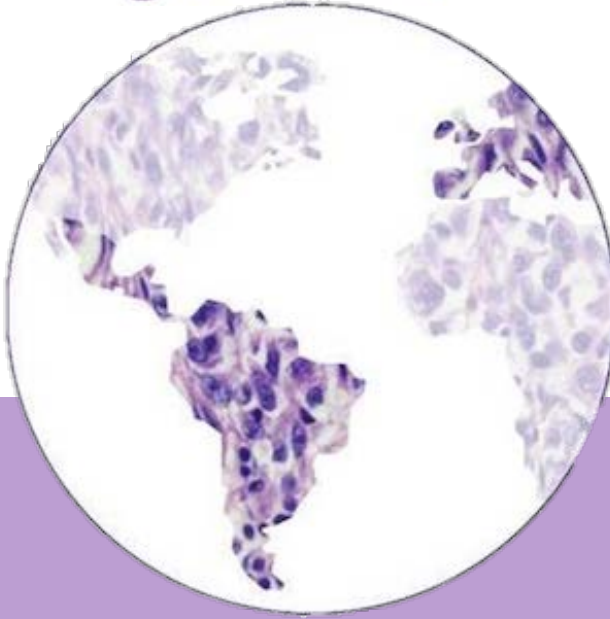


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Virtual MDT Board

25 NOVEMBER 2021

SELNET



CASE . COSTA RICA

DRA. NATALIA JIMÉNEZ
ONCOLOGÍA MÉDICA
HOSPITAL SAN VICENTE DE PÁUL
HEREDIA
COSTA RICA

DR. RODOLFO GUZMÁN
PATOLOGÍA
HOSPITAL SAN JUAN DE DIOS
SAN JOSÉ
COSTA RICA



CASE . MQV

- FEMALE, 23 YEARS OLD
- WITHOUT PREVIOUS DISEASES

FAMILY HISTORY OF CANCER: **BROTHER WITH EWING'S SARCOMA**
AT AGE 14 YEARS OLD. PRIMARY IN FEMUR.
NEGATIVE FOR OTHER FAMILY MEMBERS

- **CLINICAL PRESENTATION:** SINCE JULY 2021 WITH EPISODES OF
HEMATURIA AND PAIN IN THE RIGHT FLANK OF THE ABDOMEN



CASE . MQV

- CT IN **JULY 2021**: RIGHT KIDNEY WITH LOWER POLE MASS WITH INFILTRATIVE ASPECT AND AREAS WITH LIQUID DENSITY INSIDE IT, PROBABLY RELATED TO SPOTS OF NECROSIS. DIMENSIONS: **45 X 54 X 76 MM**, INFILTRATING THE LOWER CALICIAL GROUP. PELVIS AND SUPERIOR CALICIAL GROUPS PRESERVED.
FINDINGS COMPATIBLE WITH A T3AN0MX KIDNEY TUMOR
- ON **9/27/21** SHE IS TAKEN BY **UROLOGY** TO SURGERY AND THEY NOTE: RIGHT KIDNEY WITH MASS AT LOWER POLE LEVEL WHICH PRESENTS GIANT TENSIVE CYST WITH NECROTIC CONTENT. RENAL ARTERY AND VEIN SEEM COMPROMISED BY THE TUMOR.
- IN AN ATTEMPT TO DISSECTATE, THERE IS **RUPTURE OF THE CYST** THE KIDNEY WAS THEN COMPLETELY SEPARATED AND EXTRACTED

IMAGES FROM JULY 2021 (PRE-SURGICAL)

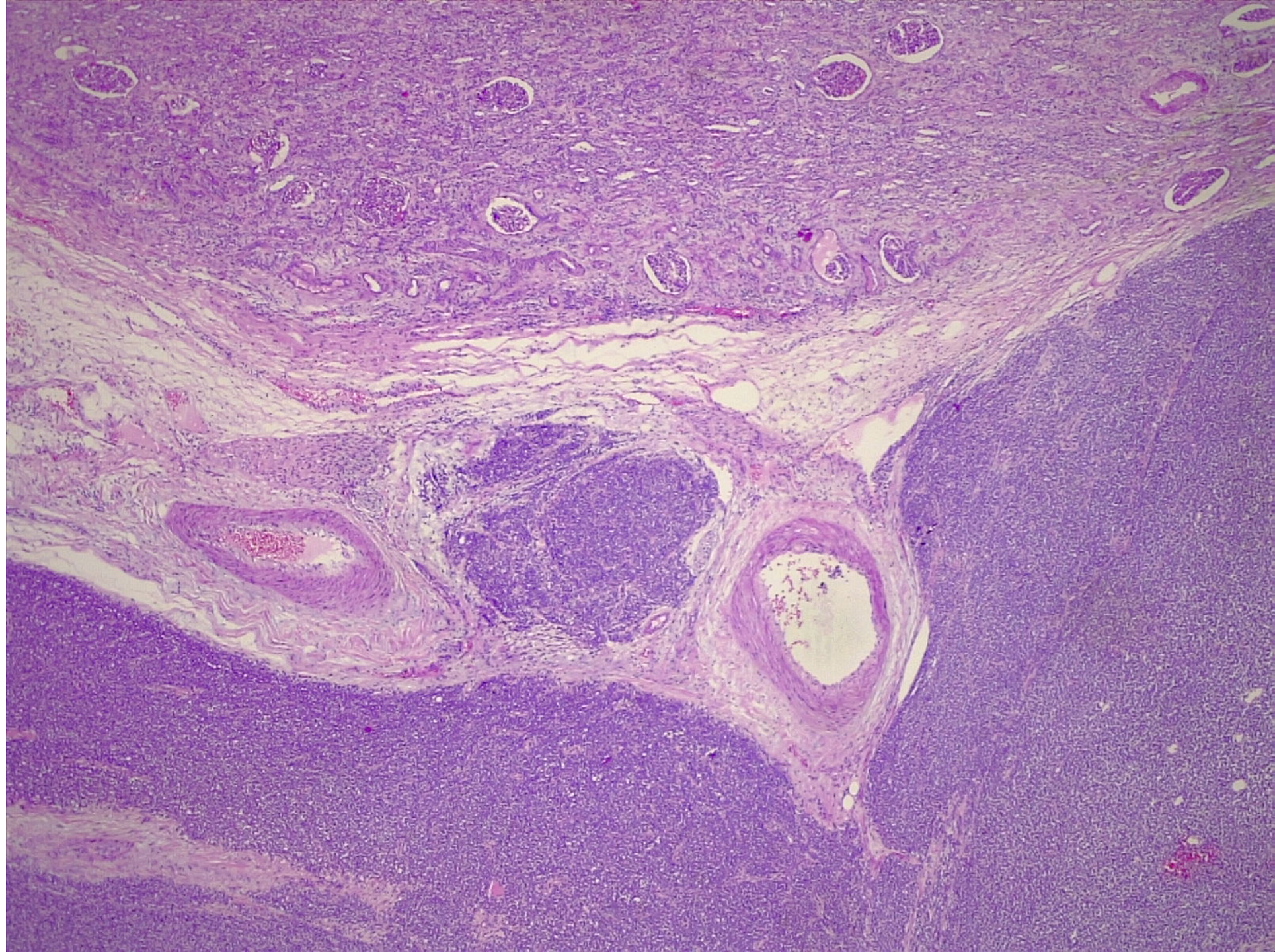


OCTOBER 2021 VIDEO (POST SURGICAL)

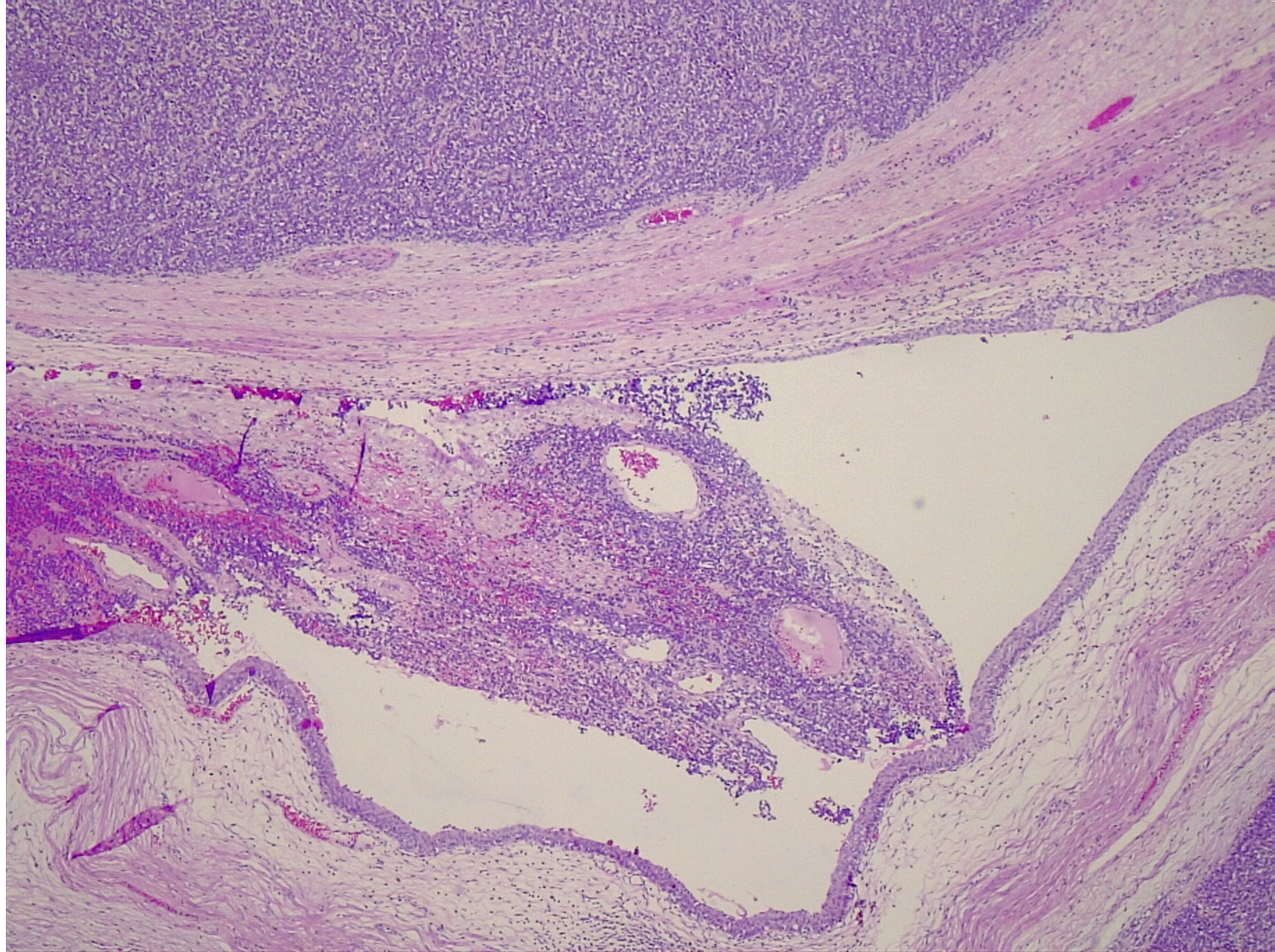


1. Collection in the right kidney bed.
2. Right adrenal collection.

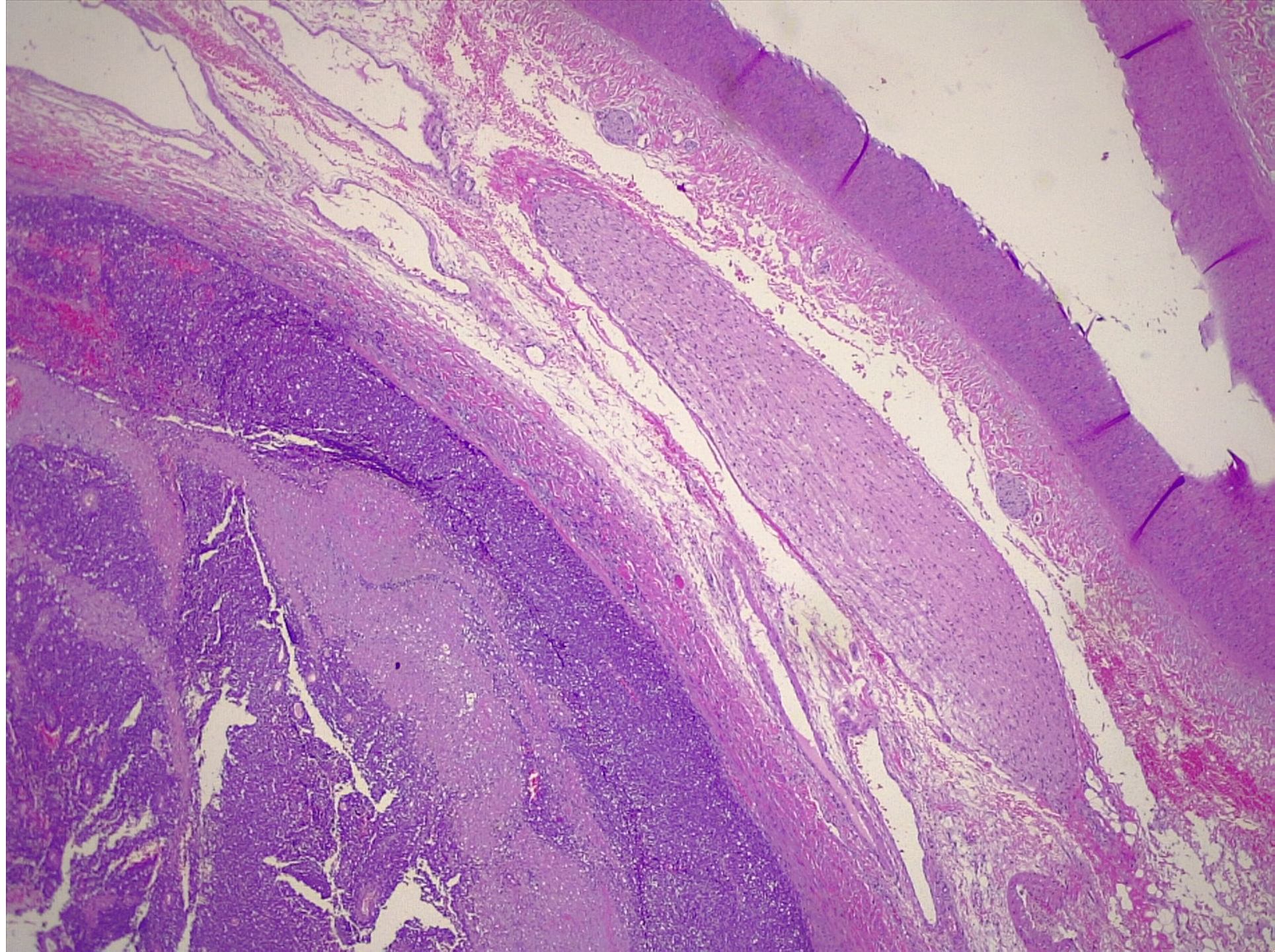
Kidney
infiltrated by
solid small
round cell
neoplasia
(Hematoxylin-
Eosin).



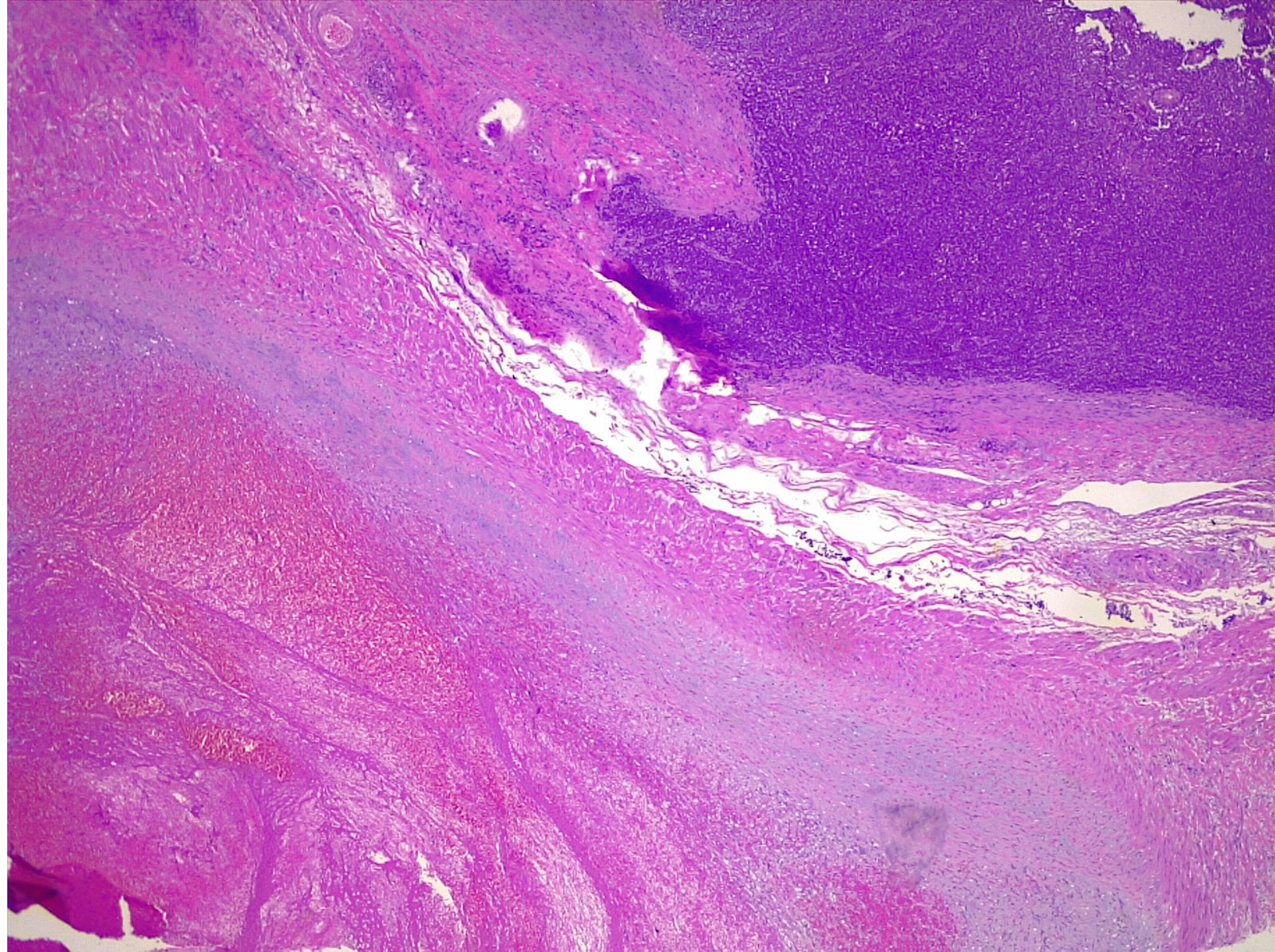
Minor calyx
infiltrated by
round cell
neoplasia
(Hematoxylin-
Eosin).



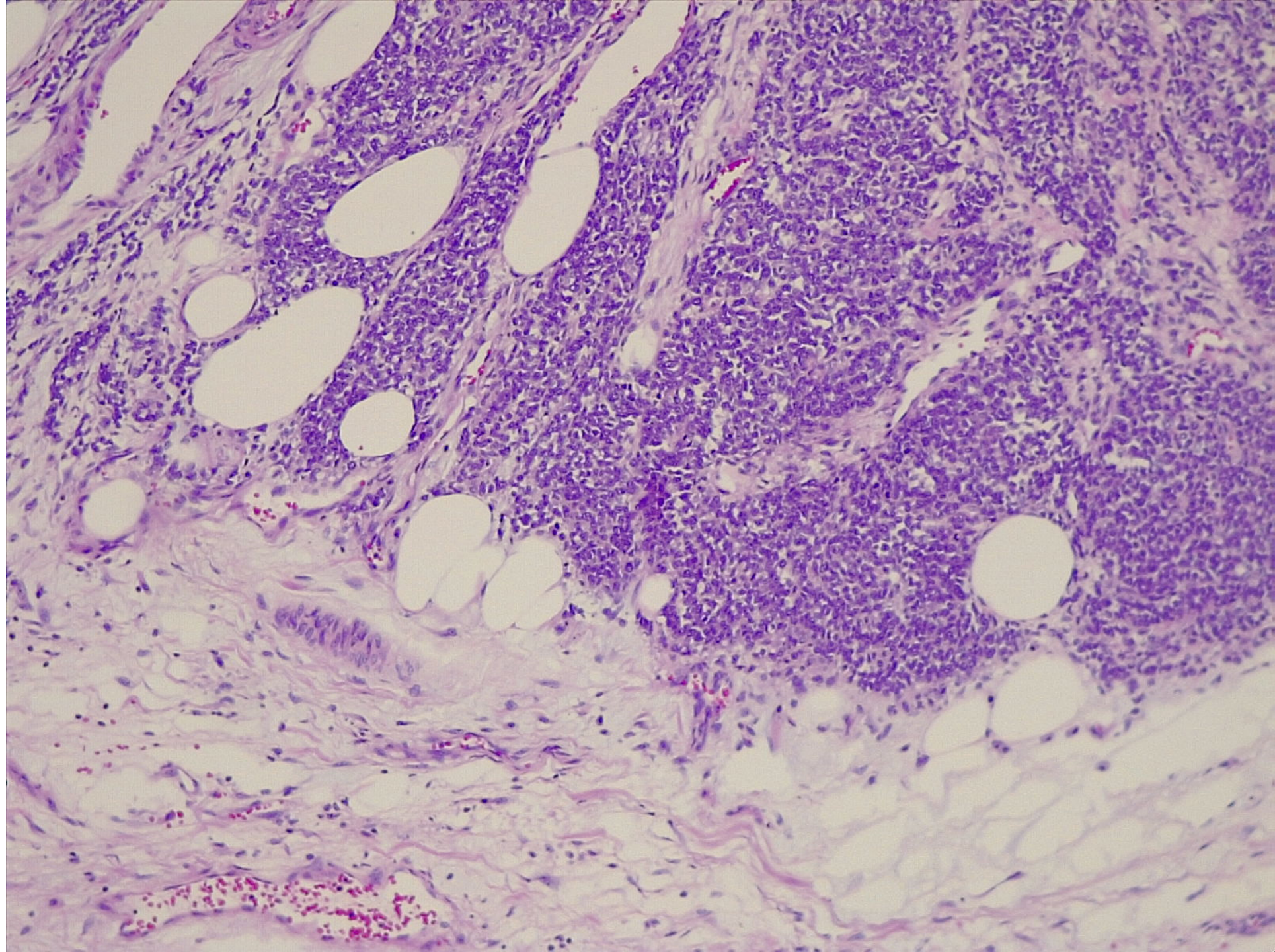
Hilar Vein
with round cell
neoplasia
(Hematoxylin-
Eosin).



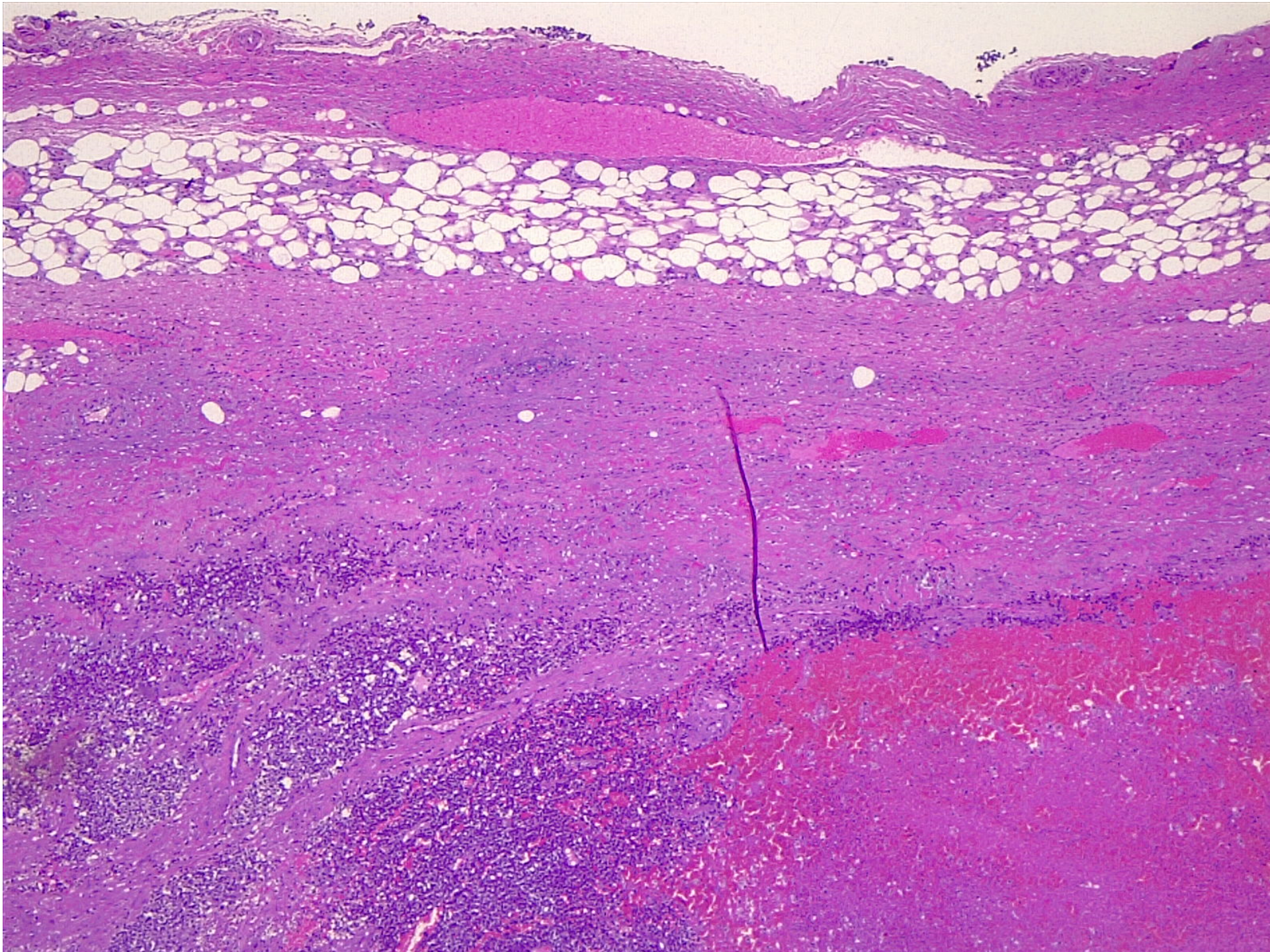
Hilar Vein
with
thrombosis
(Hematoxylin-
Eosin).



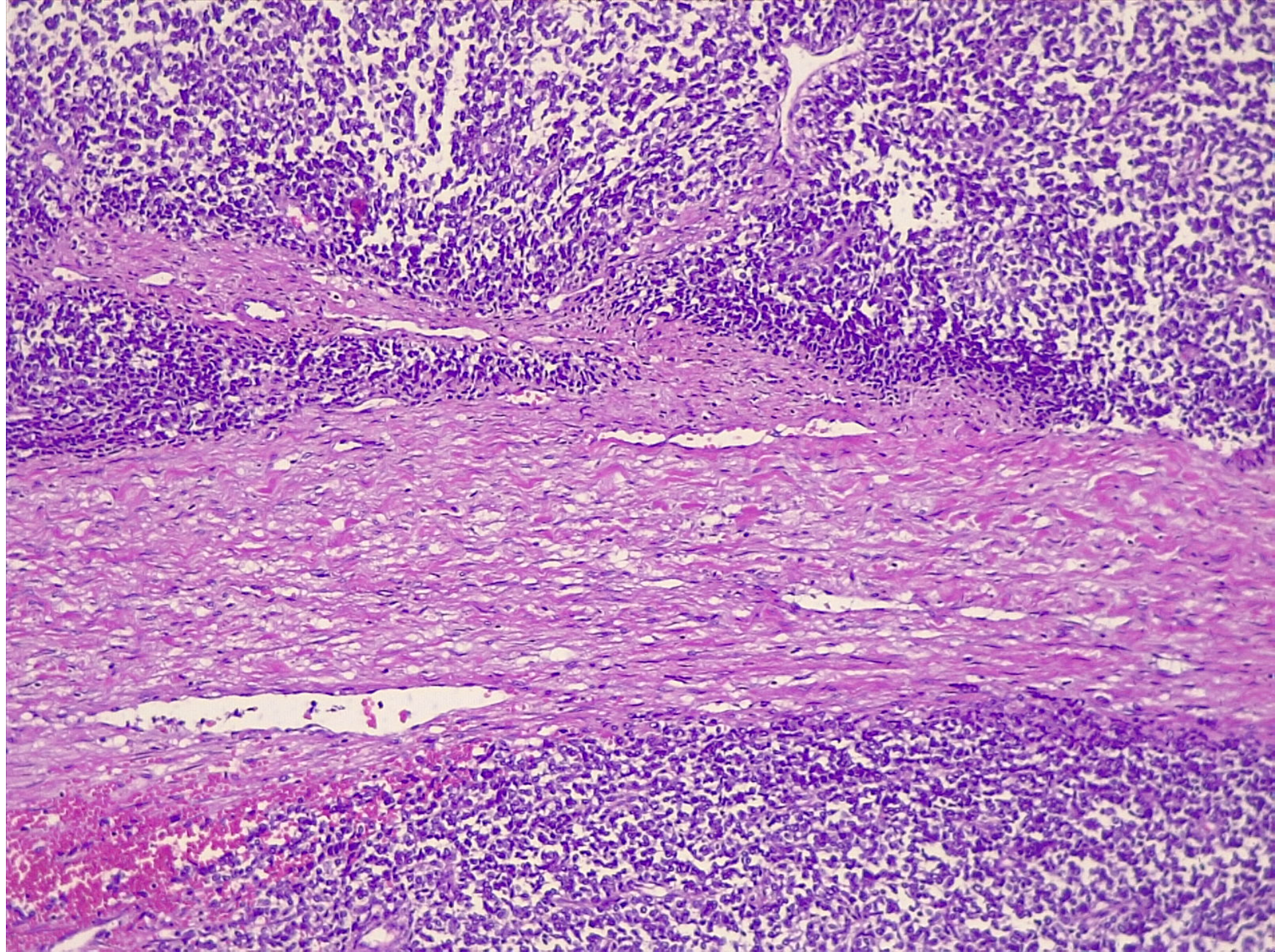
Hilar fatty
tissue
infiltrated by
round cell
neoplasia
(Hematoxylin-
Eosin).



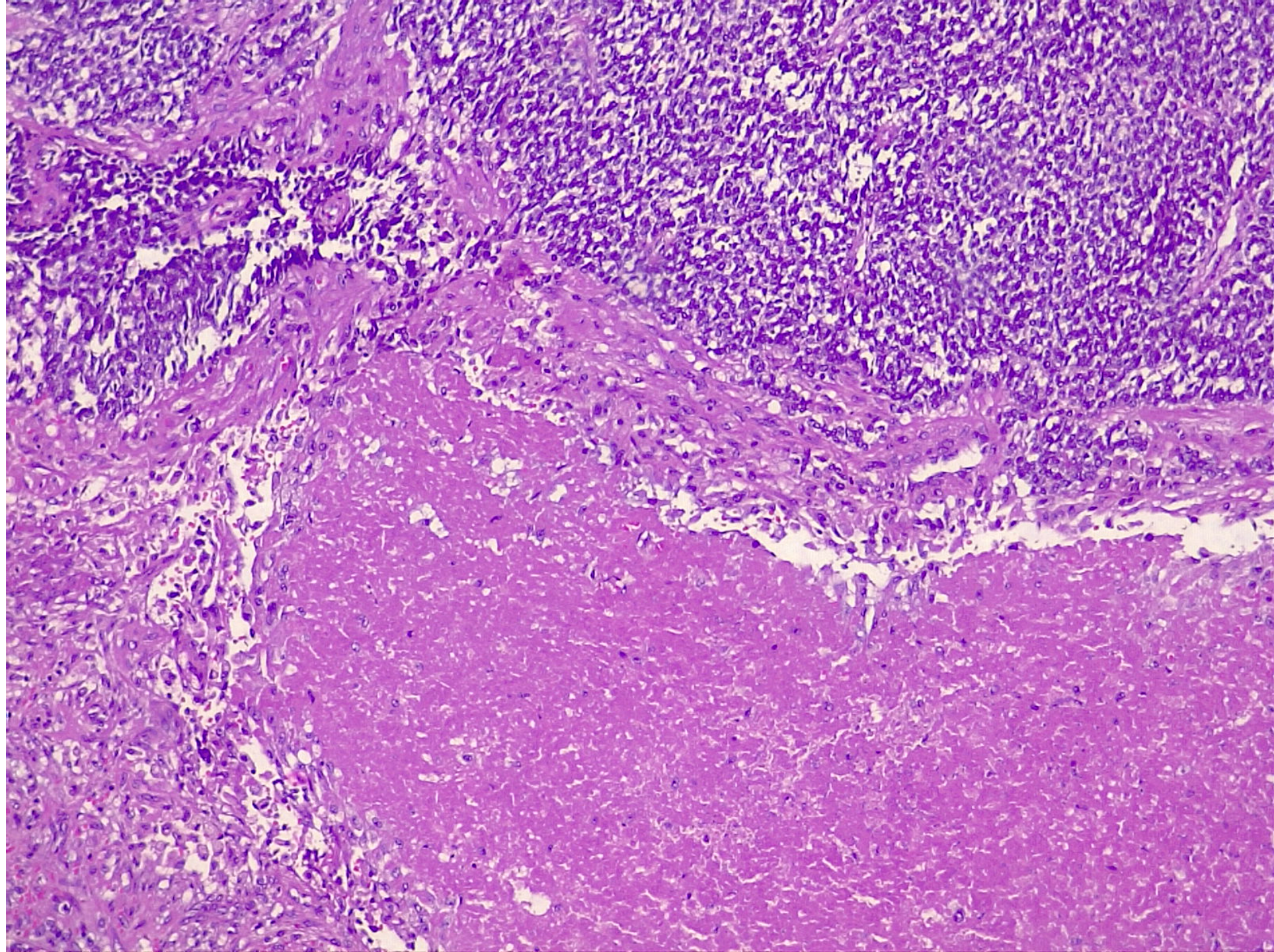
Perinephric
fatty tissue
infiltrated by
round cell
neoplasia
(Hematoxylin-
Eosin).



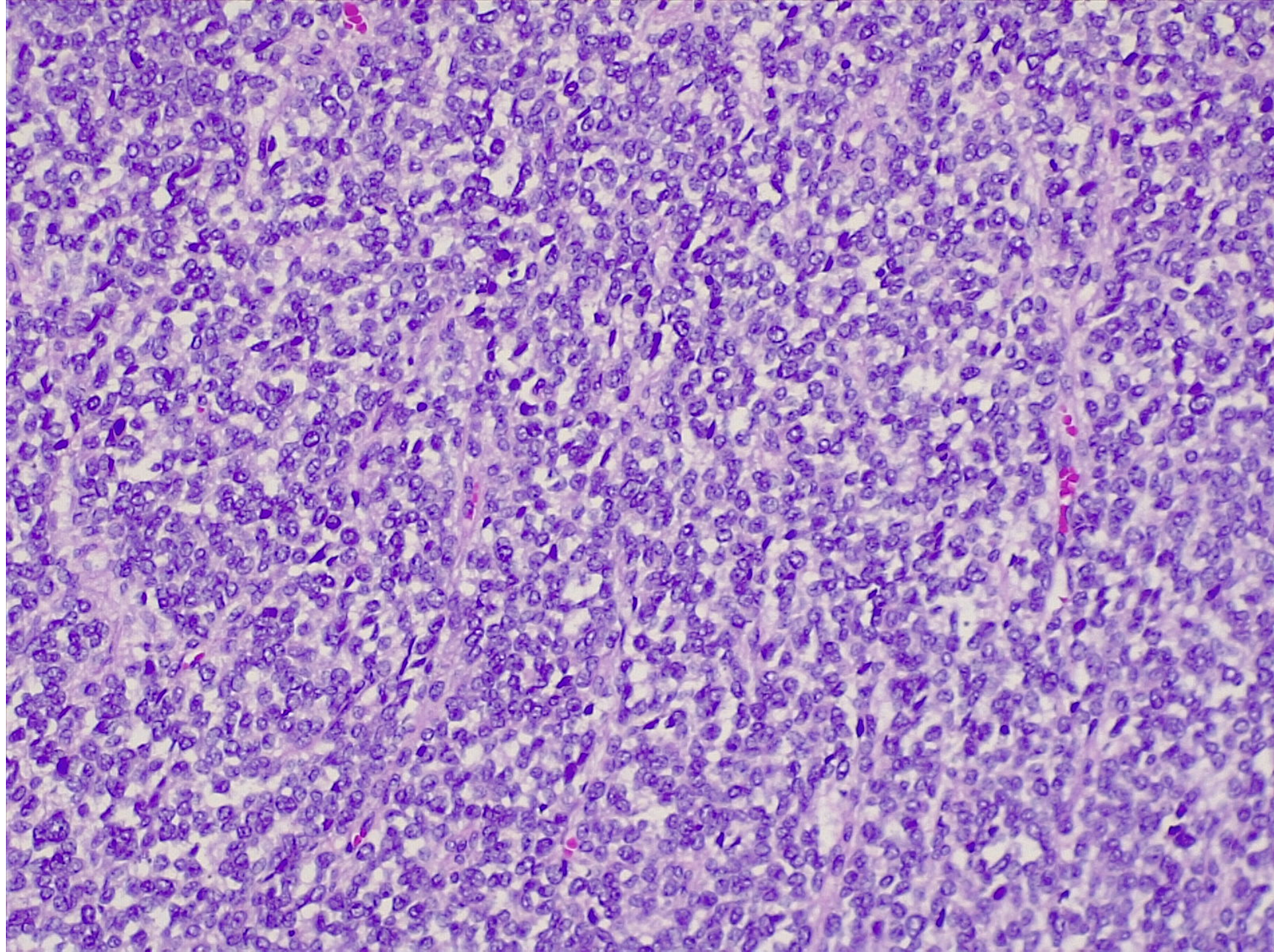
Small round cell
neoplasia
with fibrous
septae
(Hematoxylin-
Eosin).



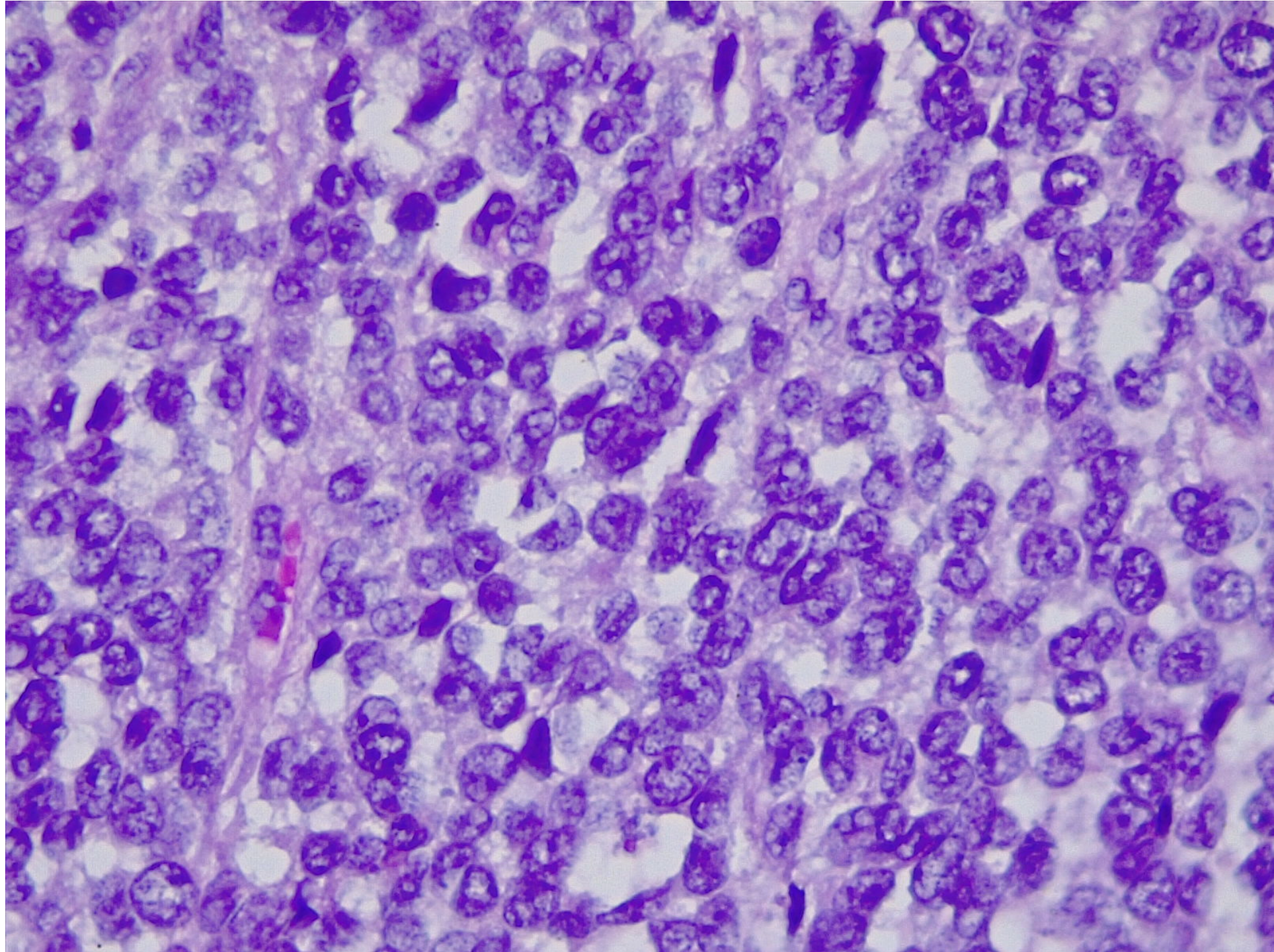
Small round cell
neoplasia
with focal
necrosis
(Hematoxylin-
Eosin).



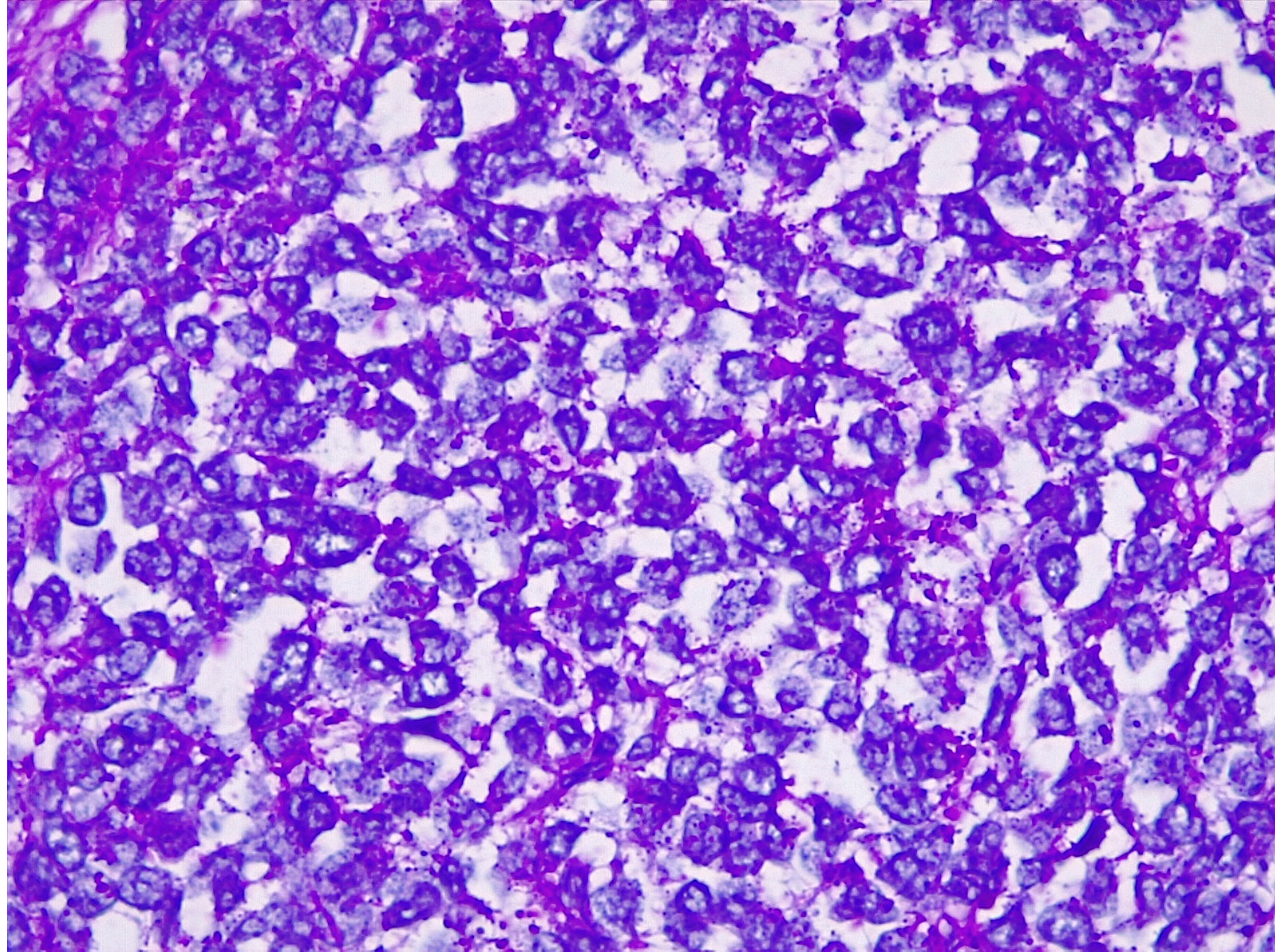
Small round cell
neoplasia
(Hematoxylin-
Eosin).



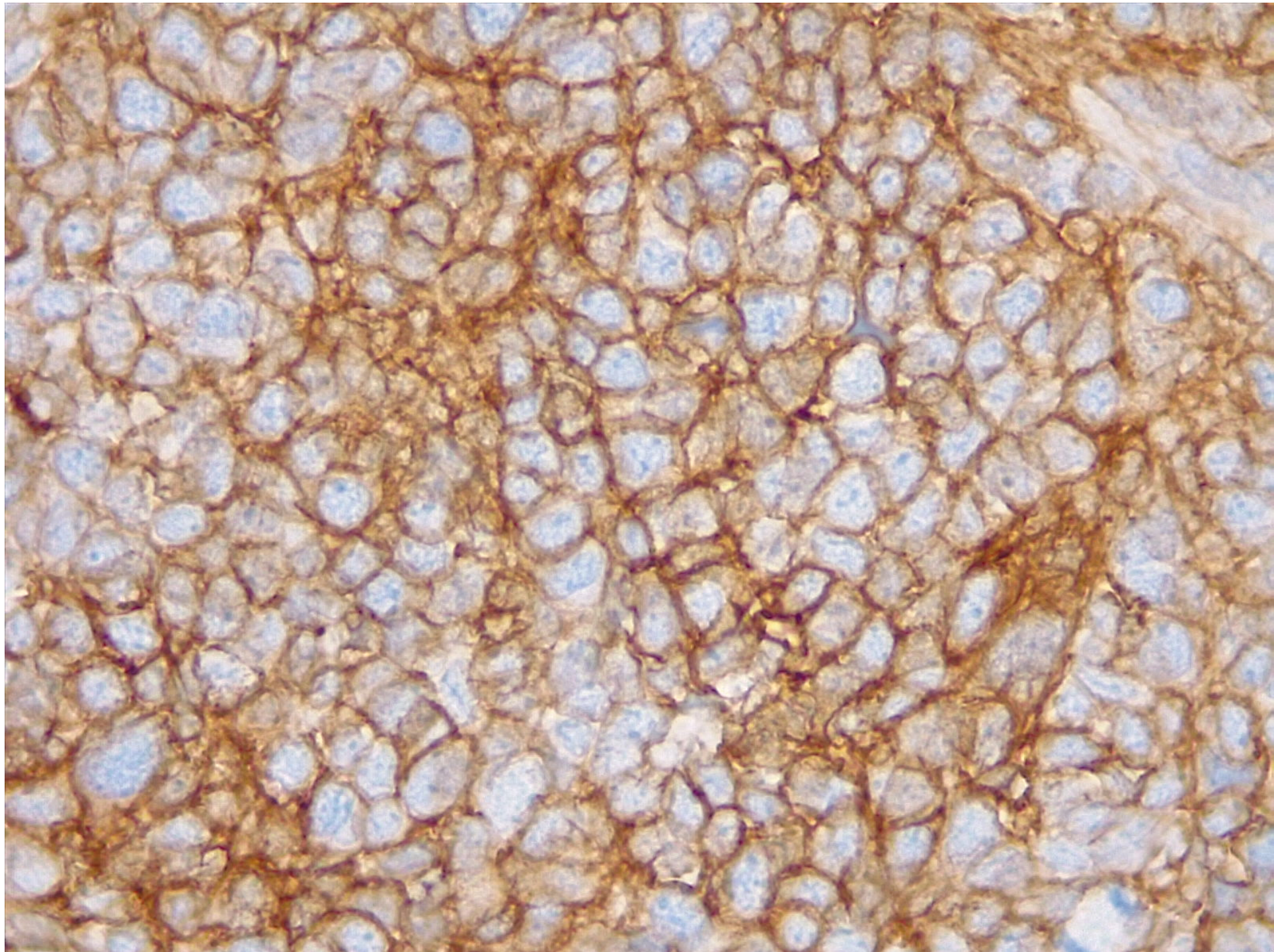
Small round cell
neoplasia
(Hematoxylin-
Eosin).



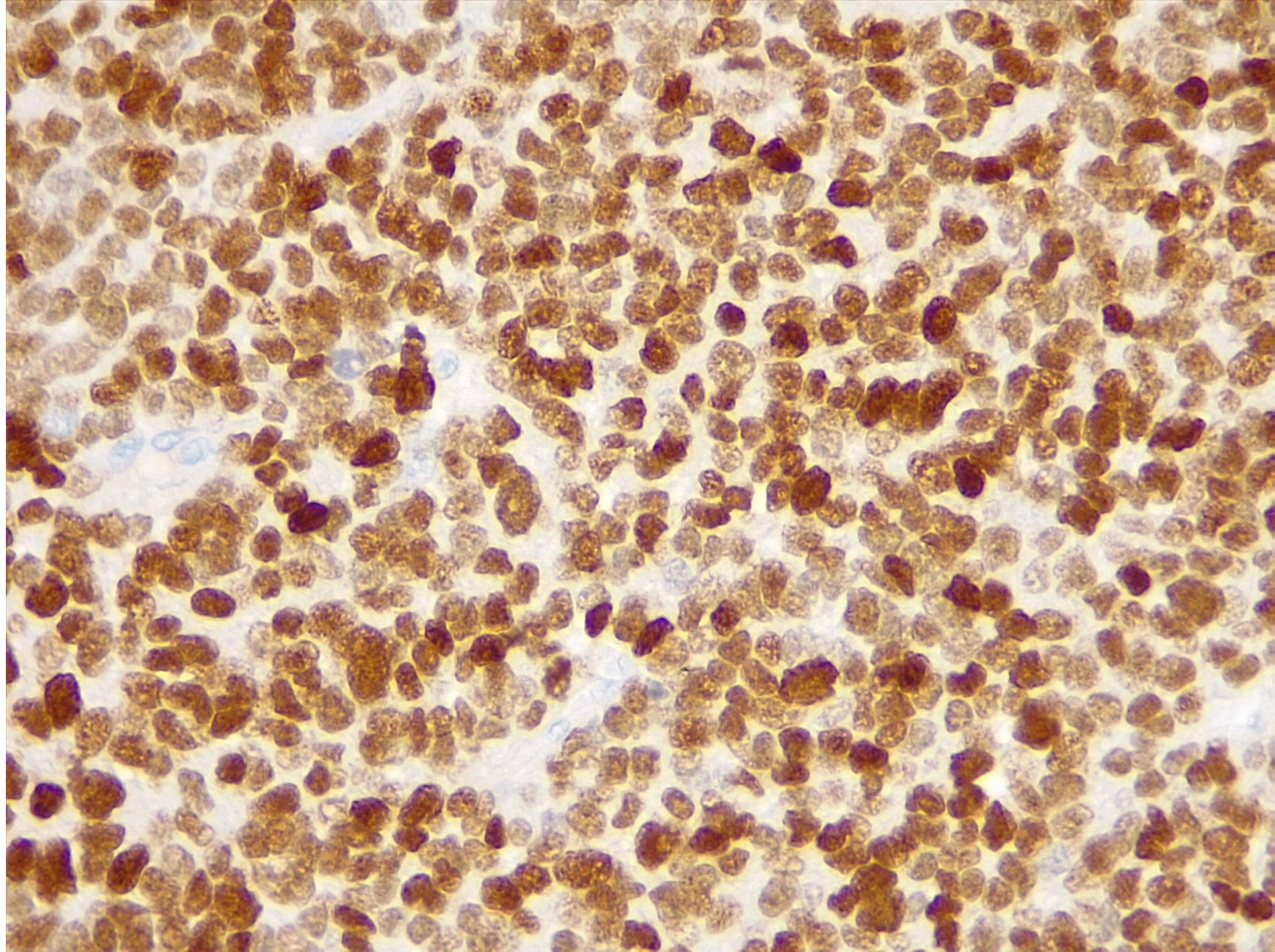
Small round cell
neoplasia
(PAS).



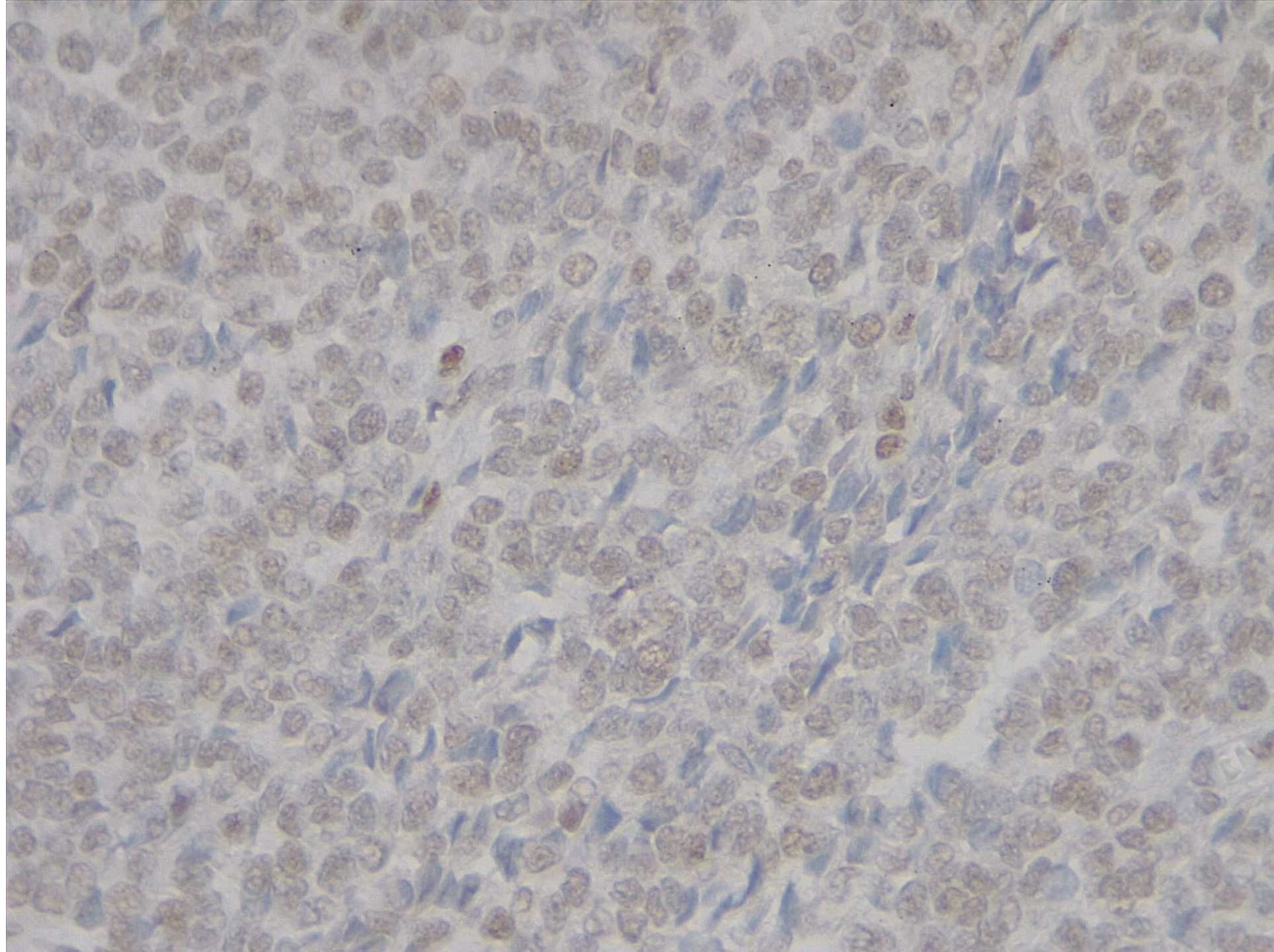
CD 99



NKX2.2



FLI1



NEGATIVE

Cytokeratin

Desmin

Myogenin

CD45

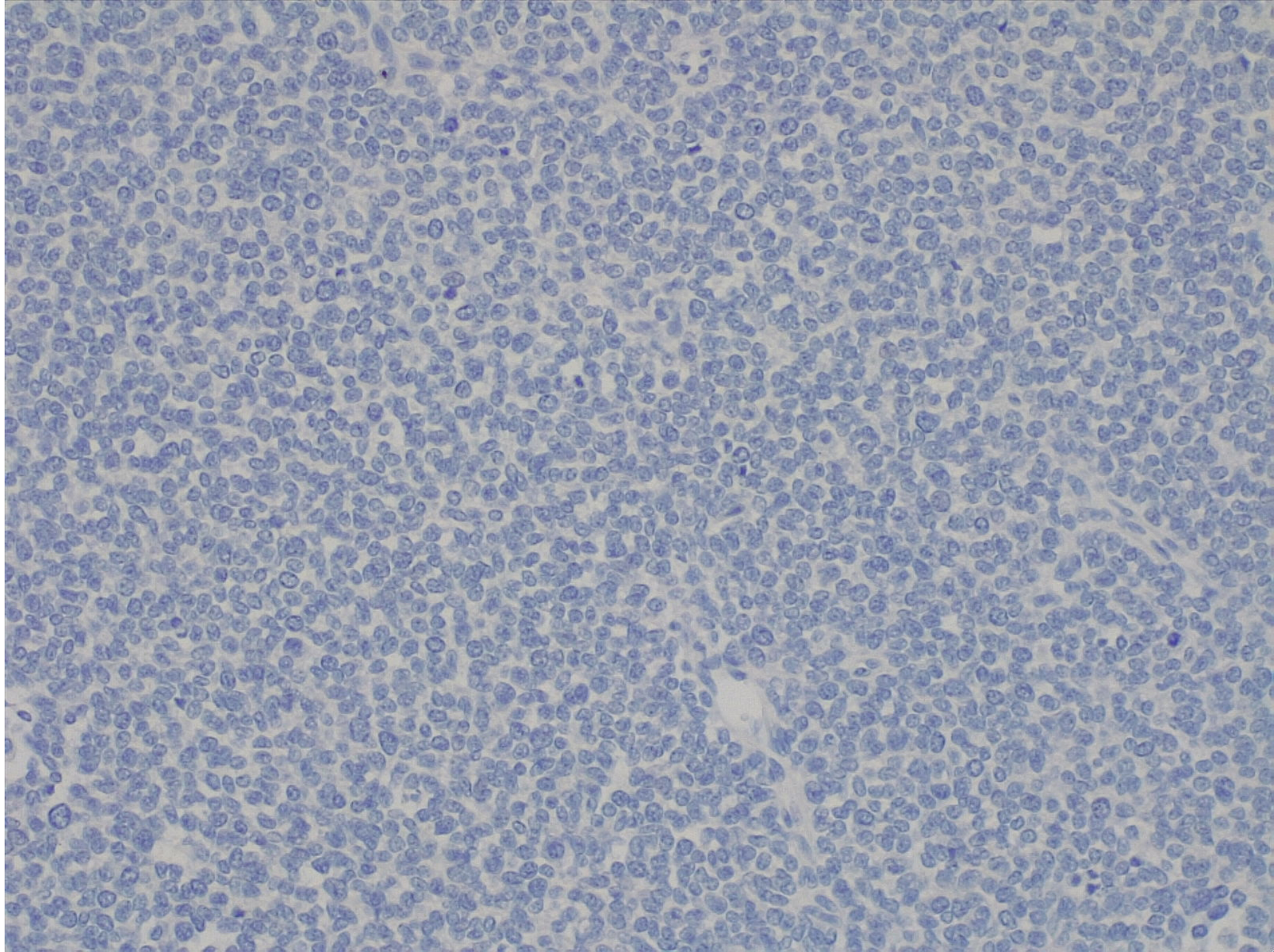
TdT

CD56

Syn

WT1

BCOR



FISH
performed at

National
Children's
Hospital

San José, C.R.

HIBRIDACION IN SITU CON FLUORESCENCIA (FISH)

| PARÁMETRO | RESULTADO | UNIDADES | INTERVALO |
|---|---|----------|-----------|
| HIBRIDACIÓN IN SITU CON FLUORESCENCIA (FISH) EN | FISH | - | |
| | nuc ish(EWSR1x2-4)(5` EWSR1 sep 3 ` EWSR1x1-2)[100] Se realiza FISH usando la sonda de ADN "Break Apart", específica para detectar rearrreglos del gen <u>EWSR1 (locus 22q12)</u> , se observa un Patrón Anormal de Señalización, compatible con un <u>rearrreglo del gen EWSR1, (22q12) en 100% de las células analizadas.</u> | | |
| | nuc ish (SYTx2-3)[100] <u>SS18 (SYT)(cromosoma18q11.2).</u> Patrón normal de señalización. En el análisis de 100 células NO se observan rearrreglos cromosómicos que involucren este gen. | | |
| | nuc ish (DDIT3x2) [100] nuc ish <u>DDIT3 (Break Apart), cromosoma, 12q13:, Patrón normal</u> de señalización. En el análisis de 100 células no se observan rearrreglos cromosómicos que involucren este gen. | | |
| | nuc ish (CEP12 x2) (MDM2 x2) [100]. Análisis de FISH utilizando la sonda específica para locus cromosoma 12q15 (gen MDM2), /cep 12 (centrómero 12p11.1-q11) muestra un patrón NORMAL; NO se observa amplificación del gen MDM2 en 100 células analizadas. | | |
| | nuc ish (FOXO1 x2-4) [100]. <u>FOXO1 (Break Apart), sonda de ADN locus cromosoma, 13q14: Patrón normal</u> de señalización. En el análisis de 100 células no se observan rearrreglos cromosómicos que involucren este gen. | | |

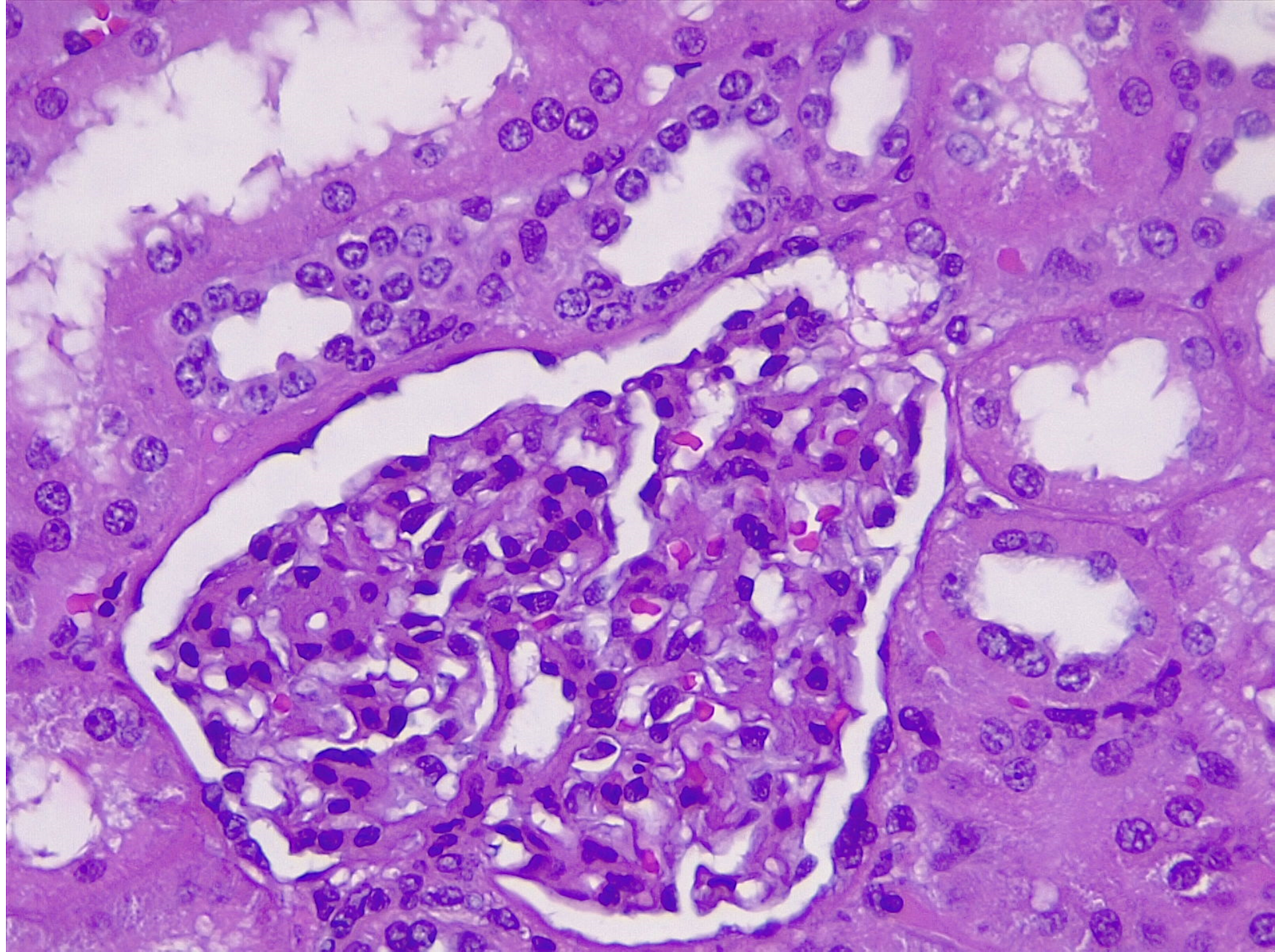
Additional
finding:

Glomerulus with
adhesion at
tubular pole

=

FSGS, tip variant

(Hematoxylin-
Eosin).





DIAGNOSIS

- Right kidney:
EXTRASKELETAL EWING'S SARCOMA
- 15 cm
- Infiltration of:
renal parenchyma, pyelocalyceal system, hilar and perirenal fat
- Mitotic activity: 23 mitoses/sq. Mm
- Necrosis: present (10%)
- Lymphovascular invasion: present
- pT3 (retroperitoneum)



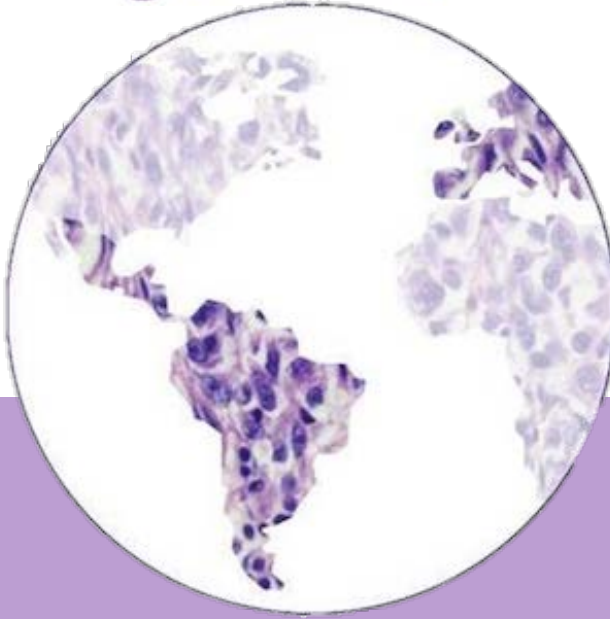
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• QUESTIONS:

CASE . MQV

- 1. The patient had a brother who died of Ewing's sarcoma. Do you think that this family has a family syndrome? What genetic studies would you carry out?
- 2. Has any of you ever had a case of Ewing Renal Sarcoma before?
- 3. Do you consider that these abdominal liquid collections could be a disease?
- 4. What chemotherapy regimen would you use in this patient? VAC-IE or just VAC?
- 5. How many cycles of chemotherapy would you give knowing that the tumor had ruptured in the abdominal cavity?

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Fernando Campos, MD
A.C.Camargo Cancer Center
Sao Paulo
Brazil

November, 2021



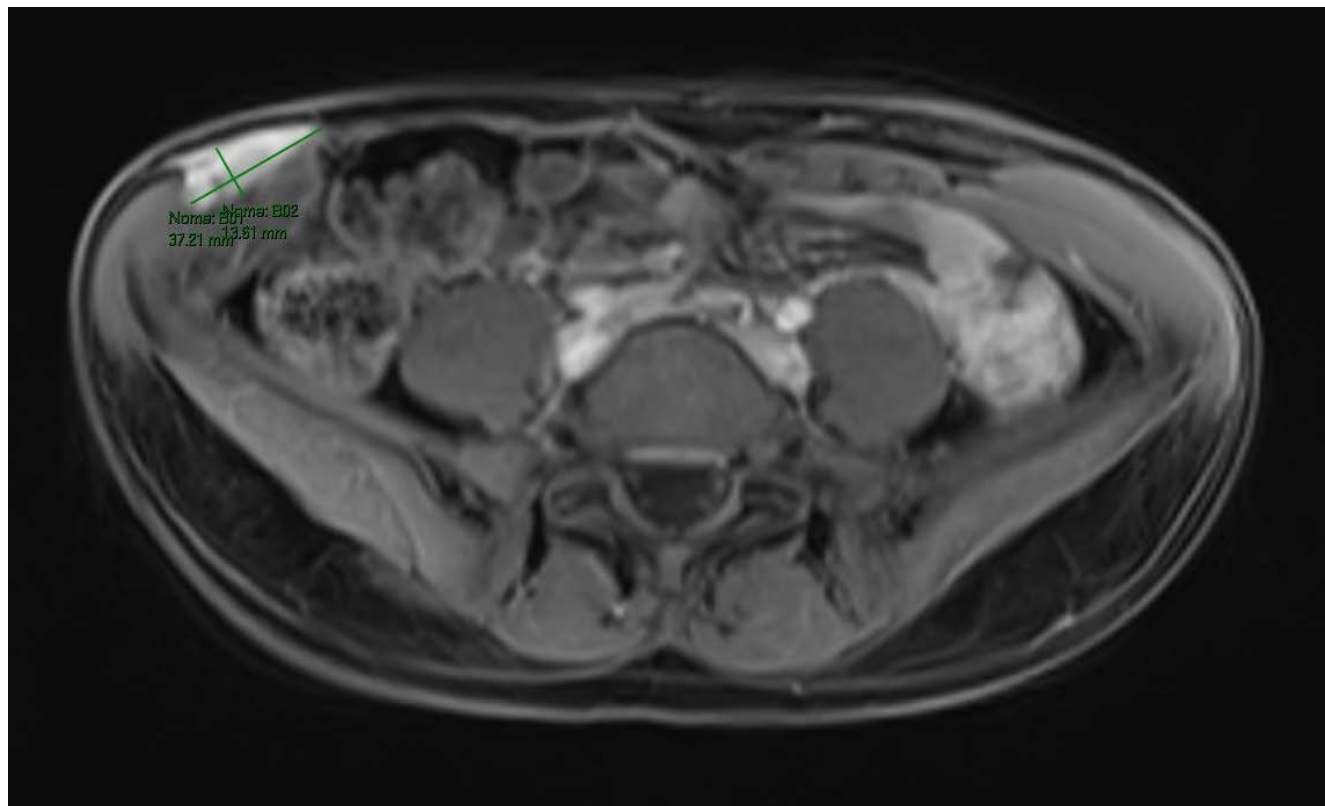
Virtual MDT Board

- **Woman, 32 yo, no comorbidities**
- 2013: Desmoid tumor in the abdominal wall – surgery
- Feb 2014: local relapse 6 months later – surgery (7 cm lesion, free margins)
- Aug 2015: relapse in the abdominal wall (Liposomal doxorubicin – SD) - Stable disease since then





Virtual MDT Board





Virtual MDT Board





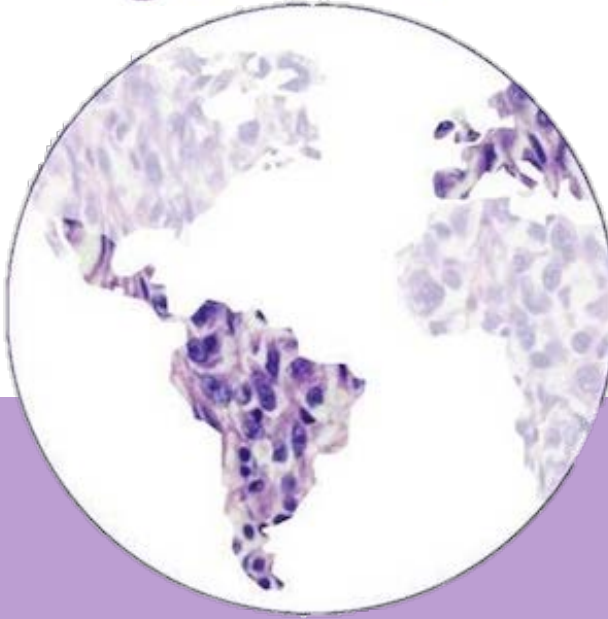
Virtual MDT Board

DISCUSSION:

32 yo woman with abdominal wall desmoid tumor. Stable disease since 2015, asymptomatic.

- The patient wishes to be submitted to plastic surgery in the abdominal wall. What would be our recommendations?
- The patient also asks about pregnancy in the future. How to guide her regarding this?

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Virtual MDT Board

Celso Mello, MD

A.C.Camargo Cancer Center
Sao Paulo
Brazil

November, 2021



Virtual MDT Board

ID: Male, 46 yo, from São Paulo

Medical History

- 2008: patient presented with pain and swelling in the left thigh. MRI showed a 3.4 x 3.8-cm mass in the deep soft tissues of his left thigh.
- 2009: resection of the lesion. Pathologic report: Endovascular papillary angioendothelioma (Dabska tumor). Negative surgical margins.
- 2010: local recurrence with painful lesion
- From 2014 to 2020 several embolizations were performed.
- After the last procedure (transarterial bleomycin embolization) in february/2020, the patient presented a worsening painful swelling in the upper left leg, associated with local bleeding and ulceration. Moreover, he related relevant weight loss and hyporexia and persistent anemia.
- MRI performed in June/2020 showed a large 20 x 14 x 11-cm mass, with an exophytic component of 4 x 3-cm in the posterior and lateral compartment of the thigh (images)



Virtual MDT Board

- MRI performed in June/2020 showed a large 20 x 14 x 11-cm mass, with an exophytic component of 4 x 3-cm in the posterior and lateral compartment of the thigh (images)
- December/2020: Considering recently growth, at a progressive rate, it suggested a component of malignant transformation - a new biopsy was indicated - confirmed Dabska tumor
- Initiated systemic chemotherapy: Weekly Paclitaxel 80 mg/m² x 12 cycles with transient improve in bleeding and pain
- March/2021: Worsening local bleeding and pain. New MRI showed signs of local progression (images) with bone destruction (femur)
- April/2021: Initiated everolimus 10 mg/dia
- Oct/2021: PD with increased bleeding, Hb 4.9 g/dl



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PMHx – non contributory

Allergy – no allergies

Habits – no smoking or drinking;

Family Hx – father with melanoma, grandfather with lung cancer, paternal uncle with lung cancer

Physical Exam – good condition, ECOG 1

Palpable mass on the left thigh, with ulceration and hemorrhagic and necrotic crusts

Diagnostic Tests

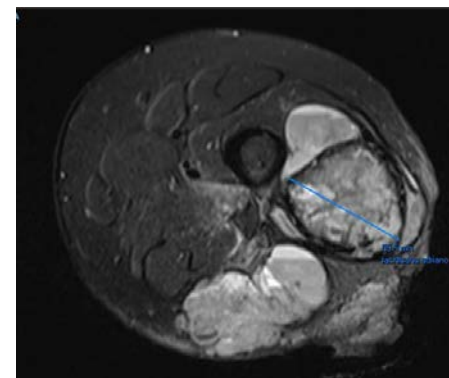
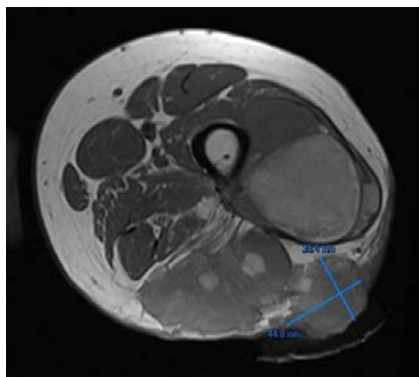
Pathologic review – Endovascular papillary angioendothelioma



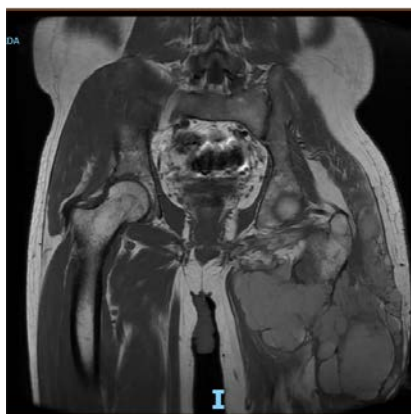
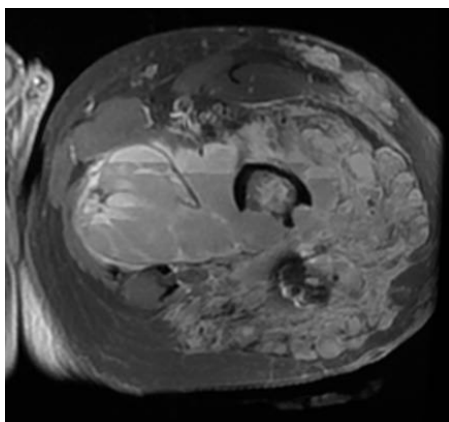
Virtual MDT Board

Images:

June/2020



March/2021





Virtual MDT Board

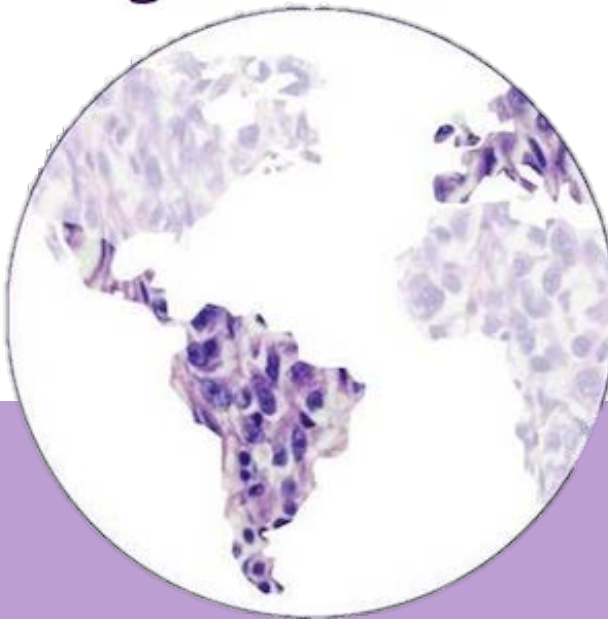
Impression:

Young adult with large PILA tumor of the right thigh, with limiting bleeding and pain, previous treatment with multiple embolization and systemic chemotherapy.

Discussion:

1. Amputation (hemipelvectomy)
2. Systemic treatment for PILA

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Hospital Oncológico de la provincia de Córdoba
Gisela German





Virtual MDT Board

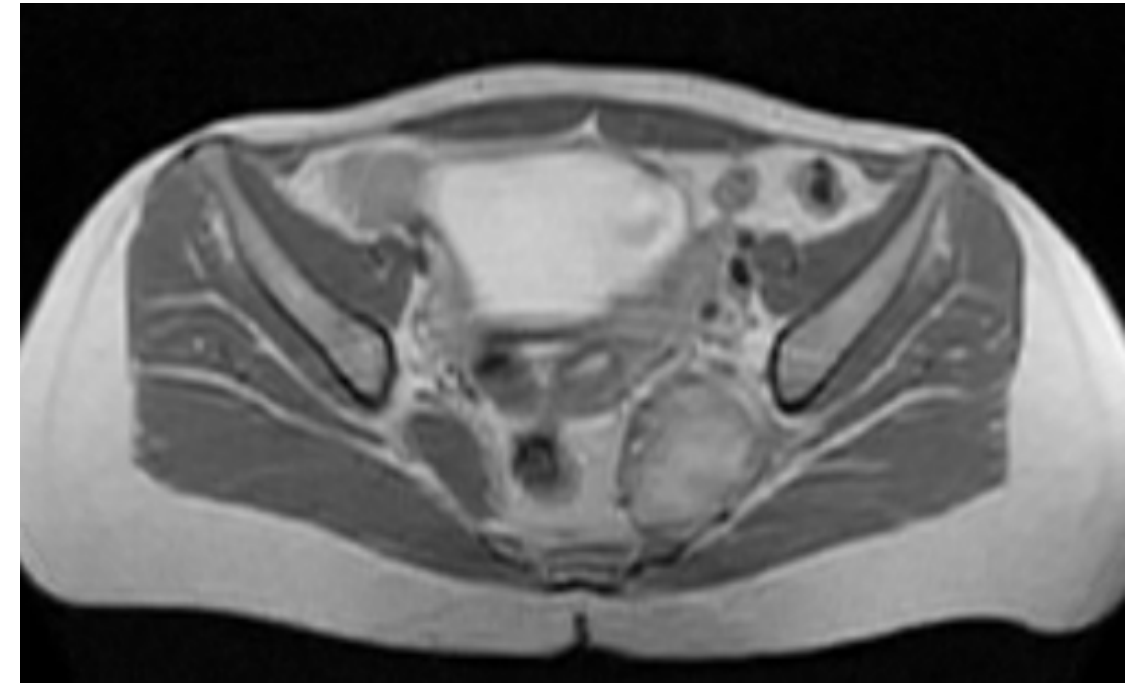
35 years old female patient.

Without pathological history.

Multiple consultations due to pain in the left lumbosacral región.

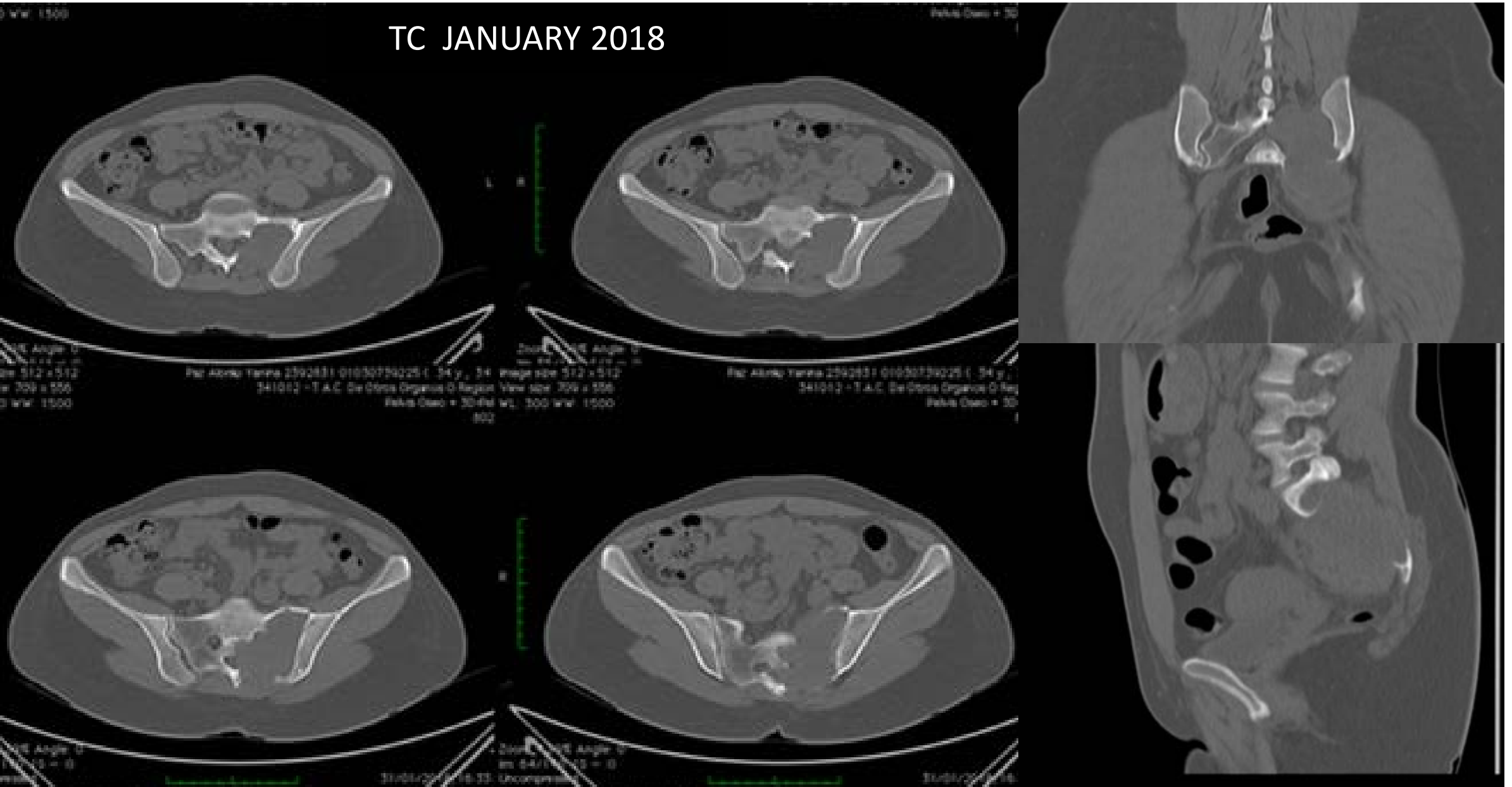


PELVIS MRI PERFORMED JAN 26th 2018:

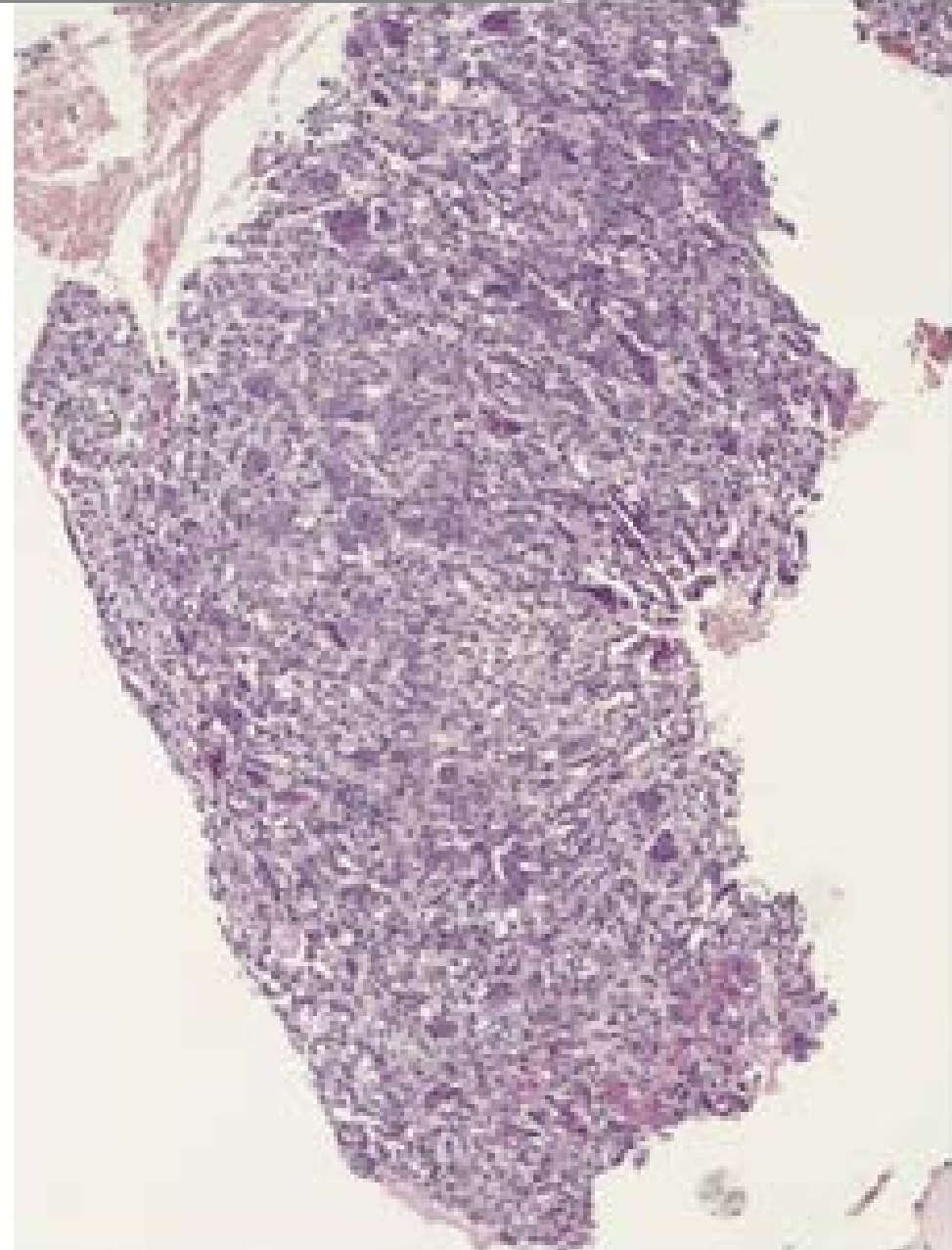
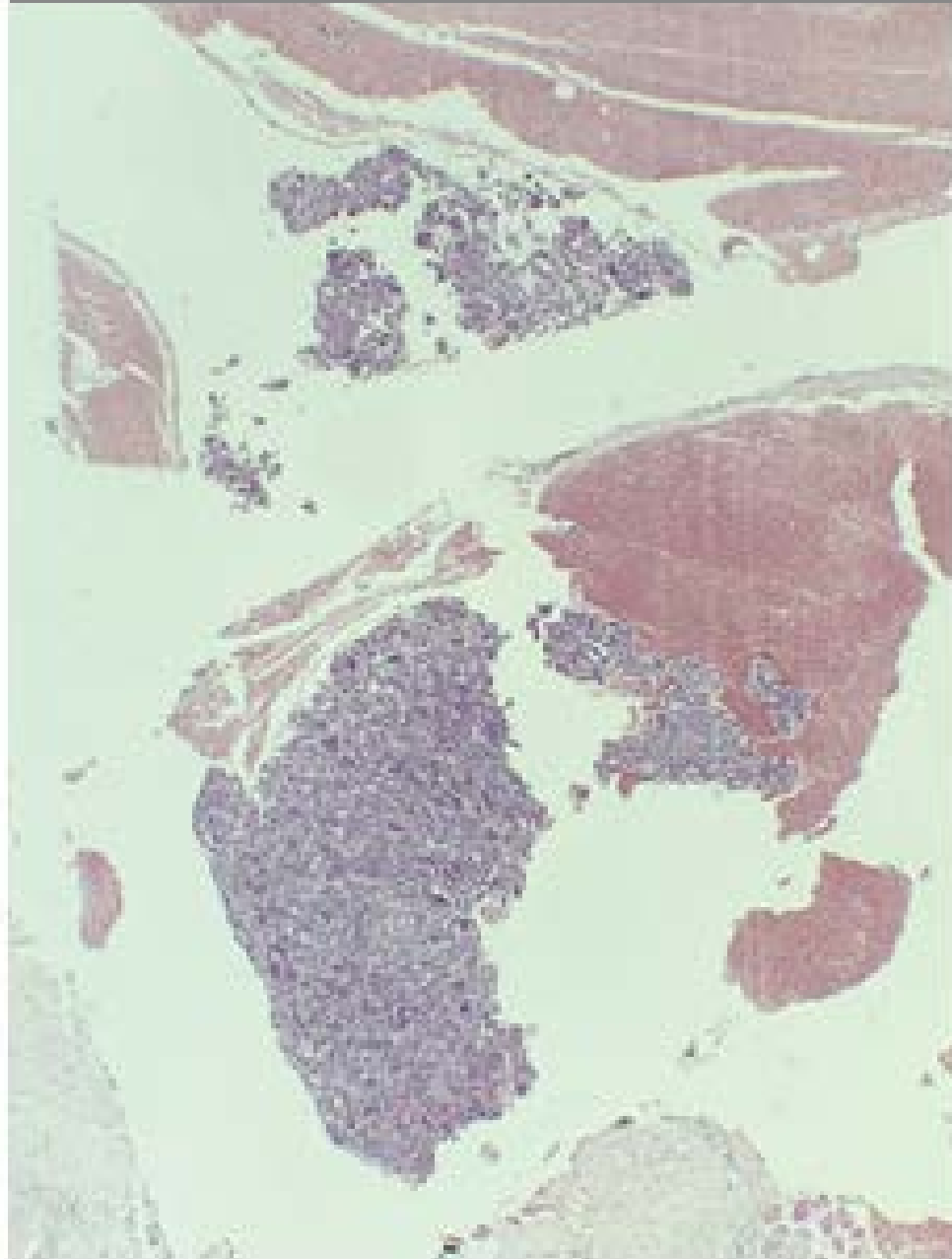


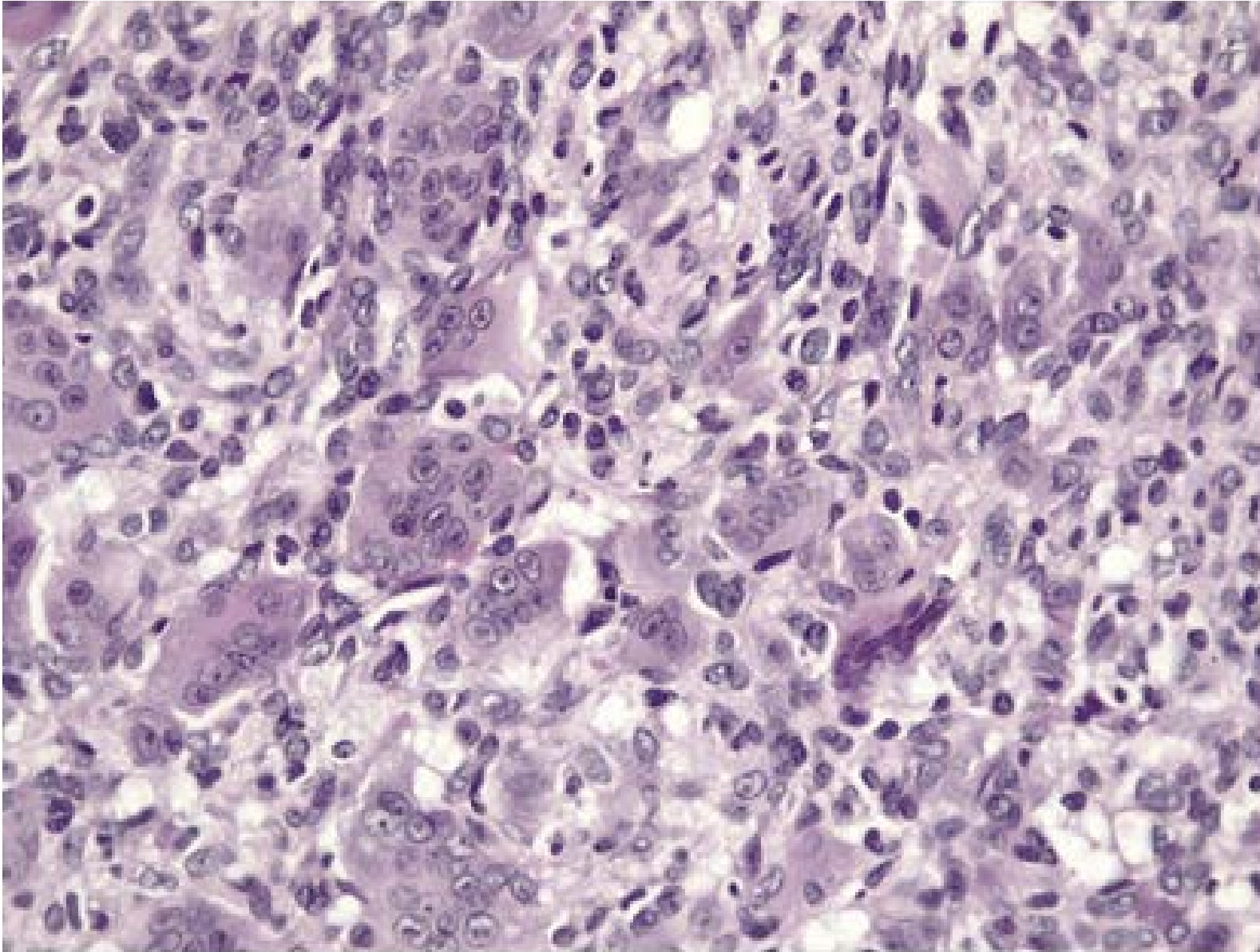
In left sacra region heterogenic image is observed. Hyperintense in STIR and T2 sequences, hypointense in T1, with heterogeneous caption to contrast. It presents apparent involvement of the left iliac bone in the inferior articular región. Presents ventral and caudal extension that displaces adjacent soft tissues. with a diameter greater than 67mm.

TC JANUARY 2018



CT-guided biopsy was performed on February 1st, 2018.





DIAGNOSTIC:

Fragment of dense fibrous tissue, periosteum, hyaline cartilage and compact bone tissue, with numerous multinucleated osteoclast-like giant cells.

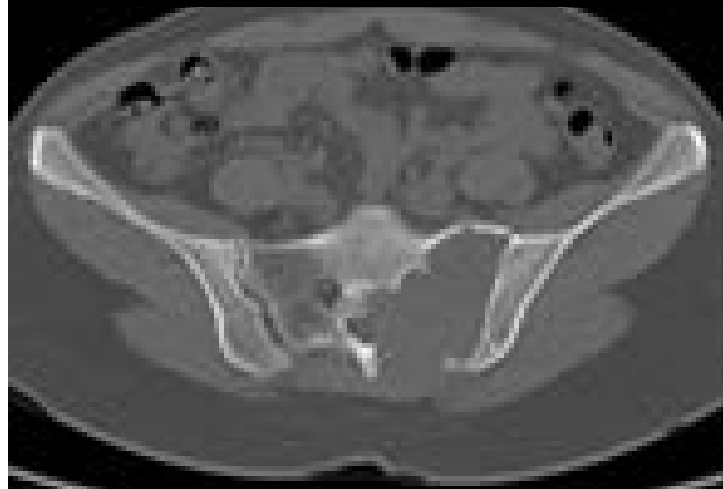
GIANT CELL TUMOUR



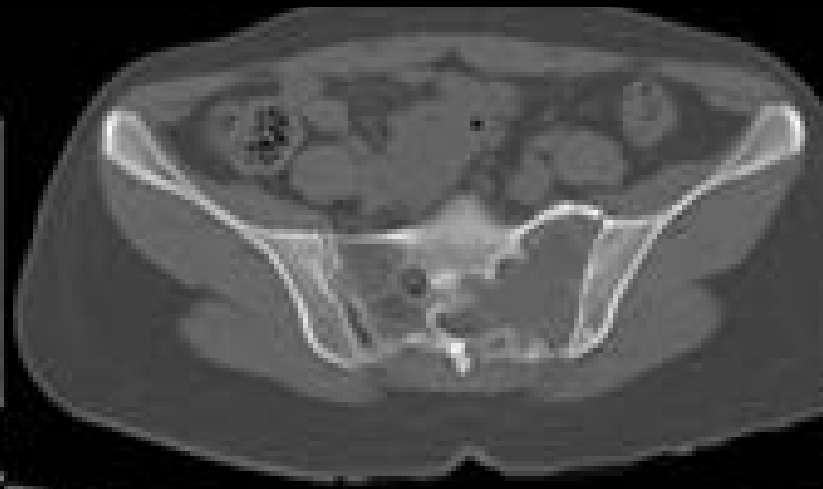
TREATMENT WITH DENOSUMAB WAS STARTED FROM MARCH 2018 TO MARCH 2019. SHE WAS TREATED ON A MONTHLY BASIS, WITH GOOD CLINICAL RESPONSE, IMPROVEMENT IN PAIN WITHOUT REQUIRING ANALGESIA, NO LIMITATION IN WALKING.

SINCE MARCH 2019 CONTINUED WITH QUARTERLY TREATMENT UP TO THE PRESENT.

JANUARY 2018



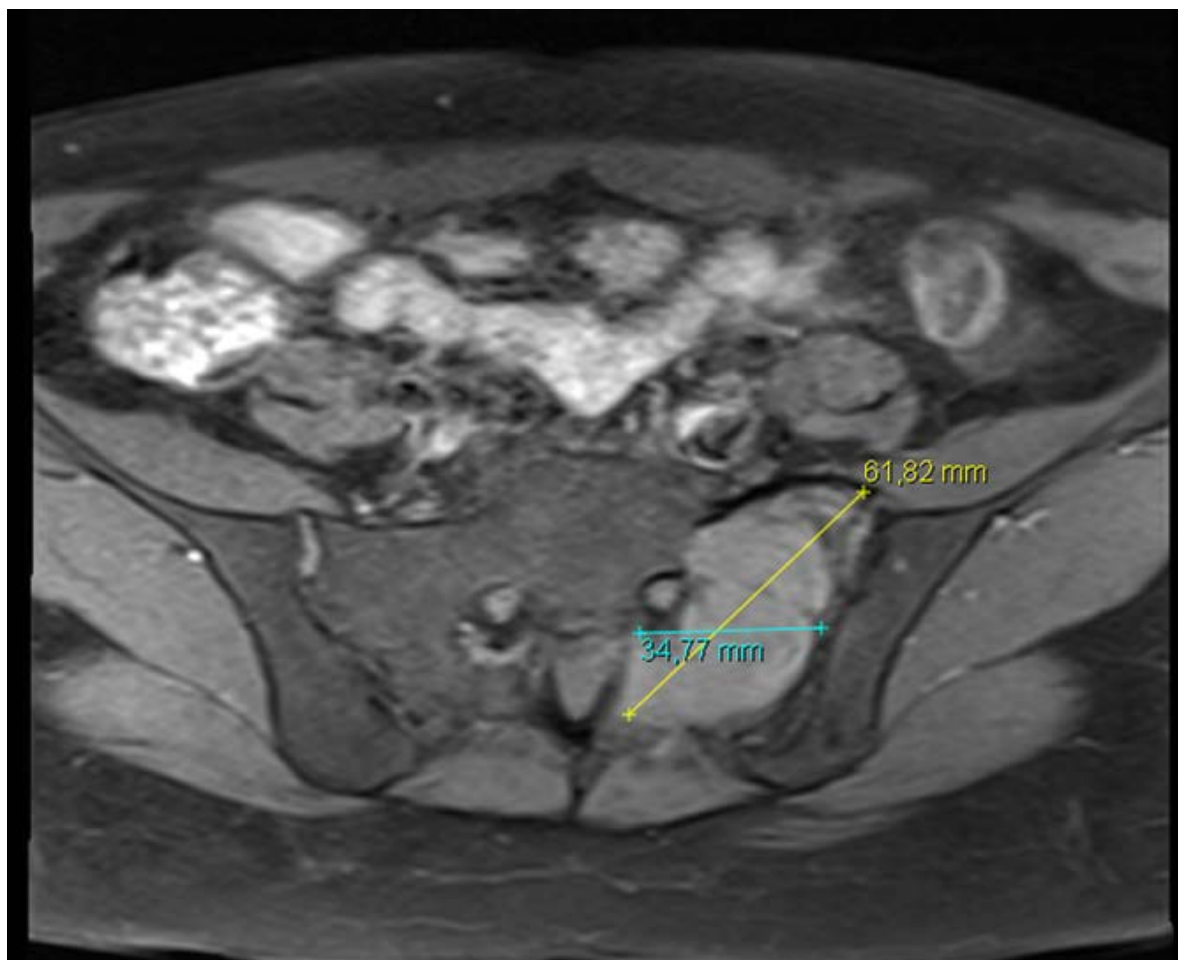
OCTOBER 2018



APRIL 2018

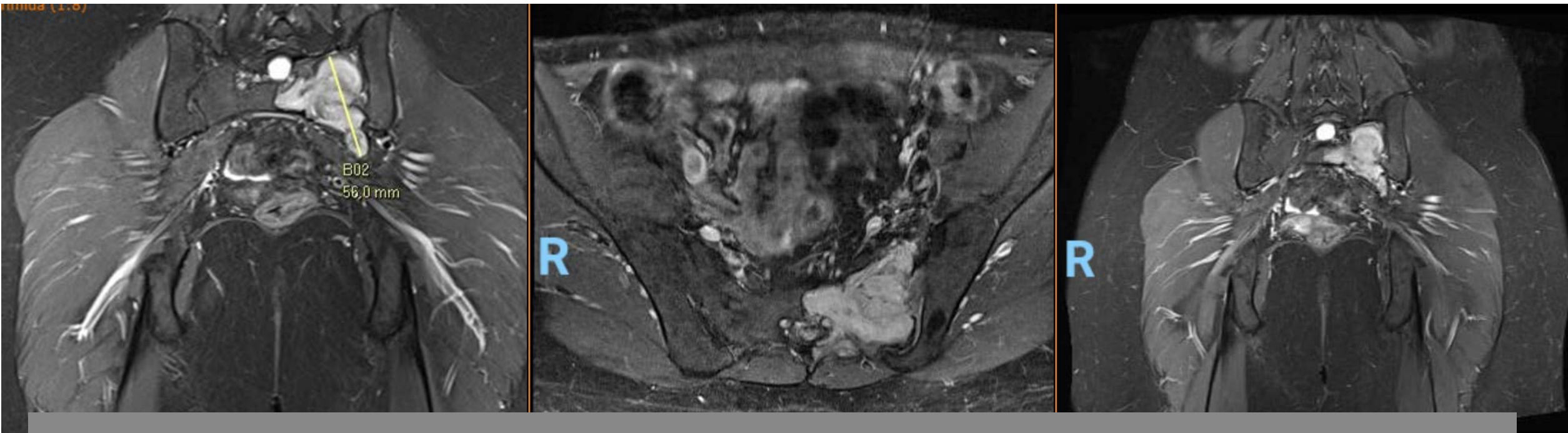


IN MAY 2019 (14th months of treatment) MRI WAS PERFORMED WITH ANGIOGRAPHY OF THE TUMOR, WITH THE INTENTION OF EMBOLIZATION, WHICH WAS DISCARDED.



IN OCTOBER 2021 (22 applications of denosumab) WITHOUT PHARMACOLOGICAL COMPLICATIONS AND ASYMPTOMATIC.

THE PATIENT EXPRESSES DESIRE FOR PREGNANCY



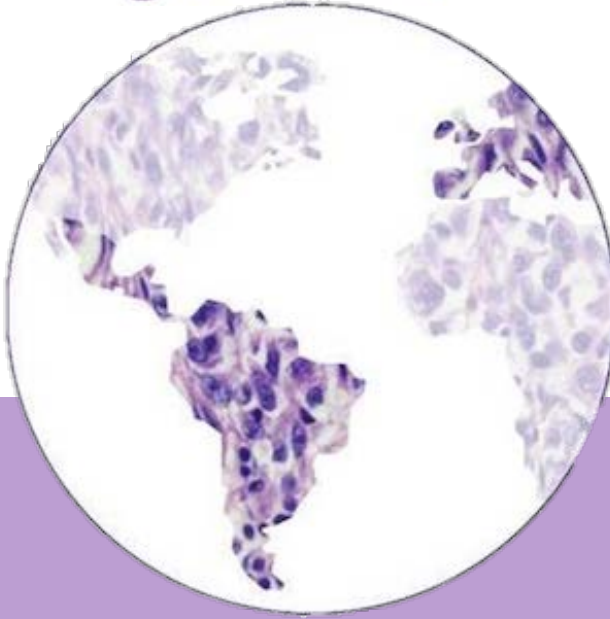
MRI

Imaging stability of the lesion located at the level of the left sacral aileron, which presents morphology and signal similar to the previous examination. It currently measures 56 mm cranio-caudally by 52 mm transversely by 46 mm anteroposteriorly.

-In the post Gadolinium injection sequence presents a moderate and homogeneous enhancement of the lesion without changes with respect to previous study.

-The ADC coefficient measurement shows similar values to those of his previous examinations, with values of 1.4, 1.2 and 1.1 x 10⁻³ mm²/sec in the upper, middle and lower thirds.

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CHALLENGING BONE TUMOR

Fundación Jiménez Díaz/HU Rey Juan Carlos
SELNET Coordination Team



Virtual MDT Board

- 23-year-old Male
- No relevant previous history
- 3 April 2021
- Emergency Department → Pain in right elbow after physical effort (over lifting an excessive load)
- Physical examination: pain in head of radius. No functional impairment. Mild swollen area



Virtual MDT Board

X Ray 3/April/2021





Virtual MDT Board

Elbow MRI (April 2021)

T1 Sequence

DP FAT SAT Sagital



Virtual MDT Board

Elbow MRI (April 2021)

Biceps tendon avulsion (probably traumatic)



MRI

FINDINGS:

- Biceps tendon avulsion (probably traumatic).
- In addition, a lytic centromedular lesion in the radius, 4.7cm, located in epiphysis-metaphysis-diaphysis with aggressive features, was found.
- The lesion do not have a soft-tissue component.
- Hypointense in T1, Hyperintense in T2, and the bone cortical was thin and even broken at some points.

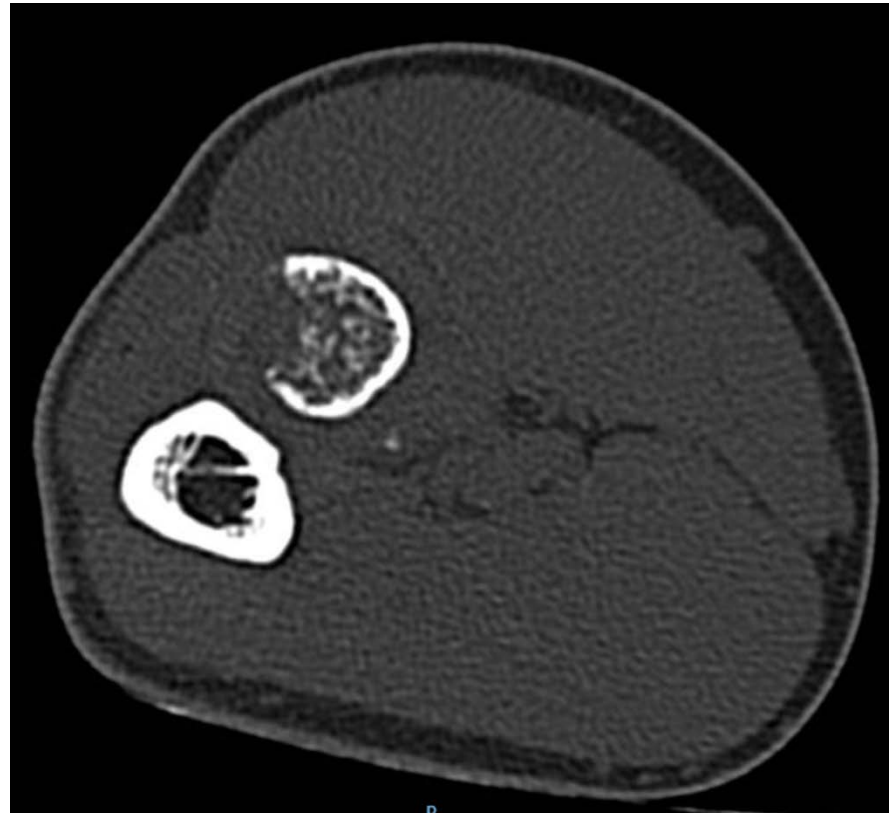
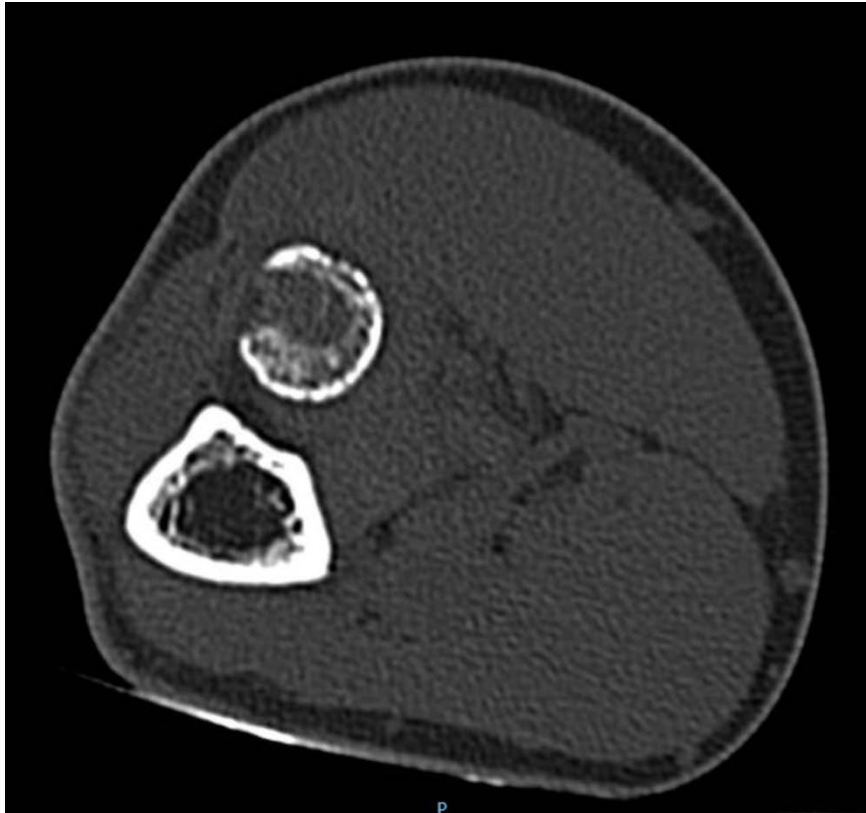


Virtual MDT Board

Reconstruction



Elbow CT (April 2021)



Lytic centromedullary lesion in proximal segment of radius, with aggressive features.



Virtual MDT Board

- MDT discussion → diagnostic Biopsy
- Biopsy: findings of a fibrotic lesion, with no atypia
- IHC for actin, desmin, Betacatenin, CKAE1-3 and CD34 were all negative.
- The most probable diagnosis on that biopsy was desmoplastic fibroma



Virtual MDT Board

- MDT discussion → SURGERY (curettage)
- Surgical findings: tumor firmly attached to the cortical endostium of the bone
- Technique: hard and meticulous resection by curettage of all the walls of the lesion, high speed burr of the cavity and filling with cancellous bone.



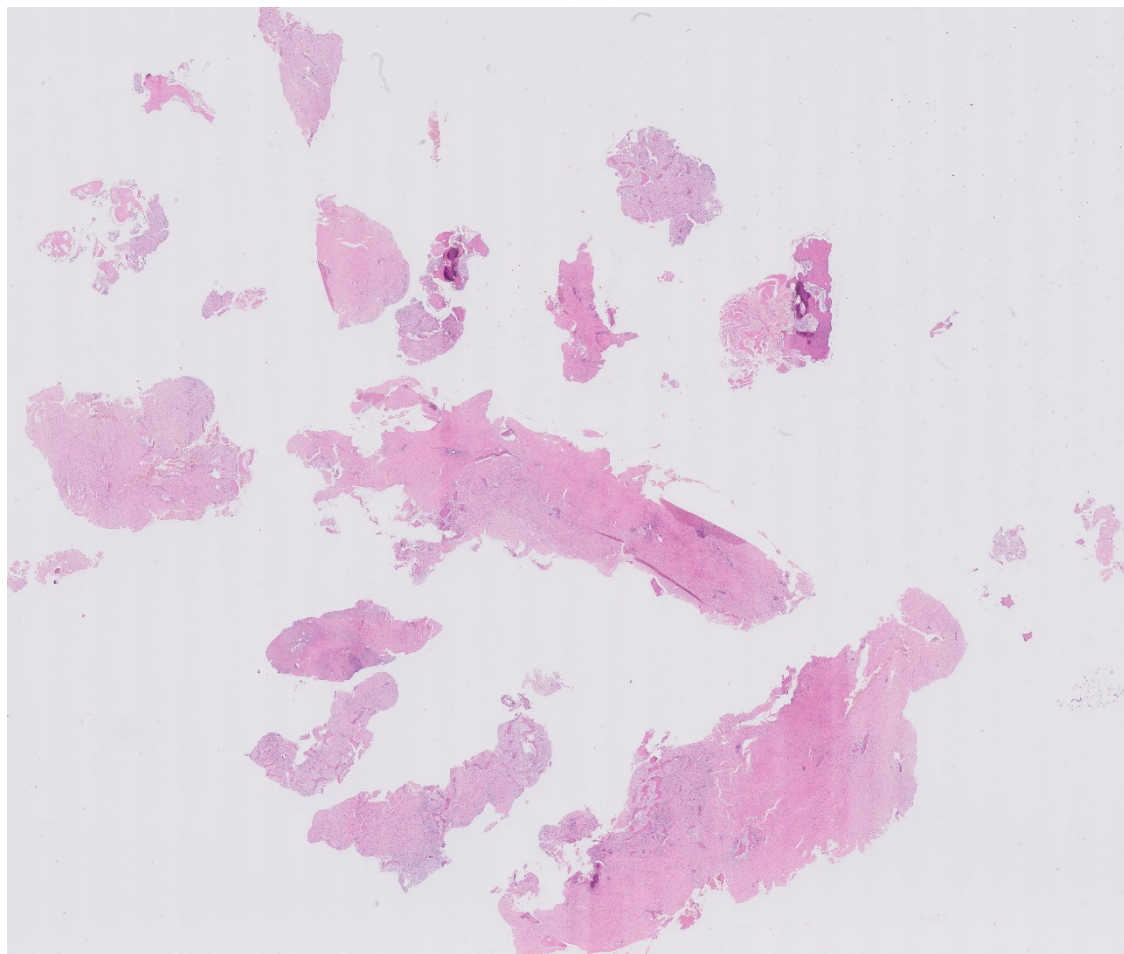
Pathology report from surgery

- Surgical specimen: intralesional resection
- Presence of a fibrous lesion
- IHC positive for SATB2
- IHC negative for MDM2, CDK4, BETA CATENINE
- Ki67 <1%
- MDM2 not amplified (FISH)



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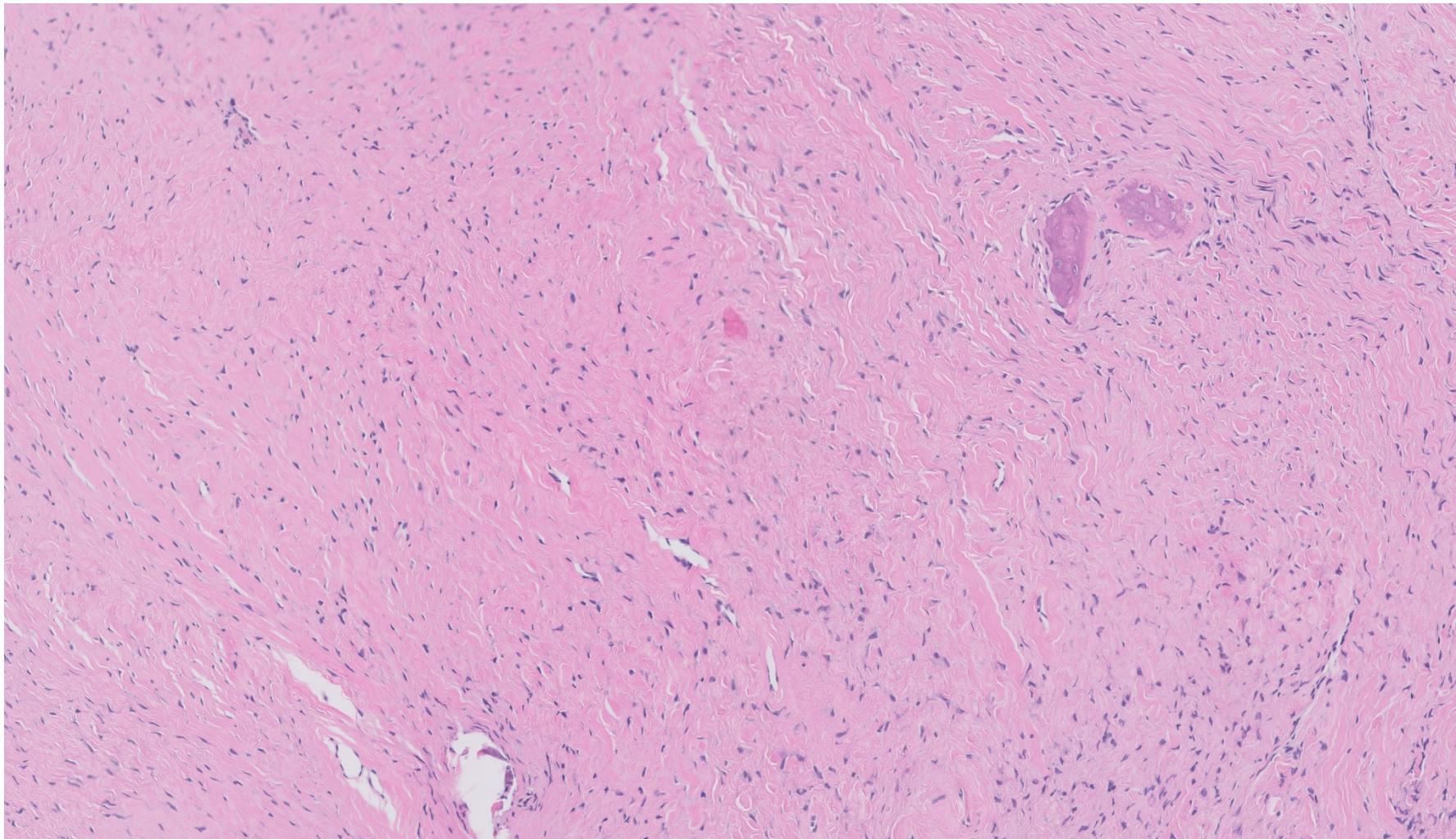
PATHOLOGICAL FEATURES





Virtual MDT Board

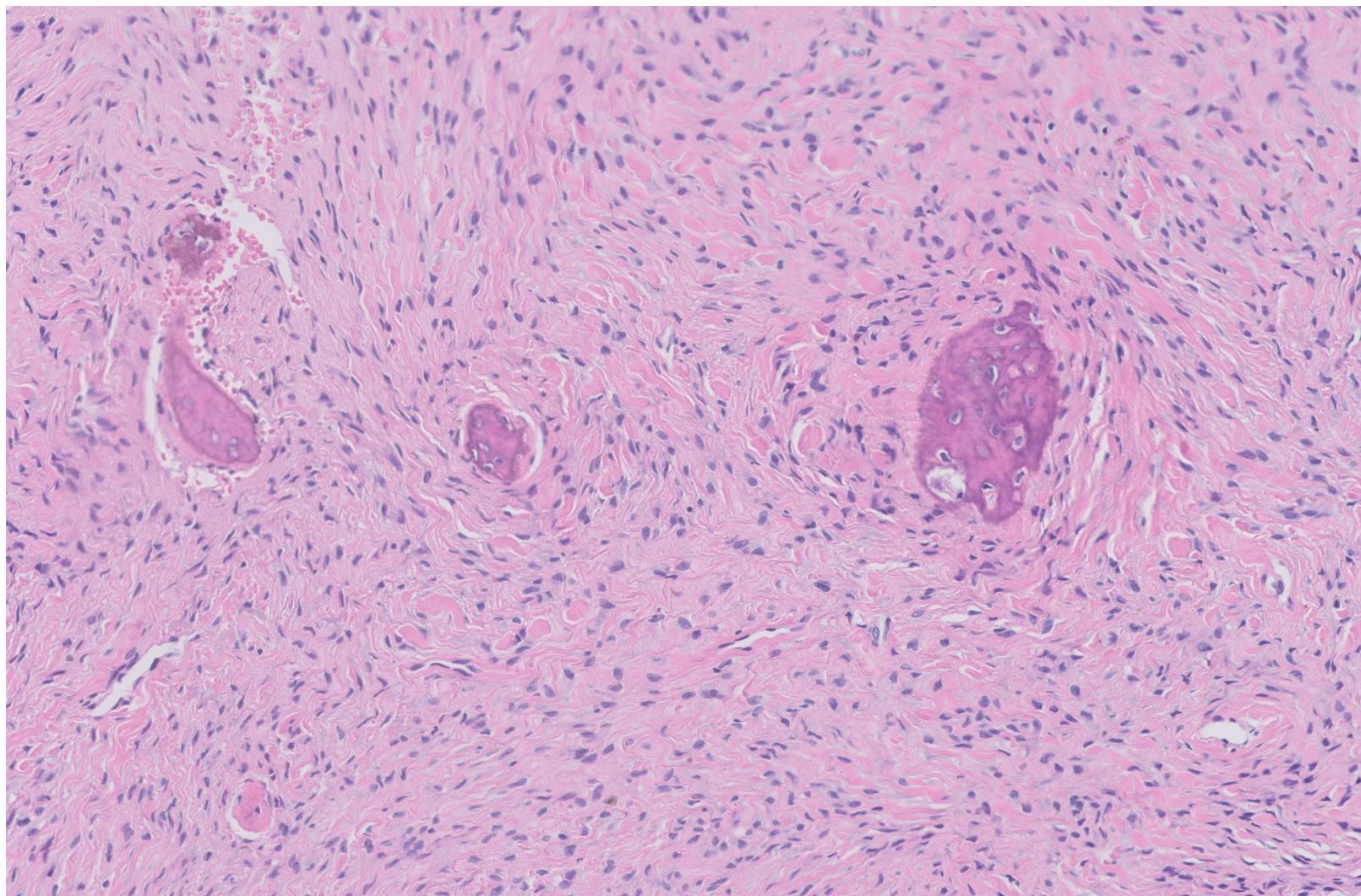
PATHOLOGICAL FEATURES





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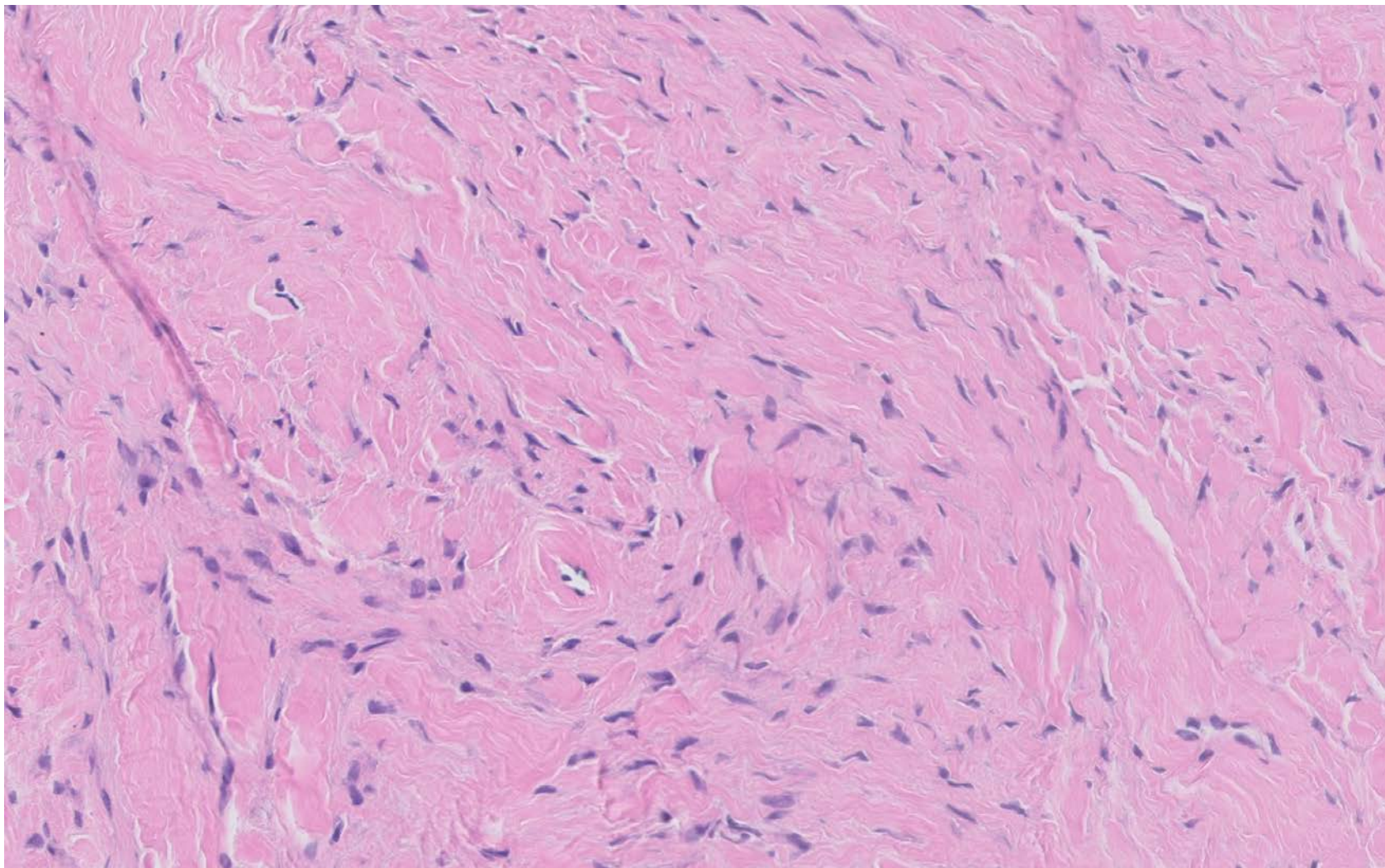
PATHOLOGICAL FEATURES





Virtual MDT Board

PATHOLOGICAL FEATURES

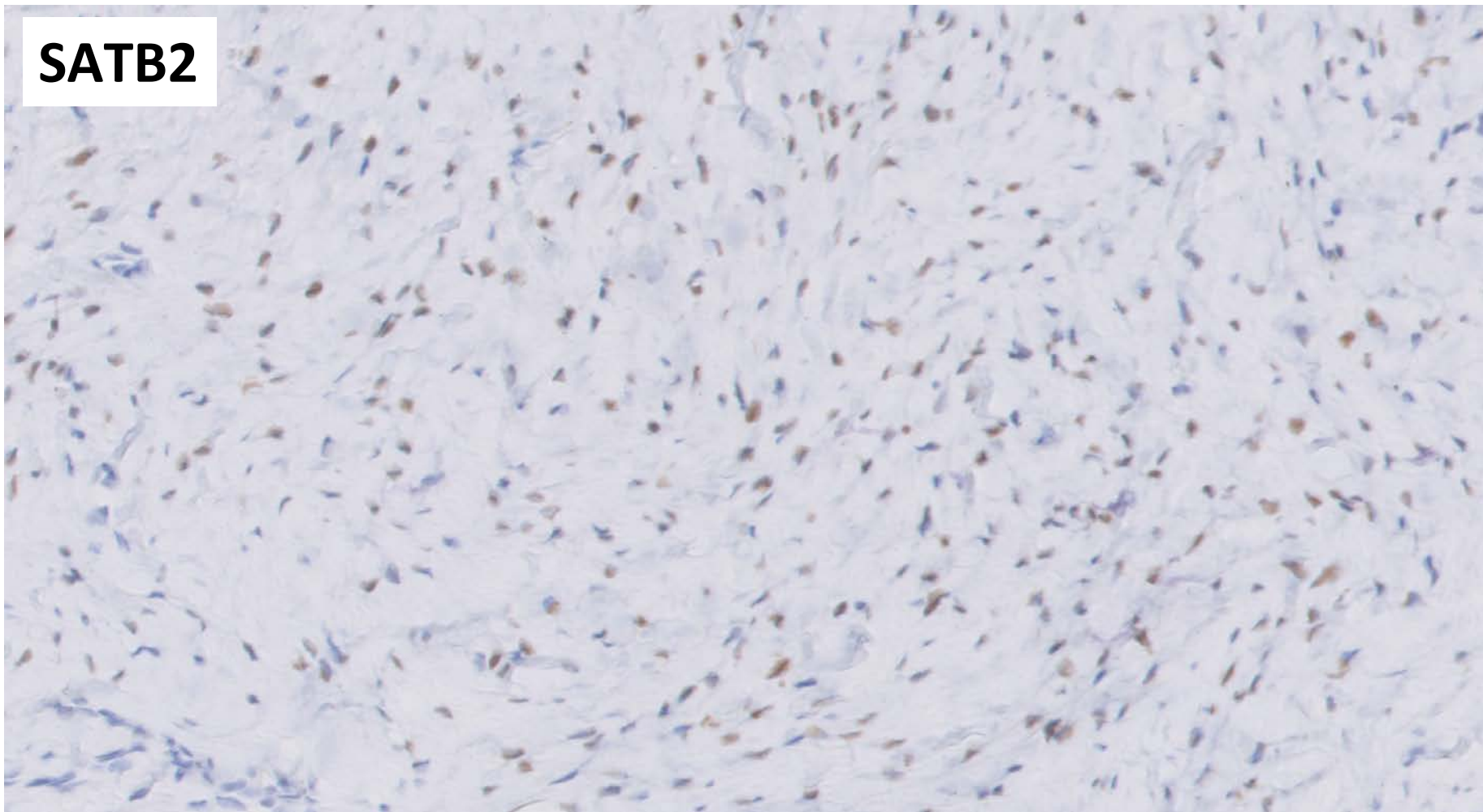




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PATHOLOGICAL FEATURES

SATB2

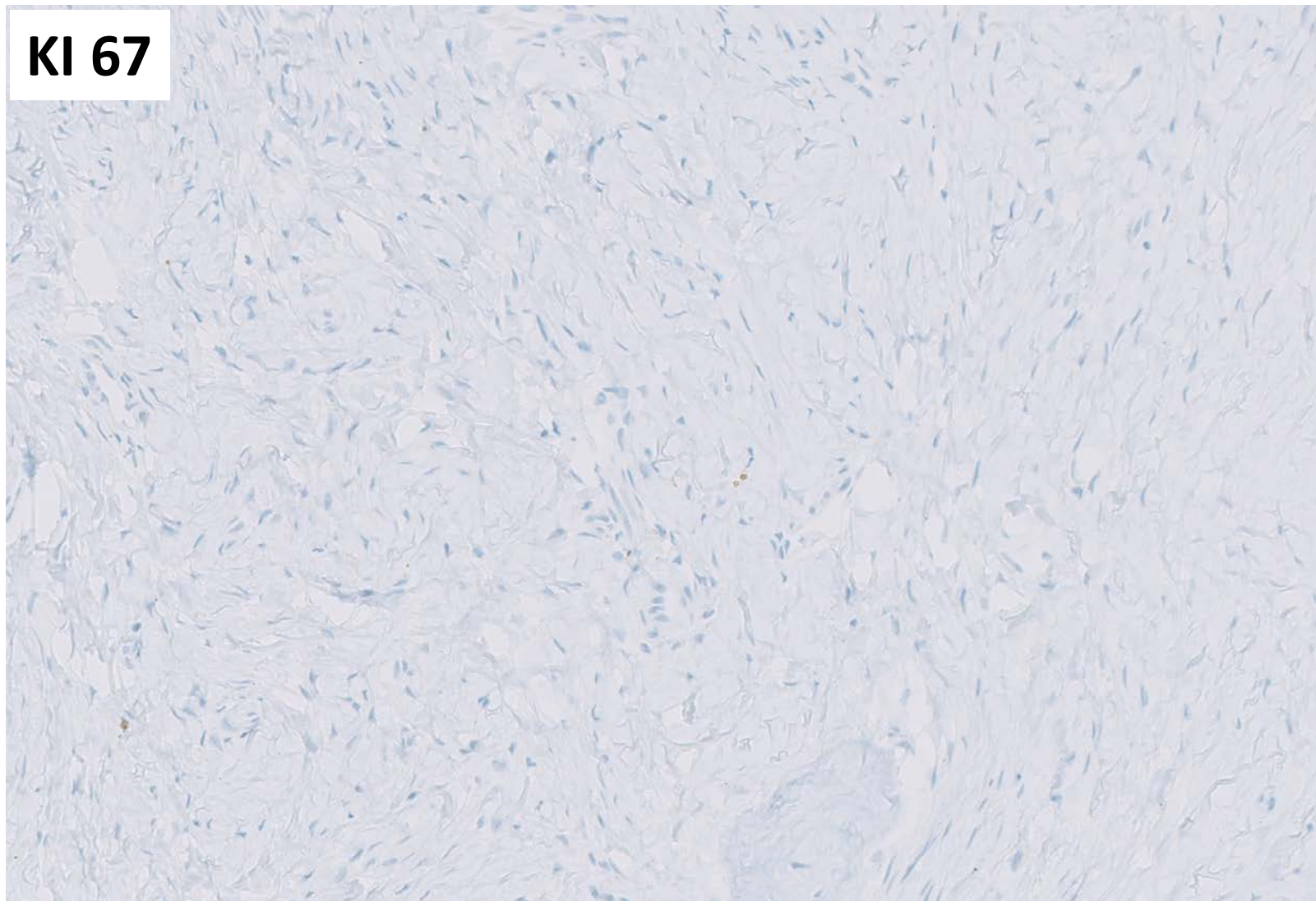




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PATHOLOGICAL FEATURES

KI 67





EVOLUTION

- Patient is doing well
- No pain
- No functional restrictions after curettage
- New MRI programmed soon



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Control X Ray October 2021

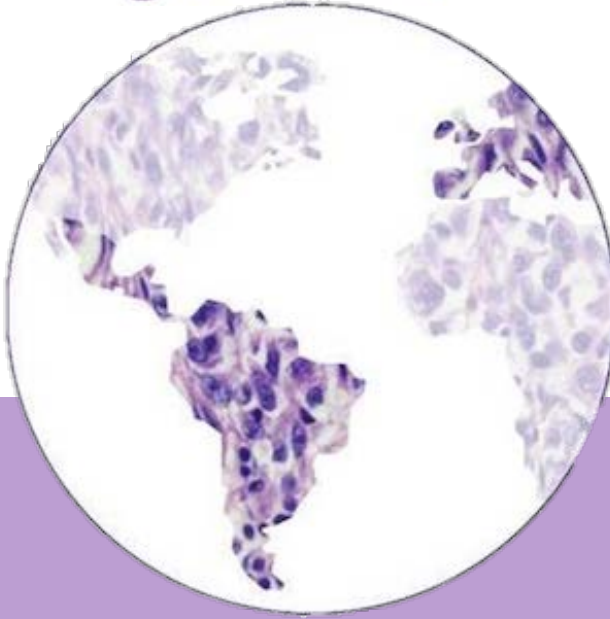




QUESTIONS

- Which is the most probable diagnosis?
- What molecular studies could help in the differential diagnosis?
- Has curettage been enough?
- In addition to a close follow-up, is there any therapeutic maneuver needed?

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THANK YOU!

NEXT MDT: **15 December 2021**, 16:00-17:00 CET
COORDINATOR: CLB