



Clinical Cases SELNET

Dr. Franklin A. Castillero R.

Oncología Médica

Instituto Oncológico Nacional de Panamá

Grupo de tumores mixtos

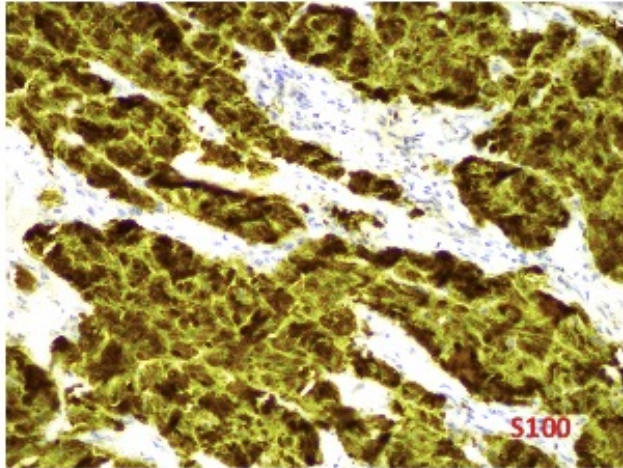


Case 1. C.R. 60y old woman

February 2018: cervical node located at occipital, level 5.

Excisional Biopsy – performed by a neurosurgeon: malignant granular cells tumor. Mitotic rate 7/10 HPF.

Pathology report

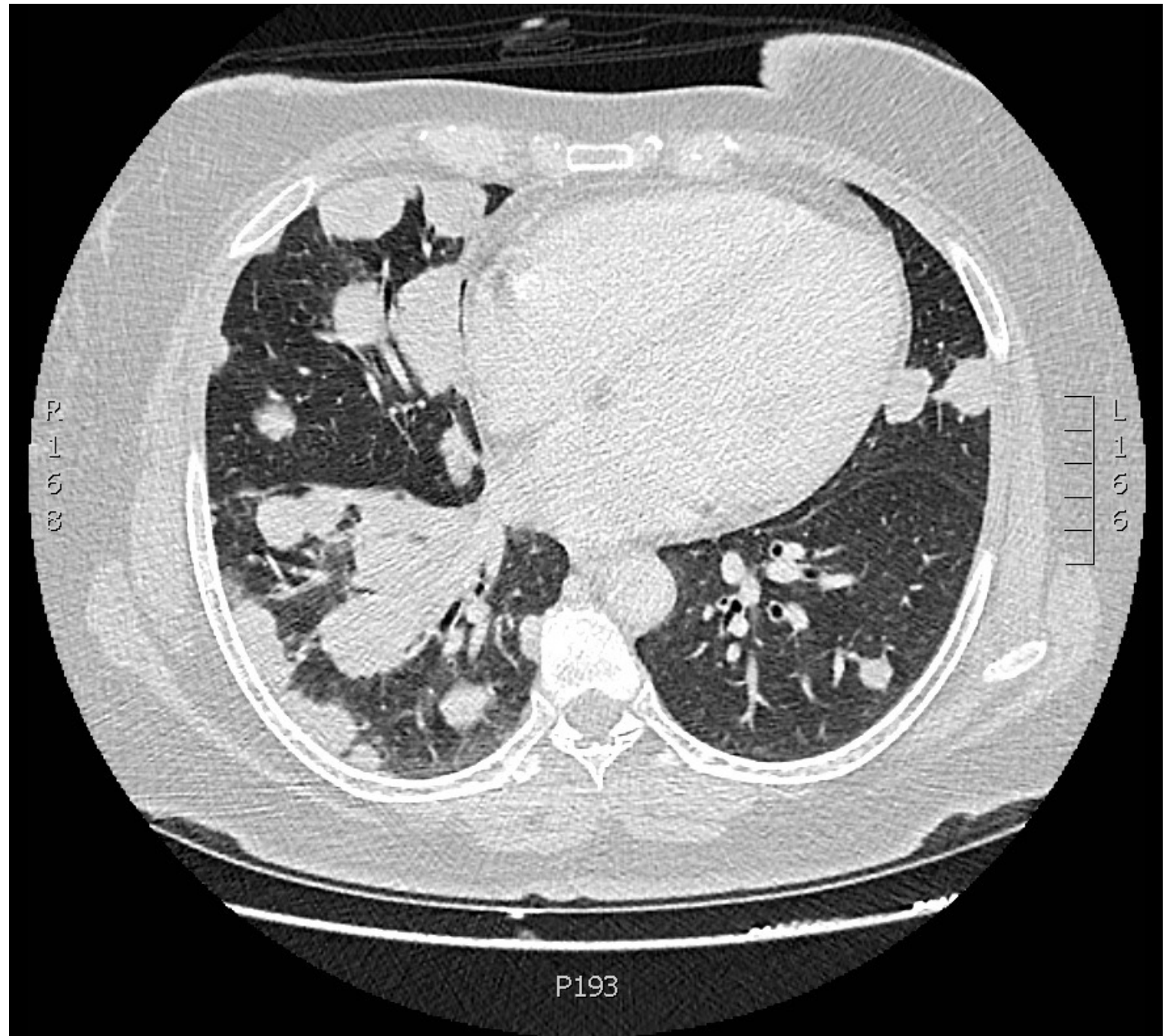


Anticuerpo	Resultados
S100	+
Inhibin	+
CD68	+
NSE	+
Melan A and HMB45	-
SMA	-
H-caldesmon	-
CD34	-
CKAE1/AE3	-

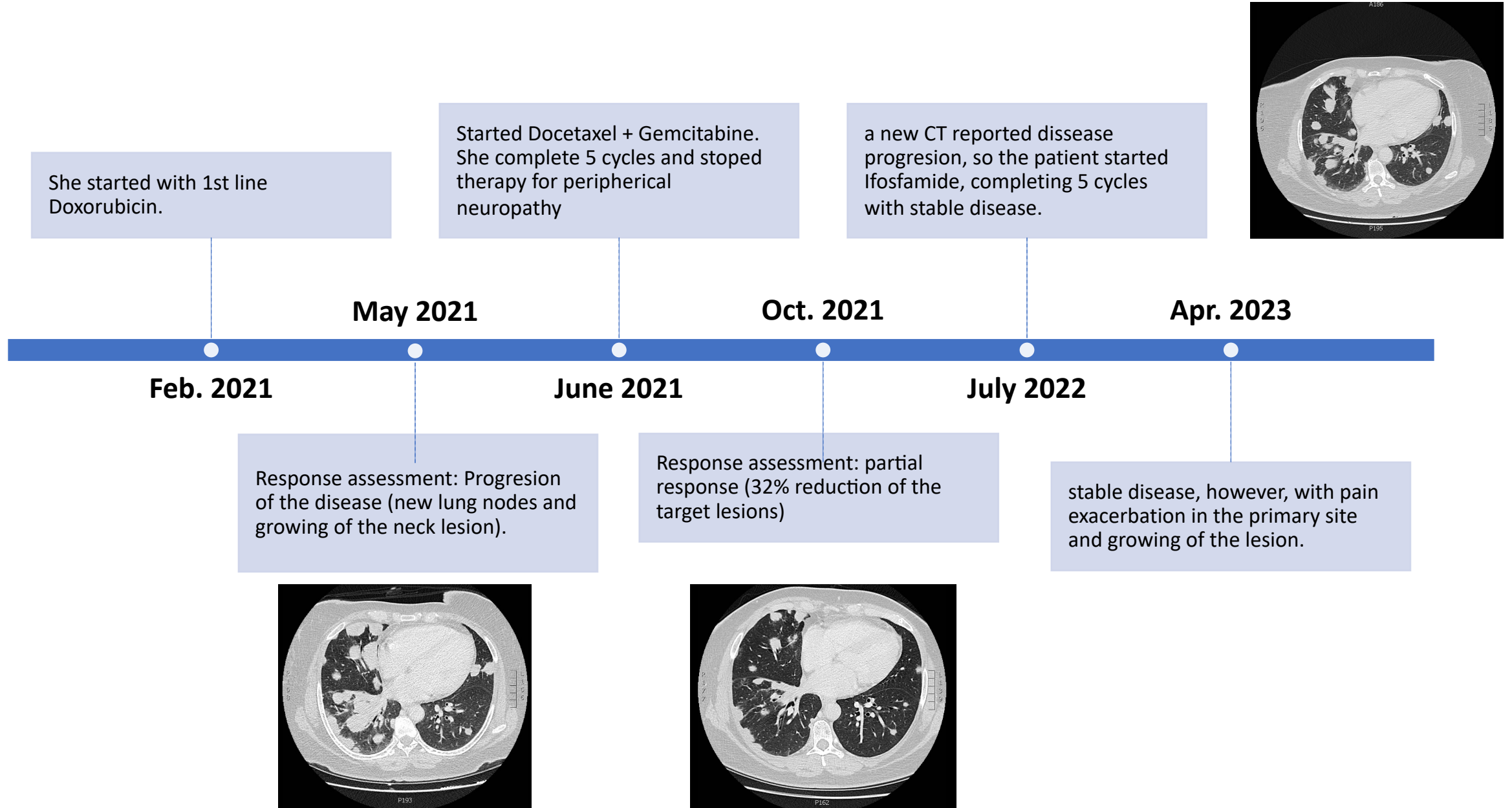
Histologic criteria of malignancy (Fanburg-Smith)	Status
Spindling	present
> 2 mitosis/10 HPF	present
Necrosis	present
High N:C ratio	present
Pleomorphism	present
Vesicular nuclei	present

Diagnosis:
Malignant granular cell tumor
Mitotic count: 7/10 HPF
Spontaneous tumoral necrosis: present

November
2020: Bilateral
Lung
Metastases



Case 1. C.R.





February 2023



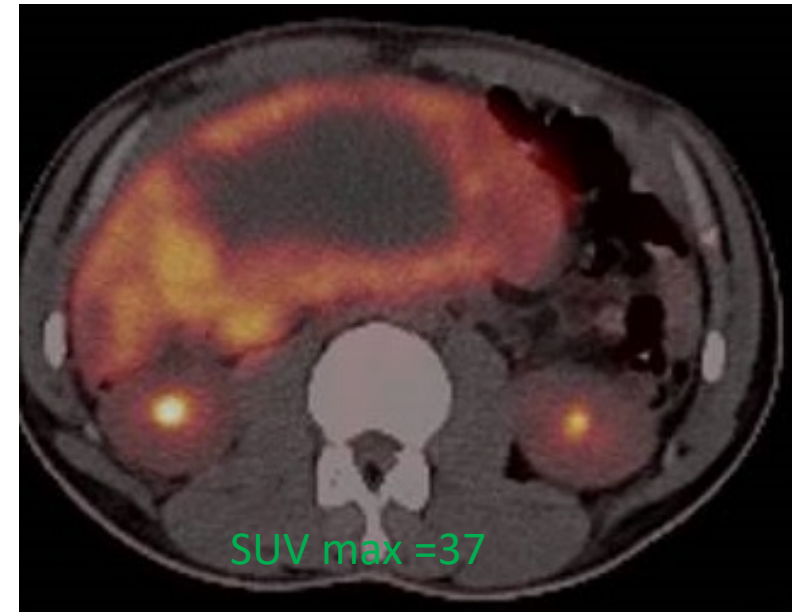
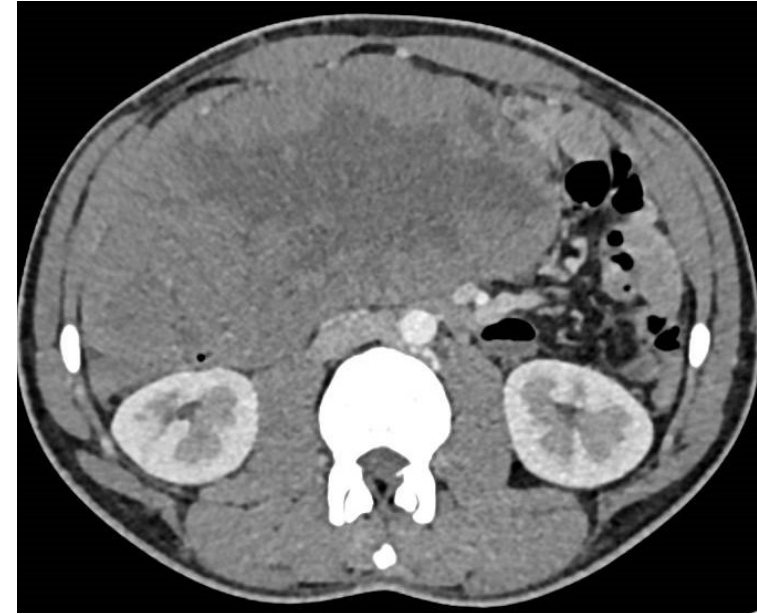
April 2023

Which would be the most appropriate step?

- Radiation therapy to the neck?
- Reexposure to Docetaxel – gemcitabine?
- TKIs? (pazopanib?)
- To perform a NGS genes panel for guiding therapy?
- Any other possible recommendation?

Present Disease and Imaging

- 28-years-old male patient
- No relevant Past Medical or Family History
- Abdominal pain for 2 months
- Body weight loss of 10 kg
- CT CAP == Large (20.6 cm) heterogeneously enhancing mass within the lower abdomen reaching up to the right subhepatic space, causing a mass effect on the adjacent bowel loops, inseparable from the adjacent large and small bowel loops, in particularly the right-sided bowel, and encasing the mesenteric vessels.
- PET-CT == Large hypermetabolic abdominal mass 20 x 14 cm, with necrosis occupying almost all right hemiabdomen



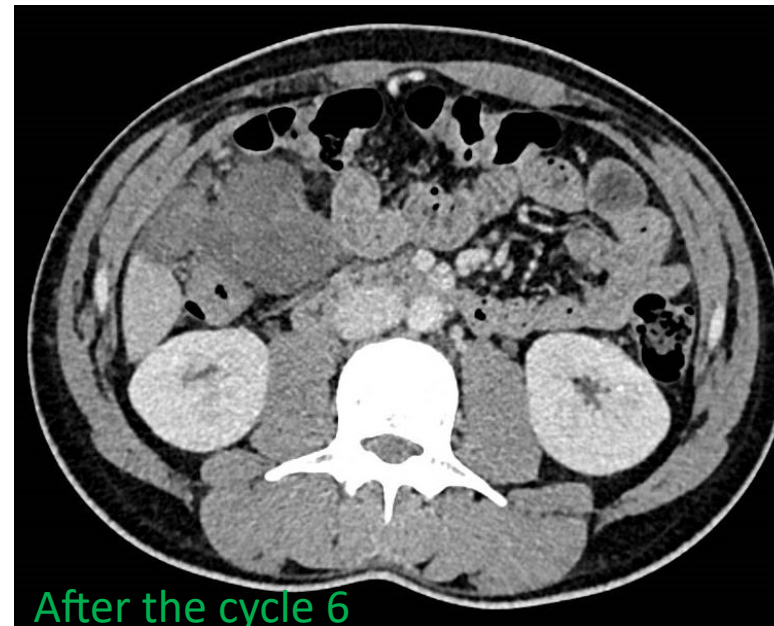
Histopathology

Biopsy done outside 13.10.2022

- Malignant neoplasm with small, round, blue cells with hyperchromatic nuclei and necrosis.
- CD99+ WT+
- CD56 -, AE1/AE3 -, LCA-, CD3-, CD20-, BCL6-, Chromogranin-
- Desmoplastic small round cells tumor is favored. Other diffs: neuroblastoma, rhabdoid tumor, Wilms tumor, rhabdomyosarcoma, synovial sarcoma
- A new Tru-cut **biopsy** was taken in our Hospital. The light microscopy was similar to the prior biopsy. However, the material was predominantly necrotic and not enough for a full IHC panel.

Treatment

- Interval compressed VDC/IE Induction Chemotherapy as per COG AEWS 1031 for 7 Cycles to PR
- Surgery: Abdominal mass resection plus Right Hemicolectomy plus HIPEC



Histopathology and NGS

- Round to spindle cell sarcomatous proliferation, residual. (7 foci)
 - Tumor site: Omentum
 - Maximum size of viable tumor focus: 0.6 cm.
 - Extent of necrosis: 95%.
 - Lymphovascular / perineural invasion: negative
 - No tumor on the inked peritumoral fatty tissue surface
 - Adjacent fatty tissue showed satellite nodules with post treatment changes and free of residual tumor.
 - The final pathological diagnosis was postponed until molecular testing results
- MDT advised against adjuvant RT and to continue the same chemo up to 17 cycles.
- He continued with Cycles 8 and 9 of VDC/IE.

Molecular testing: BCOR-MAML3 fusion transcript

Genes Tested with Pathogenic or Likely Pathogenic Alterations

Gene	Method	Analyte	Variant Interpretation	Protein Alteration	Exon	DNA Alteration	Variant Frequency %
BCOR	Seq	RNA-Tumor	Pathogenic Fusion	BCOR-MAML3	14	-	-

Unclassified alterations for DNA sequencing can be found in the MI Portal.
Formal nucleotide nomenclature and gene reference sequences can be found in the Appendix of this report.

Genes Tested with Variants of Uncertain Significance

Gene	Method	Analyte	Variant Interpretation	Protein Alteration	Exon	DNA Alteration	Variant Frequency %
TSC2	Seq	DNA-Tumor	Variant of Uncertain Significance	p.R1355G	34	c.4063A>G	41

Question to the Board:

- Should we continue the same therapy, or due to an overall better prognosis of BCOR translocated sarcoma reduce the duration or intensity of the treatment?



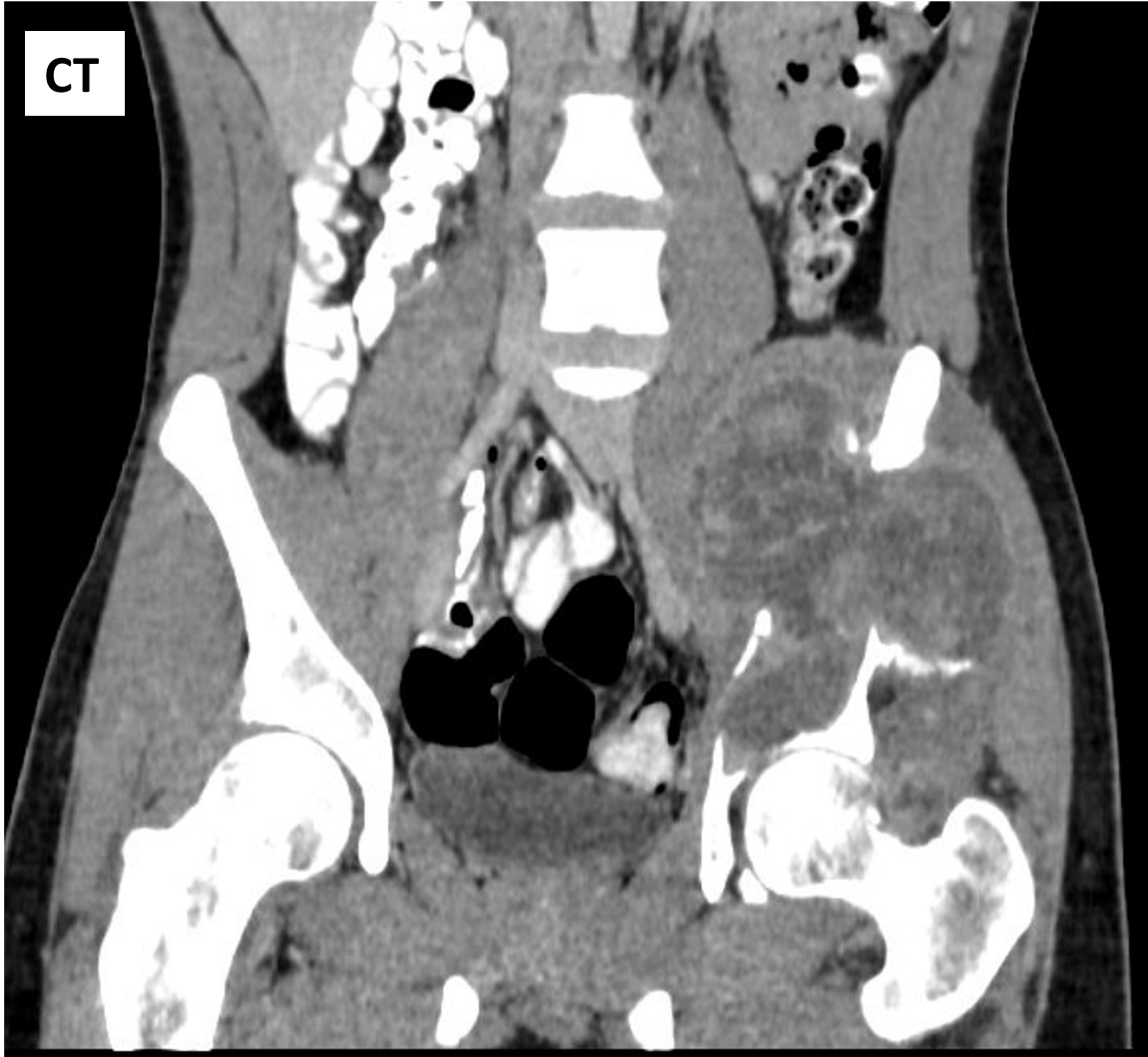
SELNET MDT

May 18th, 2023

Ronald Badilla G
Medical Oncologist
San José, CR

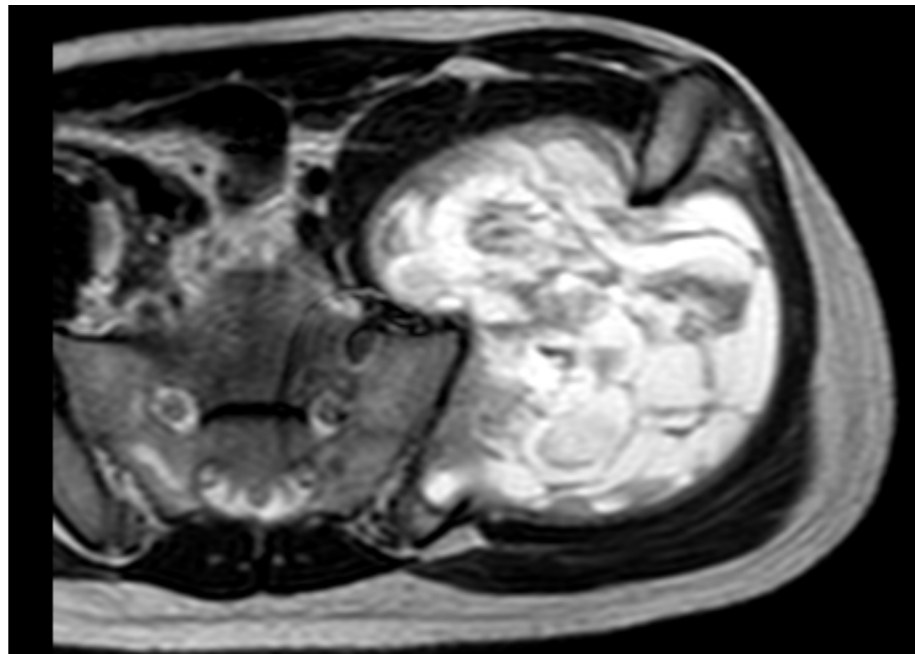
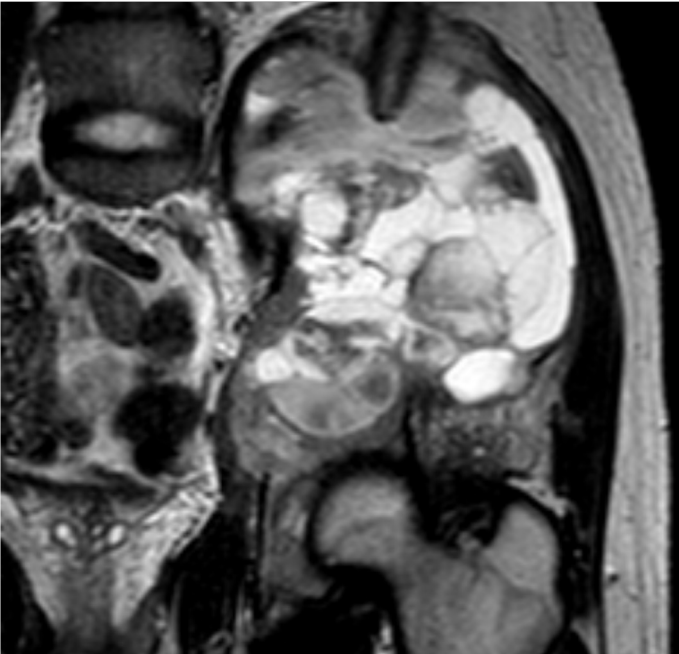
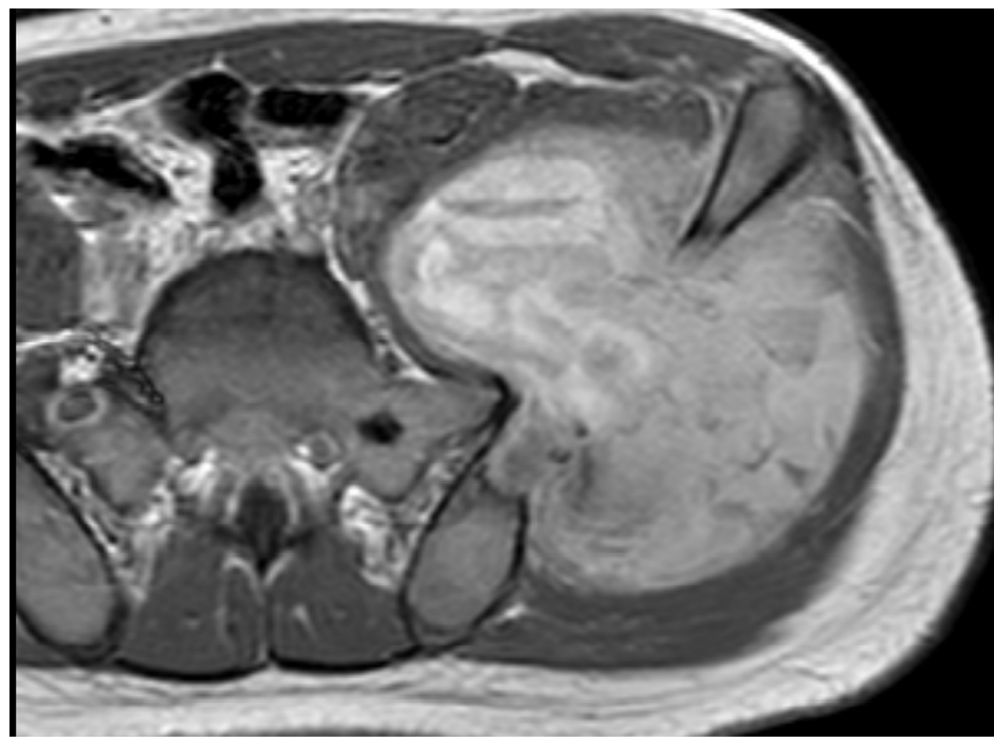
Clinical record

- 20-year-old man
- Salesman
- No comorbidities
- 2 month pelvic pain
- Pain exacerbated after a slip
- X-rays: Lytic lesion - left iliac bone



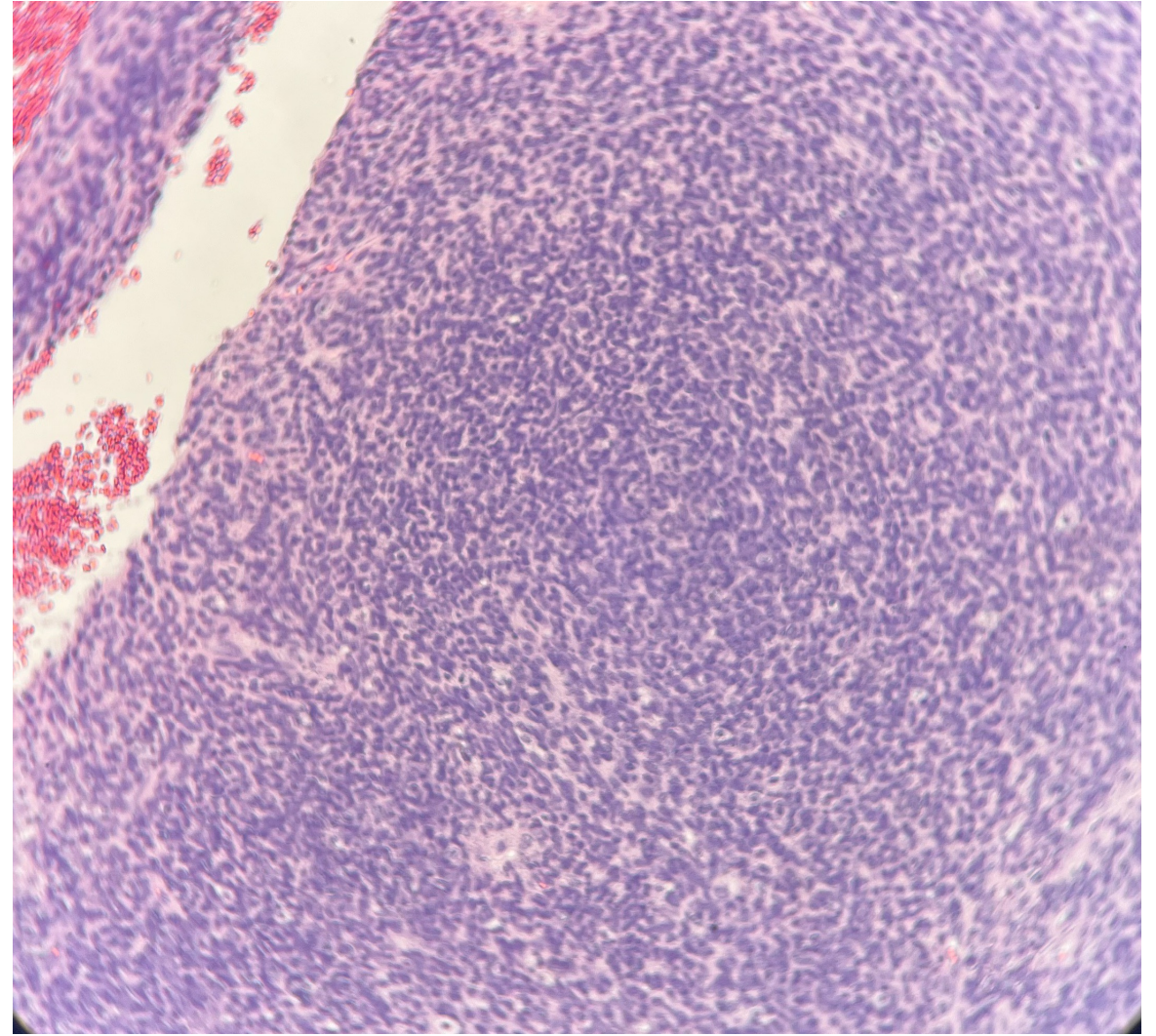
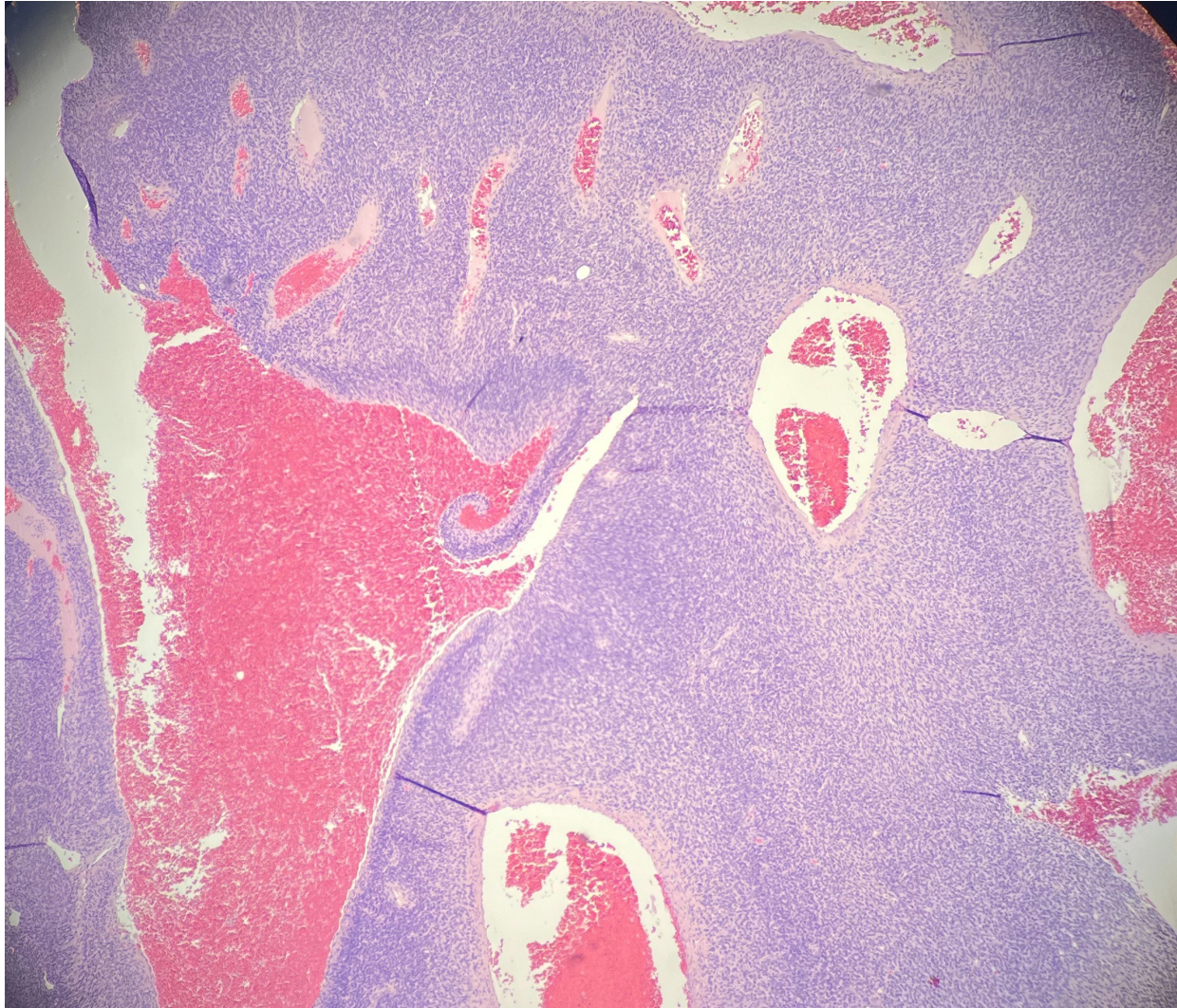
- Large, heterogeneous left iliac crest lesion
- Lytic and soft tissue component
- Septate appearance
- Mass effect on surrounding tissue

MRI

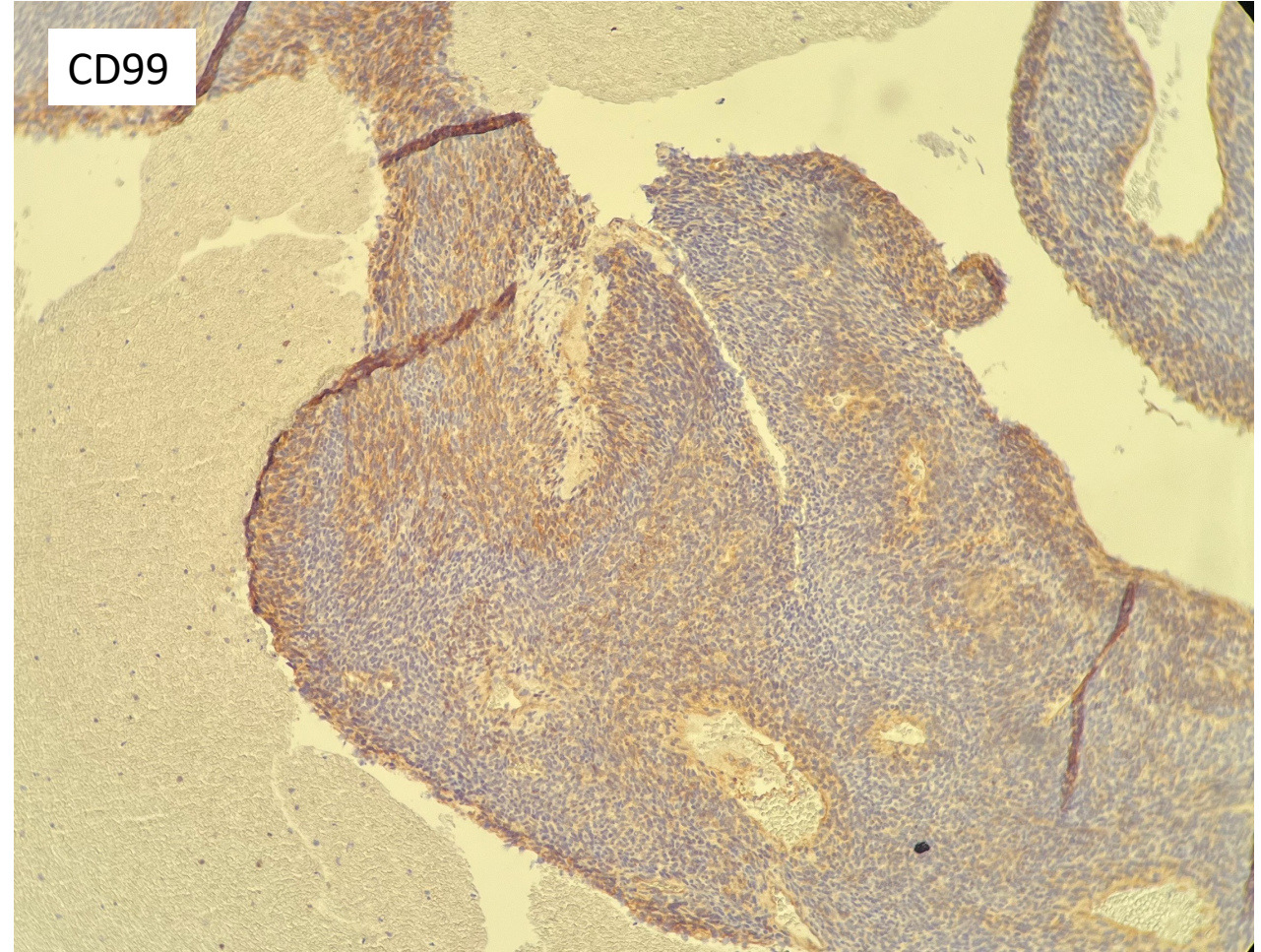
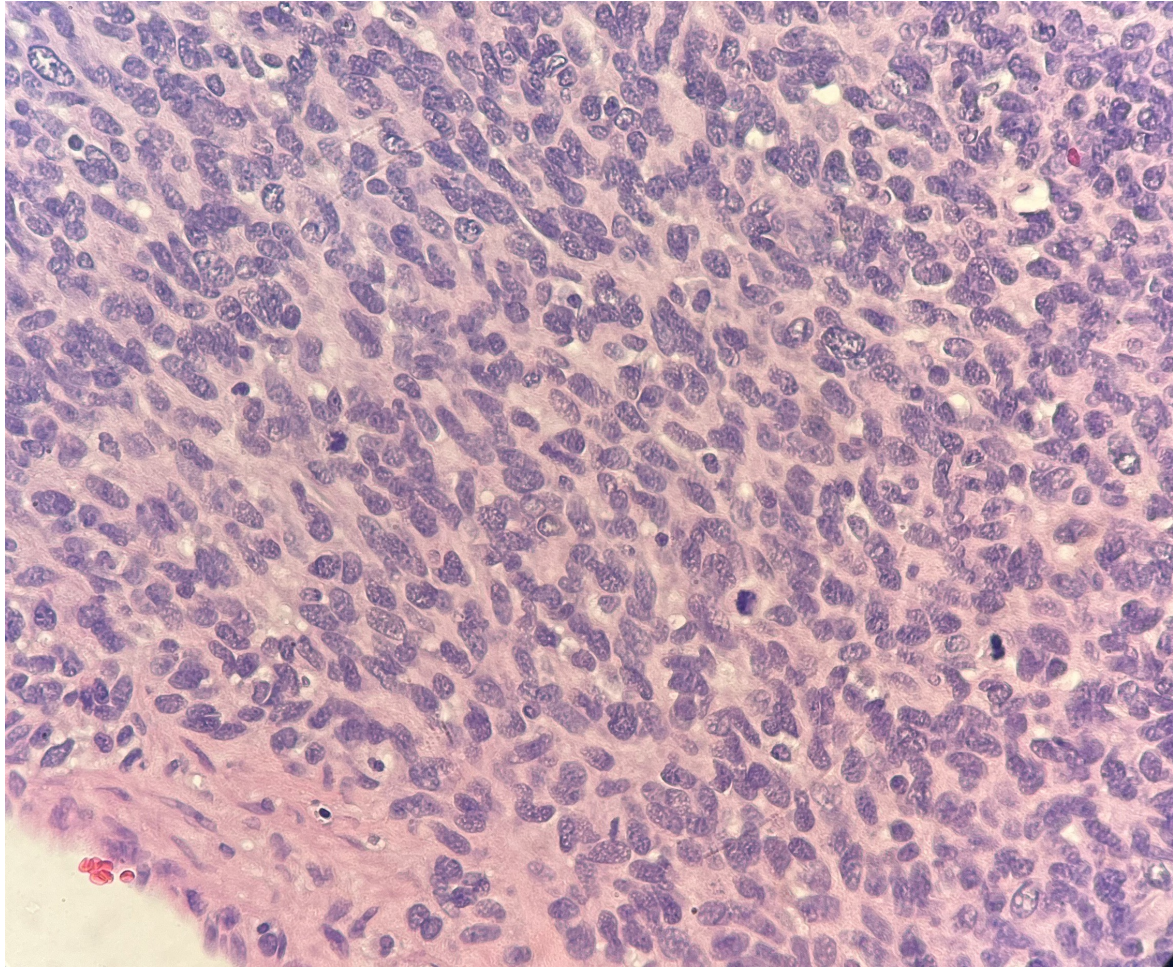


- Left iliac bone lesion
- Expansive
- Heterogeneous
- Bone destruction
- Septations
- Iliacus and gluteus minimus muscle displacement
- No femoral affection
- No joint invasion

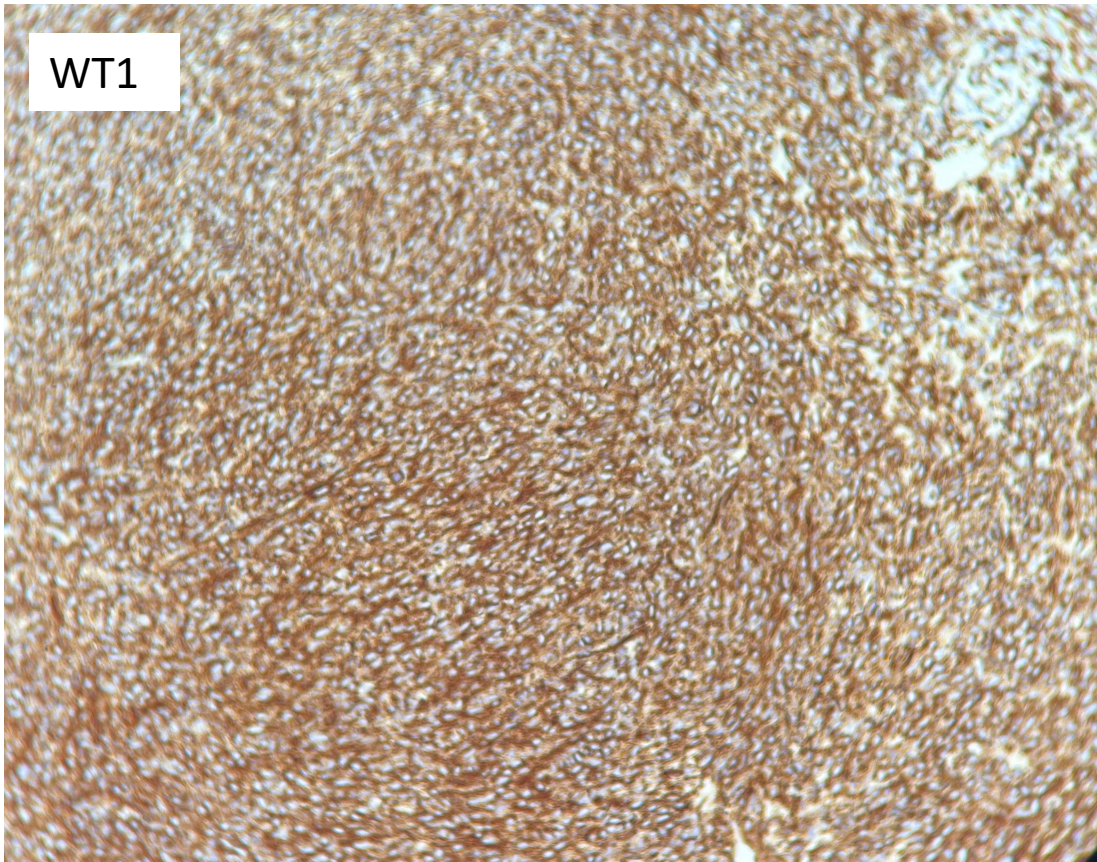
Pathology



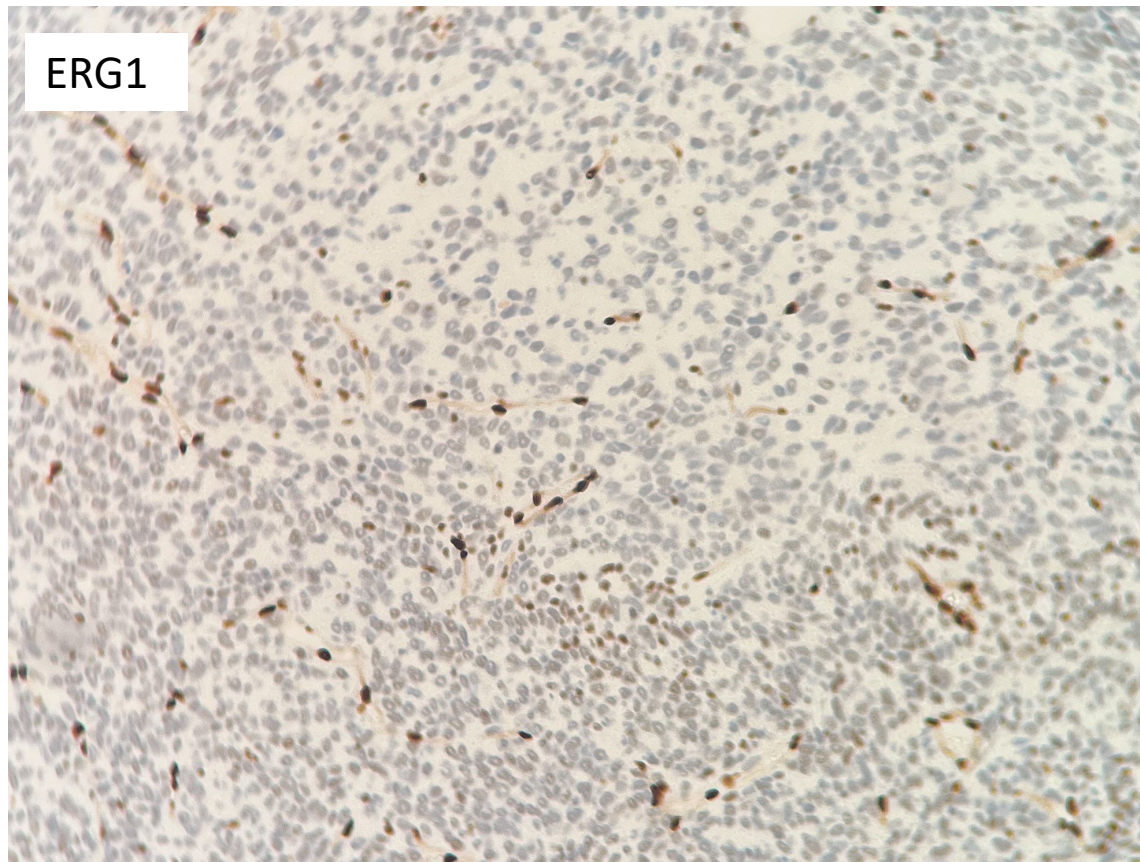
Pathologist: Tibisay Vloria



WT1

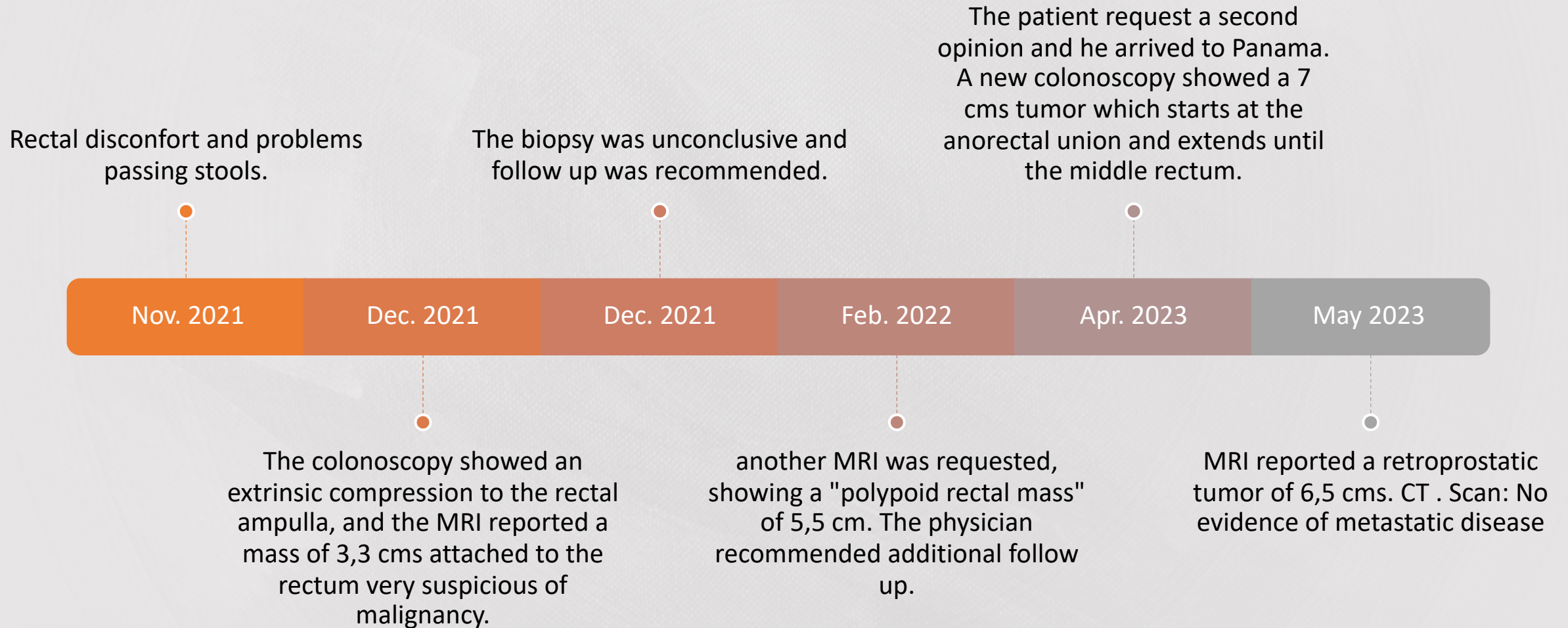


ERG1



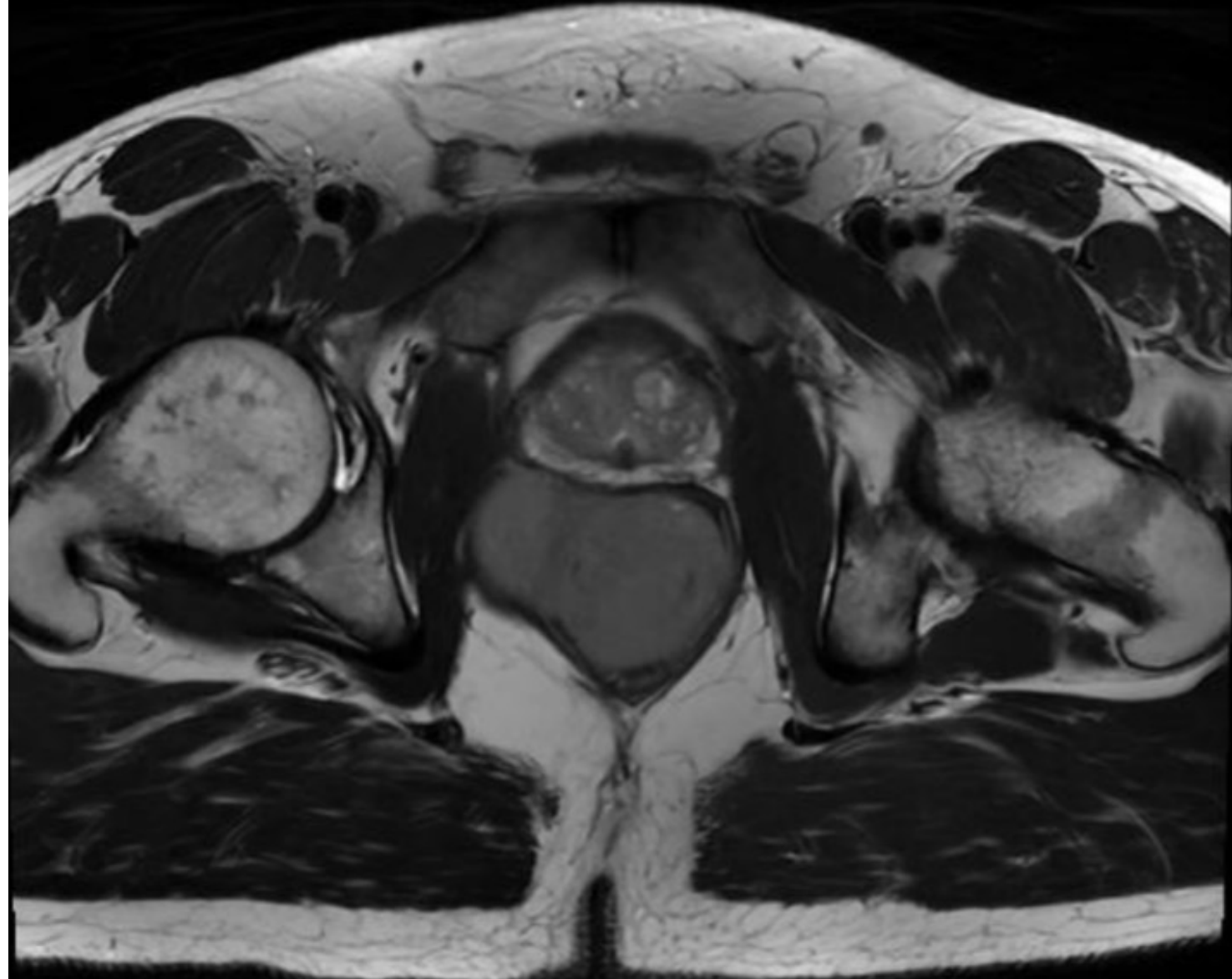
Positive markers	Negative markers
<ul style="list-style-type: none"> • CD99 (patchy) • WT1 • TLE1 (weak/focal) • ERG (weak/focal) • VIMENTIN • INI1 	<ul style="list-style-type: none"> • PANCK • S100 • DESMIN • SMA • CALDESMON • EMA • CK7 • CD34 • CD31 • BCOR
<p>CIC-DUX4 and ETV4 not available</p>	
<p>Dx: Undifferentiated small round cell sarcoma <i>CIC-rearranged sarcoma ?</i></p>	
<p>Question:</p> <p>Treat according to Ewing sarcoma approach ?</p>	

Case 2. G.S. 47 y old man; St. Marteen.



MRI

- Retroprostatic solid tumor
- Rectum displacement to the left and wall infiltration
- Contact with the posterior prostatic capsule, w/o seminal vesicles infiltration
- Levator ani muscle infiltration



INFORME HISTOPATOLOGICO

HISTORIA CLÍNICA: Masa pélvica para rectal.

DIAGNÓSTICO CLINICO:
MASA PELVICA A/D SARCOMA

TIPO DE MUESTRA:
MASA PELVICA

ENSAYO DE LA INMUNOHISTOQUIMICA:

• CD34	POSITIVO (100%)
• HHF35	NEGATIVO (0%)
• S100 (4c4.9)	NEGATIVO (0%)
• CD117:	POSITIVO (100%)
• DOG-1:	POSITIVO (100%)

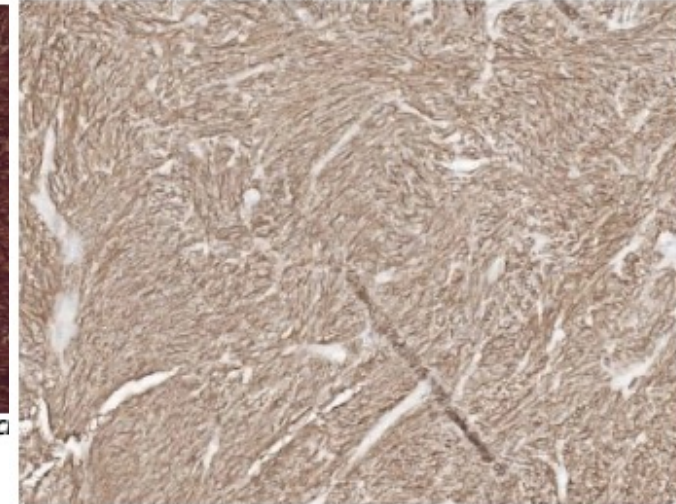
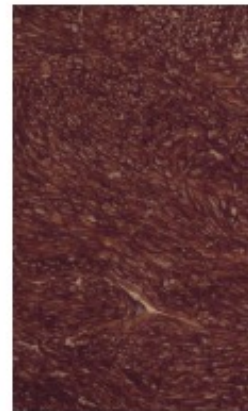
DIAGNOSTICO FINAL:

- TUMOR DEL ESTROMA GASTROINTESTINAL, FUSOCELULAR.
- HUBO EXPRESIÓN DE CD117, DOG-1 y CD34.
- CLASIFICACIÓN FINAL EN LA PIEZA DE RESECCIÓN.

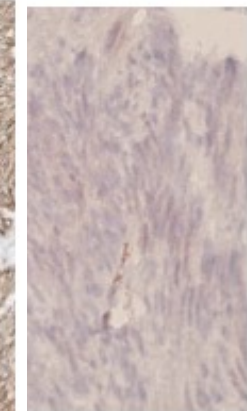
CPT: 88342

DESCRIPCIÓN MACROSCÓPICA/MICROSCOPICA

Se evalúa el tejido TPC23-1046 B. se hacen cortes en laminillas cargadas para realizar los estudios de Inmunohistoquímica solicitados en el equipo BenchMark de Ventana con controles apropiados.



CD117



Pathology
Report

Which would be the most appropriate step?

- Start neoadjuvant Imatinib
- Proceed with surgical excision and give imatinib as adjuvant therapy
- Request a NGS (repeat biopsy for tissue) for potential resistance to imatinib (PDGFR 18 D842V)
- Any further recommendation?