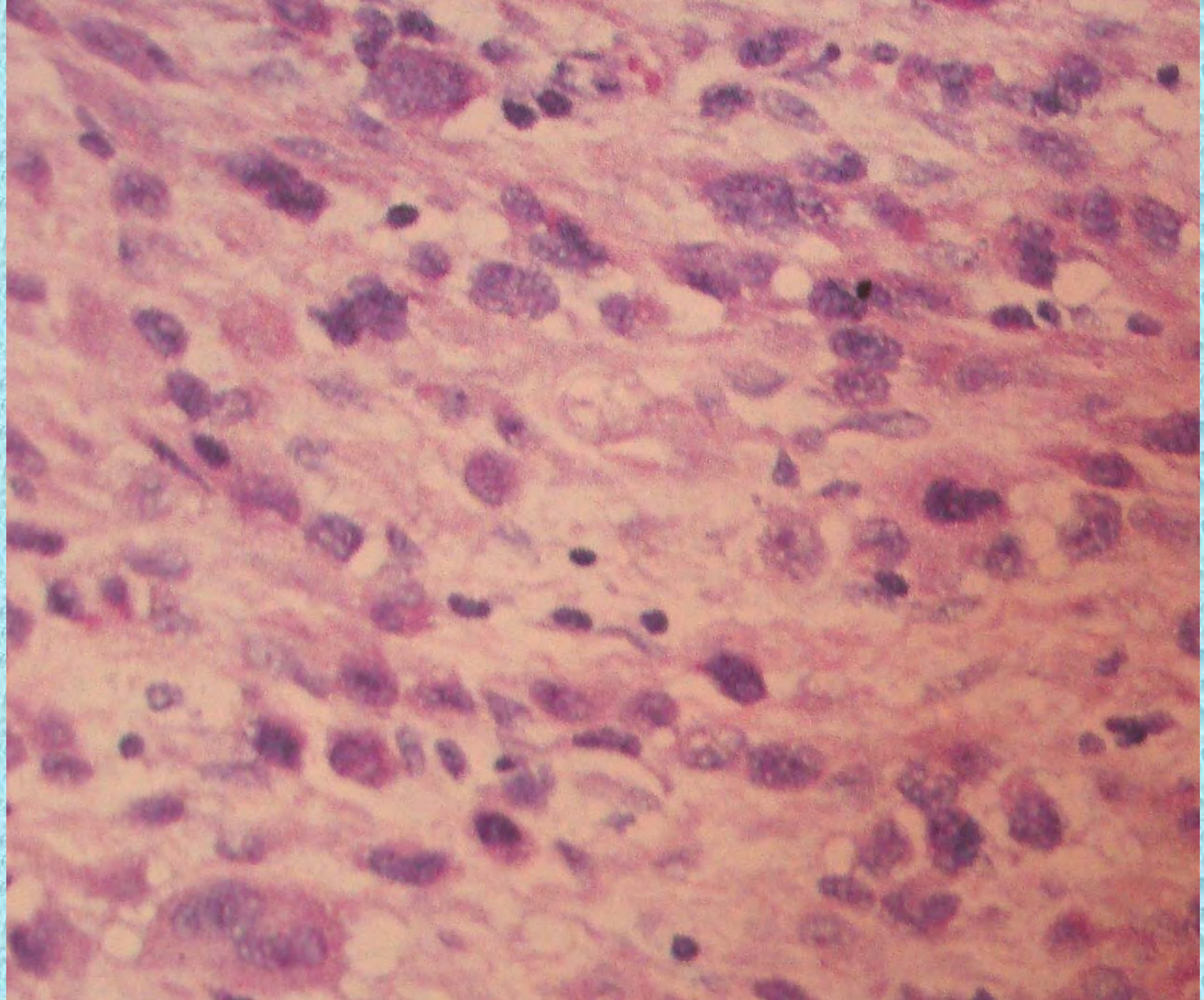
Image features

- Tumor is isointense to muscle on T1-weighted images and hyperintense on T2-weighted images.
- Just over half of the cases enhance homogeneously, and the degree of enhancement is similar to that of normal skeletal muscle.
- In most cases, tumor margin are poorly-defined.

Histopathological features

EMRS

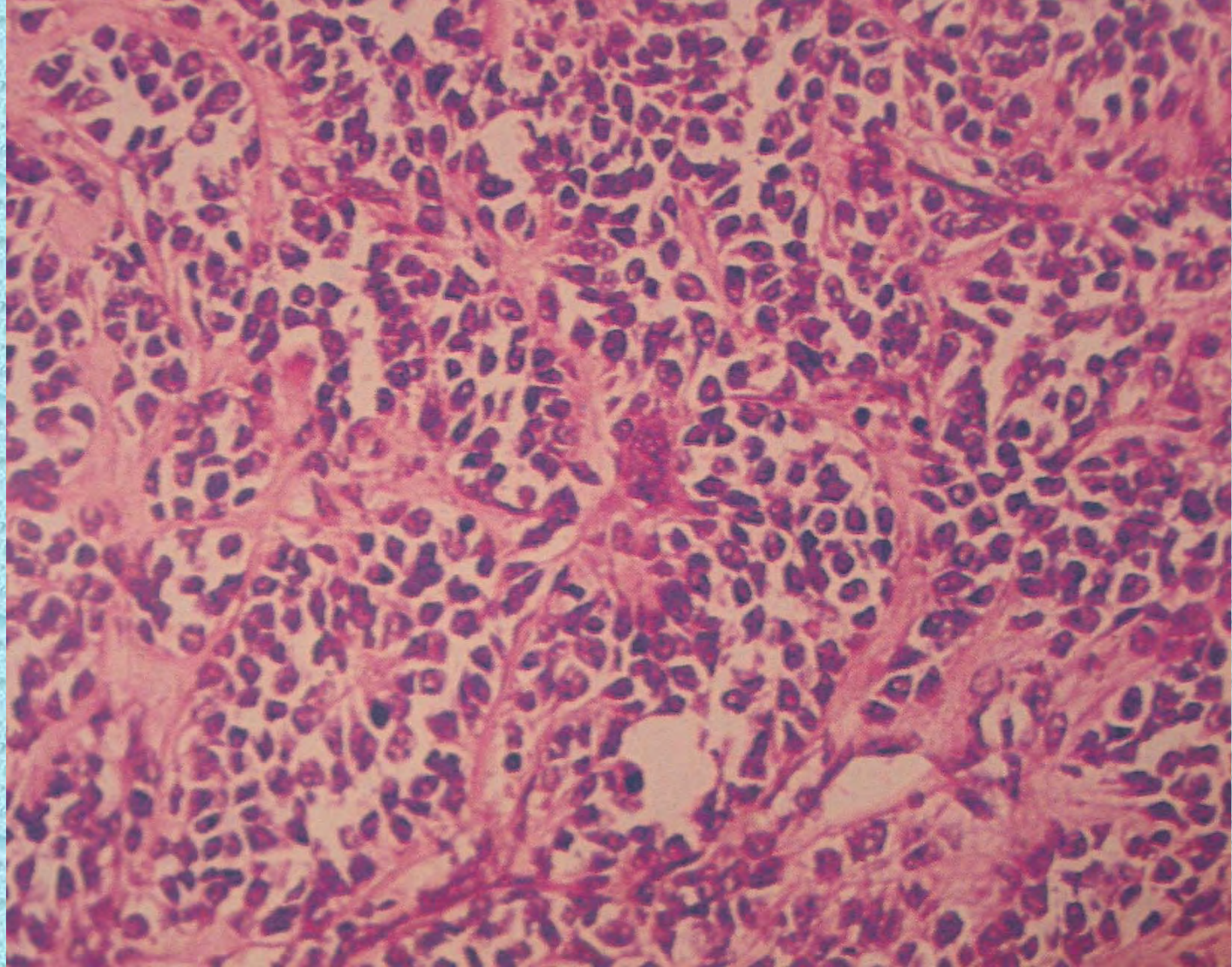
- Small round or oval cells with hyperchromatic nuclei and indistinct cytoplasm.
- Resemble the developing muscle cells of 6~8 w/o fetus.
- Cross striations are rarely found



Histopathological features

AMRS

- Aggregates of poorly differentiated round to oval cells separated by fibrous septa.
- Resemble the developing muscle cells of 10 w/o fetus.
- Mitoses are common, and multinucleated giant cells are also may be seen.



Histopathological features

PMRS

- Classic: sheets of large, atypical and often multinucleated polygonal pleomorphic rhabdomyoblasts

Histopathological features

PMRS

- **Round cell:** clusters of large, atypical PRMB throughout the lesion with a back-ground of slightly pleomorphic medium-sized, round, blue rhabdomyoblasts

Histopathological features

PMRS

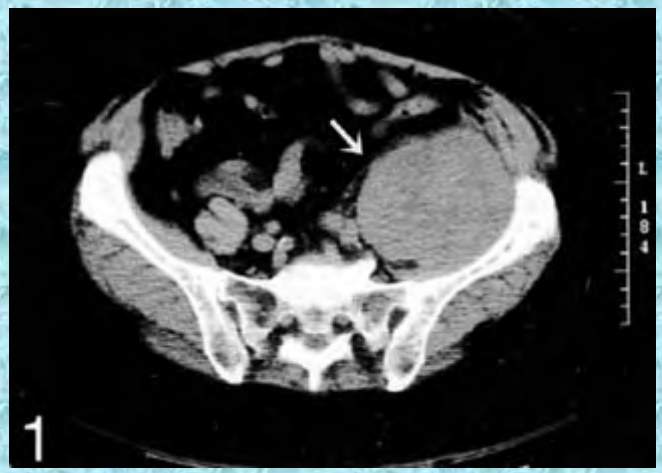
- **spindle cell:** a predominance of pleomorphic spindled RMB arranged in a storiform growth pattern with scattered large PRMB

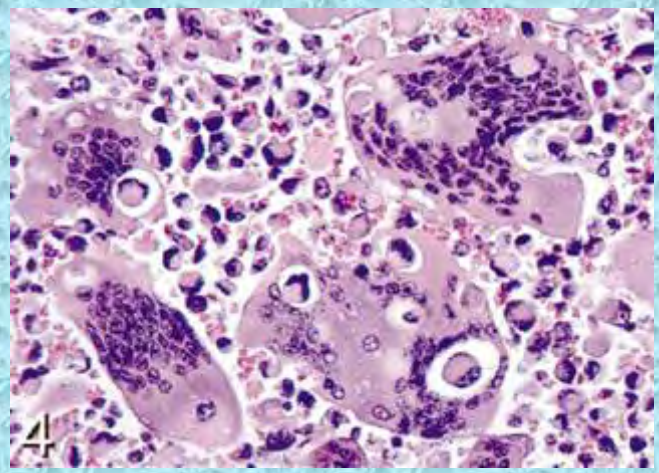
Immunoprofiles of PRMS

- At least one skeletal muscle-specific marker positive (myoglobin, nuclear MyoD1, nuclear myf4, and fast myosin)
- Myoglobin (95%) and fast skeletal muscle myosin (80%) appeared to be the most sensitive markers for PRMS.

Related article

- Pleomorphic rhabdomyosarcoma, classic variant, with reactive osteoclast-like giant cells (*Arch Pathol Lab Med-Vol 129, May 2005;703-5*, Gladell P. Paner, MD; Loyola University Medical center)





Histopathological features of the present case

- Infiltrative margin
- Small blue polygonal tumor cells with eosinophilic cytoplasm
- Prominent pleomorphism, hyperchromatism and abnormal mitosis
- Numerous multinucleated giant cells
- Chondroid, myxoid substance and metaplastic bone formation
- Numerous small blood vessles

Discussion

- general feature
  - (1) the most common soft tissue sarcoma of children (> 50 % of pediatric sarcoma)
  - (2). > 250 new cases in USA
  - (3) 25 % of pediatric rhabdomyosarcoma 發生在head and neck
- 以 soft palate 最多，其次爲 alveolar ridge (maxilla > mandible)
- 亦可出現在無肌肉處，如 gingiva.
- 常在初診斷即已 meta.

Discussion (cont.)

- age: 10 歲以下最多
- painless muscle
- if masticatory muscle s are involved → trismus is a common finding
- infratemporal fossa and orbital are common sites

Discussion (cont.)

- prognosis:
  - (1) 1970, 5-year overall survival rate < 10 %
  - (2) 1991, 5-year overall survival rate =70 %
  - (3) 早期診斷加上combination therapy → 80 %
  - (4) 50 %

Discussion (cont.)

- 治療
  - 先看是否有轉移
  - 看 resectability of the tumor
    - Group I: localized disease, completely resected → surgery only
    - Group II: compromised or regional resection (microscopic residual tumor) → surgery + 45 Gy of radiation o the tumor base
    - Group III incomplete resection → surgery + 50-55 Gy of radiation o the tumor base
    - Group IV : distant metastasis
      - multiagent chemotherapy

Discussion (cont.)

- Chemotherapy
  - multiagent chemotherapy: decrease toxicity and improved survival
  - Ex :
    - Dacarbazine + Epirubicin + Ifosfamind +Mesna

Discussion (cont.)

- adult
  - rare case
  - alveolar type 爲主
  - similar protocol of children
  - surgery + RT + chemotherapy
  - 5 year survival rate : 20 %

Thanks for your attention!