# SELNET MDT

**December 20th 2022**

Armelle DUFRESNE, Medical Oncologist
Marie KARANIAN, Pathologist
Jean-Yves BLAY, Medical Oncologist

Centre Léon Bérard
Lyon, France

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CENTRE DE LUTTE CONTRE LE CANCER LEON BERARD

# SELNET MDT - Dec. 20th 2022 - AGENDA

| Case # | Diagnosis/Hypothesis | Team | MD | Slide # |
|---|---|---|---|---|
| 1 | Leiomyosarcoma | Instituto de Cancerologia Las Américas, Colombia | Maycos Leandro Zapata M | 3 |
| 2 | High grade intimal sarcoma | Instituto de Cancerologia Las Américas, Colombia | Maycos Leandro Zapata M | 9 |
| 3 | Ewing sarcoma | A.C.Camargo Cancer Center, Brazil | Fernando Campos | 15 |
| 4 | Malignant Peripheral Nerve Sheath Tumor | Hospital Clinico San Carlos, Spain | Jorge de la Macorra Mata<br>Gloria Marquina | 26 |
| 5 | Leiomyosarcoma | Centro de Oncologia - Clinica del Country, Colombia | Juan Carlos Velásquez | 41 |
| 6 | Synovial sarcoma | Instituto Nacional de Câncer (INCA), Brazil | Bruna David | 47 |

**Case #1**

Maycos Leandro Zapata M

Instituto de Cancerologia Las Américas

Colombia

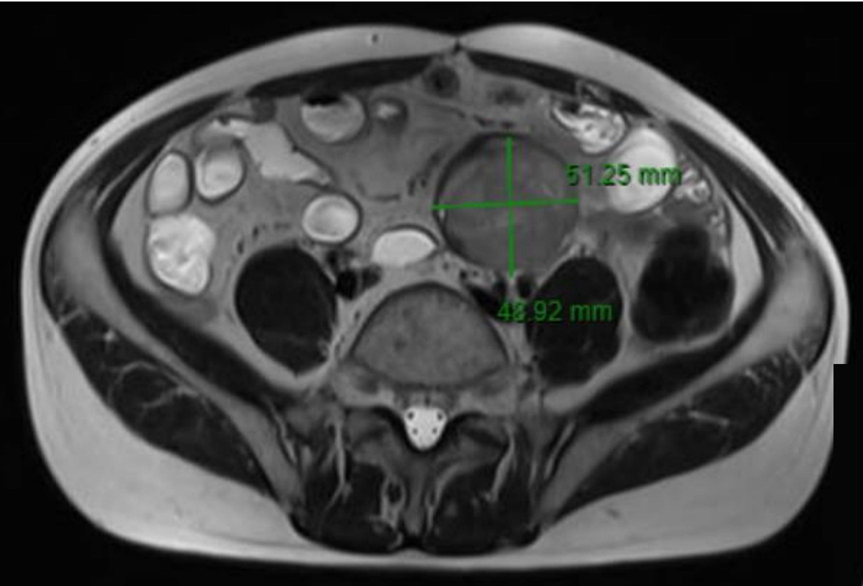
# MTB Selnet December 2022

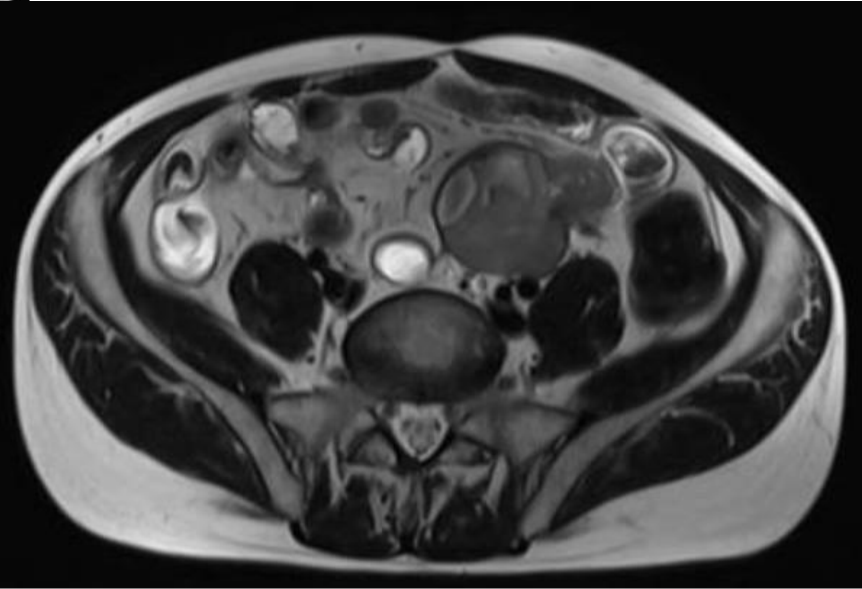
Maycos Leandro Zapata M
Medical Oncology
IDC Auna – Las Américas
Internal Medicine UdeA

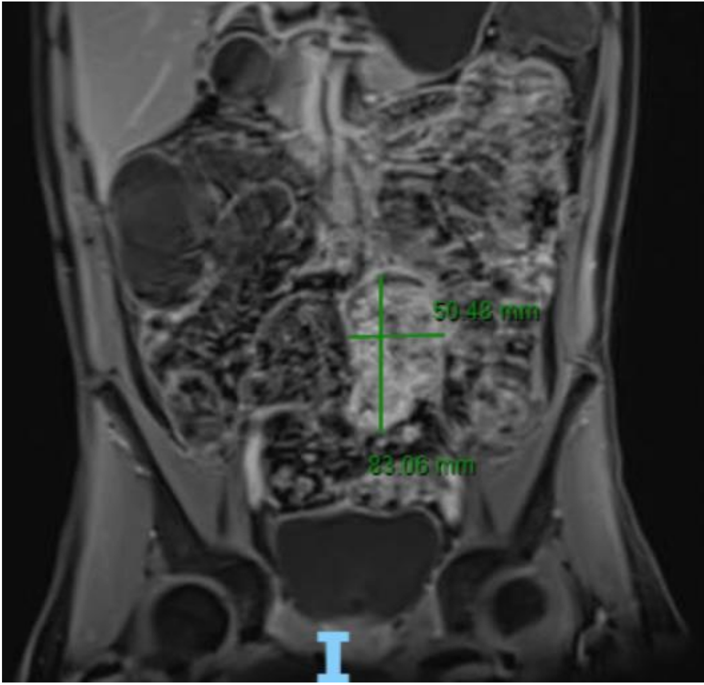
# 44yo Male

- At 19yo (1997)
  - Retroperitoneal NH Lymphoma
    - High intensity chemotherapy (with 300mg/m2 adryamicin cumulative dose)
    - Adjuvant radiotherapy. With unknown dose
- Now presented with abdominal pain in aug 2022

# 44yo Male





# 44yo Male

- A resection was performed in another center
  - 04 nov 2022: In-block with small bowel resection
  - No complications
- Pathology reports
  - “Solid lesion with well defined borders 9.5x7x6.5cm without bowel involvement”
  - “Cells with eosinophilic cytoplasm, pleomorphic nuclei. Contact with the bowel surface without invasion”
  - “High grade leiomyosarcoma: SMA positive, H-caldesmon positive, TLE-1 positive, S100 negative, DOG1 negative, HMB45 negative, CDk4 negative, MDM2 negative”

# 44yo Male

- Questions
  - Until STRASS2, the current treatment for this “high-risk” abdominal leiomyosarcoma
  - If you consider chemotherapy, any issue with a 200mg/m2 25 years ago
- institutional MTB
  - At least 3 cycles of Adryamicin 75mg/m2 plus DTIC 1gr/m2 with intensive cardiac monitoring
  - Observation
  - Alternative 4 cycles of monotherapy Ifosfamide 9gr/m2 per cycle

**Case #2**

Maycos Leandro Zapata M

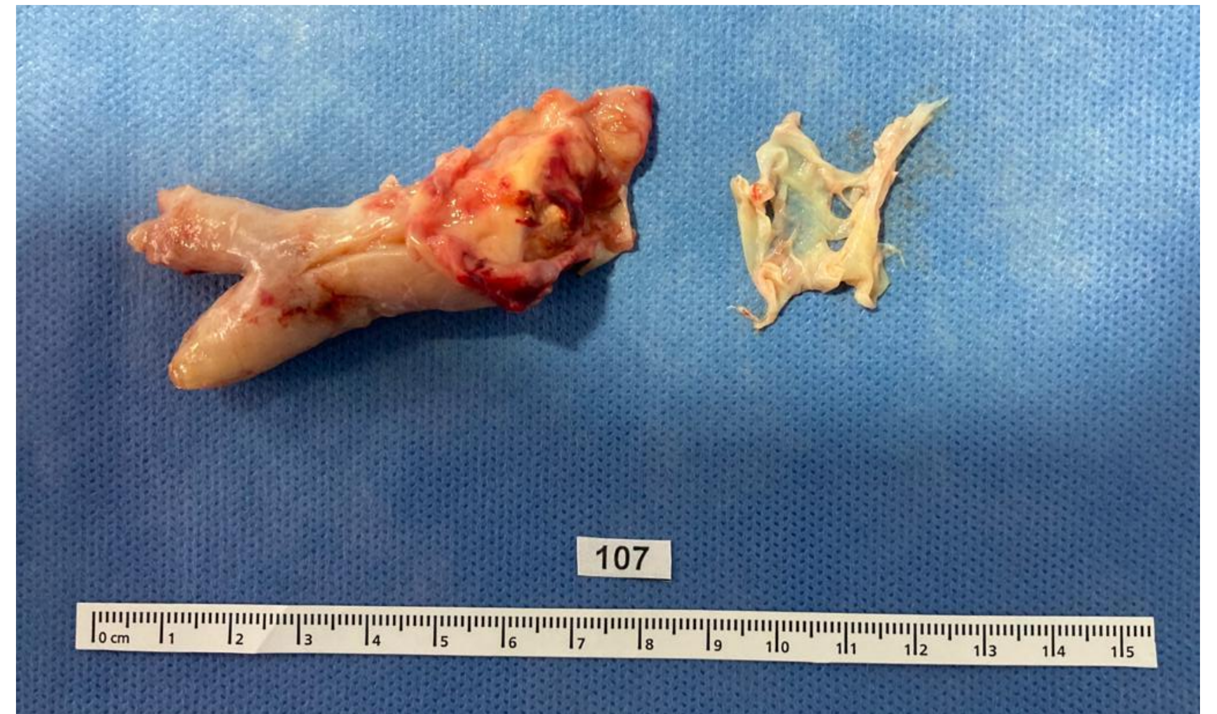
Instituto de Cancerologia Las Américas

Colombia

# 32yo Female

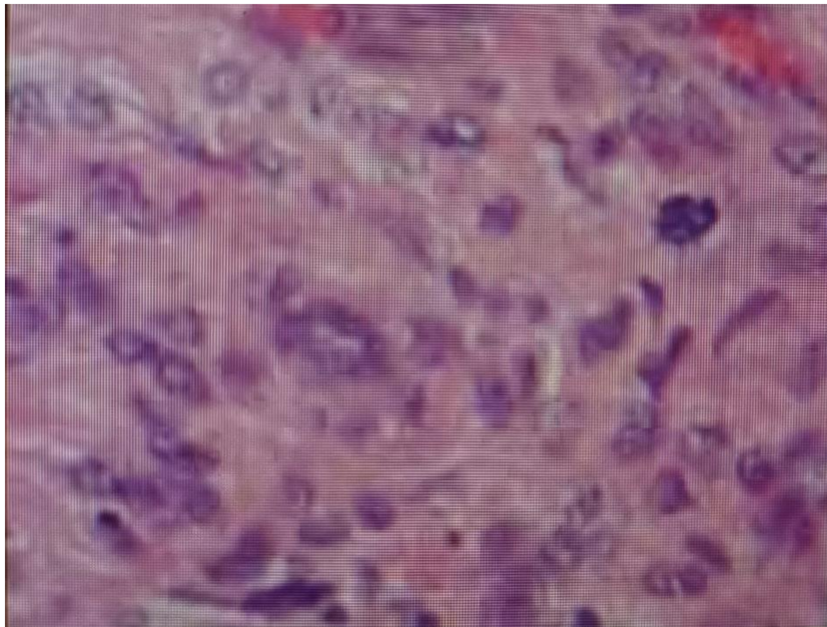
- Chronic pulmonary embolism
suspected because of dyspena
  - In context of antiphospholipid syndrome

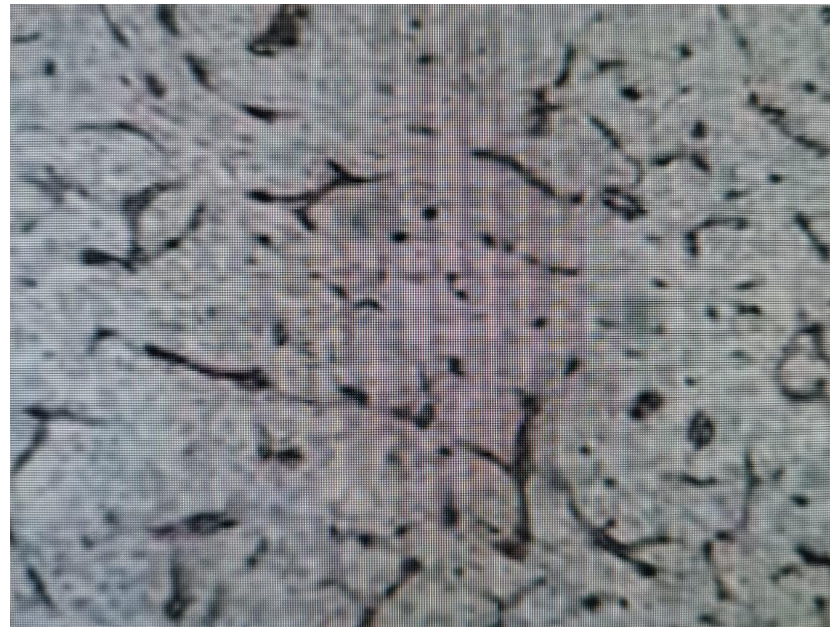
- 27/sep/2022 Pulmonary
Thromboendarterectomy
In a cardiovascular center



# 32yo Female

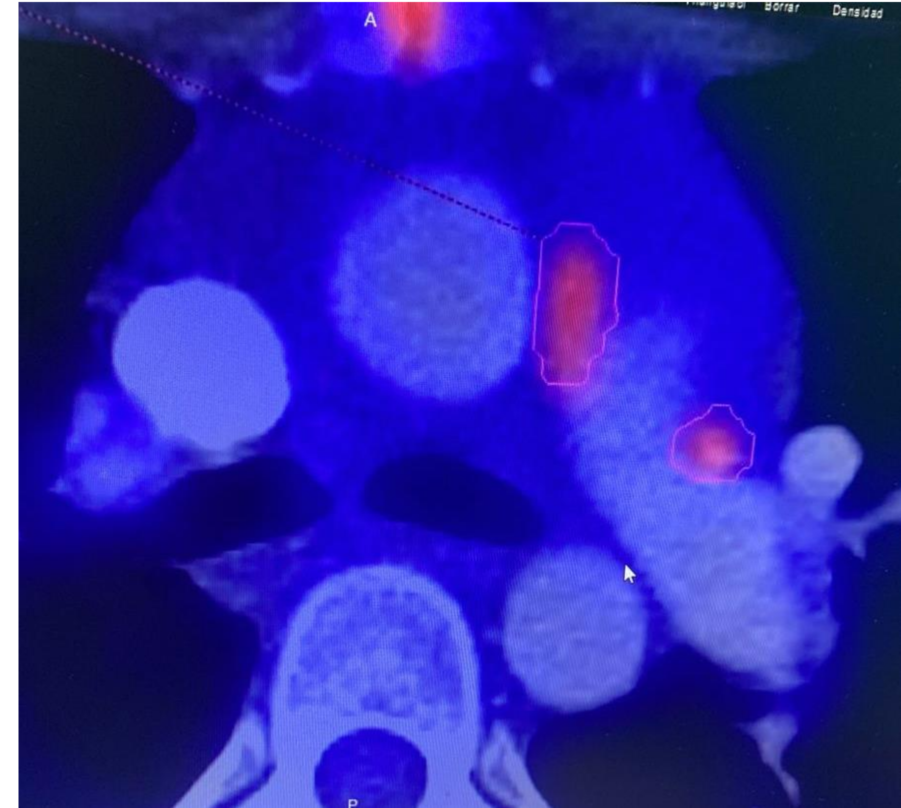
- Initial Pathology report: “intraluminal angiosarcoma”  T 6*1.7*1.6cm
- FLI1+, CD31+, CD34+, ERG+, Factor VIII+. SMA+ focal, calponina + focal, Hcaldesmon +focal, CLD2 + focal. Ki67 5%





# 32yo Female

- Expert pathologis review: High grade intimal sarcoma.
  - Vascular stains (CD34, ERG, CD31, FLI1, Fc VIII) positive in endotelial cells but negative in tumor cells.
  - Tumor cells SMA+, calponin+, BCL2+, Ki67 10%
- 10/nov/2022 PET CT: superior wall right Pulmonary A. Suvmax 4.57, metabolic volumen 5 cm3 without anatomic representation with/without IV contrast, in the place where the tumor were. Another zone suvmax 3.6 anterior Wall right pulmonary A. with same caracteristics.



# 32yo Female

Questions:

- Do you have any case similar?
- Do you consider adjuvant treatment in these patient
- Do you consider adyuvant radiotherapy

- MTB propose Adryamicin plus ifosfamide x4 cycles

# Muchas gracias

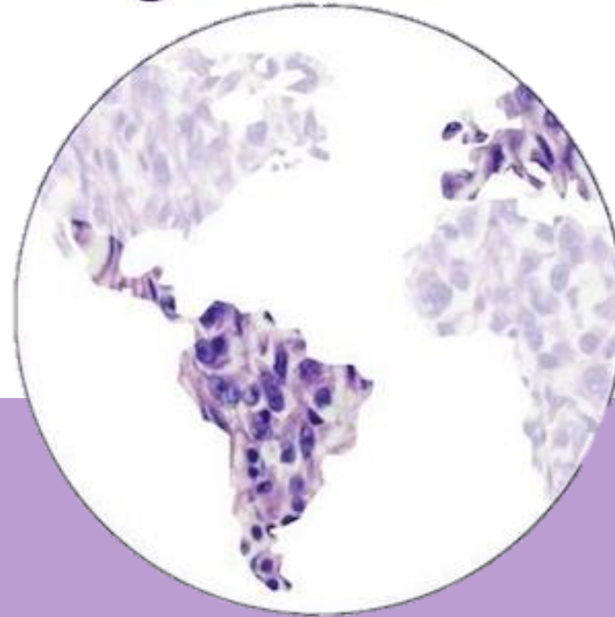
Maycos Leandro Zapata M
Oncología Clínica
IDC Auna – Las Américas
Internal Medicine Department UdeA
Maycos.Zapata@udea.edu.co

## Case #3

Fernando Campos

A.C.Camargo Cancer Center

Brazil




# Virtual MDT Board

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

December 2022

- 40 yo male

- No comorbidities. No family history of cancer.

**ONCOLOGICAL HISTORY:**

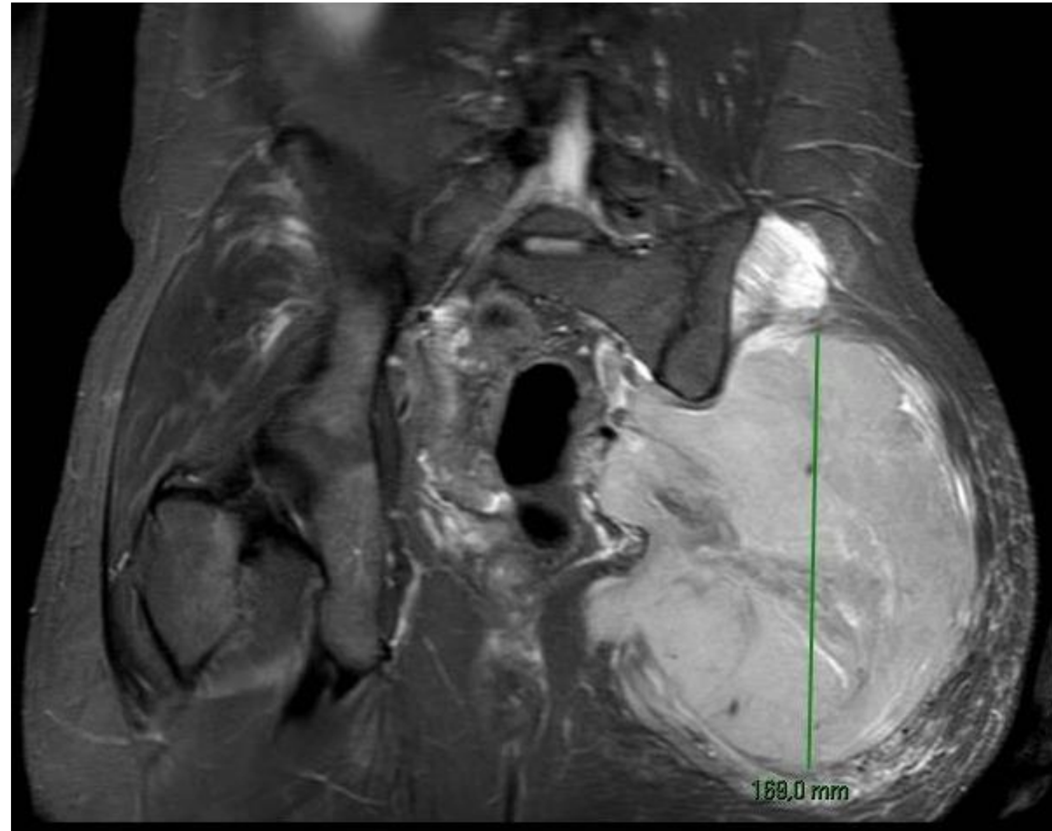
- March 2020: Mild pain in the left gluteal region, which progressively intensified, associated with local swelling. Evolved with functional impairment and weight loss in February 2021.

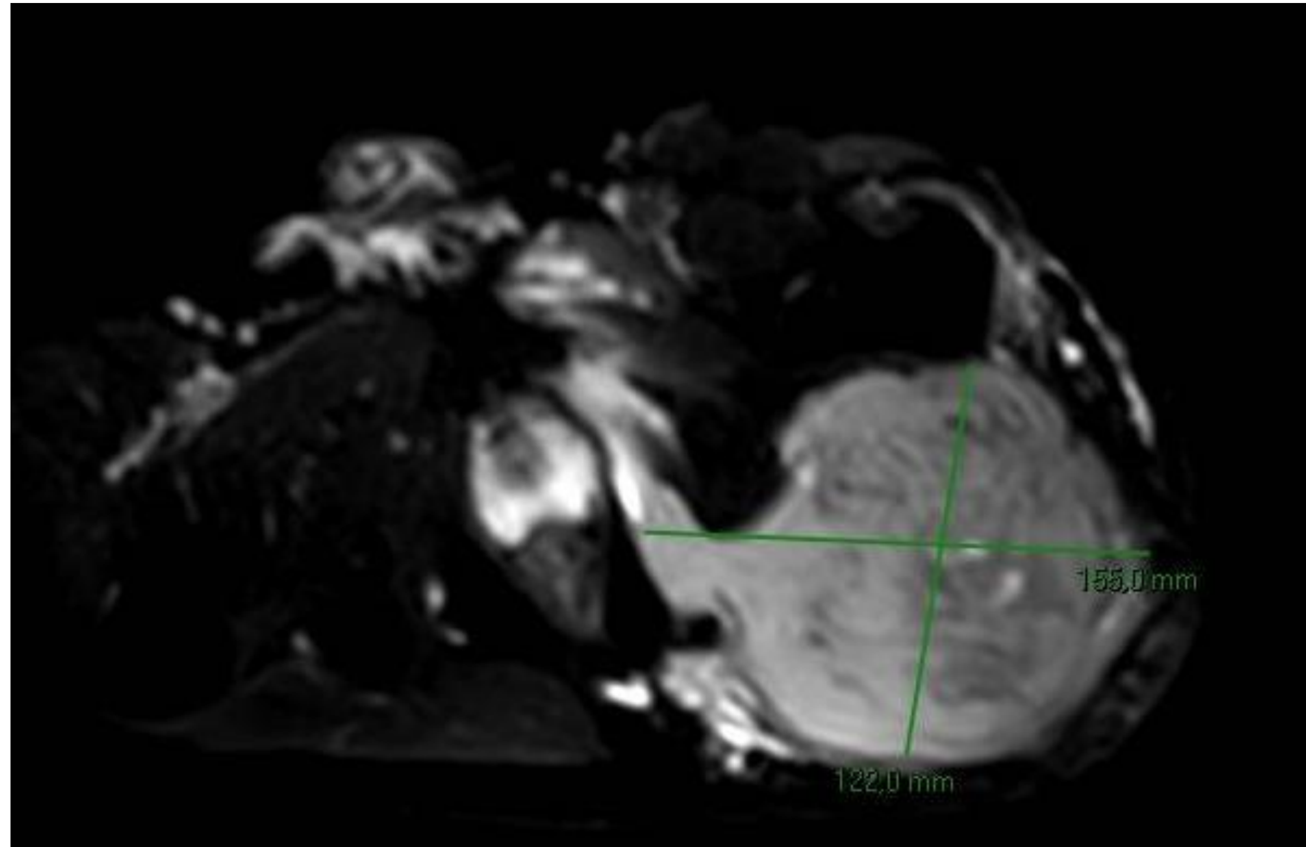
- March 2021: Pelvic MRI - mass in the musculature of the left gluteal region.

- April 2021: Core biopsy: **Ewing Sarcoma**

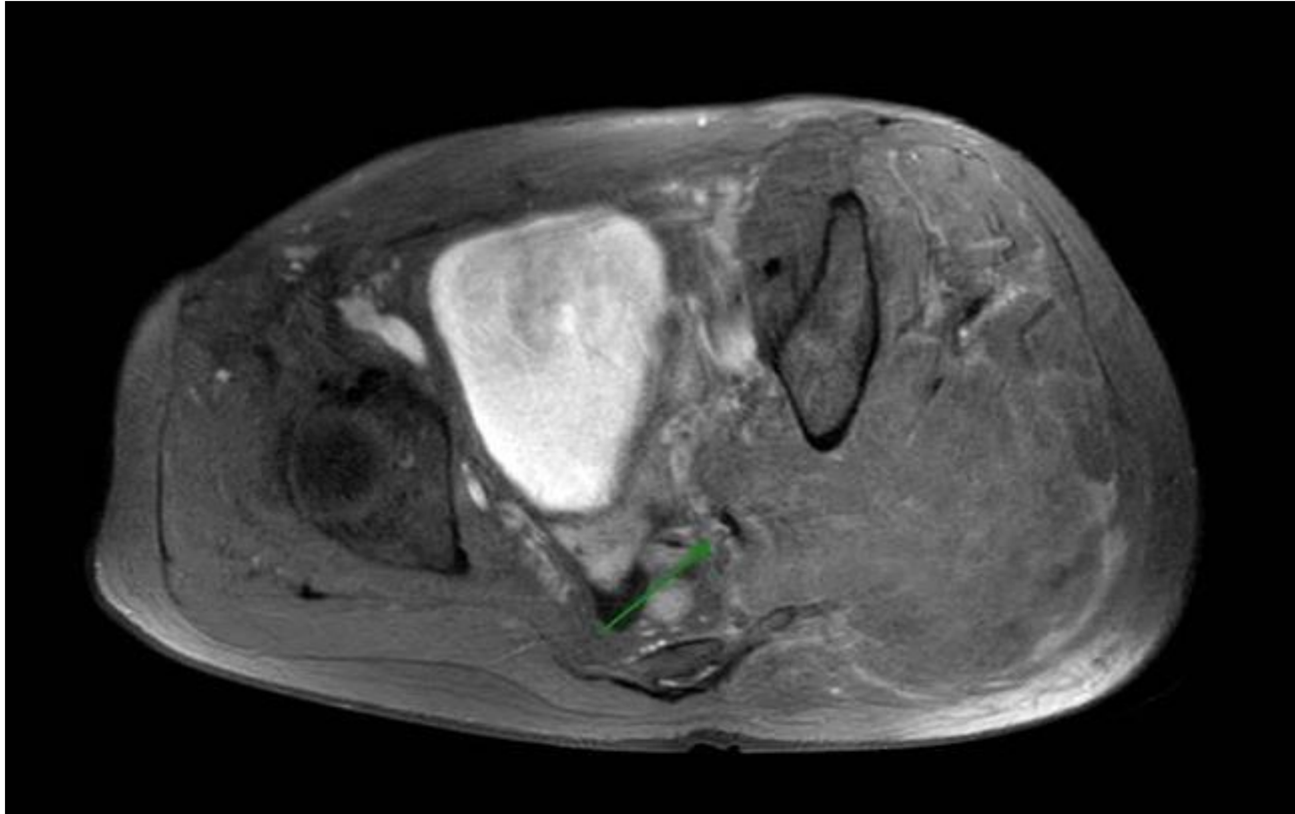
- May 2021: Pelvic MRI – **Mass in the soft tissues of the left gluteal region**, affecting the gluteus maximus, medius, minimus, piriformis and internal and external obturators muscles, extending into the left pararectal pelvic cavity, through the sciatic foramen and in the ischiorectal fossa region , involving the sciatic nerve and in close contact with the left internal iliac vessels. It has intimate contact with the ischial tuberosity, iliac body, posterior segment of the acetabulum, proximal femur up to the subtrochanteric region and sacrum on the left from S3. This lesion presents with hyposignal on T1, hypersignal on T2, heterogeneous contrast enhancement and diffusion restriction, **measuring approximately 178x146x144mm (CC x LL x AP)**

# Virtual MDT Board





155,0 mm
122,0 mm

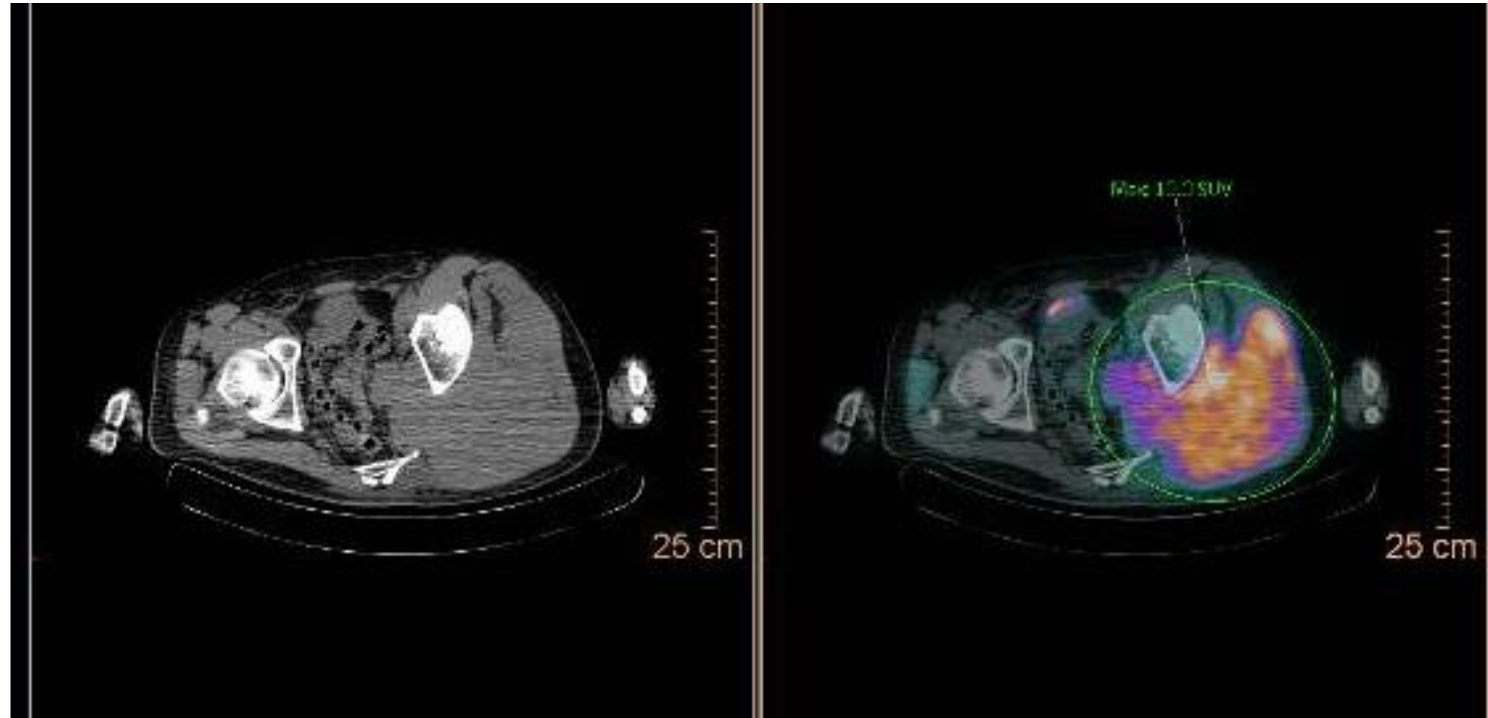


# Virtual MDT Board

- April 2021: Chest CT – no metastasis

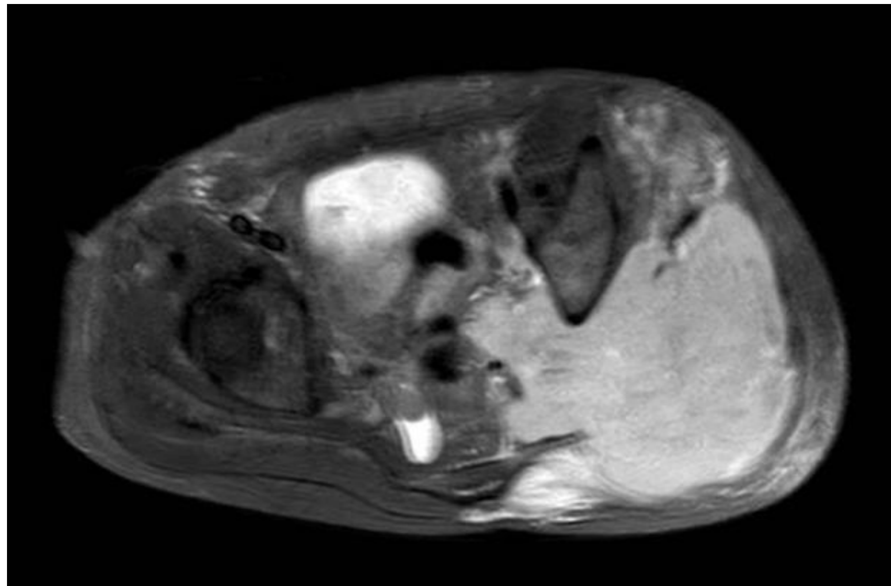
- May 2021: Tumor Board – to perform PET Scan, and if localized disease, induction chemotherapy, local treatment with radiotherapy, and consolidation chemo.

- May 2021: PET Scan – localized disease

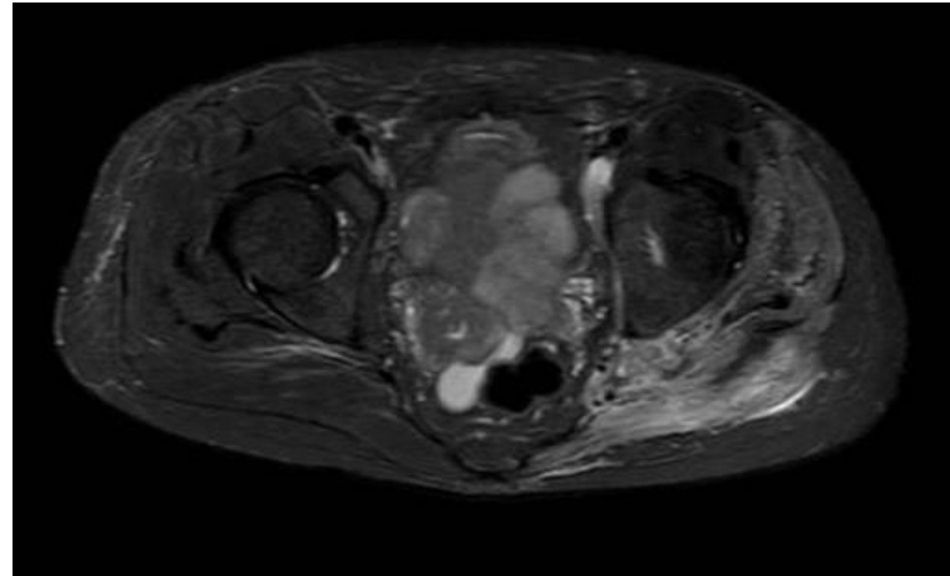


- May to Aug 2021: 4 cycles VAC/IE (3 weeks interval)
- After cycle 4: partial response

**05/04/2021**



**08/16/2021**



# Virtual MDT Board

- Aug to Oct 2021: + 3 cycles VAC/IE (3 weeks interval)

- October 2021: IMRT 45Gy (25 x 180 cGy) in pre chemo volume + 14,4Gy (8x180cGy) in affected muscle group.

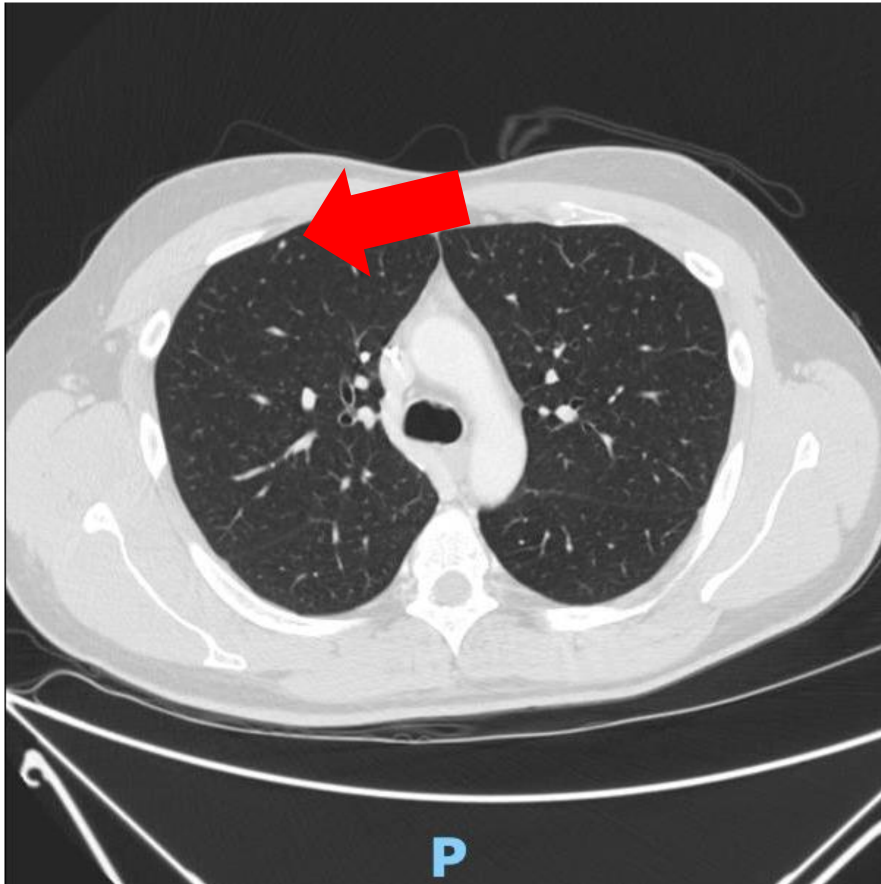
- May 22: completed 17 cycles VAC/IE >> Follow up

- November 22: lung nodule. Asymptomatic.
  No metastatic disease in abdomen CT and bone scan. No local relapse.

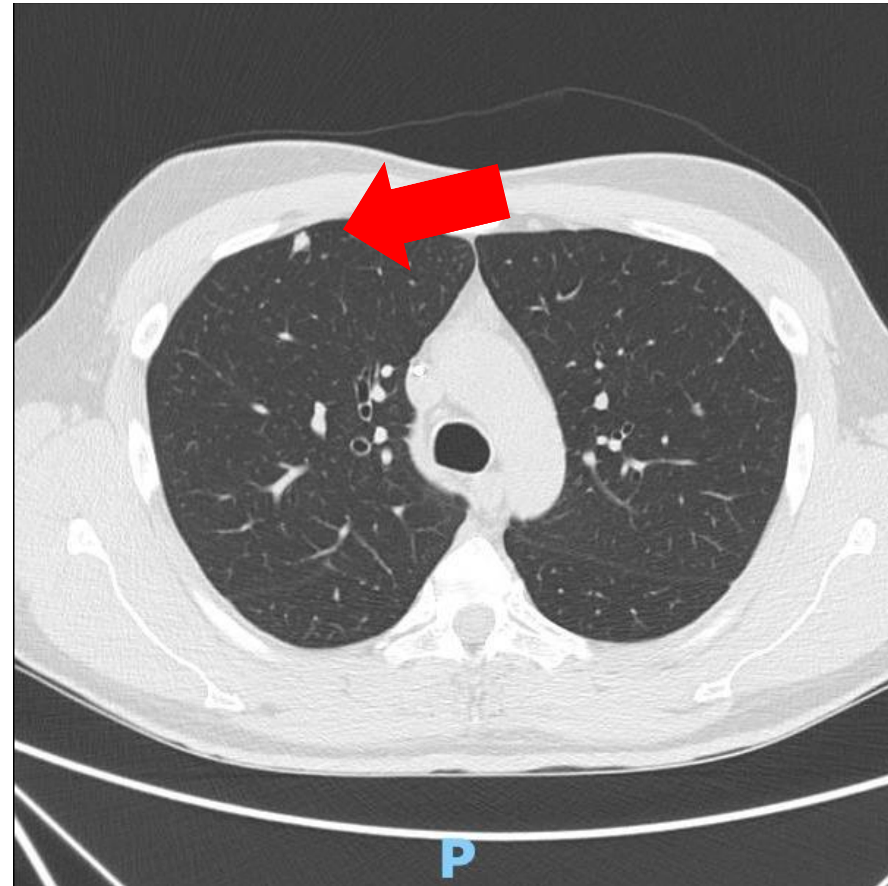
- December 22: lung biopsy – Ewing sarcoma metastasis

# Virtual MDT Board

**August 2022 (4 mm)**




**November 2022 (9 mm)**



**DISCUSSION: TREATMENT**

HD ifosfamide?

Total lung irradiation if response?

PET Scan?

**Case #4**

Jorge de la Macorra Mata / Gloria Marquina

Hospital Clinico San Carlos

Spain

# JMGG

Dr Jorge de la Macorra Mata / Dr Gloria Marquina

Resident in training / MD, PhD

Medical Oncology Department

SELNET MDT 20th December 2022

Hospital Clínico San Carlos
SaludMadrid

European Reference Network for rare or low prevalence complex diseases
Network Adult Cancers (ERN EURACAN)

IdISSC INSTITUTO de INVESTIGACIÓN SANITARIA Hospital Clínico San Carlos

# JMGG

68-year-old male

## Past medical history

**Stage … c**olon cancer in 2010 treated with surgery.

## Social history

ECOG 1 (Post-surgical right elbow **paresia**).

# Oncological history

## 1. Local hospital

Study workup of right cervical discomfort:

- **CT scan (03/May/22)** : 4 cm right cervical tumor.
- **Biopsia via right cervicotomy (06/June/22):** Malignant Peripheral Nerve Sheath Tumor, epithelioid variant.
- **Cervical MRI (30/June/22):** 36 x 29 x 54 mm (T x AP x CC) right cervical mass which affected / invaded / infiltrated the C4-C5 junction foramen.

# Oncological History

## 2. Transfered to another institution (not sarcoma referral centre)

**Surgery (01/August/22):** excision of the tumor lesión
with right cervical dissection.
The lesion infiltrated the right spinal nerve, cervical sympathetic nerve,
and roots of the brachial plexus at C5-C6 junction foramina.
**R1 surgery.**

MALIGNANT PERIPHERAL NERVE SHEATH TUMOR WITH SOFT TISSUE INFILTRATION.
2 **OUT OF 16 LYMPH NODES AFFECTED**

# Oncological History

## 3. Hospital Clínico San Carlos National referral centre for sarcoma(CSUR)

**MULTIDISCIPLINARY COMMITTEE (08/Sept/22) :**

Re-staging

Sequential chemo-radiotherapy.

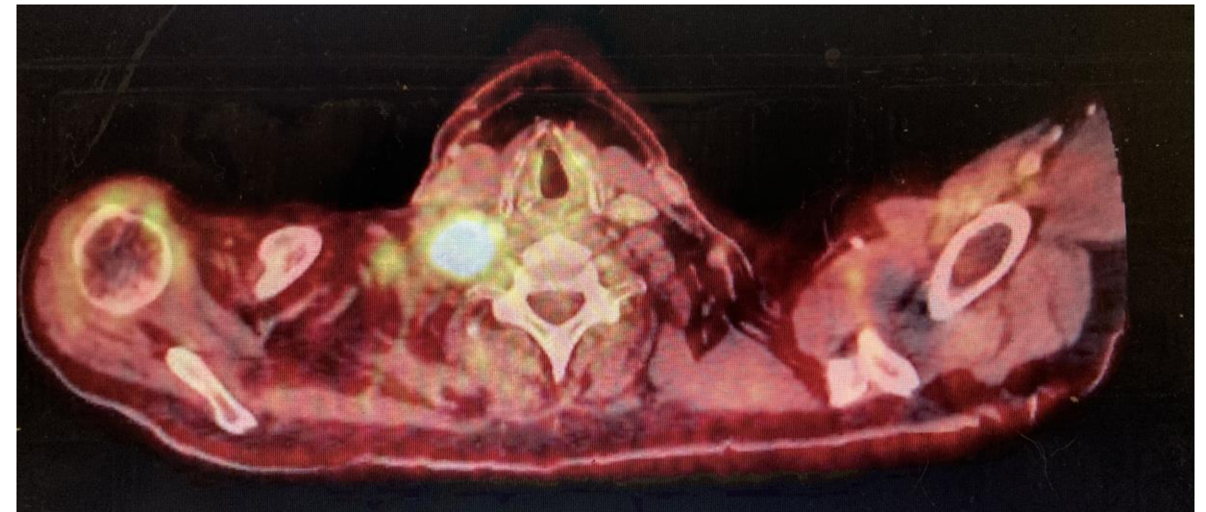
# Oncological History

## 3. Hospital Clínico San Carlos National referral centre for sarcoma (CSUR)

**MULTIDISCIPLINARY COMMITTEE (08/Sept/22)** :

Re-staging

Sequential chemo-radiotherapy.



**PET-CT (Sep/22)**:

persistence of local tumor disease.

**MULTIDISCIPLINARY COMMITTEE (06/Oct/22)** :

Chemotherapy --> surgery → radiotherapy

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# Oncological History

## 3. Hospital Clínico San Carlos National referral centre for sarcoma (CSUR)

**Cycle 1 (10-12/Oct/22):** EPIRRUBICIN (60 mg/m2 days 1-2) with IFOSFAMIDE (3000 mg/m2 days 1-3).

- **Toxicity: G1** Cheilitis, **G1** Asthenia, **G1** Encephalopathy.

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- **Toxicity: G1** Cheilitis, **G1** Asthenia, **G1** Encephalopathy.

**Cycle 2 (8-9/Nov/2022 - Hospitalized):** EPIRRUBICIN (60 mg/m2 days 1- 2) with IFOSFAMIDE (3000 mg/m2 days 1-3).

- **Toxicity:** G3 Anaemia, G4 Thrombopenia, G4 Neutropenia, G3 Encephalopathy, G2 Diarrhoea.

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# Oncological History

## 3. Hospital Clínico San Carlos National referral centre for sarcoma (CSUR)

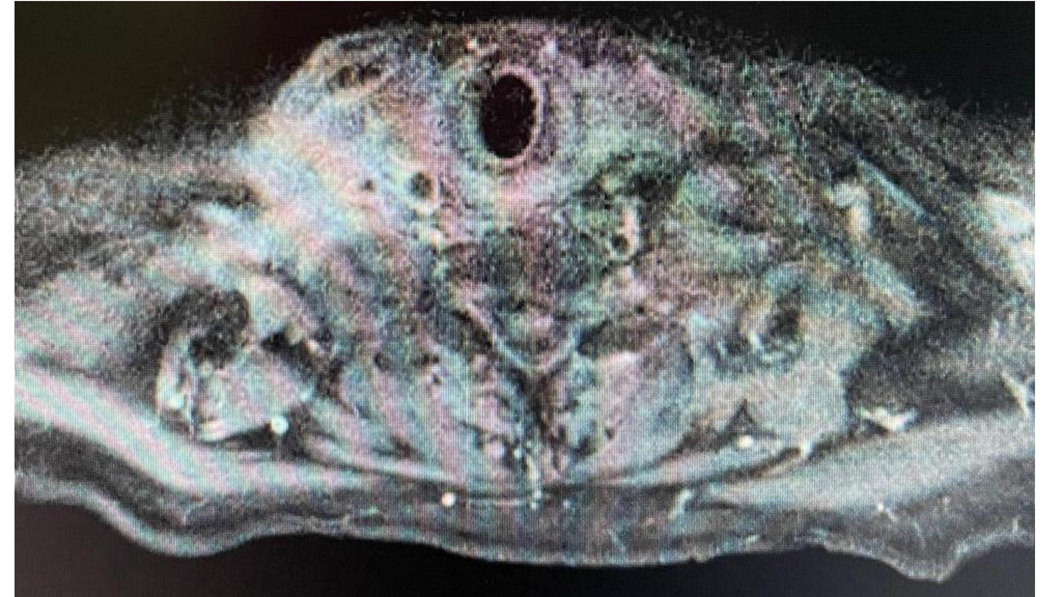
- Prolonged hospital admission (08/Nov/22 - 13/Dec/22)
- Severe Sars-CoV-2 infection – transferred to ITU (20/Nov/22 – 28/Nov/22) high-flow oxygen requirements.

- Once discharged from ITU, the patient was re-staged: cervical MRI and PET-CT.
- Discharged from Medical Oncology Department on 13/Dec/2022
- He currently presents post-Covid pulmonary fibrotic changes and requires daily home oxygen therapy (3-4 litres per minute).

# Oncological History

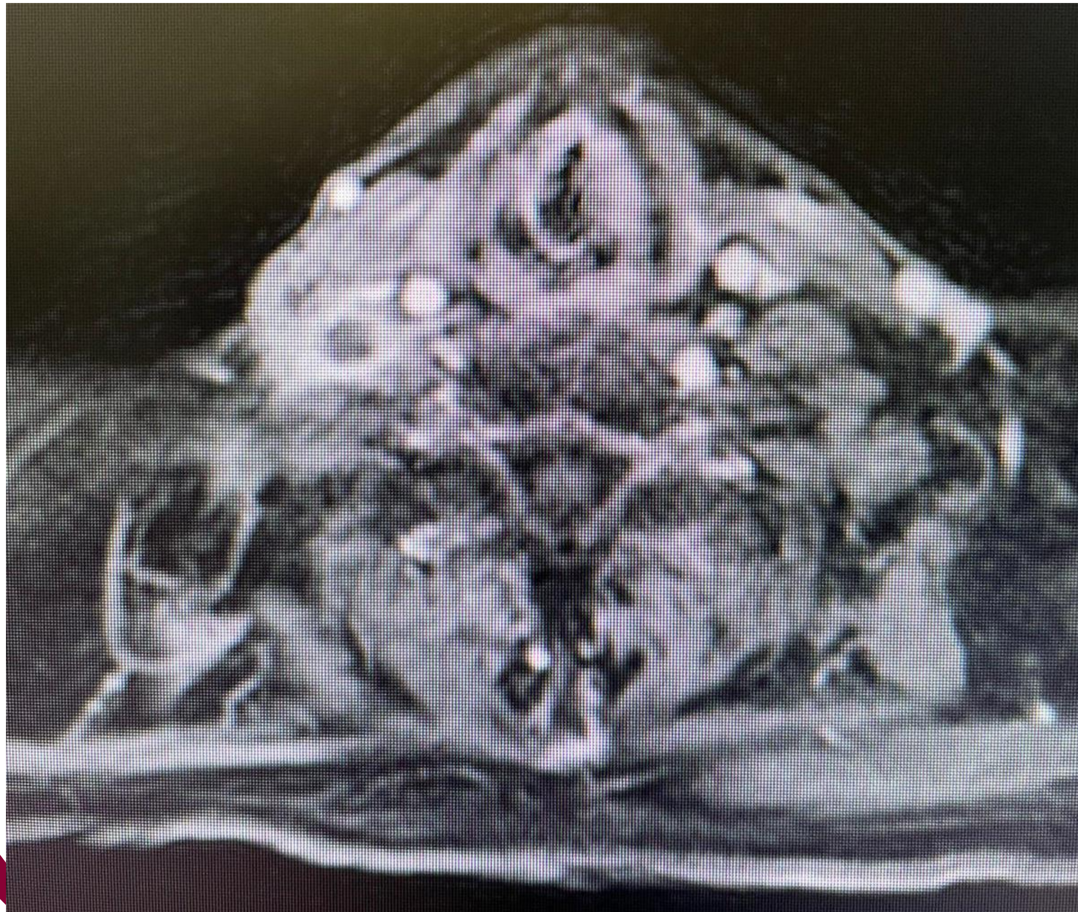
## 3. Hospital Clínico San Carlos National referral centre for sarcoma (CSUR)

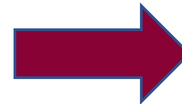
**Cervical MRI (01/Dec/22):** Decreased size (3 cm now) of the right laterocervical lesion compared to PET-CT performed in September 2022

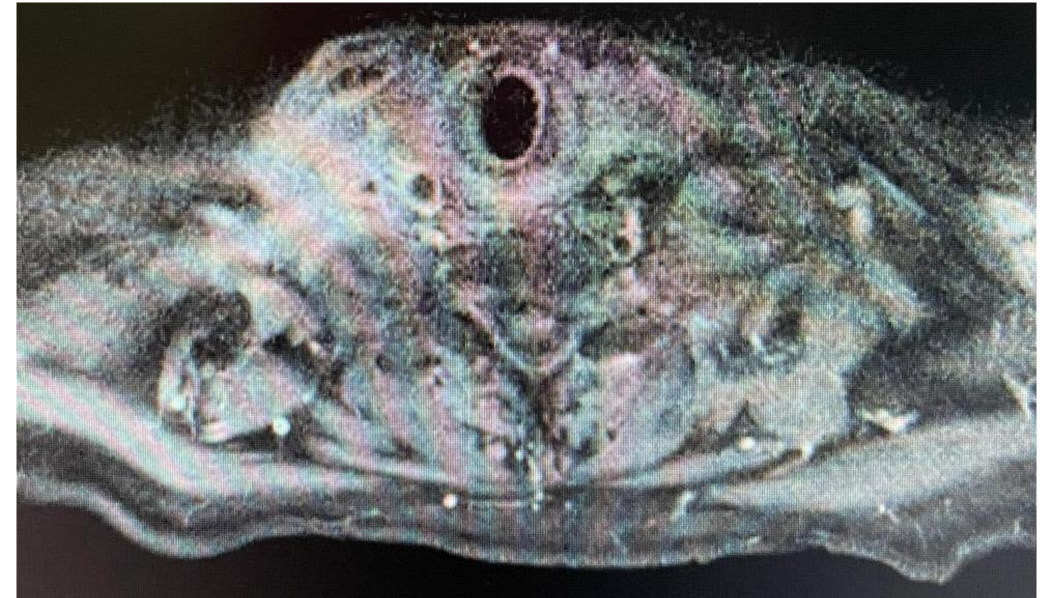


# Oncological History

## 3. Hospital Clínico San Carlos National referral centre for sarcoma (CSUR)







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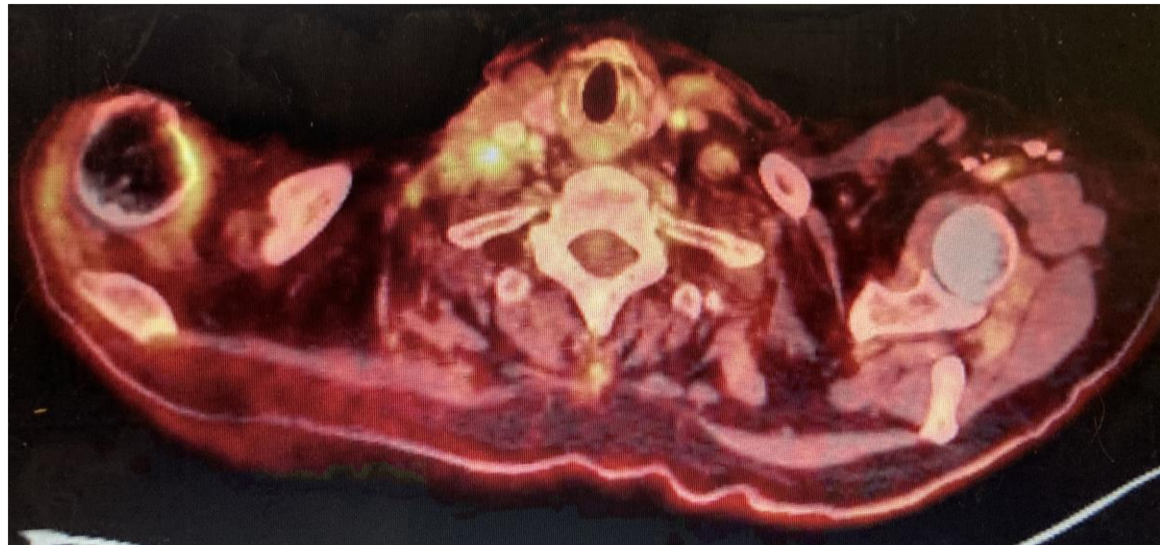
# Oncological History

## 3. Hospital Clínico San Carlos National referral centre for sarcoma (CSUR)

**PET-CT (09/Dec/22):** Significant decrease in the size and the uptake of the right laterocervical adenopathy. 2.3 x 1.6 cm, SUVmax of up to 6.1.

Sars-CoV-2 pulmonary changes still present.



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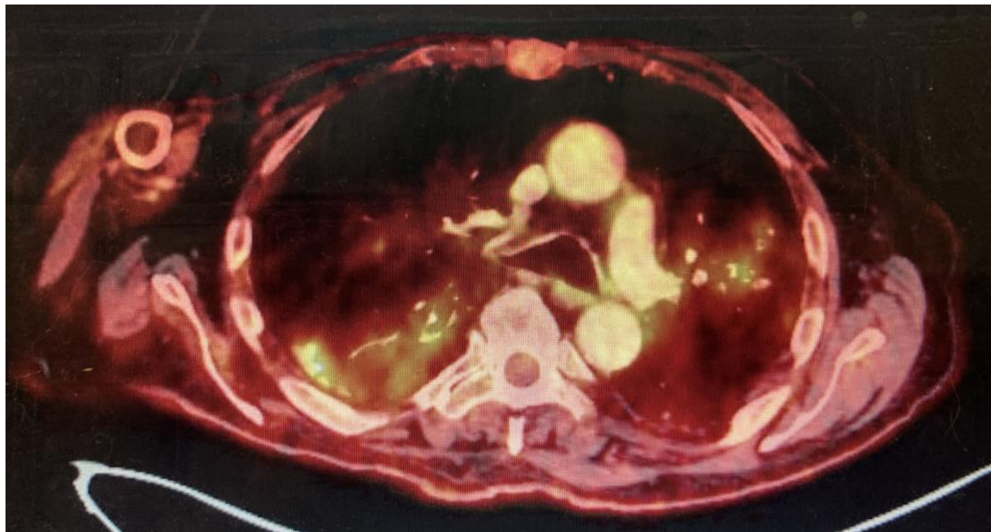

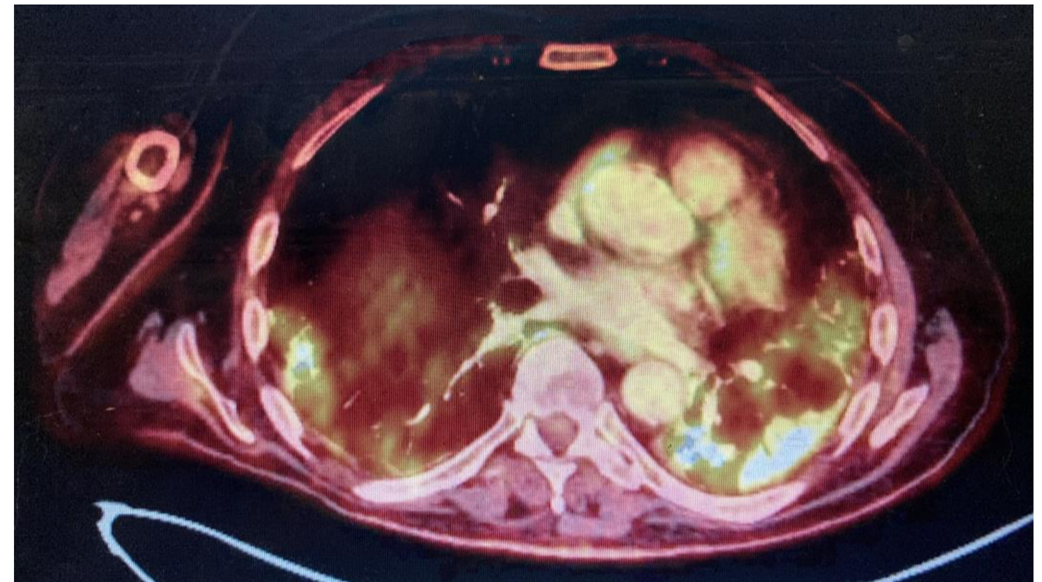

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# Oncological History

## 3. Hospital Clínico San Carlos National referral centre for sarcoma (CSUR)

Sarcoma MDT 14/Dec/22: Postpone surgery (patient is not fit at the moment… wait at least 8 weeks from Sars-CoV-2 infection…

Selnet MDT question: What should we offer to the patient?
Doxorrubicin monotherapy for additional 2 cycles until the patient is fit for surgery?
Wait for surgery without administering any systemic treatment?

**Case #5**

Juan Carlos Velásquez

Centro de Oncología -  Clínica del Country

Colombia

# Clinical case

A 57-year-old female patient started with colicky abdominal pain radiated to the in the right dorsal region, and to the right iliac fossa, predominantly at night, associated with weight loss (-8 kg) since April 2022.

In November 2022, a MRI revealed a 28 mm x 20 mm mass in the inferior vena cava, below the renal veins, with no other lesions.

18 FDG PET /CT comfirmed the presence of a hypermetabolic mass in the infrarrenal inferior vena cava (SUV 6.66), with no other abnormalities

Vascular resection was performed on 11/21/2022, and a latter reintervention was needed because a 1000cc retroperitoneal hematoma on 11/24/2022.

## Pathology report, Fundación Cardioinfantil, Bogotá, Colombia – Dr Hugo Herrera, on 11/21/2022

Moderately differentiated leiomyosarcoma.
-Malignant spindle cell neoplasia tumor of probable mesenchymal lineage of the vascular wall.
-Size: 3.2 x 2.8cm
- Necrosis. Focus present 10%
- Mitotic activity more than 30 mitoses in 10 high power fields.
- Grade 2
- Extension of the invasion: vascular wall and focally to the soft tissue of the adventitia.
- Free edges
- External surface edge of the vena cava: focally in contact with the tumor.
- 8 pericave lymph nodes, all free of tumor

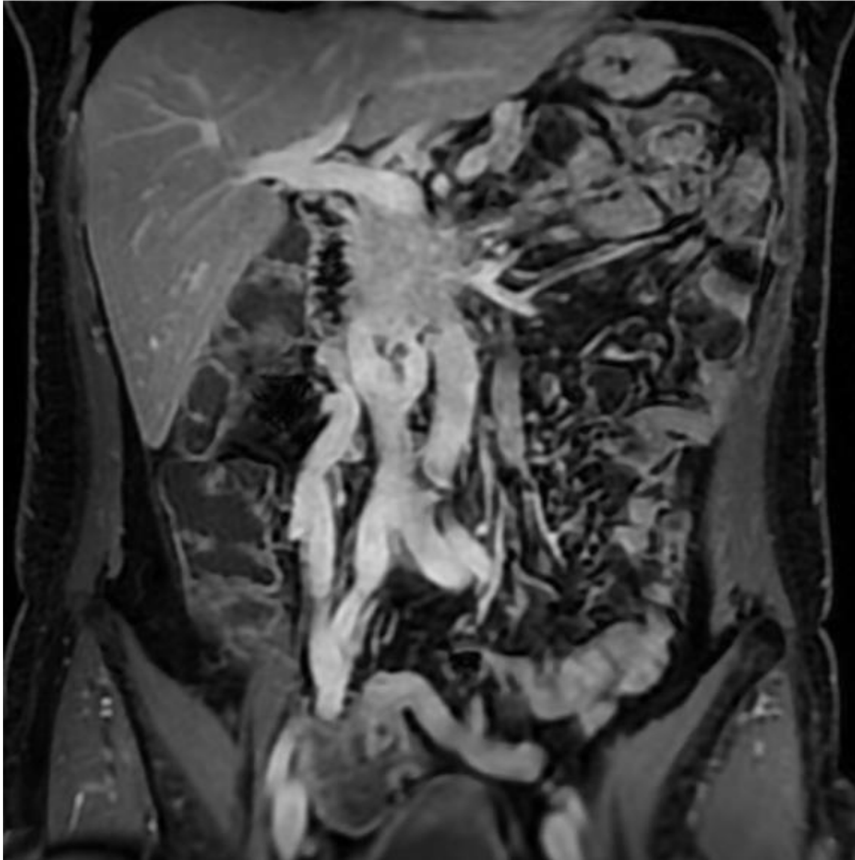
Immunohistochemistry:
(+): Caldesmon, SMA, focal with desmin
(-): MDM2, CDK4.

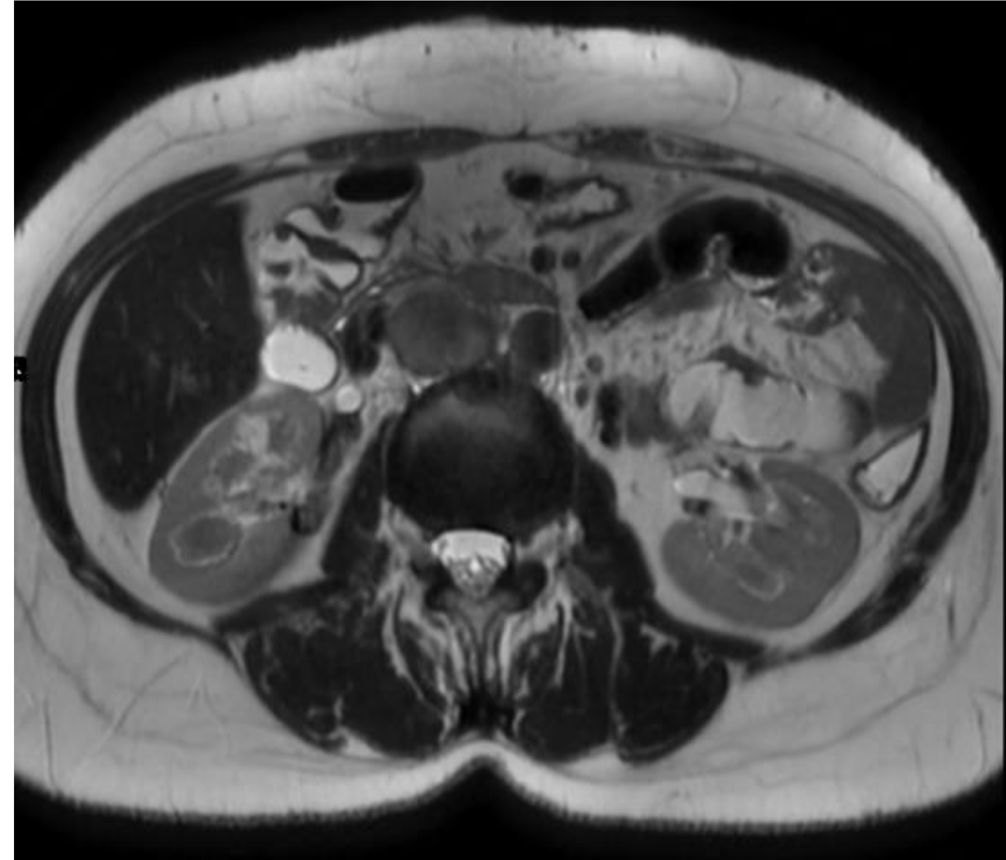
## Revision, Hospital Universitario San Ignacio, Bogotá, Colombia, Dr. Oscar Messa, on 15/21/2022

Inferior vena cava tumor
Histological grade conventional leiomyosarcoma (French Federation of Cancer Centers Sarcoma Group [FNCLCC]): Grade 2 (High grade) (D: 2 M: 2 N: 1 5/8)
Tumor size: 3 cm
Necrosis present: 30%
Mitotic activity more than 6 mitoses in 10 high power fields.
No evident vascular invasion
Margins without tumor involvement
Reactivity in tumor cells with Caldesmon, focal SMA with desmine
No reactivity in tumor cells with MDM2, CDK4
KI67: 30%
8 lymphatic nodes without tumor involvement (0/8)

Clínica del Country | Clínica La Colina

# Entero resonance, Fundación Santa Fe, 24.10.2022:

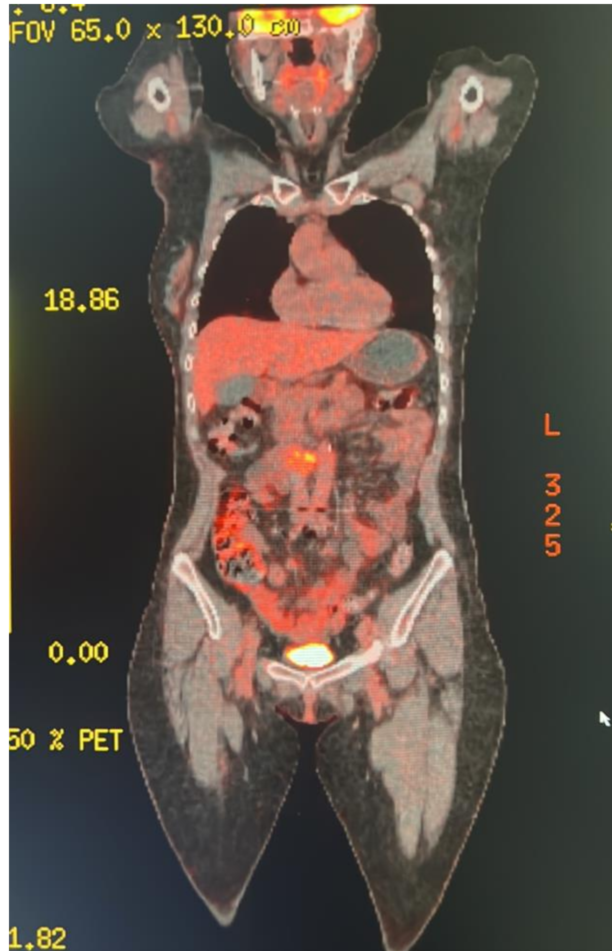


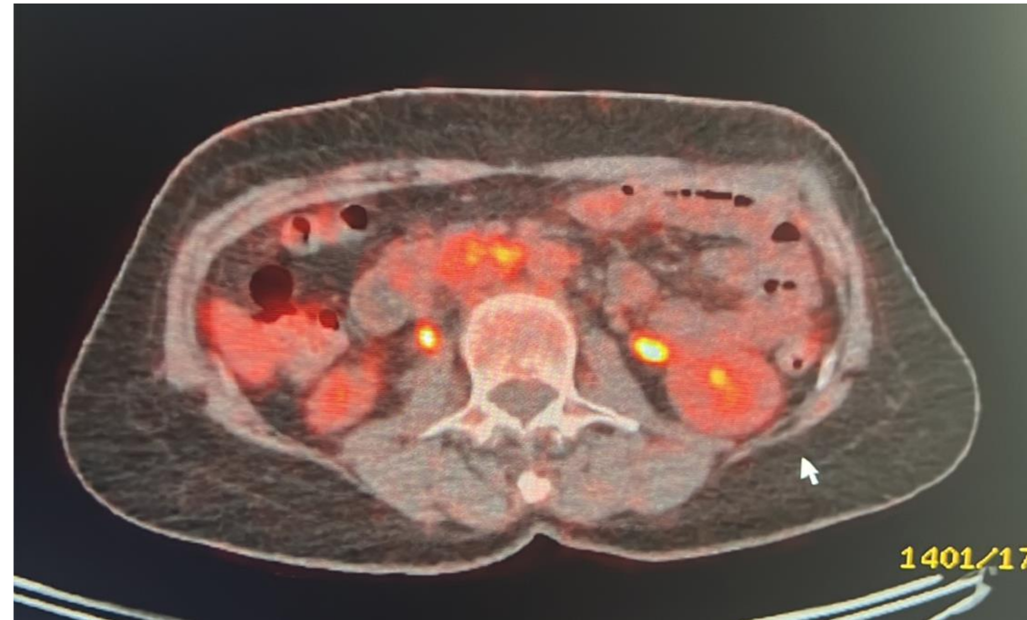


Solid, moderately vascularized, intraluminal lesion, in the inferior vena cava, infrarenal portion, located 4 cm distant from the renal veins, oval morphology, well-defined contours, with heterogeneous post-contrast enhancement, without endoluminal thrombi. Lesion measures 28x20mm in axial plane diameter, longitudinal diameter of 33mm.

Clínica del Country | Clínica La Colina

# PET CT, Fundación Santa Fe: 11/15/2022:




Slightly hypermetabolic lesion in cava topography (SUV 6.66), with no other foci.

# DISCUSSION

- Is there any role for adjuvant chemotherapy or radiotherapy for this 57-year-old female patient, diagnosed with a type I - inferior vena cava leiomyosarcoma, pathological stage I (pT1pN0G2R0) AJCC 8th edition (November 2022), treated with vascular resection on 11/21/2022?

- Very rare disease
- Low quality evidence
- Aggressive disease
- Observation vs MAI scheme for 3 to 4 cycles.

**Case #6**

Bruna David

INCA

Brazil

Male, 27 years, no comorbidities, no family history of cancer.

**-Jul/22:**

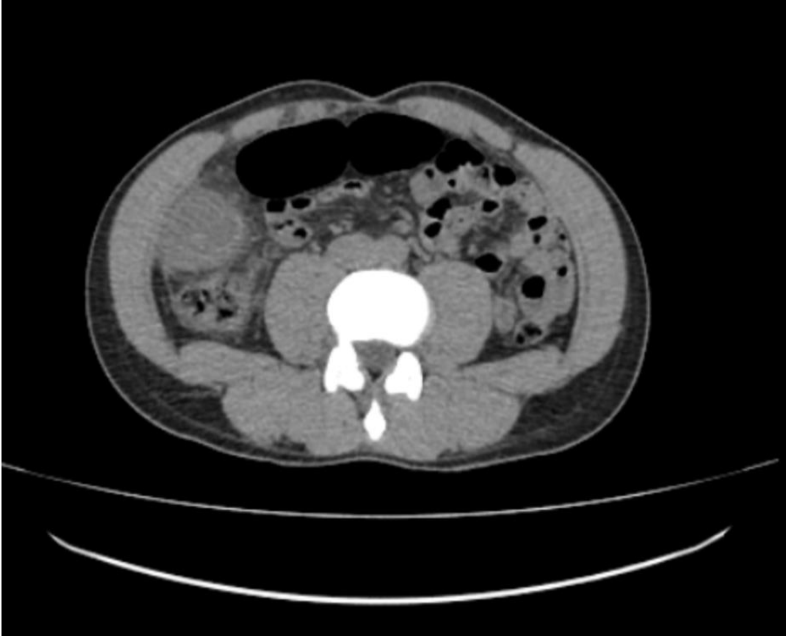
Mild abdominal pain, without any other symptoms.

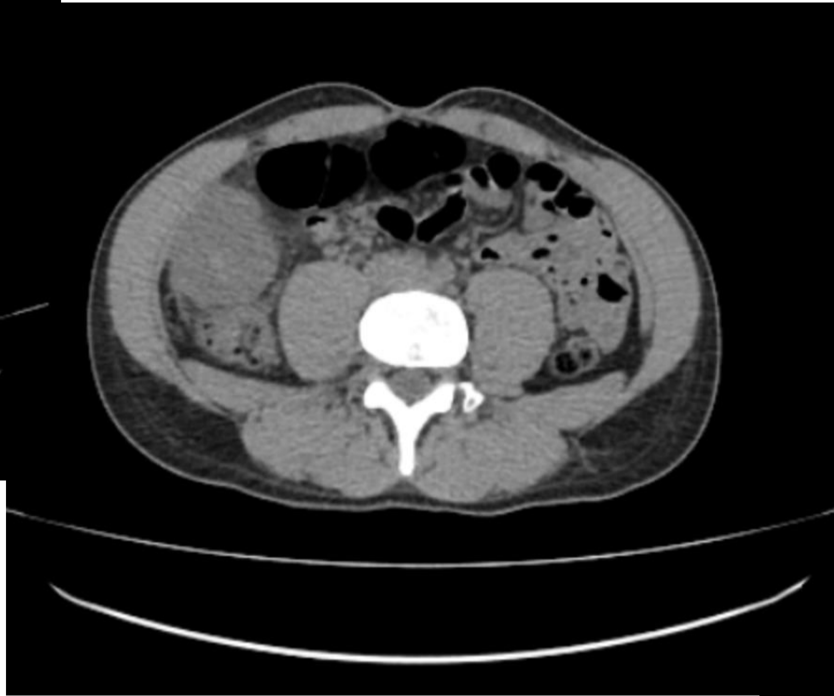
**-Aug/22:**

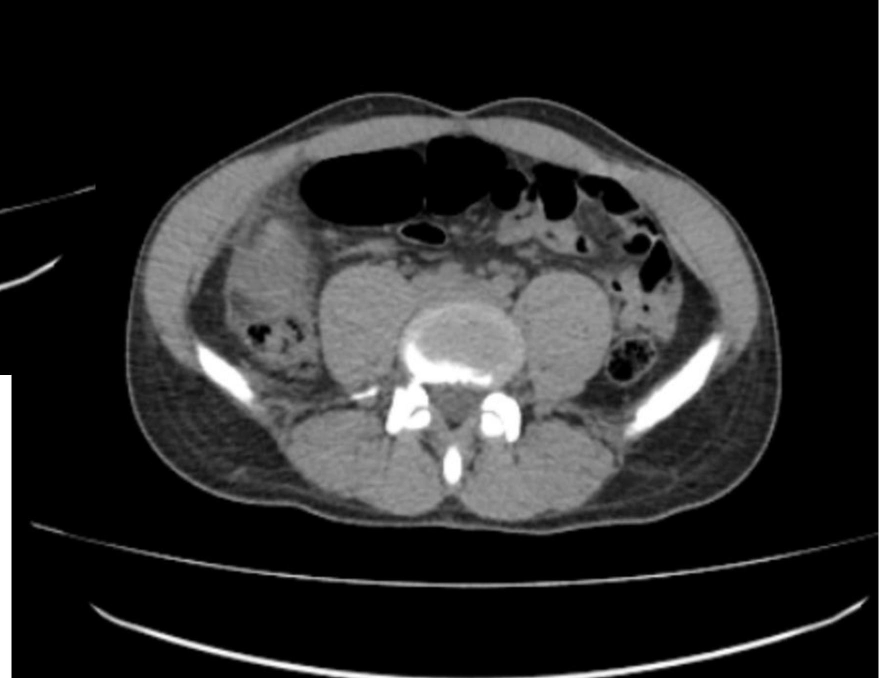
ER – Abdominal CT scan:

Heterogeneous and predominantly hypodense oval expansive formation, with areas spontaneously hyperdense intermingling, measuring about 5.1 x 4.4 cm in the largest axes axial, located in the transition of the flank with the iliac fossa on the right, next to the ascending colon, of unspecific aspect (hematoma? primary lesion?)

- CT scan Aug/2022:







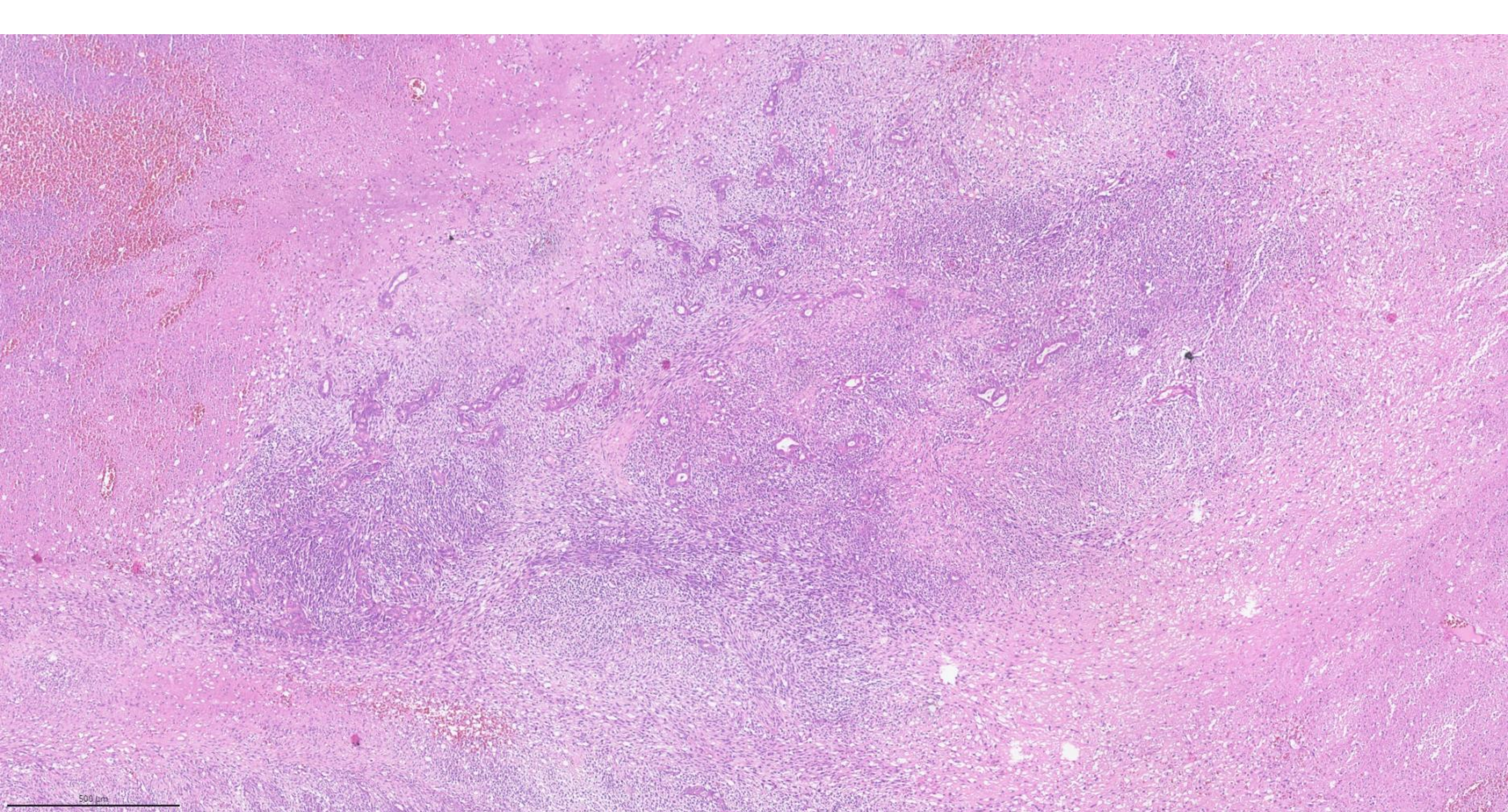
**-Sep/2022:** Lumpectomy without prior biopsy.

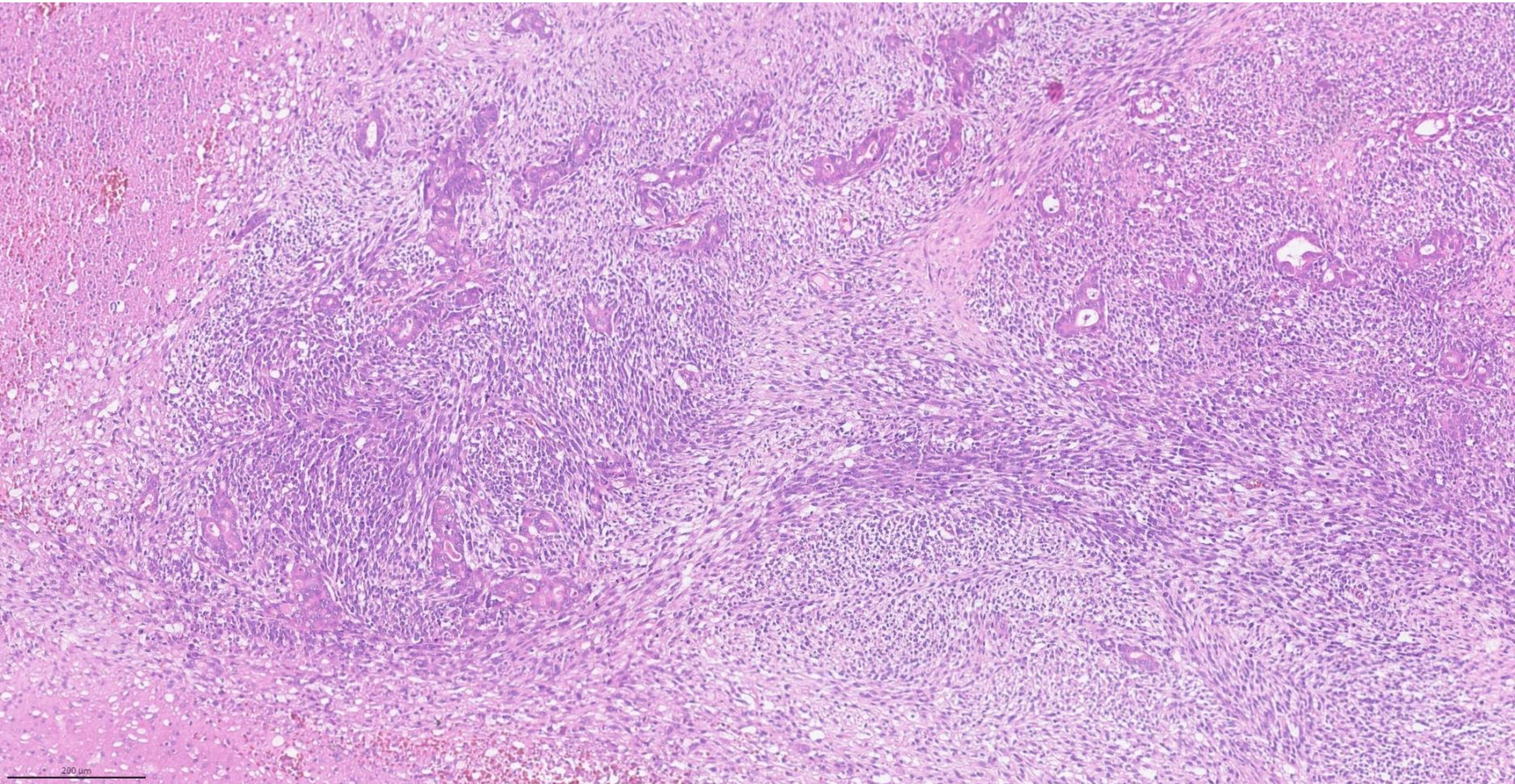
-Pathology report:

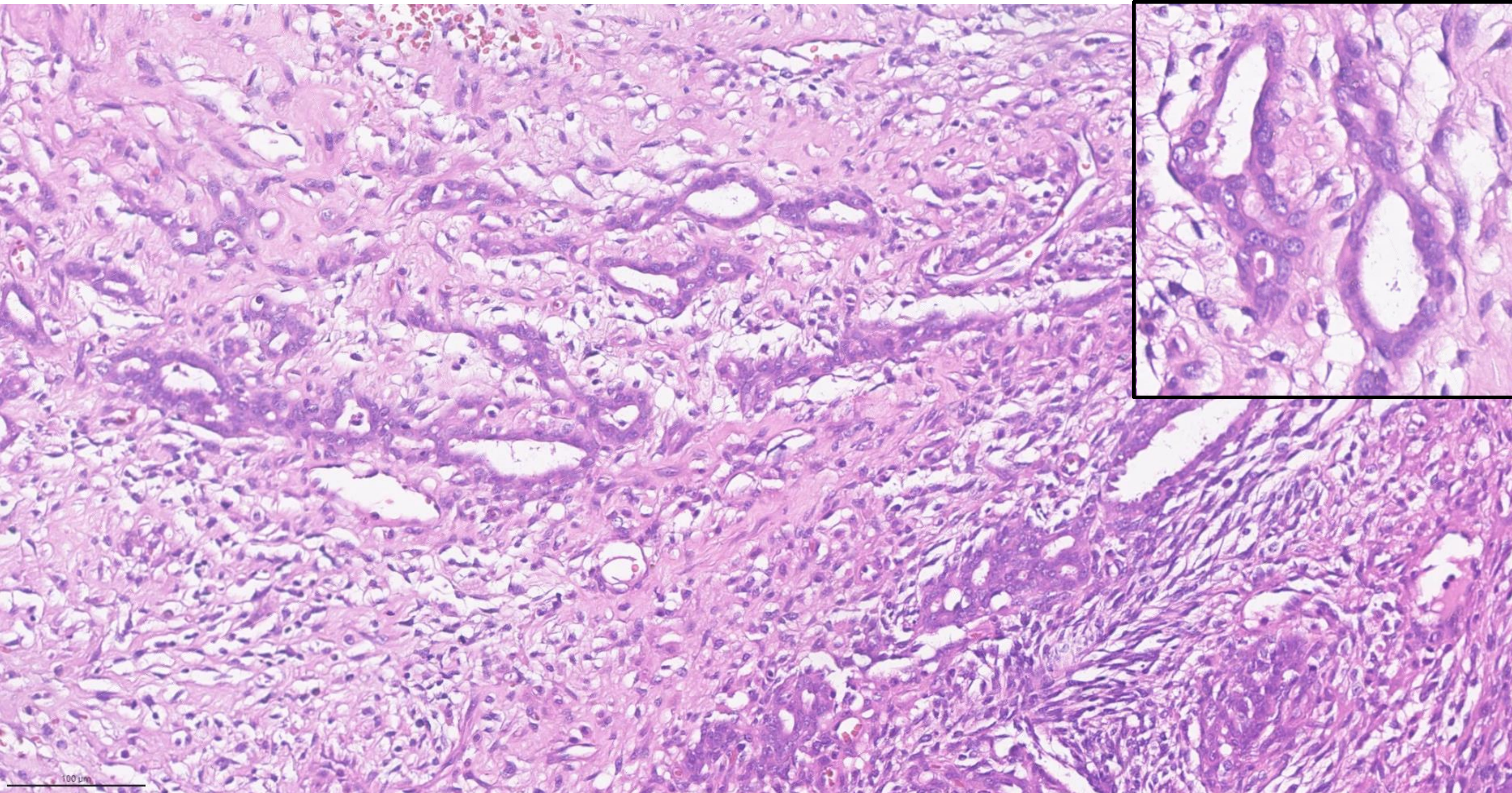
Positive microscopic radial margin

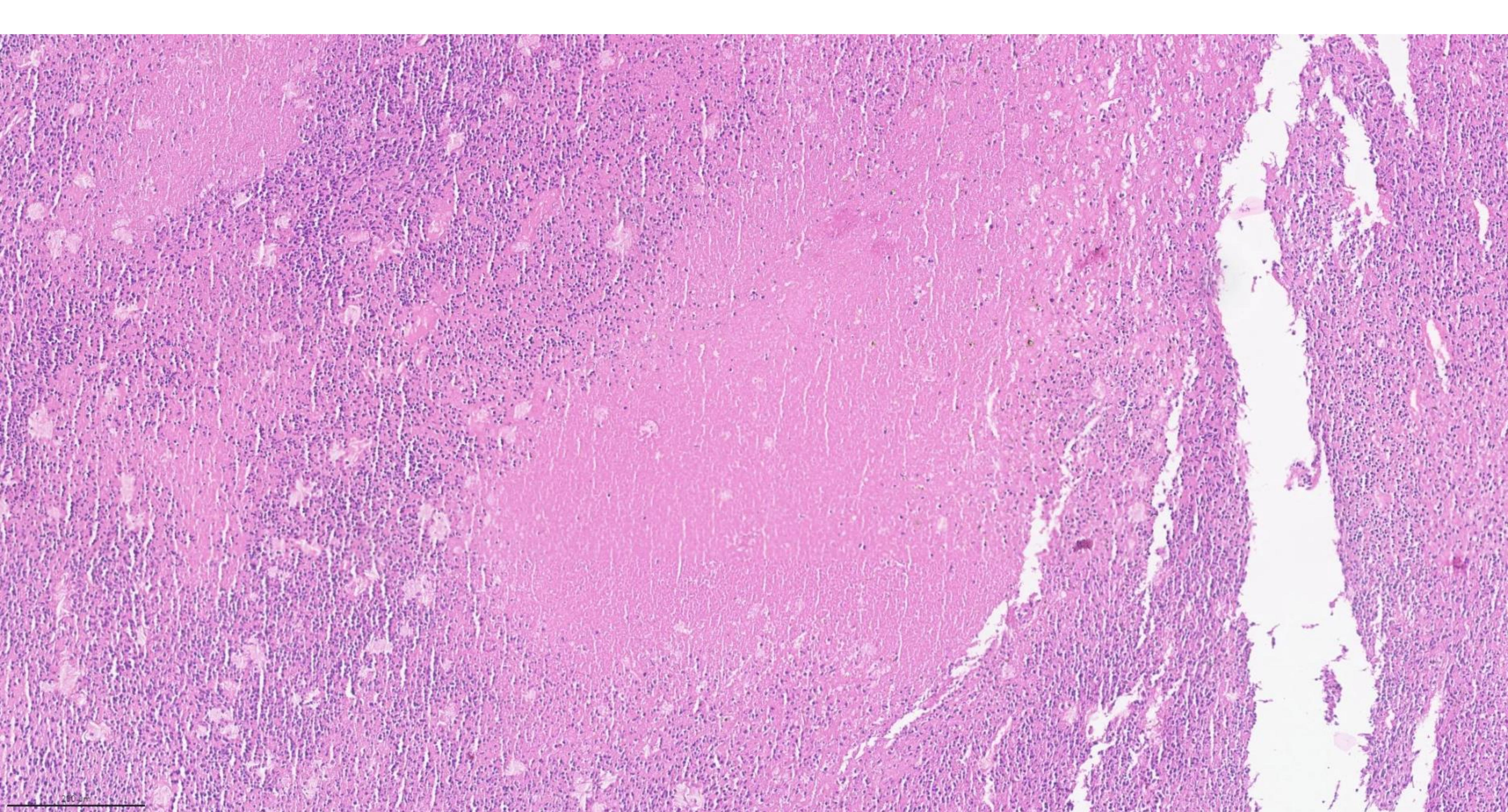
| Material | Marcador | Clone | Resultado | Uso |
| --- | --- | --- | --- | --- |
| 1-A Bloco de Parafina | AE1/AE3 | AE1/AE3 | Positivo focal nas células de interesse | |
| 1-A Bloco de Parafina | CD10 | 56C6 | Negativo | |
| 1-A Bloco de Parafina | PAX-8 | MRQ-50 | Negativo | |
| 1-A Bloco de Parafina | RE | EP1 | Negativo | |
| 1-A Bloco de Parafina | DESMINA | D33 | Negativo | |
| 1-A Bloco de Parafina | EMA | E29 | Positivo focal nas células de interesse | |
| 1-A Bloco de Parafina | AML | 1A4 | Negativo nas células de interesse (controle interno reagente) | |
| 1-A Bloco de Parafina | CD34 | QBEnd10 | Negativo nas células de interesse (controle interno reagente) | |
| 1-A Bloco de Parafina | TLE1 | 1F5 | Positivo focal e com fraca intensidade | |
| 1-A Bloco de Parafina | SOX10 | EP268 | Negativo | |
| 1-A Bloco de Parafina | P63 | DAK-p63 | Negativo | |
| 1-A Bloco de Parafina | SALL4 | 6E3 | Negativo | |
| 1-A Bloco de Parafina | TTF-1 | 8G7G3/1 | Negativo | |
| 1-A Bloco de Parafina | CDX2 | DAK-CDX2 | Negativo | |
| 1-A Bloco de Parafina | KI-67 | MIB-1 | Positivo em até 90% das células neoplásicas | |
| 1-A Bloco de Parafina | SINAPTOFISINA | DAK-SYNAP | Positivo focal nas células neoplásicas | |
| 1-A Bloco de Parafina | DESMINA | D33 | Negativo | |
| 1-A Bloco de Parafina | S100 | Policlonal coelho | Positivo nas células de interesse | |
| 1-A Bloco de Parafina | MIOGENINA | F5D | Negativo | |
| 1-A Bloco de Parafina | CD57 | TB01 | Positivo nas células de interesse | |

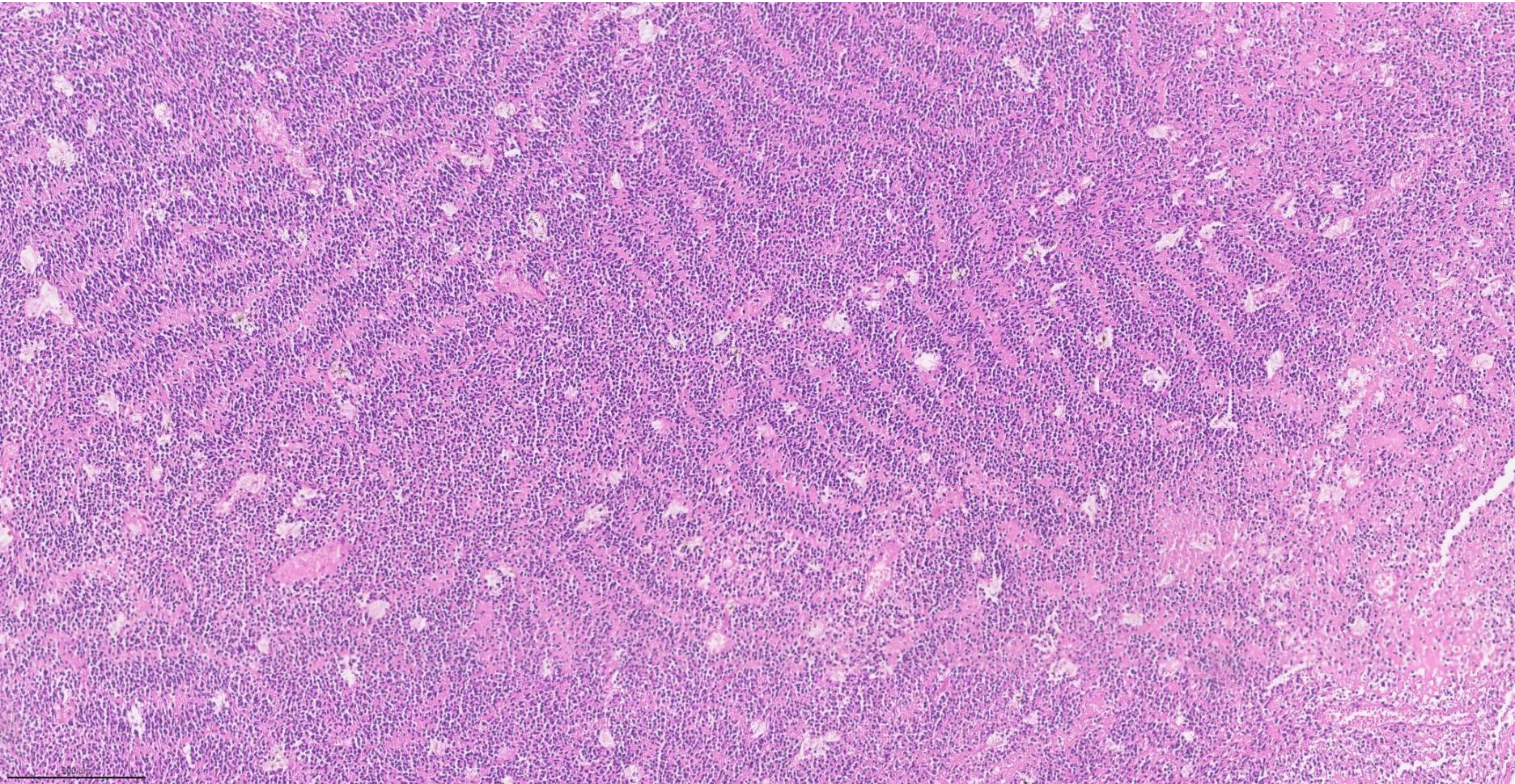
**Conclusion: Malignant peripheral nerve sheath tumor with heterologous elements**

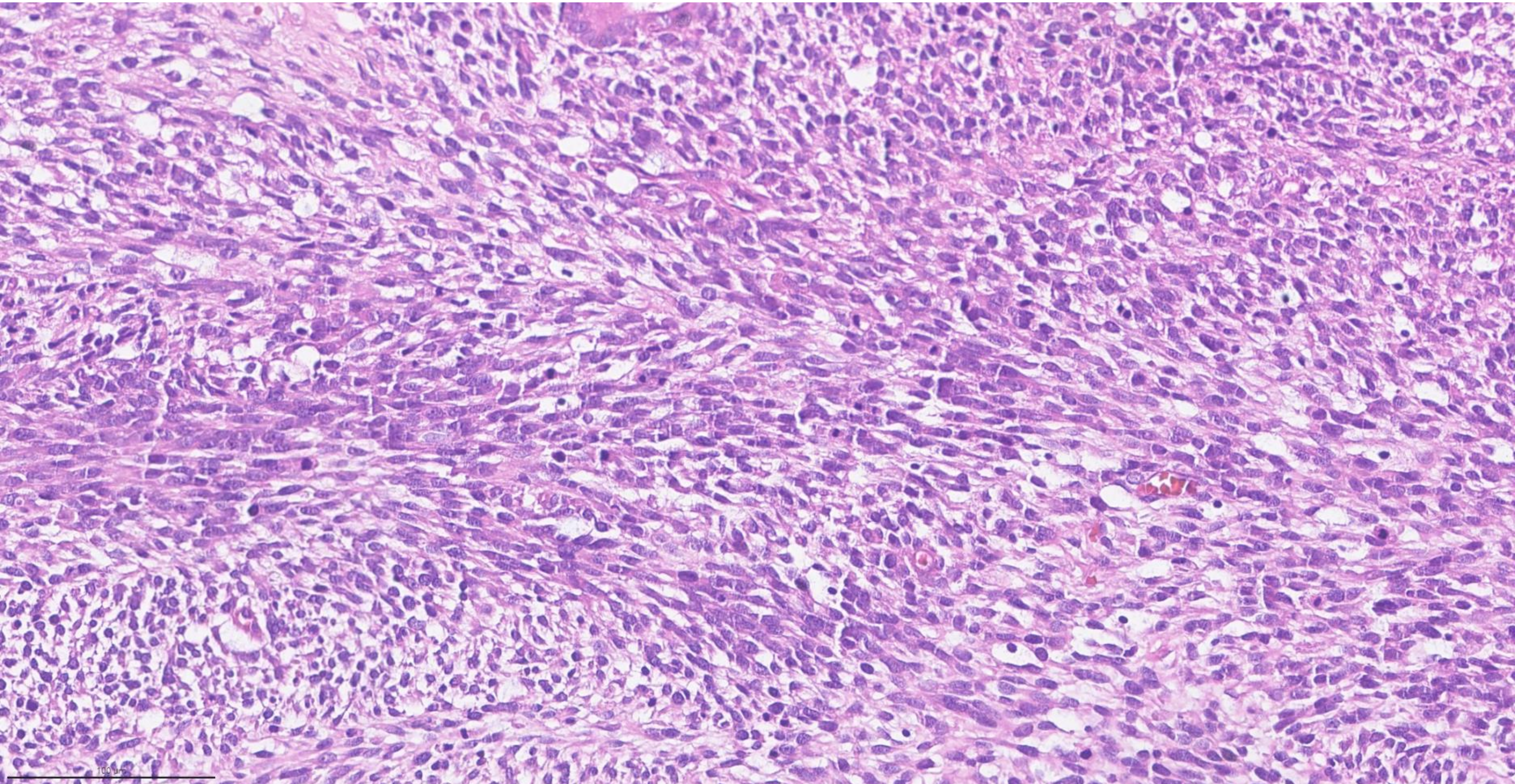

500 µm

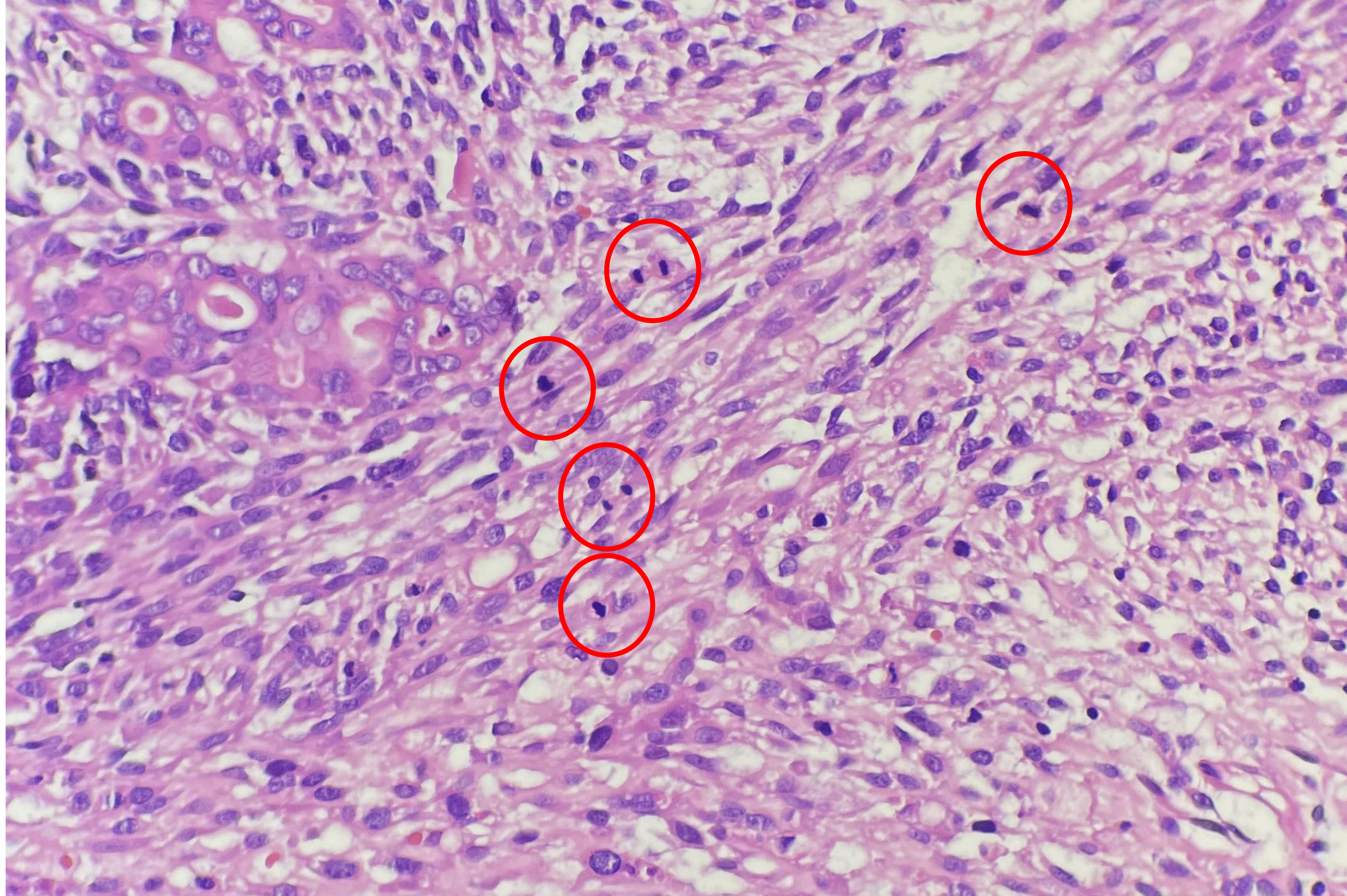

200 µm

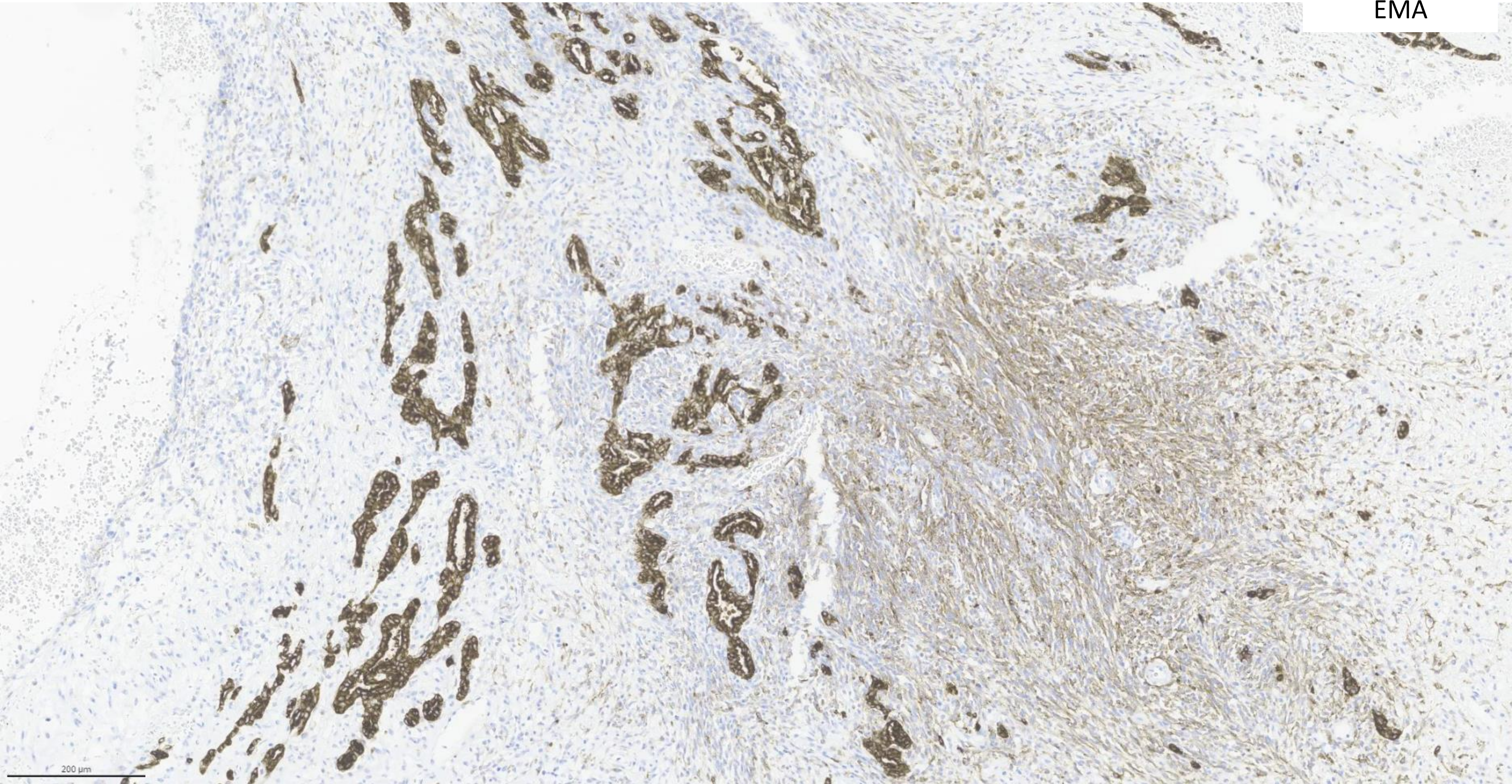

100 µm

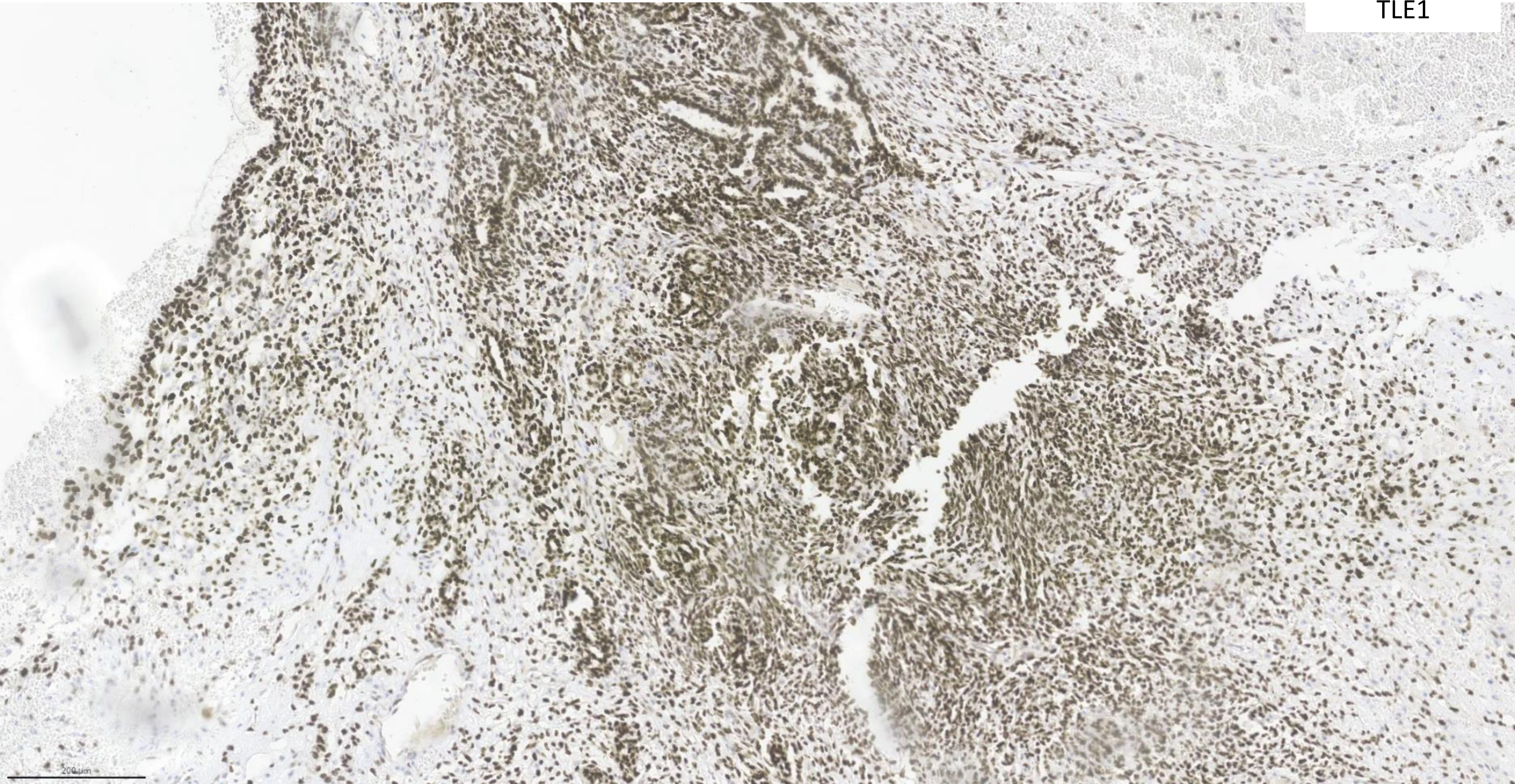


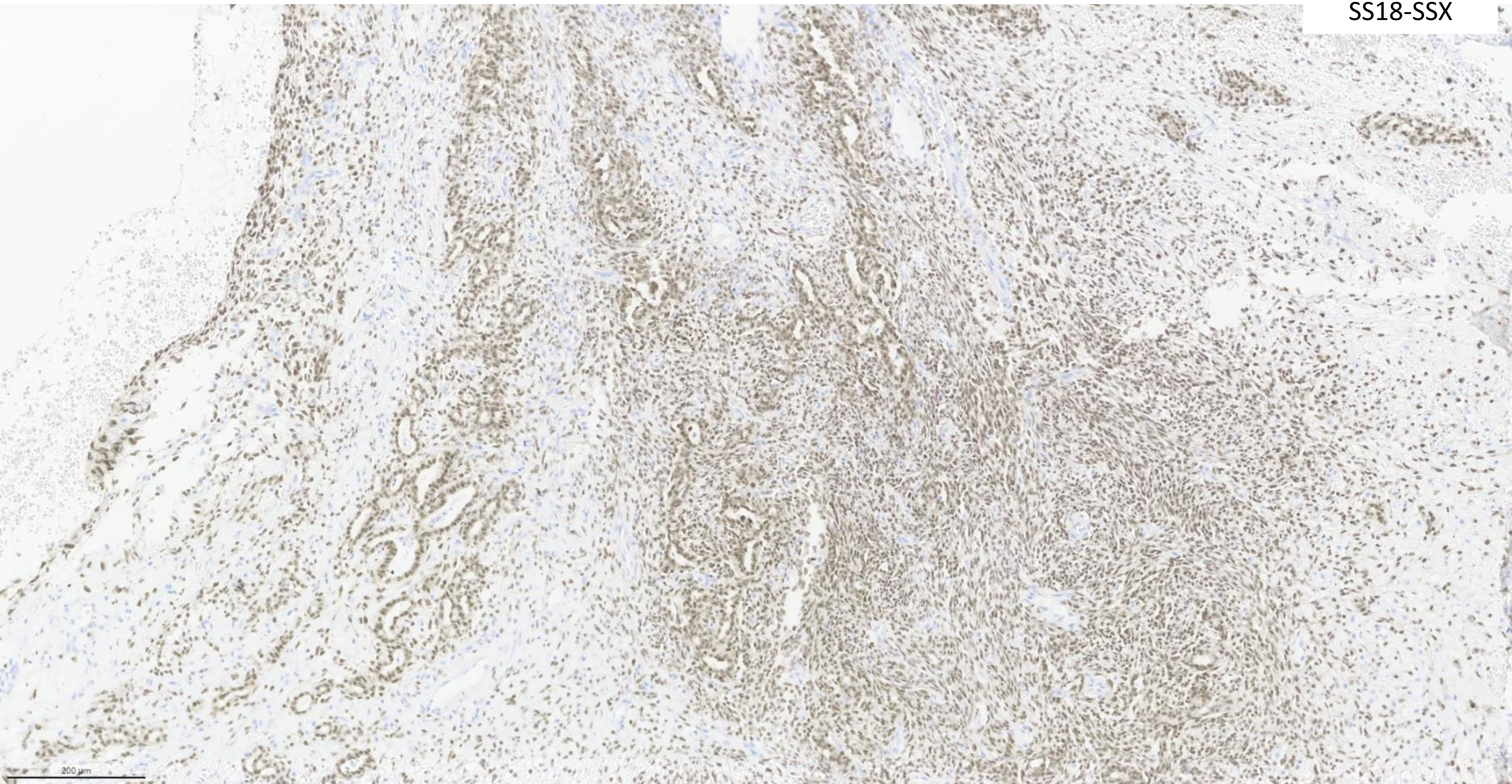


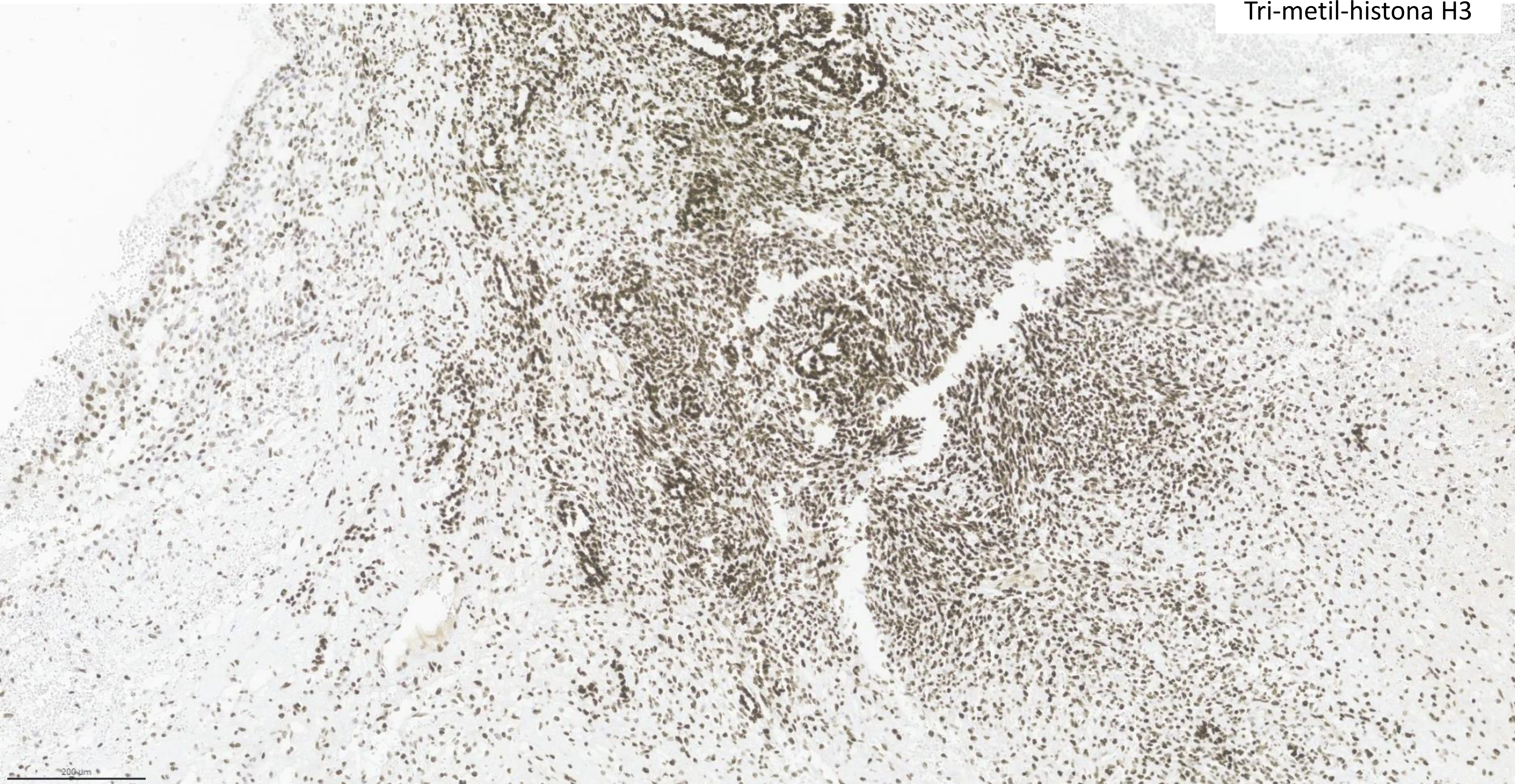

100 µm

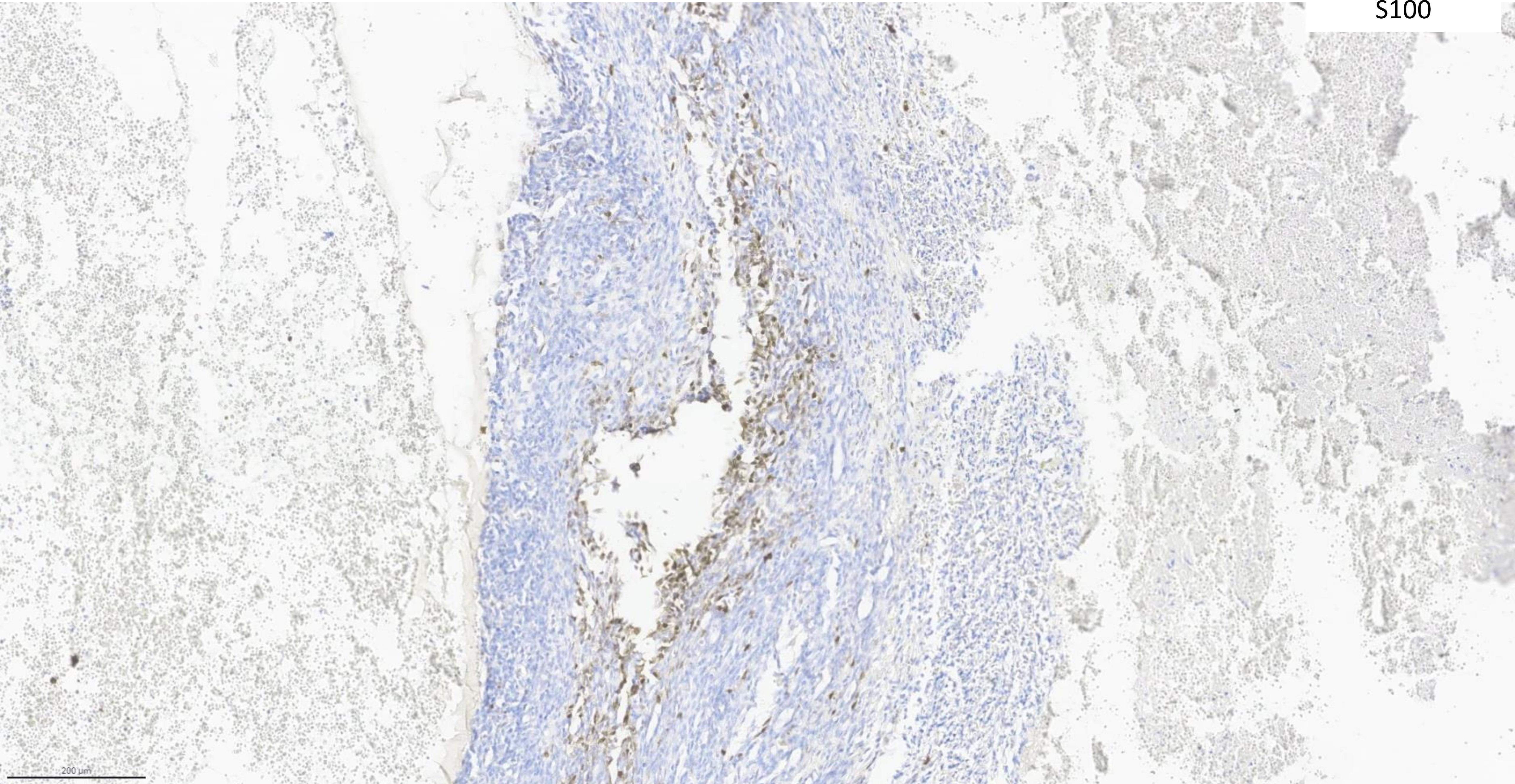


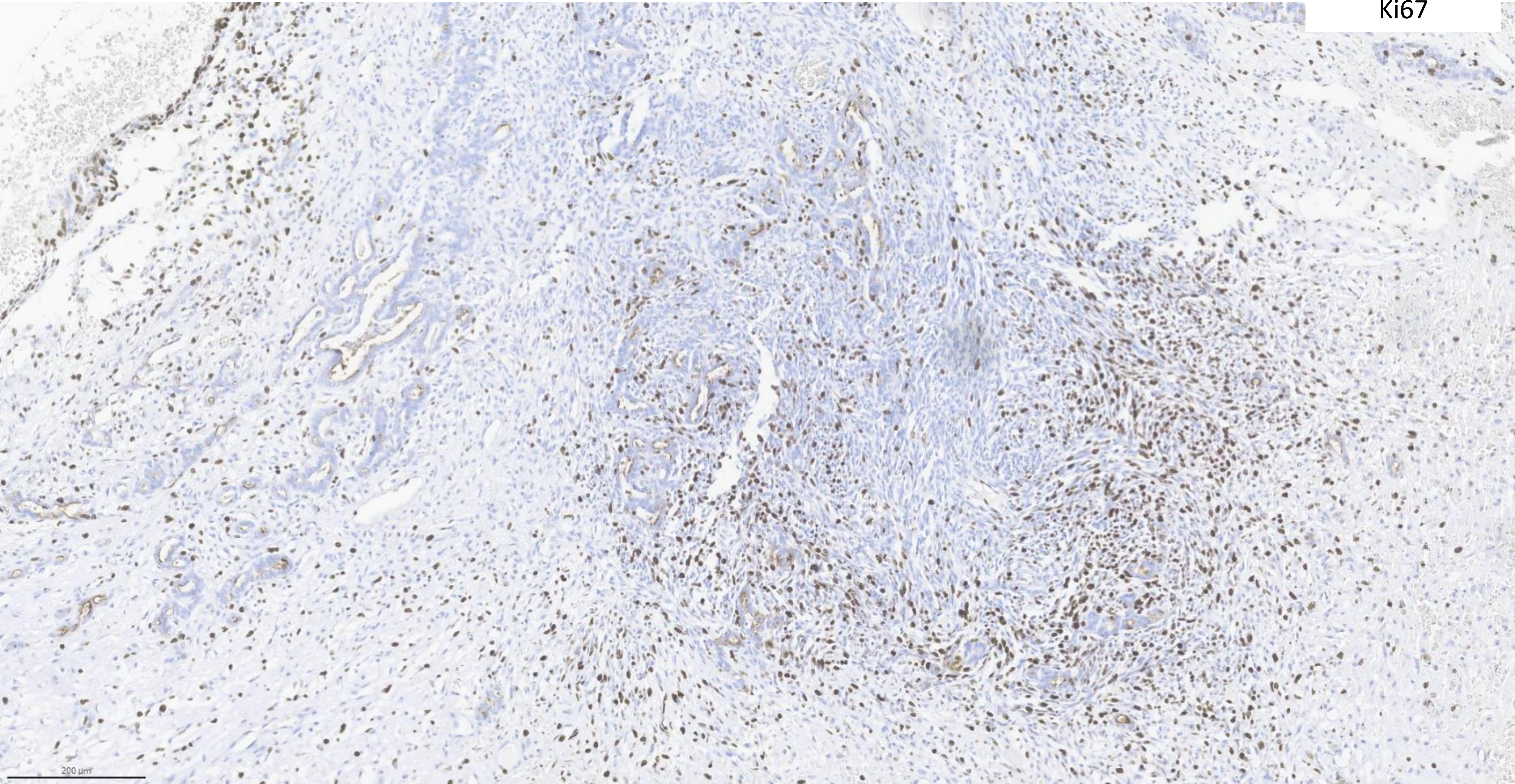

EMA
200 µm


TLE1
200 µm


SS18-SSX
200 µm


Tri-metil-histona H3
200 µm


S100
200 µm


Ki67
200 µm

- Pathology report – second opinion

| Sequência | Antígenos (clone) | Resultados |
|---|---|---|
| 1 | Citoqueratinas AE1/AE3 (AE1/AE3) | Positivo em áreas |
| 2 | EMA (antígeno de membrana epitelial) | Positivo em áreas |
| 3 | Citoqueratina 7 (CK7) (OV-TL 12/30) | Positivo em áreas |
| 4 | TLE1 (1F5) | Positivo |
| 5 | SS18-SSX (E9X9V) | Positivo |
| 6 | Tri-metil-histona H3 (Lys27) (C36B11) | Positivo (expressão preservada) |
| 7 | Proteína S100 (policlonal) | Positivo focal |
| 8 | Ki-67 (MIB-1) | Positivo em 60% das células neoplásicas |
| 9 | CDX2 (DAK-CDX2) | Negativo |
| 10 | CD34 (QBEnd 10) | Negativo |
| 11 | CD31 (JC70A) | Negativo |
| 12 | Desmina (D33) | Negativo |
| 13 | Miogenina (F5D) | Negativo |
| 14 | MYO-D1 (5.8A) | Negativo |
| 15 | SOX-10 (EP268) | Negativo |
| 16 | GFAP (policlonal rabbit) | Negativo |

- **Nov/2022:** first medical appointment at our clinics. No symptoms.

Pathology report second opinion:

**- BIPHASIC SYNOVIAL SARCOMA:**

. SIZE (CENTIMETERS): **6.0 cm** (MACROSCOPIC MEASUREMENT IN THE ORIGINAL LABORATORY REPORT).

.GROWTH PATTERN: PREDOMINANTLY EXPANSIVE WITH INFILTRATIVE FOCIUMS.

.CELLULARITY: EXTENDED.

.PRESENCE OF FUSIFORM AREAS WITH SHORT CELL BEAM.

.PRESENCE OF AREAS WITH NUCLEAR PALICATIONS IN NEOPLASIA.

.PRESENCE OF HYPOCELLULAR FOCUS IN THE PERIPHERY OF THE NEOPLASM.

**.HISTOLOGICAL GRADE: 3** {French Federation of Cancer Centers Sarcoma Group (FNCLCC)}.

.MITOTIC INDEX: 59 MITOSES / 10 HIGH-POWER FIELDS (23 MITOSES / mm2).

.TUMOR NECROSIS: PRESENT (70% OF THE NEOPLASTIC SURFACE).

.ANGIOLIPHATIC NEOPLASTIC INFILTRATION: NOT DETECTED.

**.MARGINS OF SURGICAL RESECTION: FOCALLY COMMITTED (MARGINAL RESECTION).**

.PATHOLOGICAL STAGING (pTNM): pT2.

**-Nov/2022:**

Chest CT and Abdominal MRI: no evidence of disease.

**-Sarculator risk:**

- Retroperitoneum:

-Complete resection: 7y OS = 51%

-Incomplete resection:  7y OS = 31%

- ESTS:

-5y OS = 78%

- **Questions:**

1. New surgical procedure?
2. High risk? Neo/Adjuvant chemotherapy?