

SELNET MDT

Jan 30, 2025

Dr. Luis Matamala

Dr. Nicolas Devaud

Cases

- Case 1 - Instituto Nacional del Cancer, Chile
- Case 2 – Fundación Arturo López Pérez, Chile
- Case 3 – Instituto Valenciano de Oncología, España
- Case 4 – Hospital Nacional de Niños, Costa Rica
- Case 5 – Instituto Alexander Fleming, Argentina

Case 1

Instituto Nacional del Cancer, Chile

CASO CLINICO

Dra. Javiera Diaz Espinosa

Residente Segundo Año

Cirugía Oncológica Mención Digestivo y Partes Blandas



Facultad de Medicina
Clínica Alemana - Universidad del Desarrollo



33 AÑOS, FEMENINO. SIN ANTECEDENTES MORBIDOS.

Historia de paresia progresiva de EEl izquierda desde febrero 2024, evolucionando en marzo con lumbalgia aguda, por lo cual finalmente se realiza estudio.

RMN 15.03.2024:
Gran masa tumoral de aspecto solida, con matriz condroide a nivel de pelvis izquierda con características de condrosarcoma.



TC Pelvis 02.04.2024:
Lesión ósea exofitica dependiente del pilar anterior del acetábulo izquierdo, con voluminoso componente hipodenso asociado a multiples componentes cálcicos distróficos de distribución difusa.
20x13.7x13.5 cms



TAC TAP 14.04.2024:
Sin evidencias de lesiones secundarias.



IMAGENES

BIOPSIA

Proliferación condroide con leve atipia.

S-100 (+), SOX 10 (-), D2 40 (+)

Conclusión: Tumor condroide atipico compatible con condrosarcoma de bajo grado.

OPCIONES

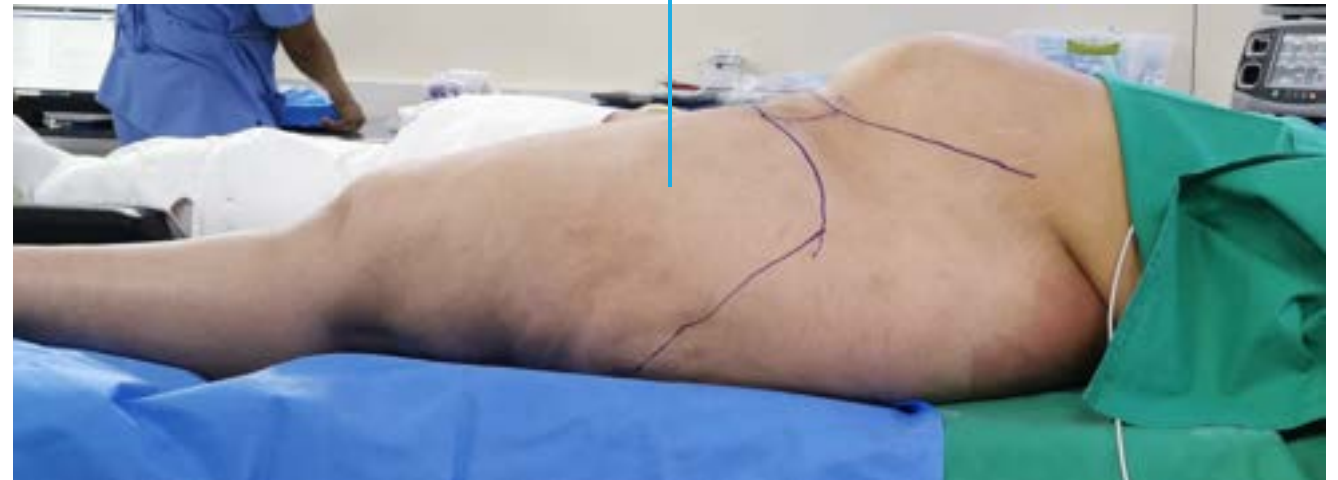
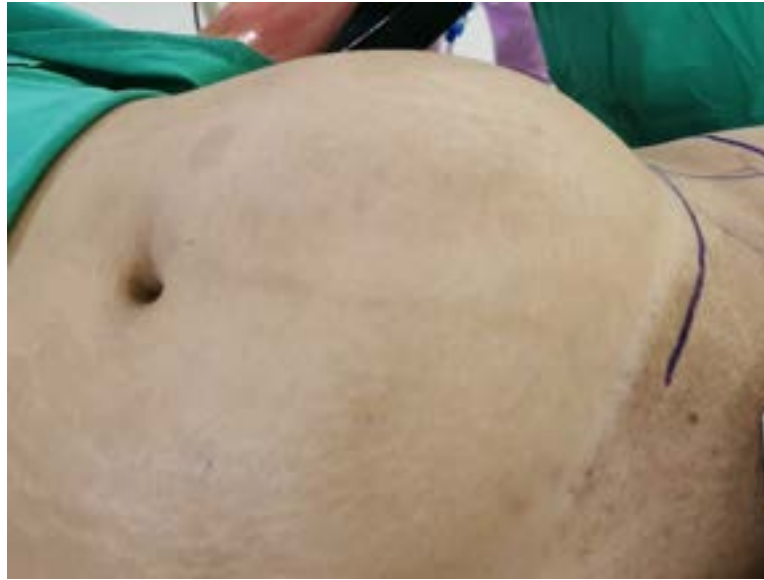
A) RT+ cirugía

B) Cirugía mediante tumorectomía

C) Hemipelvectomia

D) Cuidados Paliativos

CIRUGIA





CIRUGIA

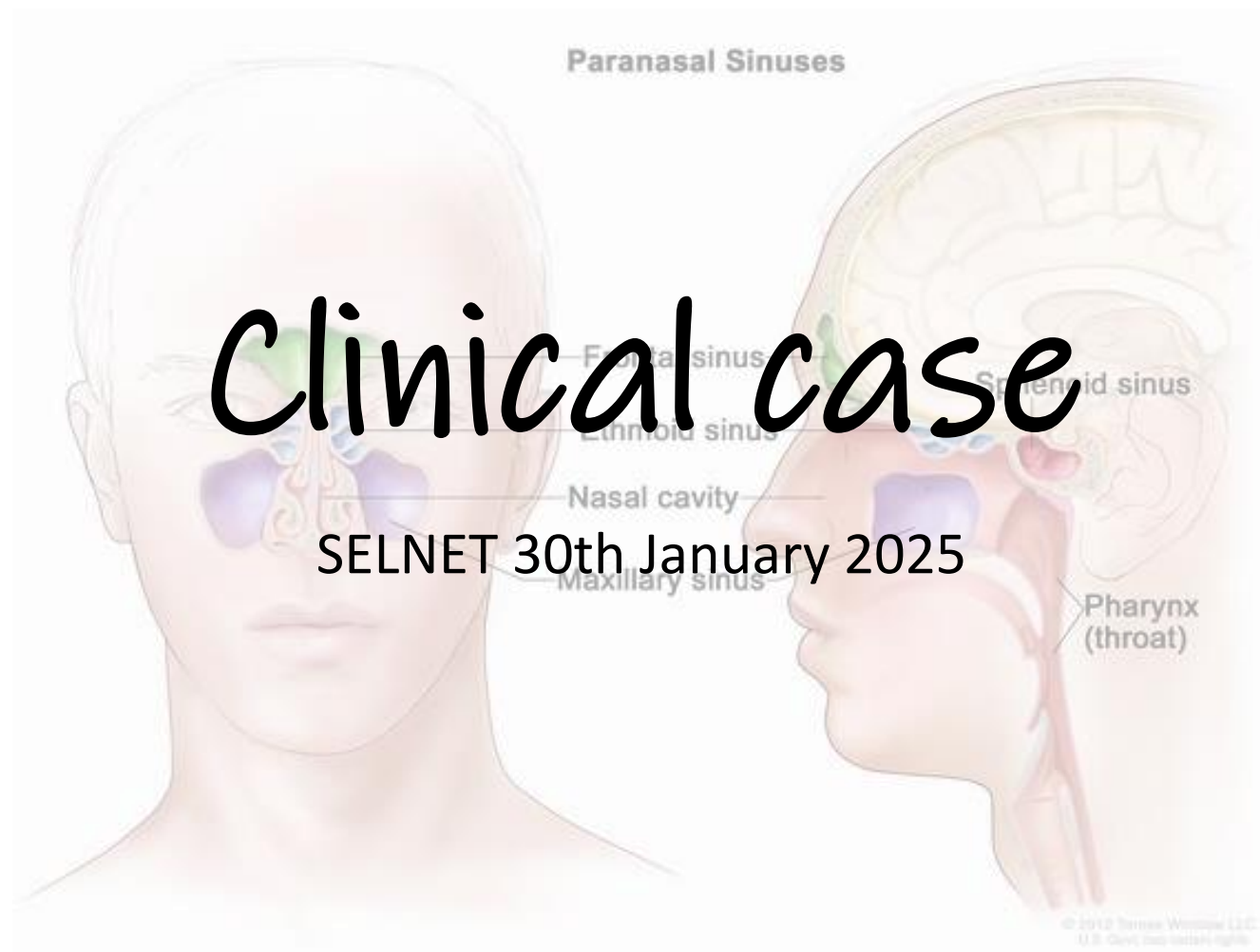
14 DIAS POST OPERATORIO

Paciente en buenas condiciones generales,
deambulando con carro.



Case 2

Fundacion Arturo Lopez Perez, Chile



28
years
old,
male

Medical history (-)

Prior surgery (-)

Current medications (-)

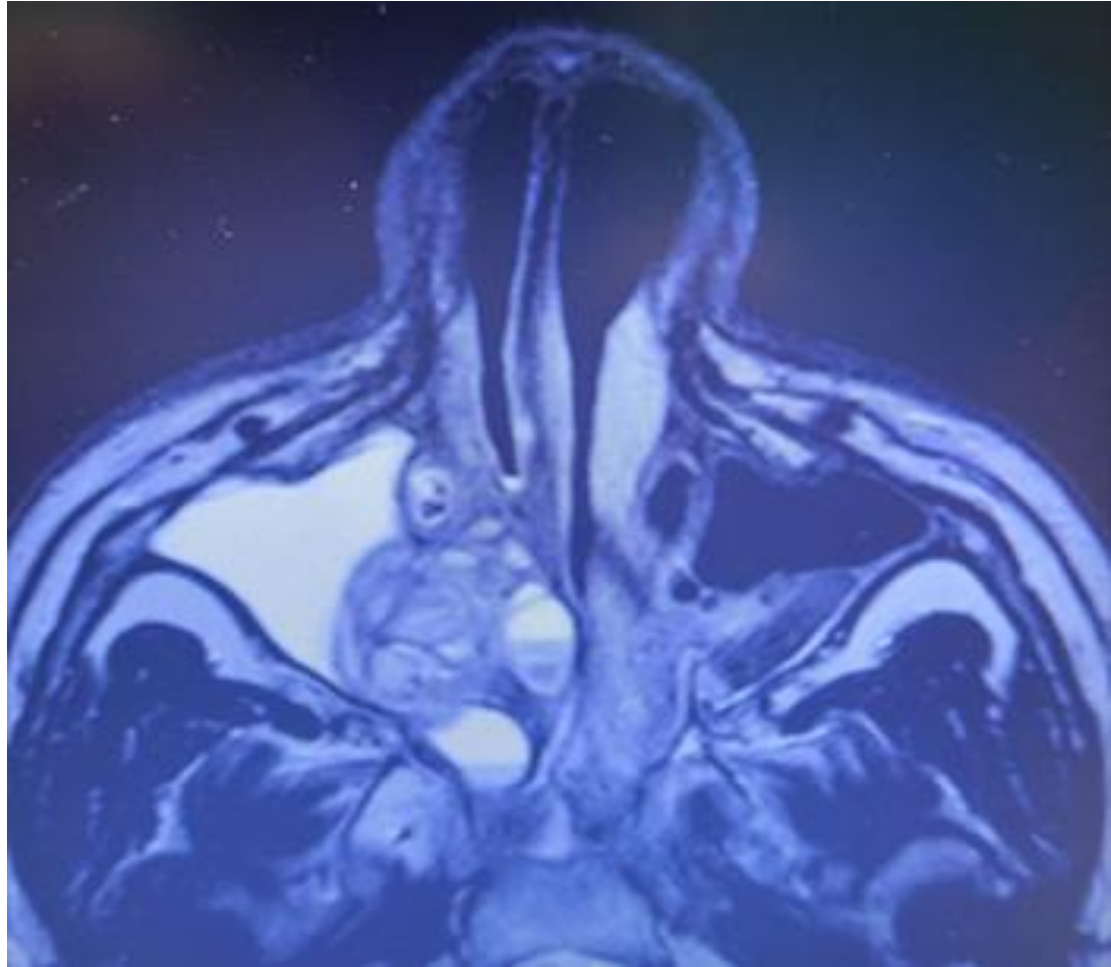
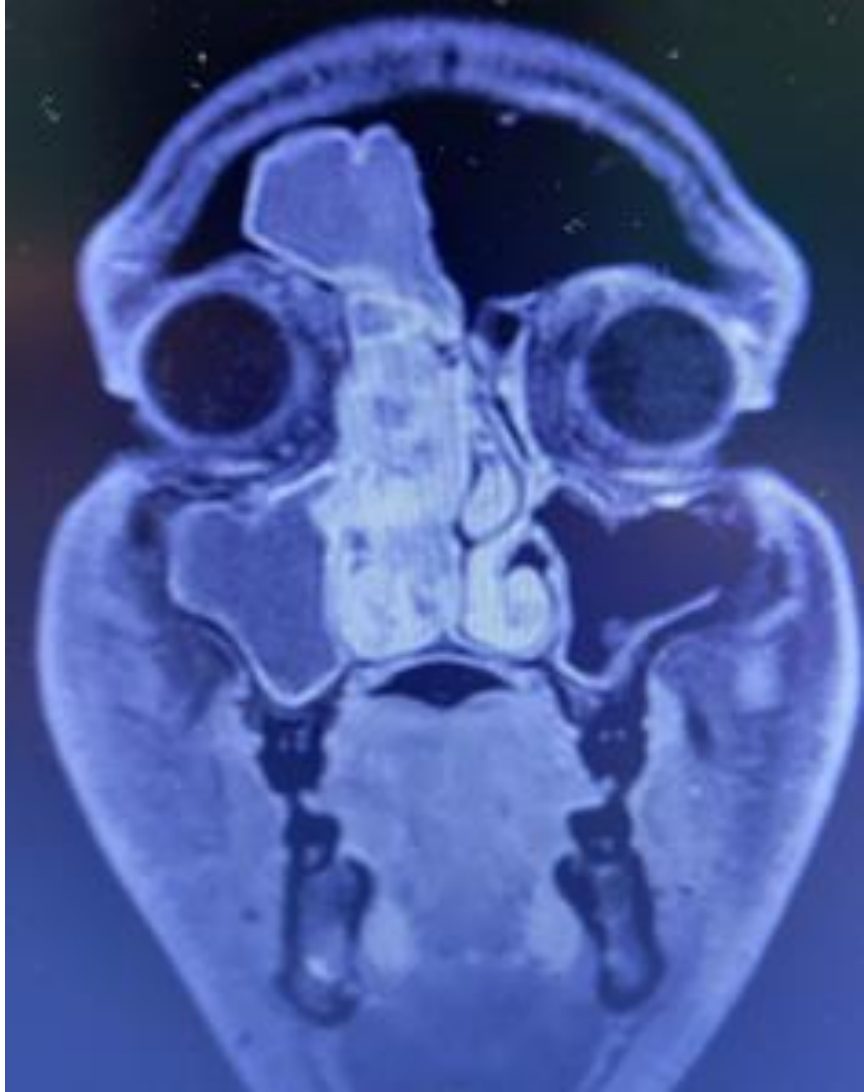
Smoking (-)

Alcohol or drugs consumption (-)

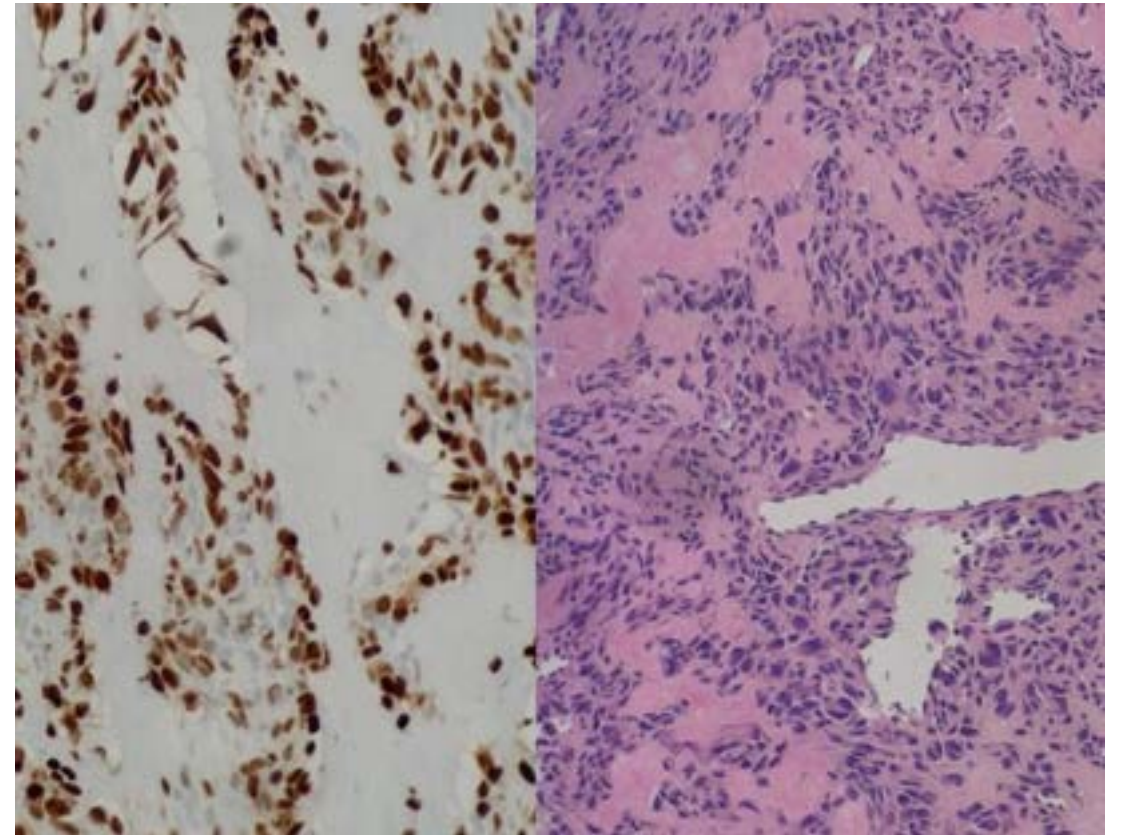
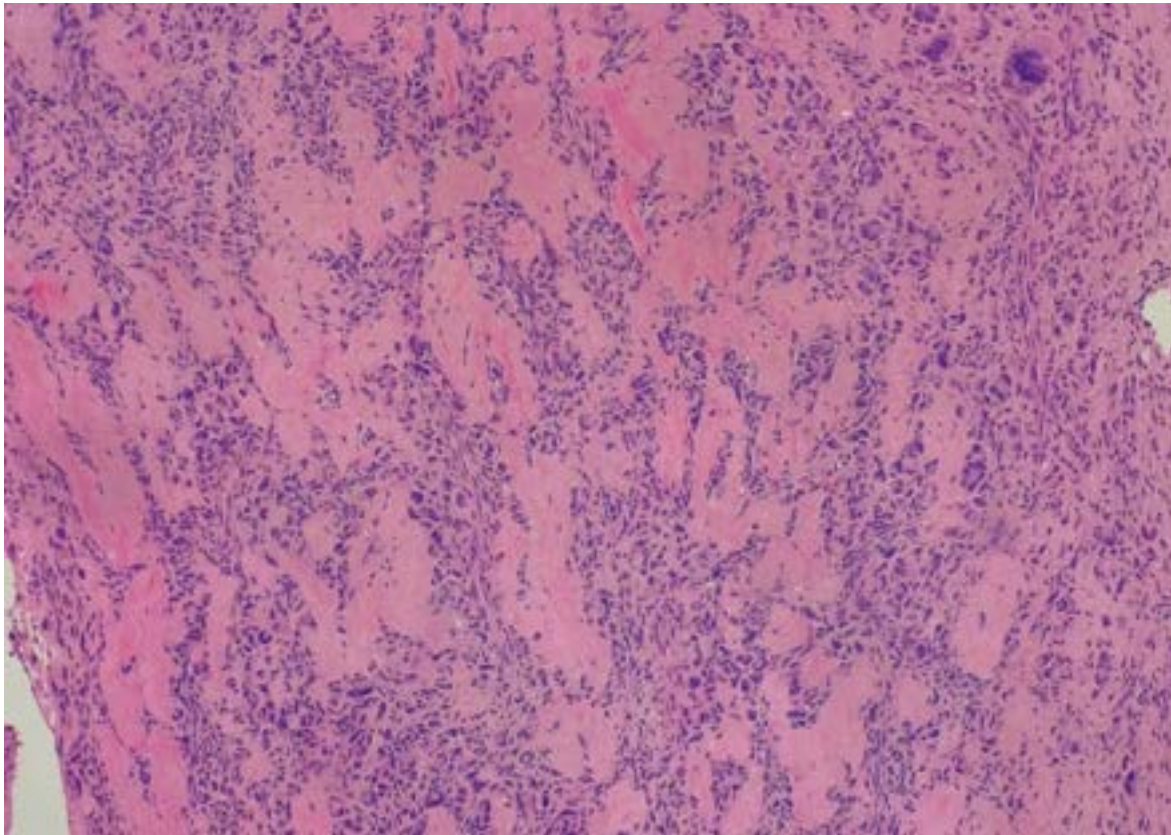
No family history for cancer



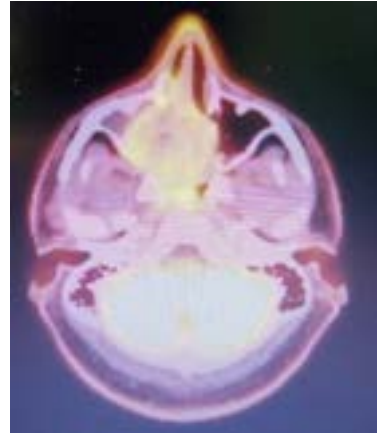
- **April 2024:** Persistent Flu-like symptoms
- **July 2024:** Massive epistaxis. Requires anterior and posterior tamponade
- **31.07.24 Paranasal sinuses MRI:** Expansive process with a neoplastic appearance centered in the right nasal cavity, involving the ethmoid and the right maxillary ostium, measuring 48x32x39mm. It appears to infiltrate the middle third of the nasal septum. There is no orbital involvement.
- **08.08.24 Chest abdomen and pelvis CT:** no evidence of metastasis



- First Biopsy **12.08.24**: Mucosa infiltrated by poorly differentiated malignant neoplasia compatible with fusocellular and pleomorphic sarcoma rich in high-grade multinucleated giant cells.

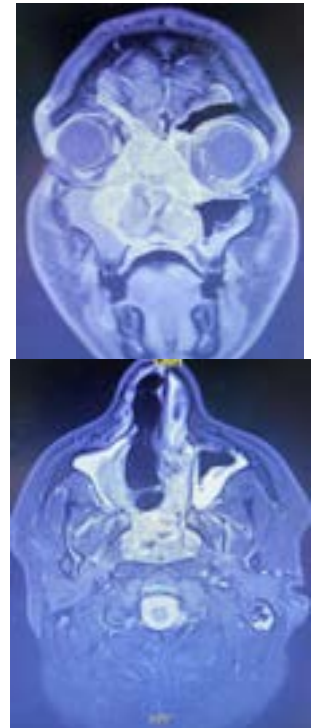


- **03.10.24 PET/CT:** Expansive solid hypermetabolic mass in the right nasal cavity, locally infiltrative, compatible with a primary reference tumor. Slightly hypermetabolic sclerotic rib lesions, with an unspecific appearance.



- New biopsy **8.10.24:** Convencional central Osteosarcoma G3

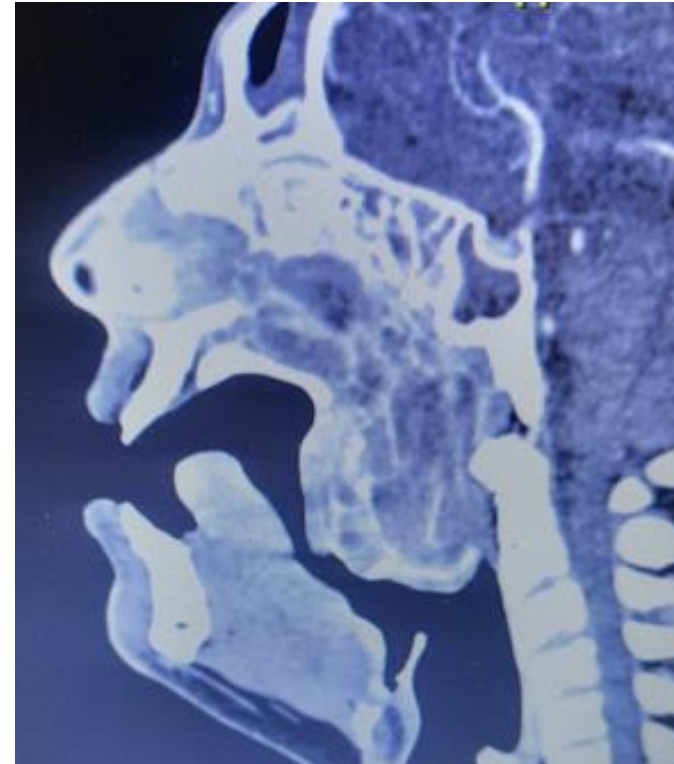
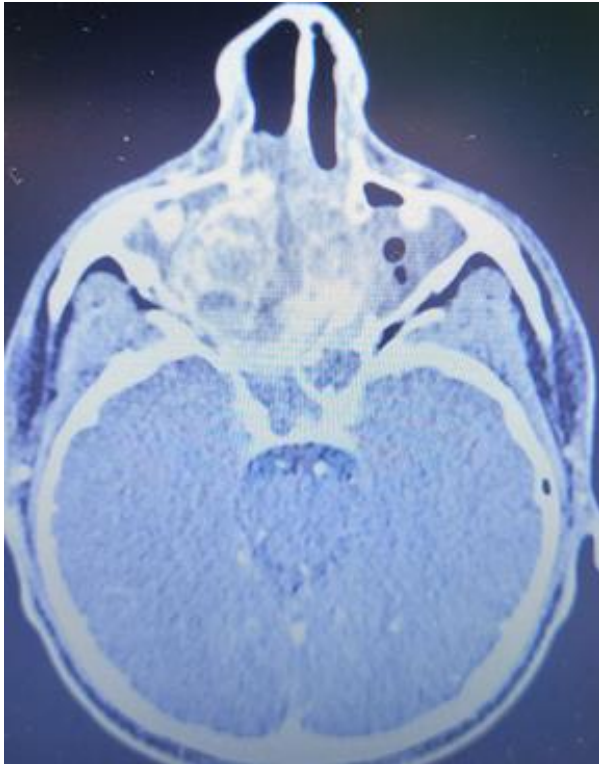
- **16.10.24 MRI Paranasal sinuses:** Solid mass in the right nasal cavity with extensive extension into the left nasal cavity, with an obstructive appearance, extending towards the right orbit, right maxillary sinus, sphenoidal sinus, and extensively towards the nasopharynx.



Discussed in oncology committee **22.10.24:**

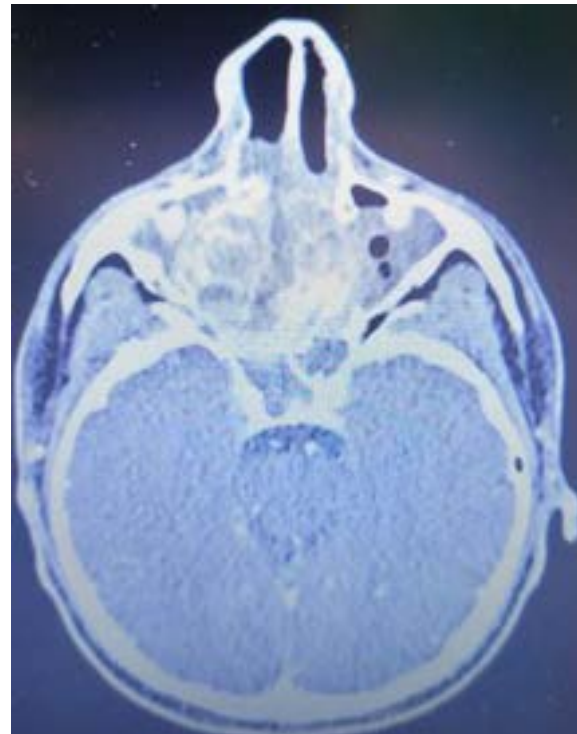
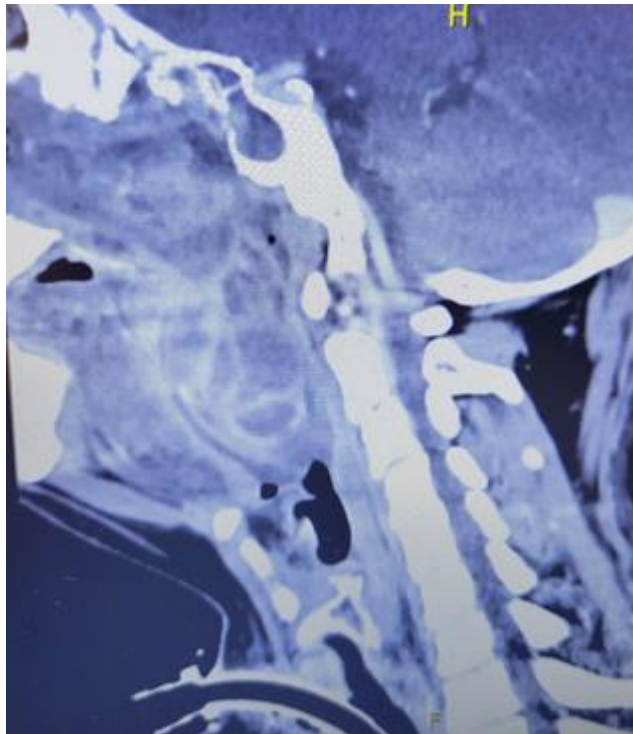
- Osteosarcoma cT3N0M0, stage II
- Neoadjuvant chemotherapy with MAP x2C, and new evaluation for possible resection.
- **24.10.24:** Begins 1st cycle, well tolerated. Discharged 3 days after.
- **01.11.24:** Recurrent epistaxis → New hospitalization
- **02.11.24:** No possible surgical intervention nor embolization.
Hemostatic RT (8Gy in 1Fx)

- **02.11.24 PNS CT:** Solid hypervascularized lesion in the nasal cavities with involvement of the medial and posterior walls of the right maxillary sinus, the medial wall of the left maxillary sinus, and the anterior wall of the sphenoidal sinus



- Physical examination → partial obstruction of aerodigestive pathways
- **06.11.24:** profilactic Traqueostomy + gastrectomy
- **14.11.24:** C1 D22 → 1st HD MTX
- **15.11.24:** New episode of epistaxis → Radiotherapy 25Gy in 5Fx
- **24.11.24:** C1D29 HD MTX → Delayed clearance

03.12.24 PNS CT: Expansive tumor process occupying the nasal cavities. Posterior and caudal extension, reaching the epiglottis, obstructing the airway, with significant expansive involvement. Infiltration of structures in the right orbit, nasal septum, and right pterygomaxillary fossa. Shows **growth** compared to the previous study from 02.11, particularly **caudal extension**



- **04.12.24** New discussion in MDT board
 - C2 MAP suspended for toxicity
- **17.12.24** Surgery with tumor resection + bifrontal craniotomy + right orbital exenteration + right maxillary segmental osteotomy + flap reconstruction
 - Bp: high grade osteoblastic osteosarcoma R1/R2, LN 0/5 group II, ILV (-), necrosis 80%

Questions to the board:

Is it a new PD?

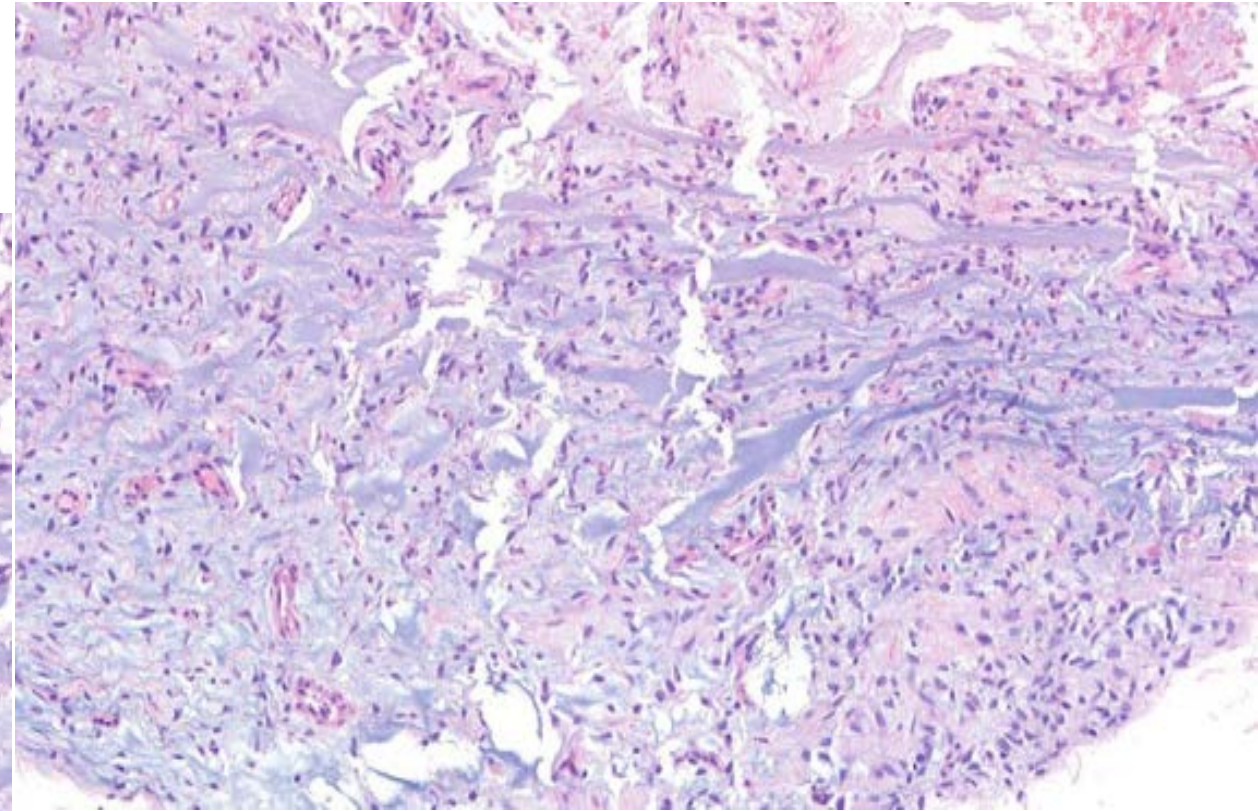
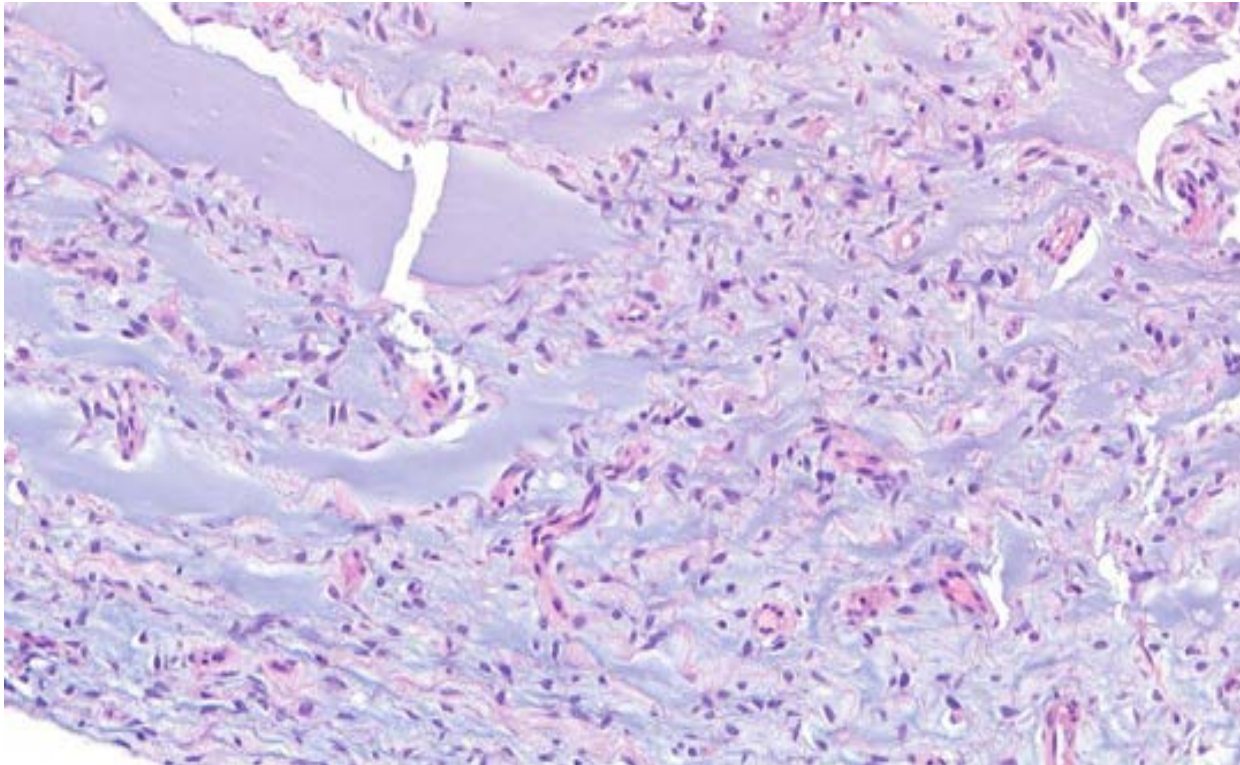
If not, should we continue systemic therapy?

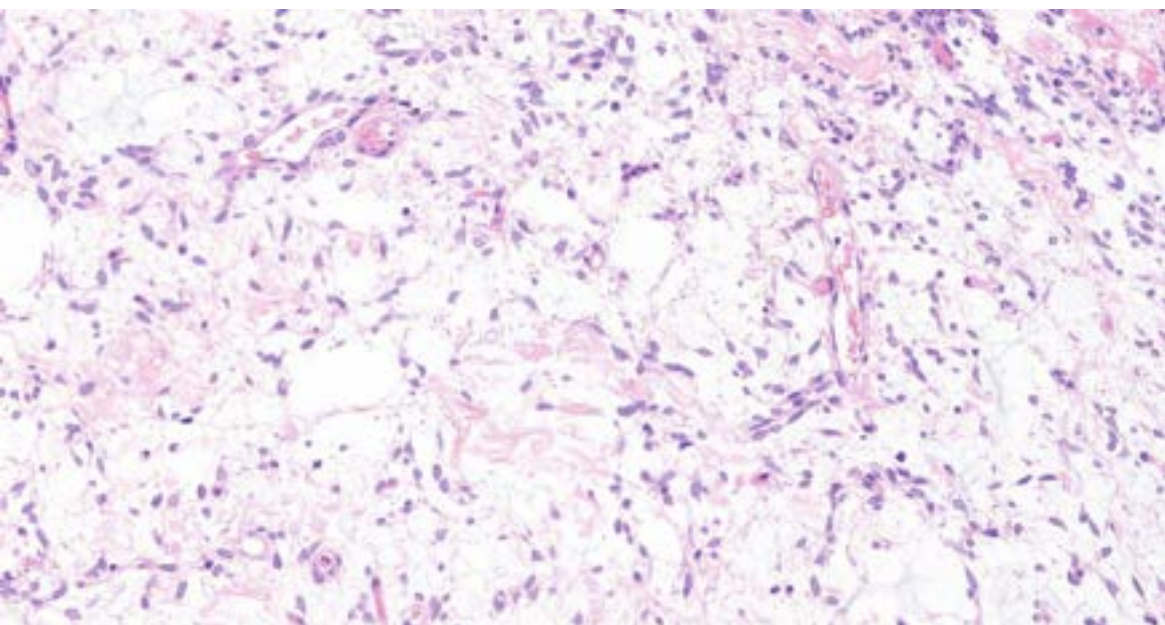
Case 3

Instituto Valenciano de Oncología, España

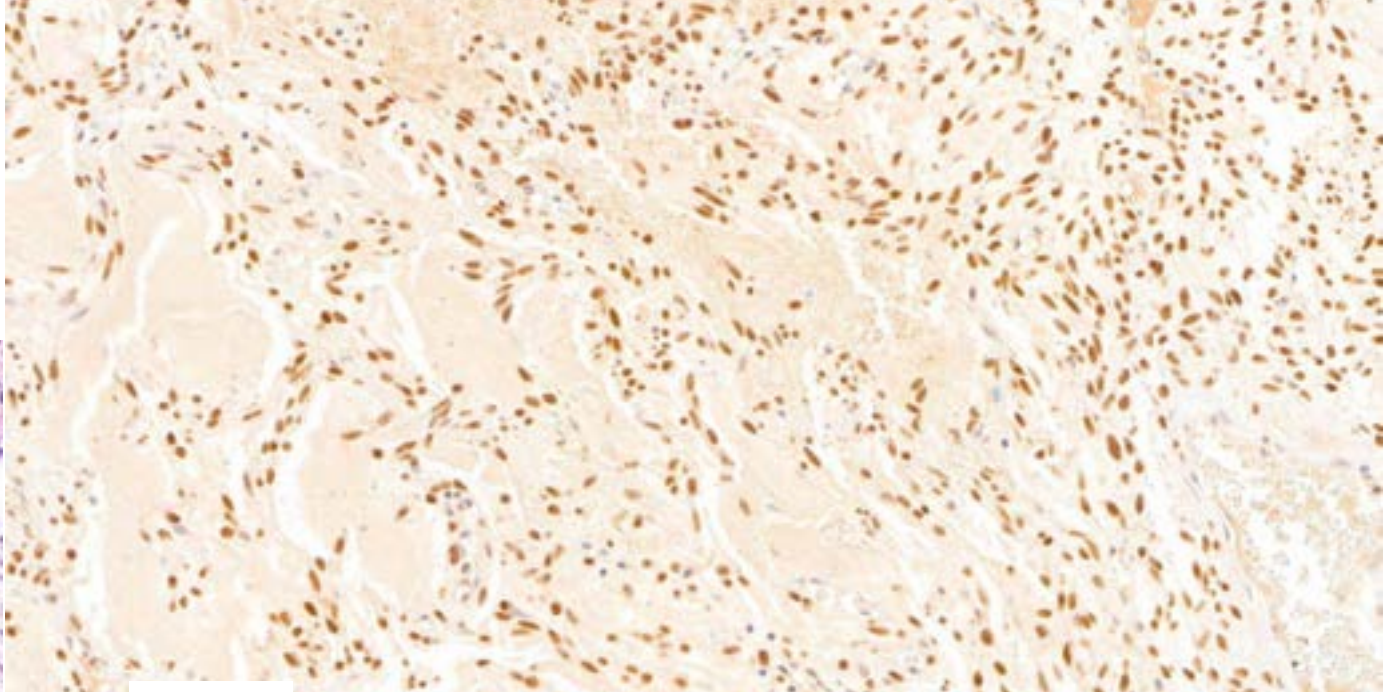


A 59-year-old male with a pleural tumor (4cm) underwent surgical tumor removal, and a diagnosis of myxoid liposarcoma was established (CD99: positive, CD34: negative, S100: focal). Immunohistochemical markers for epithelial, vascular, and myogenic differentiation were negative. FISH analysis revealed EWSR1 negativity and no MDM2 amplification. Neither FUS and DDIT3 studies nor NGS were available. One surgical margin was positive, and the patient subsequently received radiotherapy. Eleven years later, the patient experienced a tumor recurrence, which was treated with surgical resection.

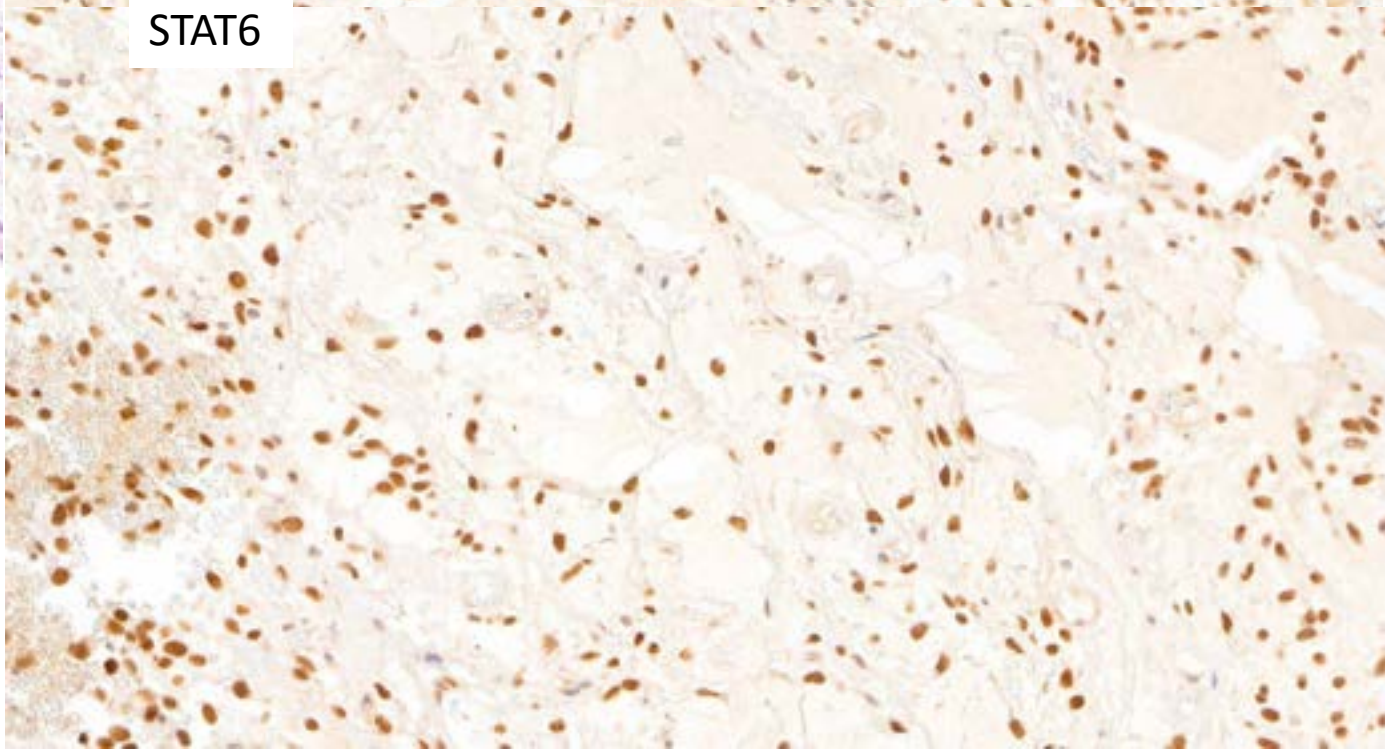




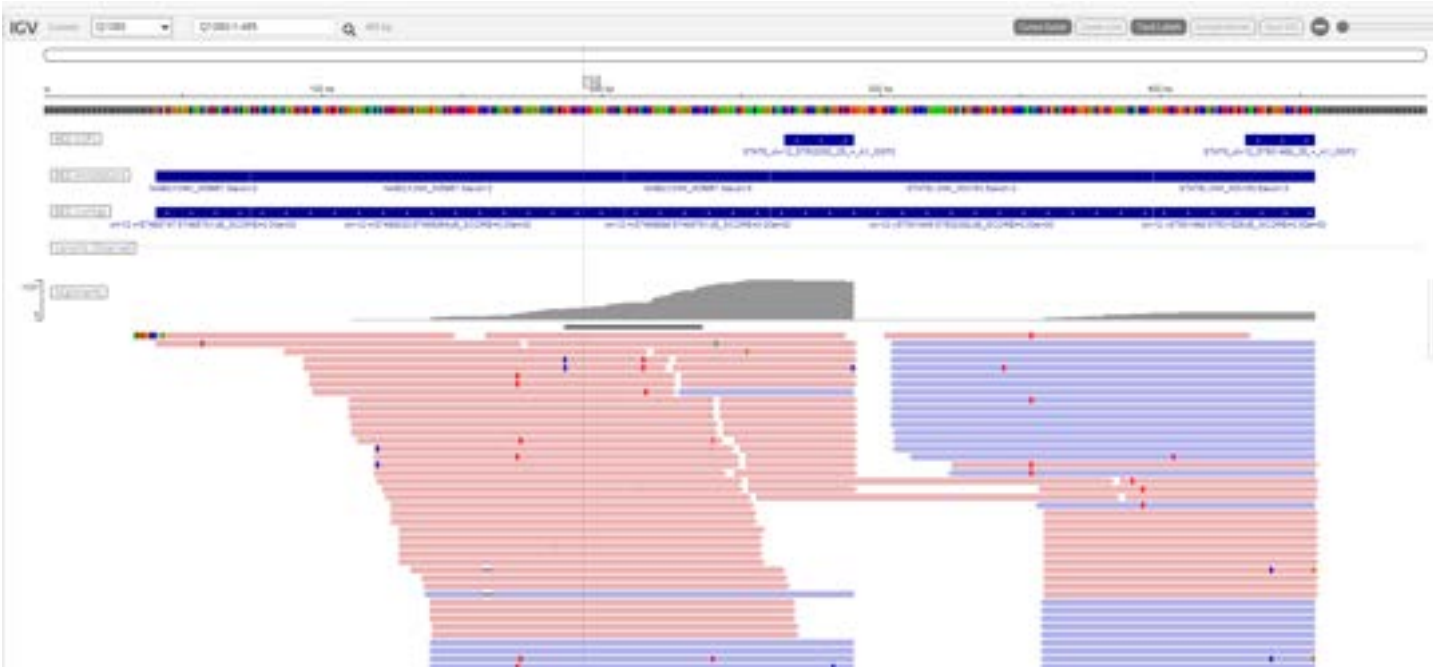
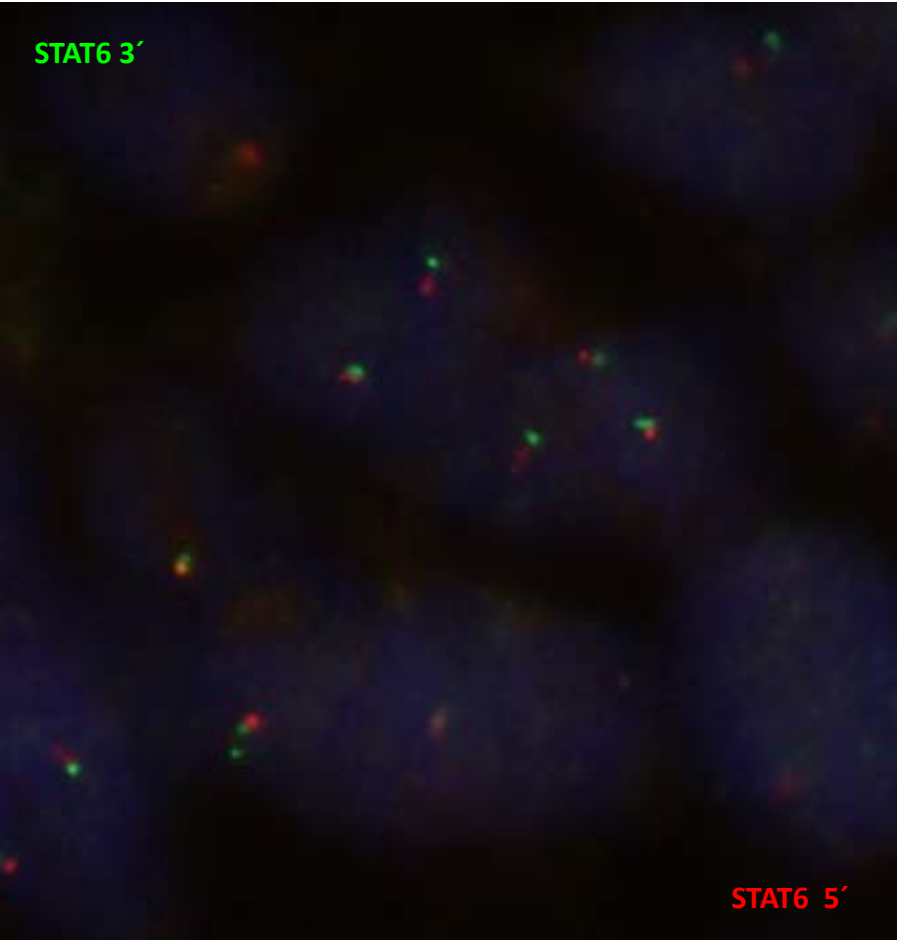
Recurrence

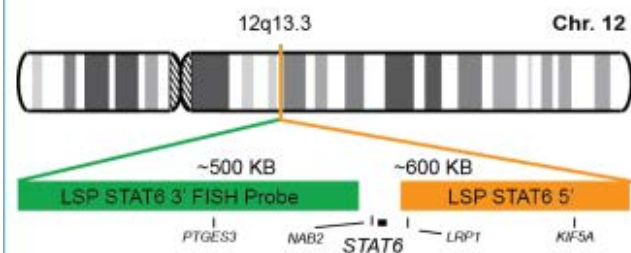


STAT6

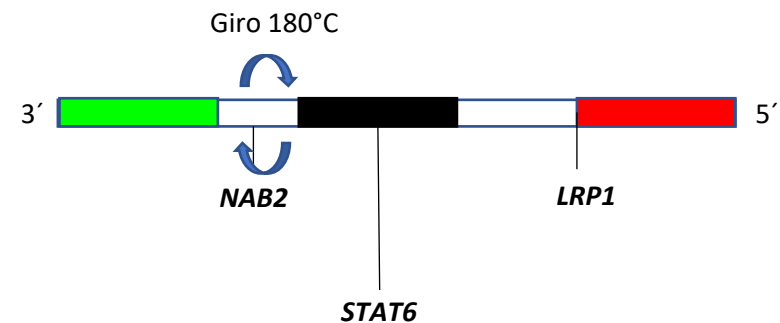
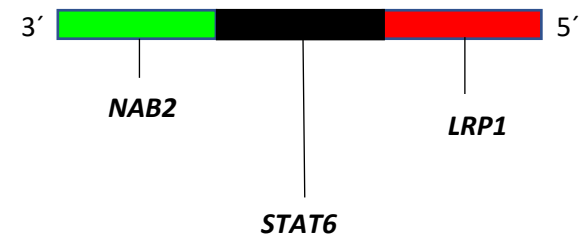
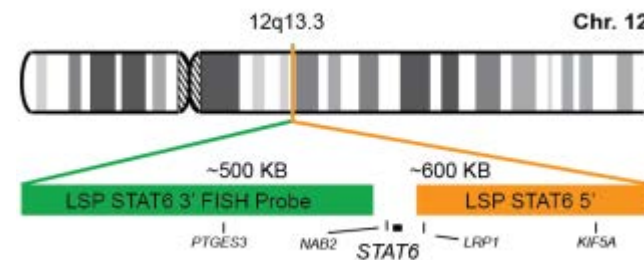


STAT6- present case (BREAK APART PROBE FISH)

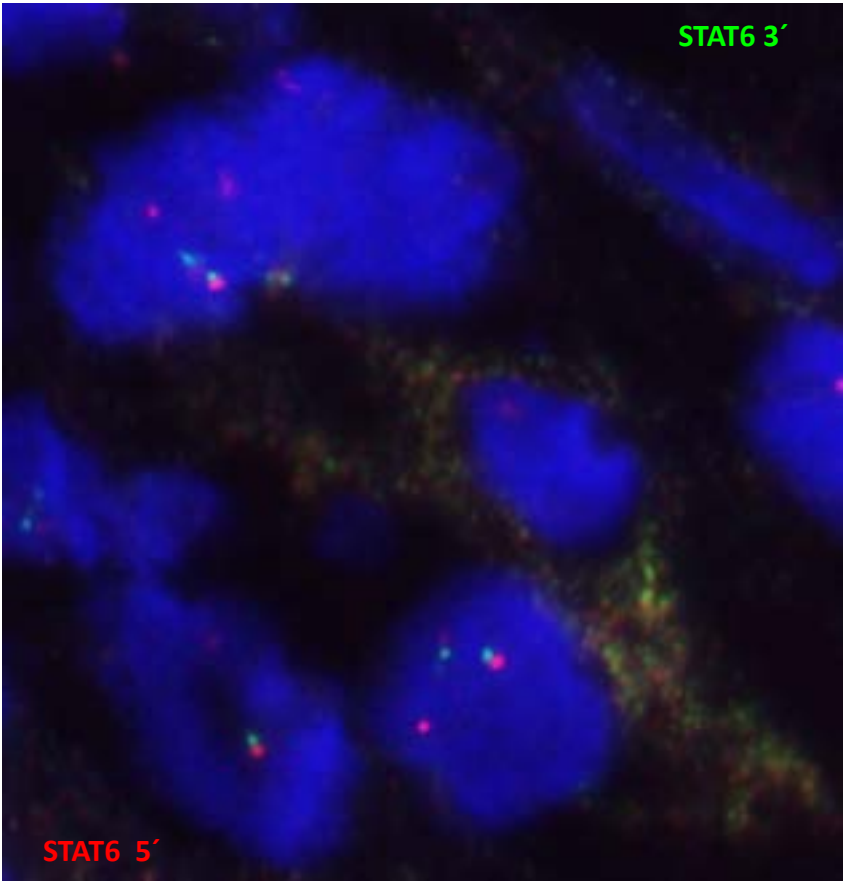




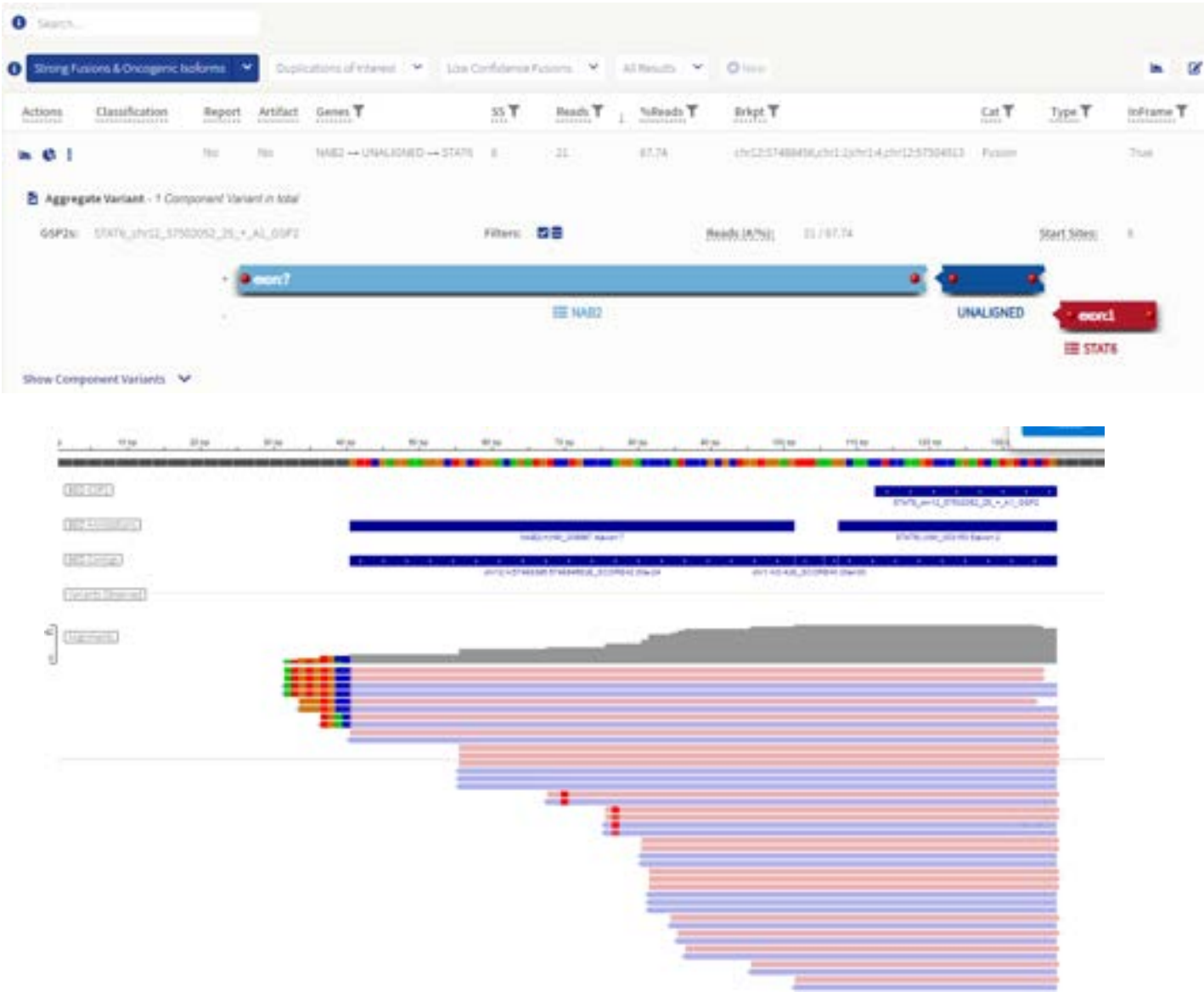
LSP *STAT6* 5' FISH Probe covers a genomic area upstream of the 5' (start) portion of the *STAT6* locus and some adjacent genomic sequences. LSP *STAT6* 3' FISH Probe covers sequences adjacent to the 3' (end) of the gene. The two probes are flanking sequences across the *STAT6* locus in which variable breakpoints have been observed.



STAT6- other case (BREAK APART PROBE FISH)



STAT6 translocation by FISH



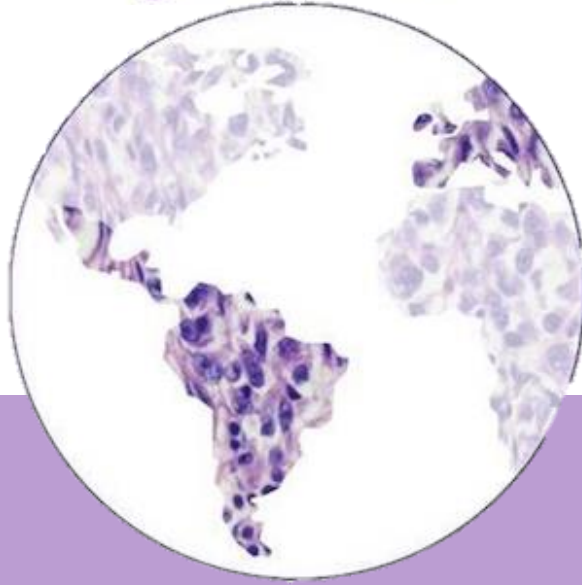
Summary Points

- The STAT6 FISH result may vary (positive or negative) depending on the breakpoint and the design of the probe.
- STAT6 IHC is a very good surrogate with high specificity and sensitivity (Differential diagnosis: DDLPS or *GLI*-amplified neoplasms).
- Be cautious of myxoid SFT mimicking Myxoid liposarcoma.
- All myxoid lung tumors should be tested for STAT6.

Case 4

Hospital Nacional de Niños, Costa Rica

SELNET



MDT

**DR AMANDA LEIS
PEDIATRIC ONCOLOGY
HOSPITAL NACIONAL DE NIÑOS
COSTA RICA**

January 2025

Medical history

- Female, current age 14 yo
- Diagnosis: Spindle cell synovial sarcoma of the right cervical region (involving nerve root C6-C7)
 - Initial diagnosis January 2023
 - Age at diagnosis 12 yo
- Initial CT scan December 2022:
 - Solid mass in the supraclavicular area, initiates at the level of transverse process of C6 and extends to the lung apex, encompassing the subclavian artery, dimensions 39 x 39 x 50 mm.
 - Solid nodule in the lateral segment of right middle lobe of 6x4 mm.
- Initial surgery: Enucleation of tumor in January 2023
- Pathology report: Synovial sarcoma - spindle cell type
 - Histological grade 2, with compromised margins.
 - FISH: SSY18 (SYT) gene in Cr 18q11.2: chromosomal rearrangements in > 90% of analyzed cells.

Details of diagnosis and treatment

- Excisional biopsy of residual tumor in May 2023
 - Pathology report was negative
- Protocol COG NSRTS 0332 – High risk
 - Ifosfamide 9g/m²/cycle total 6 cycles
 - Doxorubicin 75mg/m²/cycle total 5 cycles
 - January to Sept 2023
- IMRT: post surgical
 - Boost tumor bed. Total dose 55.8 Gy.
 - June to August 2023
- End of treatment scans
 - CT: small solid residual lesion considered scar tissue
 - MRI: no lesions or masses

Treatment Arm D – Neoadjuvant Chemoradiotherapy							
Induction Phase				Surgery Phase	Continuation Phase		
Week 1	Week 4	Week 7	Week 10	Week 13	Week 16	Week 19	Week 22
I	I	I	I	Surgery	I	I	
D	D				D*	D**	D
Radiotherapy†					Radiotherapy†		

Details of relapse

11 months follow-up

- MRI August 2024: nodular image in right supraclavicular area of 1.3 x 0.9 cm, that shows continuation with nerve root C6, posterior to subclavian artery.
- MRI November 2024: same nodular image 3.8 x 1.5 x 2.2 cm.
- Lung CT scan Dec 2024: Solid nodule in the lateral segment of right middle lobe has remained stable. No new nodules.

Navegación 1

Ser: 2
Ubic: 3.2, 11.8, 0.0 mm
50 / 350

HOSPITAL NACIONAL DE NIÑOS

10-Dic-2024 7:29:25 A.M.

TF: 23/Paso: 0.6
kv 120/mA 95/500mseg
Inclinación: 0.0
Espesor 3 mm/FOV 29.4 cm
7:35:43 A.M./C+

Navegación 3

Ser: 2
Ubic: 0.0, 0.0, 54.0 mm
50 / 350

HOSPITAL NACIONAL DE NIÑOS

10-Dic-2024 7:29:25 A.M.

TF: 23/Paso: 0.6
kv 120/mA 95/500mseg
Inclinación: 0.0
Espesor 3 mm/FOV 29.4 cm

Navegación 2

Ser: 2
Ubic: -40.0, 11.0, 0.0 mm
50 / 350

HOSPITAL NACIONAL DE NIÑOS

10-Dic-2024 7:29:25 A.M.

TF: 23/Paso: 0.6
kv 120/mA 95/500mseg
Inclinación: 0.0
Espesor 3 mm/FOV 29.4 cm
7:35:43 A.M./C+

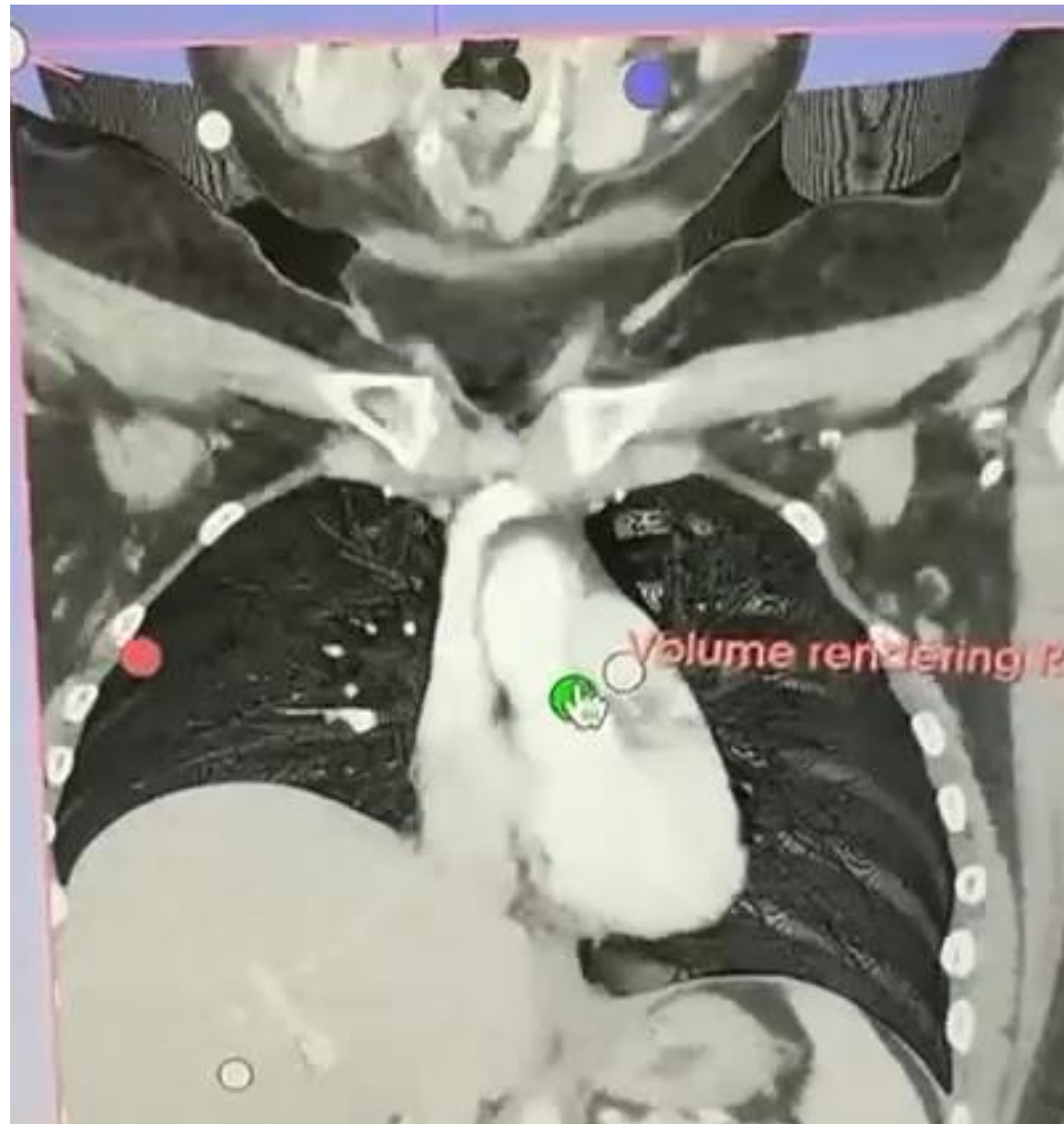
Resultados

Ser: 2
Ubic: 3.2, 11.8, 0.0 mm
50 / 350

HOSPITAL NACIONAL DE NIÑOS

10-Dic-2024 7:29:25 A.M.

TF: 23/Paso: 0.6
kv 120/mA 95/500mseg
Inclinación: 0.0
Espesor 3 mm/FOV 29.4 cm



Questions for discussion

Considered **inoperable** by pediatric and adult surgical teams

Re-irradiation therapy is possible

- Second line chemotherapy ?
- Other suggestions ?

Case 5

Instituto Alexander Fleming, Argentina

Female
30 Y/O

Current disease

Control

2020

begins with sports-related pain

01/2022

MRI: 4.5cm injury to the coccyx bone

03/2022 Surgery in Chaco

- Pathology report: Cordoma, 7cm
- Review at reference center: Conventional cordoma

07/2023

MRI: PD, 2 nodes in gluteo area

RT in the pelvic (sep-oct 2023)

Clinical progression with pain
(without neurological symptoms)

Female
30 Y/O

Current disease

07/2023

MRI: PD, 2 nodes in gluteo area

RT in the pelvic (sep-oct 2023)

Clinical progression with pain
(without neurological symptoms)



Palliative surgery with good
(pain) response

03/2024

Hemicorporectomy (with cure
intention)

1th time in Fleming: we decided
Imatinib

Female
30 Y/O

Current disease

03/2024

1th time in Fleming: we decided
Imatinib

08/2024

PD (with numer and size)

Female
30 Y/O

Current disease

03/2024

08/2024

1th time in Fleming: we decided
Imatinib

PD (with numer a

Main in/exclusion criteria



Inclusion criteria

- Locally advanced or metastatic, pathologically proven, EGFR expressing chordoma, amenable for local therapies
- Progression of disease according to RECIST 1.1 in 6 months before study entry
- Age 18 years and up
- Availability of archival tumor material or new biopsy
- Adequate bone marrow, liver and renal function

Exclusion criteria

- ECOG performance status >2
- Known history of interstitial lung disease, poorly controlled gastro-intestinal disorder, clinically relevant cardiovascular abnormalities

Female
30 Y/O

Current disease

03/2024

08/2024

1th time in Fleming: we decided
Imatinib

PD (with nur

De-differentiated Chondrosarcoma (DDCS)

Targeted / Immune-checkpoint Inhibitors in advanced / metastatic DDCS

Ref	Drug	No. pts	Time on Rx weeks / PFS (mth)	BEST ORR (RECIST) n, (%)
Targeted agents				
Bui	Pazopanib/	6	11 (6 - 20)	SD
	Regorafenib/ sorafenib/ Everolimus/ temozolimus	2	16 (12-20)	SD
Immune-checkpoint inhibitors				
Bui	Ipi/nivolumab	4	11 (1-24)	SD
	Pembrolizumab	5	21 (12-48)	PR
	Atezolizumab	1	12	PD
Salken ¹	Atezolizumab	9	9 weeks	SD
Paoluzzi ⁵	nivolumab	1	> 13 months	PR
Singh ⁶	Pembrolizumab	1	24 months	CR

Immune-landscape not well understood

- PDL1 +: 42-52%¹
- High T-cell infiltration²
- Hints of activity in small numbers of pts / case reports

→ Disruption of angiogenesis could enhance the efficacy of immune-based cancer therapies³

Female
30 Y/O

Current disease

03/2024

1th time in Fleming: we decided
Imatinib

08/2024

PD (with numer and size)

10/2024

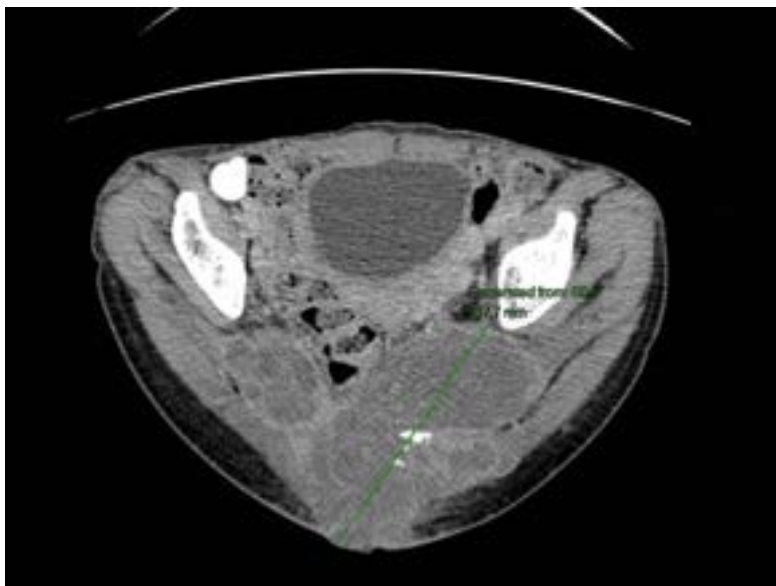
Pembrolizumab

Female
30 Y/O

Current disease

01/2025

Pembrolizumab 5 cycles



Questions for de MDT

Next step?

- **Other TKI**
- **Chemotherapy**
- **Surgery (hemicorporectomy? Family support?)**
- **Clinical trial? (in Argentina its not an option)**