

SELNET MDT 23 May 2024





| Case | Diagnosis | Site | Country |
|------|--------------------------------------|---------------------|-----------|
| 1 | Locally Advanced Left Leg Myxoid LPS | Dr. Boris Itkin | Omán |
| 2 | Desmoplastic small round cell tumor | Dr. Fernando Campos | Brazil |
| 3 | Maxillary osteosarcoma | Dr. Soulé Tomás | Argetnina |
| 4 | Angiosarcoma | Dra Eliza Ramirez | Paraguay |
| 5 | GCT | Dr. Pablo Segura | Argentina |



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SELNET

Fernando Campos, MD

Medical Oncologist A.C.Camargo Cancer Center Sao Paulo, Brazil

May 2024

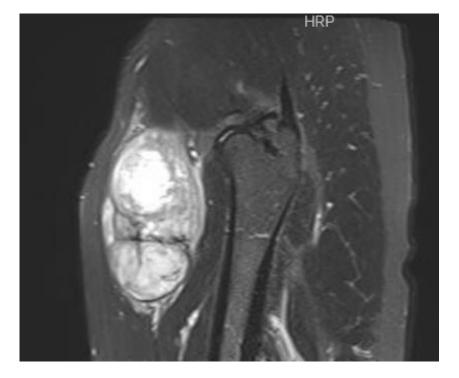


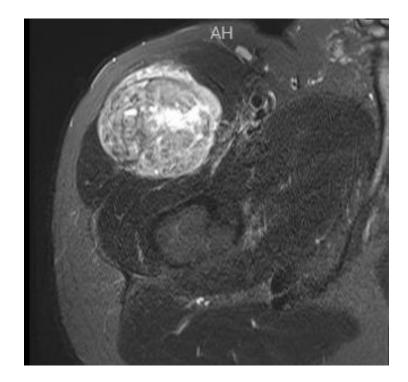
- 52 yo male
- No comorbidities

ONCOLOGICAL SUMMARY:

- Sept 2023: Progressively growing mass in the right thigh. He reports mild discomfort in the area. No other symptoms.
- Feb 2024: MRI Solid and heterogeneous expansive formation, centered in the intramuscular planes of the proximal third of the thigh, next to the muscular belly of the rectus femoris, with heterogeneous contrast enhancement, measuring approximately 12.5 x 7.0 x 6.5 cm (CC x LL x AP). It presents some areas of spontaneous high signal on T1, which may represent hematic content/necrosis. There is also a slight edema of the adjacent muscular planes.
- April 2024: Chest CT no metastasis
- April 2024: PET-CT FDG uptake only in the thigh mass SUV 16.3.







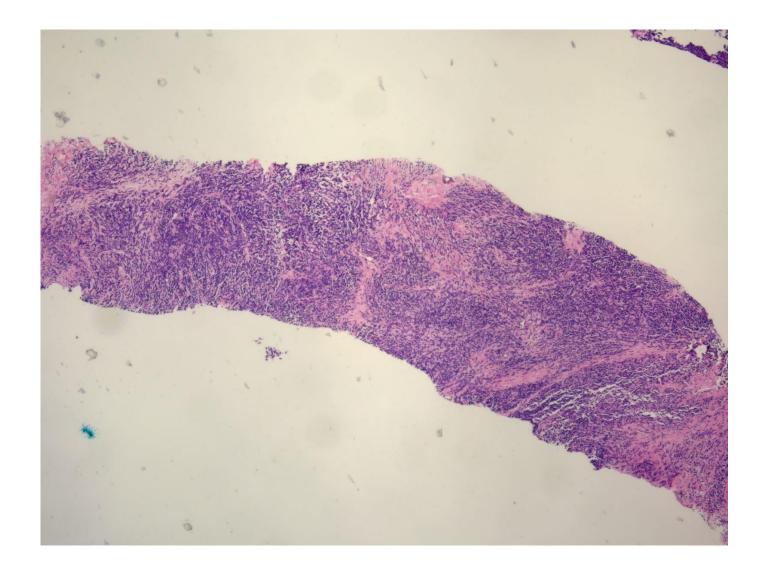


• March 2024: Biopsy – Undifferentiated small cell sarcoma, which may correspond to desmoplastic small round cell tumor.

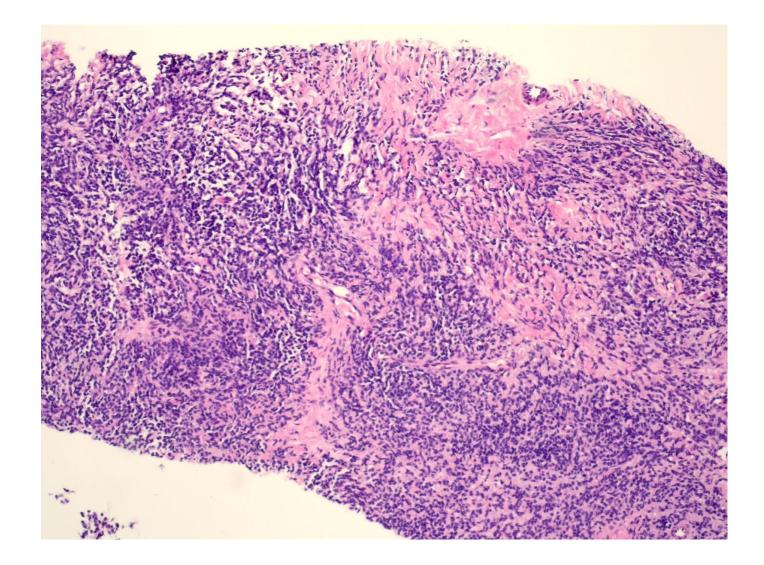
- Patient was being treated in another hospital – a DNA methylation profiling test was requested

- April 2024: patient come to our hospital for 2nd opinion and treatment. Pathology diagnosis review was performed, also corroborating desmoplastic small round cell tumor.
- April 2024: DNA methylation profiling test consistent with DSRCT.

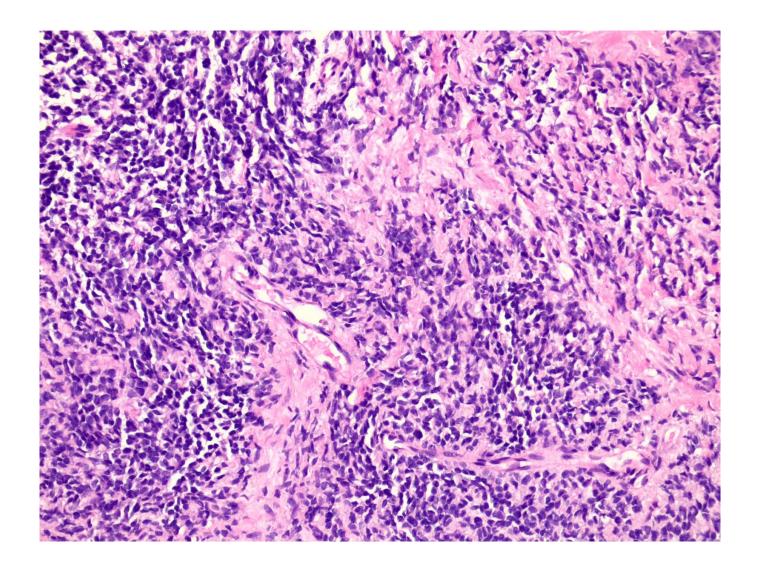




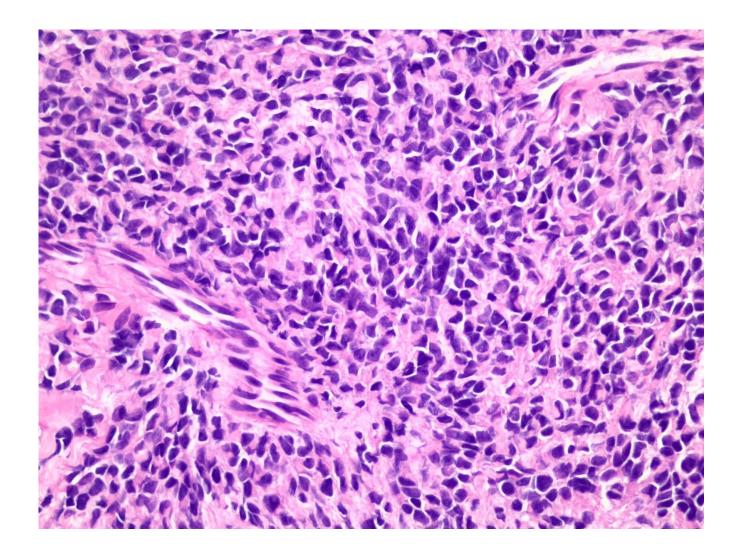




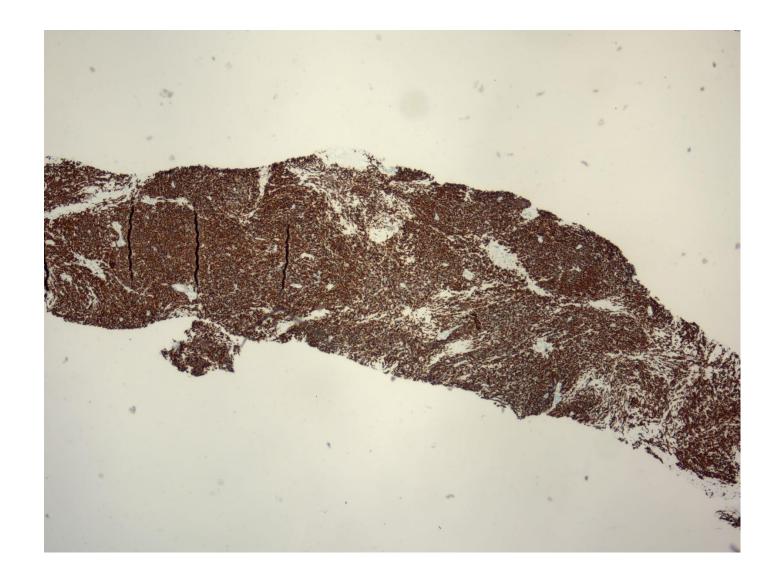




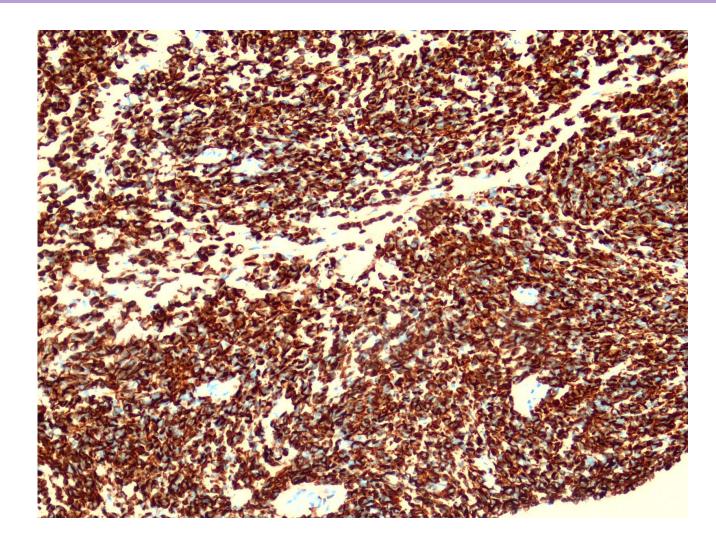






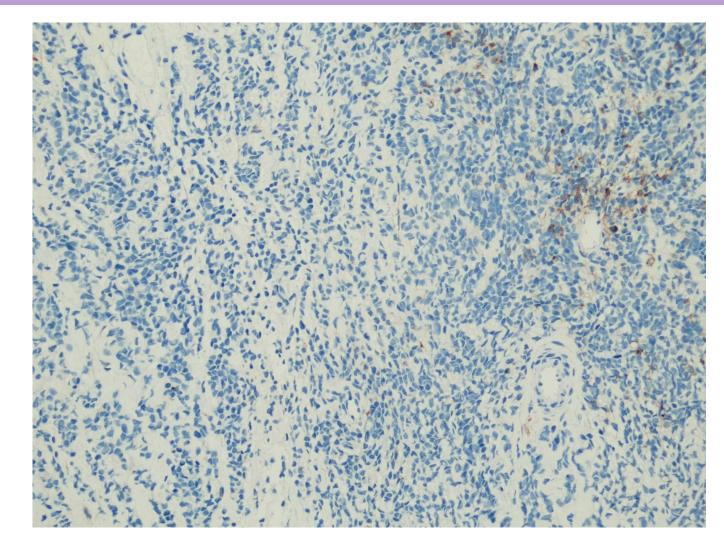






Desmin







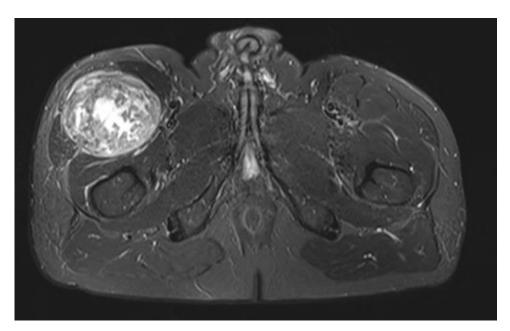
Immunohistochemistry

Anticorpo (Clone-Fabricante): Resultado

Actina de músculo liso (1A4-Cell Marque): negativo nas células neoplásicas. Desmina (DER11-Ventana): positivo forte e difuso nas células neoplásicas. Miogenina (F5D-Cell Marque): negativo nas células neoplásicas. MyoD1 (5.8A-Dako): negativo nas células neoplásicas. Citoceratinas AE1/AE3 (AE1AE3/PCK26-Ventana): negativo nas células neoplásicas. EMA (E29-Ventana): positivo focal nas células neoplásicas. CD34 (Qbend10-Ventana): negativo nas células neoplásicas. CD45/LCA (RP2/18-Ventana): negativo nas células neoplásicas. WT-1 (6F-H2-Cell Marque): negativo nas células neoplásicas. Cromogranina A (LK2H10-Ventana): negativo nas células neoplásicas. Sinaptofisina (SP11-Cell Margue): negativo nas células neoplásicas. Proteína S-100 (4C4.9-Ventana): negativo nas células neoplásicas. CD99 (O13-Ventana): positivo fraco e focal nas células neoplásicas. NKX2.2 (EP336-Cell Margue): negativo nas células neoplásicas. BCOR (C-10-Santa Cruz): negativo nas células neoplásicas. INI1 (BAF47-BD): expressão preservada nas células neoplásicas. TLE-1 (1F5-Cell Margue): positivo fraco nas células neoplásicas. SSX (E5A2C-Cell Signaling): negativo nas células neoplásicas. Ki-67 (MIB-1-Dako): positivo em 40% das células neoplásicas.









DISCUSSION

Unusual presentation of a DSRCT located in the thigh in a 52 yo male patient. No metastasis.

How would you plan the treatment of this patient?

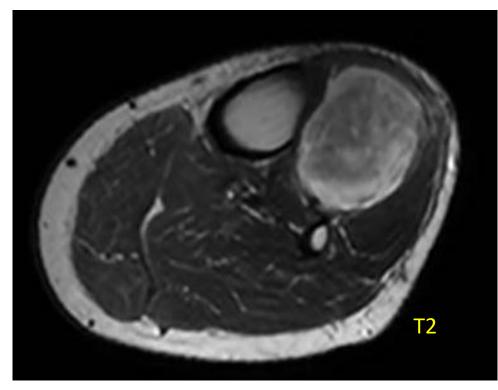


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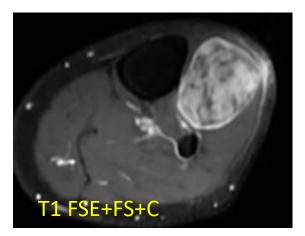
Male, 58

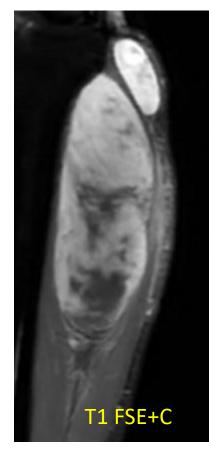
- Diagnosis: Locally Advanced Left Leg Myxoid LPS
- Comorbidities:
 - DM, insulin dependent since the age of 40
 - Never smoker
- Present Illness:
 - Left leg swelling for five months, progressively. Recently, he started noticing a deformity in the form of mild rotation of the left leg. The knee function is not affected but it is tender on moderate exertion or climbing stairs. No associated pain or limitation of movement.
 - Investigated outside:
- PS-0 ECOG

 25 March 2024: MRI left leg revealed a well-defined fusiform lesion, 14 x 13 cm, heterogeneous appearance on T2. On T1 there is focal areas of fat signal intensity at the caudal part of lesion. One subcutaneous component showing post contrast enhancement mostly cystic. Located in tibialis anterior muscle, closely abutting anterior Tibial artery and Nerve tibia but no infiltration of bone at this level









- Staging PET-CT:
 - FDG-avid mass
 - Systemic staging is negative



- Summary: 58 y.o male with Myxoid LPS of the left leg T3 N0 M0 G3
- Sarculator
 - 5-year OS 74%
 10-year OS 63%
 5-year DM 37%
 - ≻10-year DM 41%
- Question to the Board? What is the best initial management?

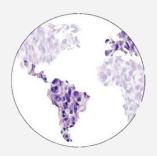
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• What is the best initial management?



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35 y/o male No medical history



11/2023

Notice a small nodule below the lower dental arch

He says that it is something similar to a grain of rice.

12/2023

He reports rapid growth of the lesion, so he consults his dentist.



01/2024

Biopsy: maxillary osteosarcoma with predominance of osteoblastic and fibroblastic pattern, infiltration of oral mucosa

35 y/o male No medical history



- 01/2024

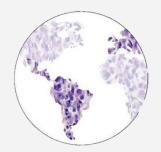
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02/2024

1th first visit in Fleming: - New CT, MRI and



35 y/o male No medical history

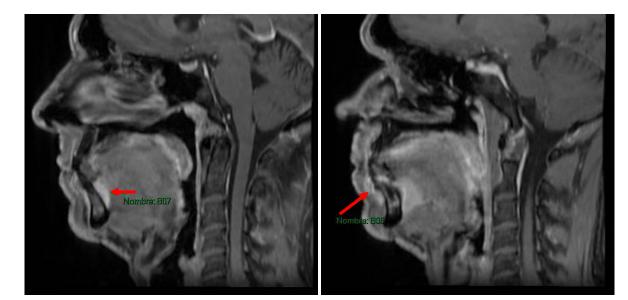


02/2024

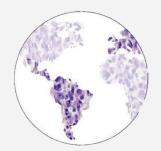
1th first visit inFleming:New CT, MRI andPathology review





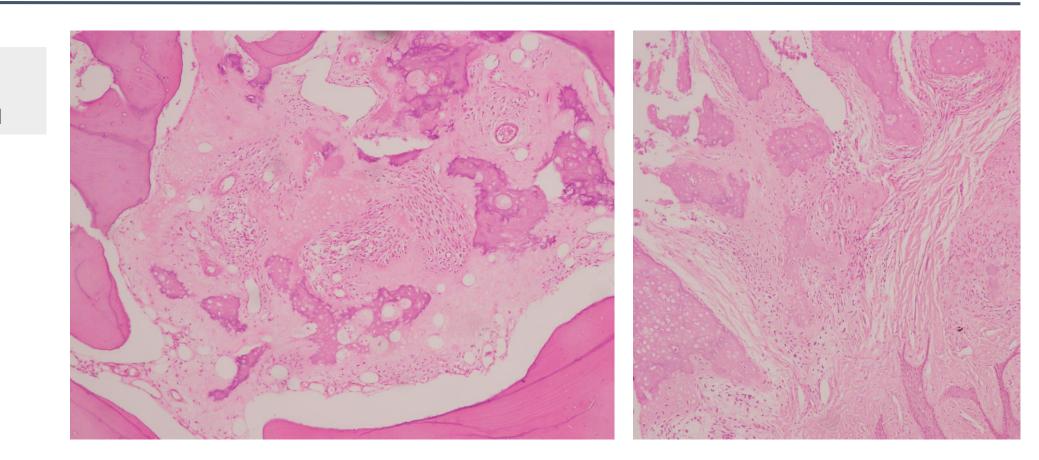


36 y/o male No medical history

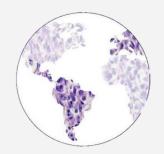


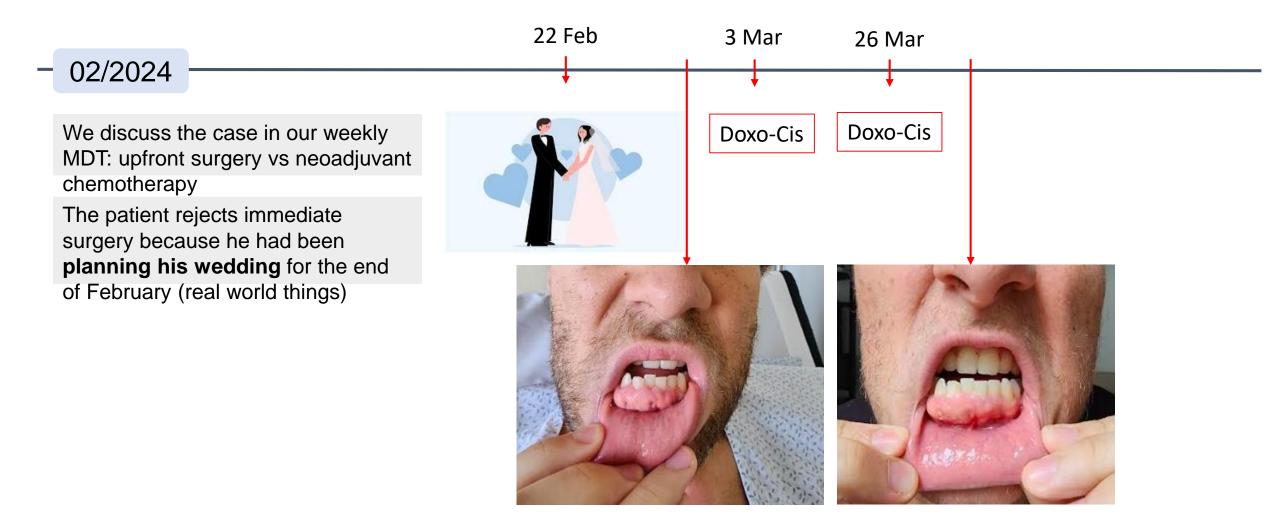
02/2024

1th first visit inFleming:New CT, MRI andPathology review



35 y/o male No medical history





35 y/o male No medical history



11/04/24

The surgery was planned





35 y/o male No medical history



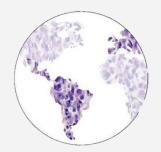
11/04/24

The surgery was planned





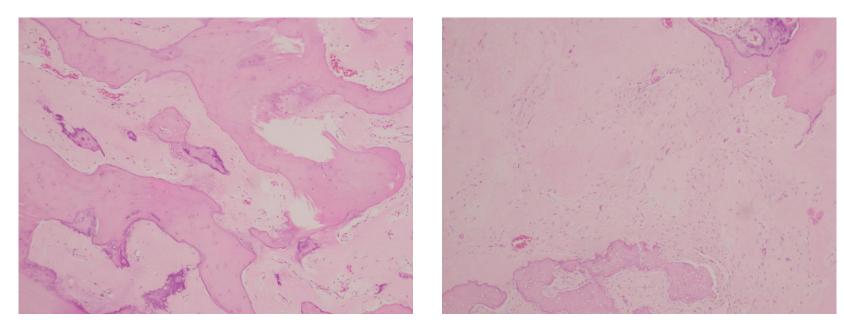
35 y/o male No medical history



11/04/24

Pathology report:

- Osteosarcoma with >90% of necrosis



35 y/o male No medical history



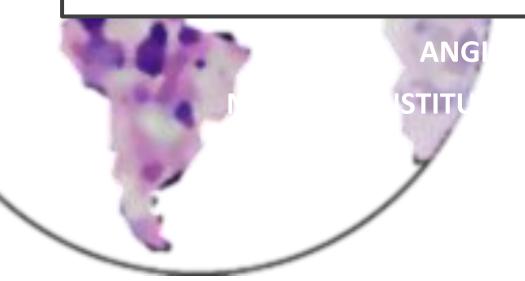
Questions to the MDT:

- experiences with these tumors
- adjuvant radiotherapy?
- control?



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SELNET INTERNATIONAL TUMOR BOARD – MAY 2024.



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CLINICAL HISTORY

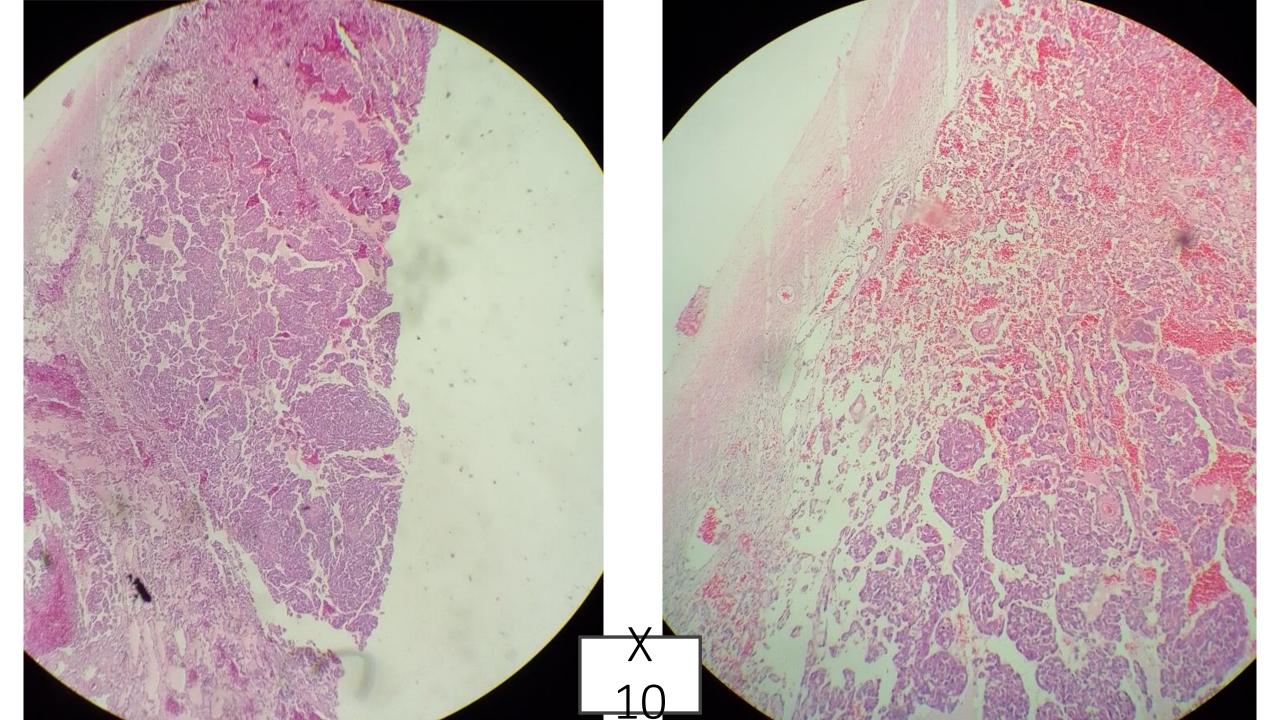
- 28 yo female
- ECOG 2
- The patient came to our center on 03/01/2023 due to a 6-month history of a growing mass in the right breast.
- On physical examination, a 5 cm mass of solid consistency was found at the junction of the upper external quadrants, it was painful on palpation and no lymphadenopathy was found.

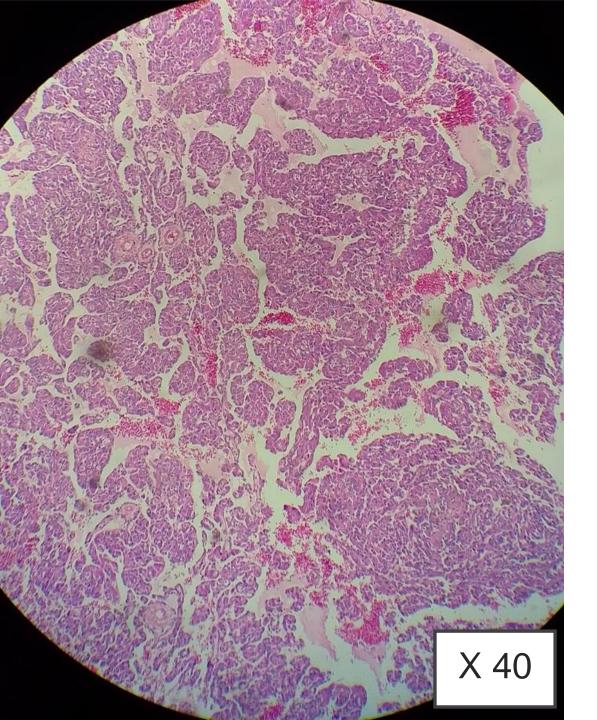


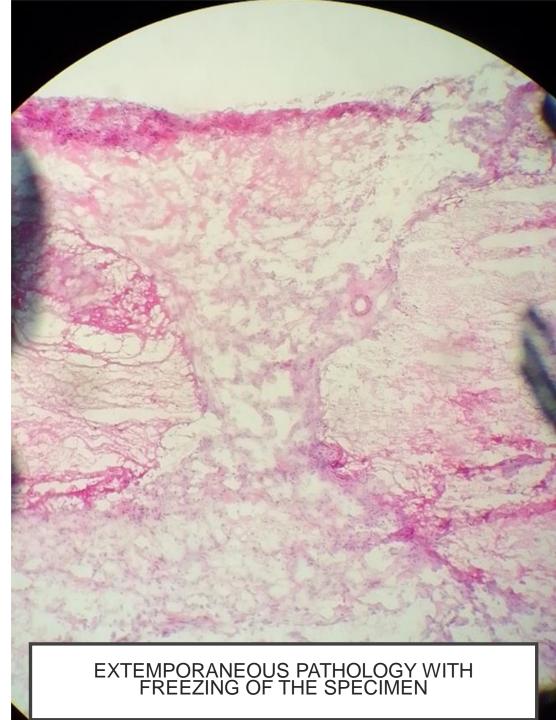
- An ultrasound-guided biopsy of the right breast mass was performed.
- Pathology Report:
- Vascular proliferation covered by endothelium showing minimal nuclear atypia, with anastomosing channels and areas of proliferation of the papillary endothelium.
- **<u>IHC:</u>** AE1/AE3 **NEGATIVE** CD31 **POSITIVE**
- DX: Low grade angiosarcoma of the right breast.
- We discuss the case in multidisciplinary board at our center and decided a surgery consistent with a simple mastectomy performed in 25/05/23.

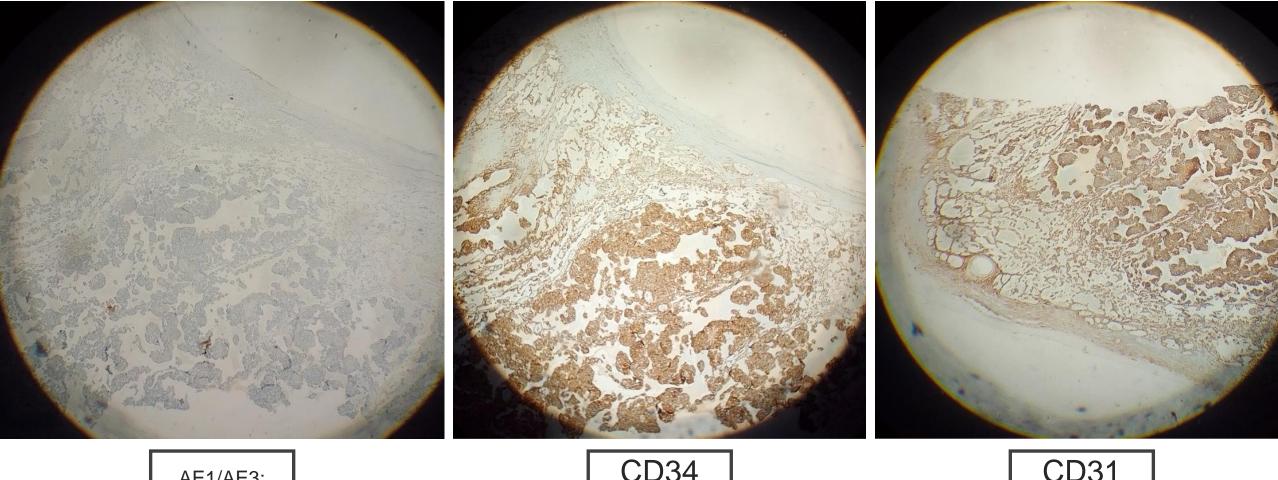


- After this surgery, the patient came again for follow-up and a CT scan was performed, observing a 140 mm mass replacing the right ovary and another of 95 mm replacing the left one, poorly defined, with diffuse images and compression of the uterus, suggested as secondary. In addition, osteolytic images were found in the spine at the dorsal and lumbar column.
- The case is discussed again in our sarcoma board and surgery is decided to excise the mass. Complete hysterectomy was performed on 12/15/2023.
- Pathology Report of the Hysterectomy:
- Ovaries: Metastasis of moderately differentiated <u>angiosarcoma</u> in both ovaries, with extensive infarct-like ischemic necrosis. It invades the hilum of both ovaries but does not invade the uterine tubes. Uterus and cervix without alterations. Findings <u>similar</u> to the known primary breast tumor.
- IHC: Negative for AE1/AE3, D240. Positive for CD31, CD34. Ki-67: 40%.

















• The patient underwent a bone scintigraphy which revealed multiple active bone lesions in the skull, the entire extension of the spine, bilateral costal arches, epiphysis of both humeri, sternal manubrium, sacrum and right sacroiliac joint.

• In the last consult, the patient presented a significant --!-/**-** * $_{3}/dL$), probably ັ້^{ຼຸ}ກoglobin ' decre bone ma assoc)n.

DISCUSSION

• What can we offer to this patient as a treatment ?



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F. Pablo Segura, MD

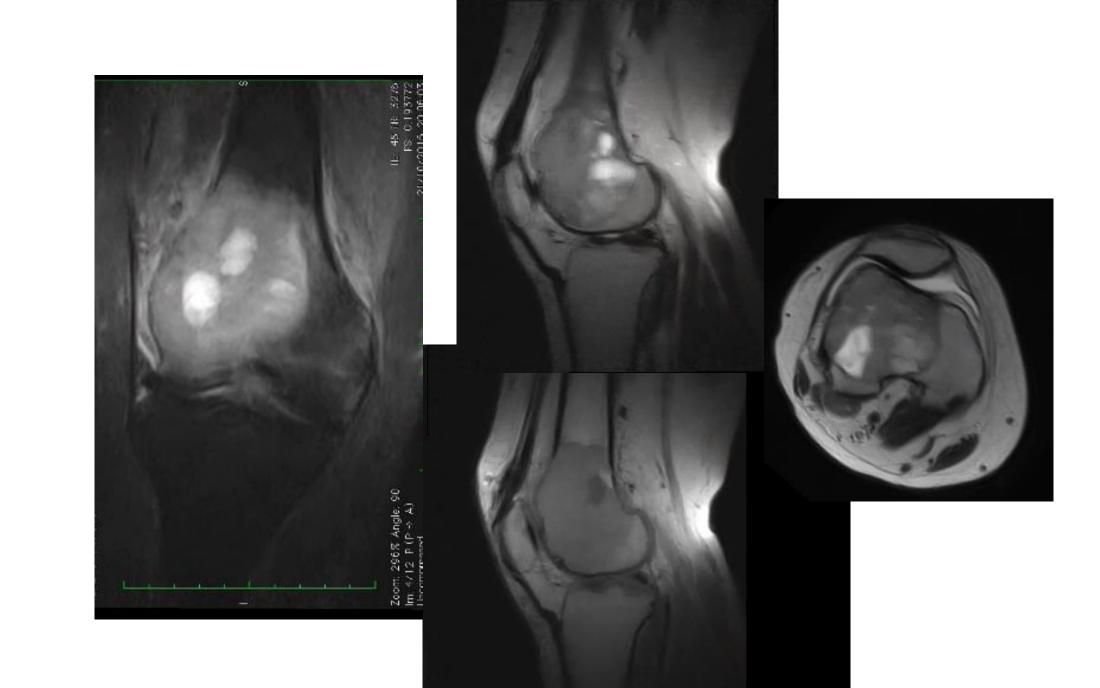
Orthopaedic Surgeon Unidad de Tumores Musculoesqueleticos Córdoba, Argentina



- 35 yo male
- No comorbidities

- 1st visit to clinic November 2016
- History of chronic right knee pain. He reports mild discomfort since 2014, progressively growing during last 3-4 months until 1 week before, when he refers sudden and acute pain exacerbation after minor trauma
- Xray and MRI: distal femur pathologic fracture in an osteolitic (agressive) epiphiseal lesion.







- 35 yo male
- No comorbidities

- 1st visit to clinic November 2016
- Biopsy report: giant cell tumour of bone

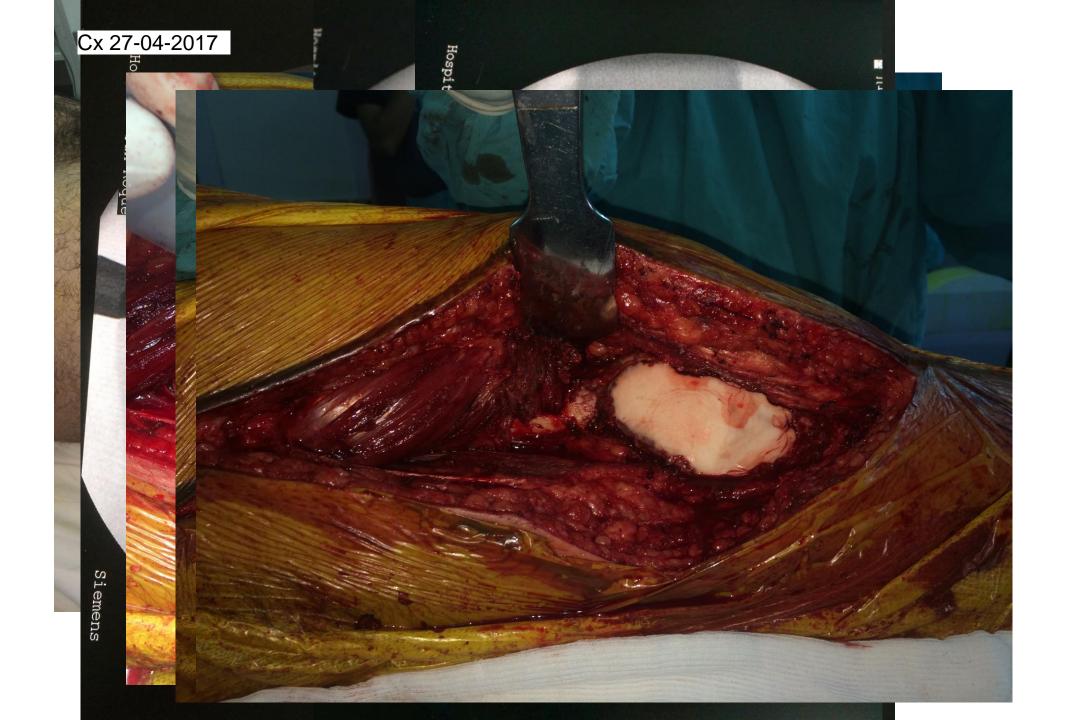
| Material: | LESION TUMORAL. | | |
|--|--|---|--|
| | últiples fragmentos tisulares irregulares que miden entre 2x0,8x0,8 con áreas pardas y otras con aspecto de coágulo. Al corte de similar | | |
| Examen Microscópico Los cortes histológicos muestran proliferación tumoral caracterizada por dos poblaciones celulares, unas mononucleares con núcleos redondeandos y ovales y citoplasma fusado, abuntantes células gigantes multinucleadas de tipo asteoclásticas. Se identifican escasas mitosis. Se observan además sectores de hemorrágia, histiocitos con hemosiderina como signo de hemorrágia antigua. | | | |
| Diagnóstico TUMOR DE (| CELULAS GIGANTES OSEO (Fragmentos tisulares) | - | |

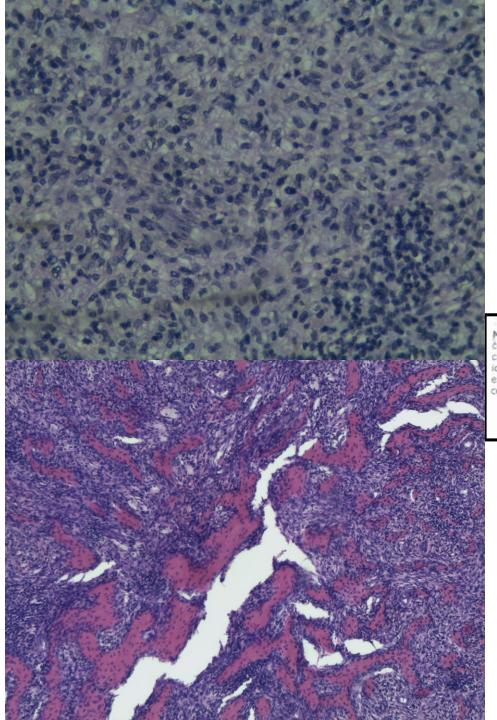


- 35 yo male
- No comorbidities

- 1st visit to clinic November 2016
- Treatment decision: neoadyuvant Denosumab and intralesional surgery
- Xray after 5 months

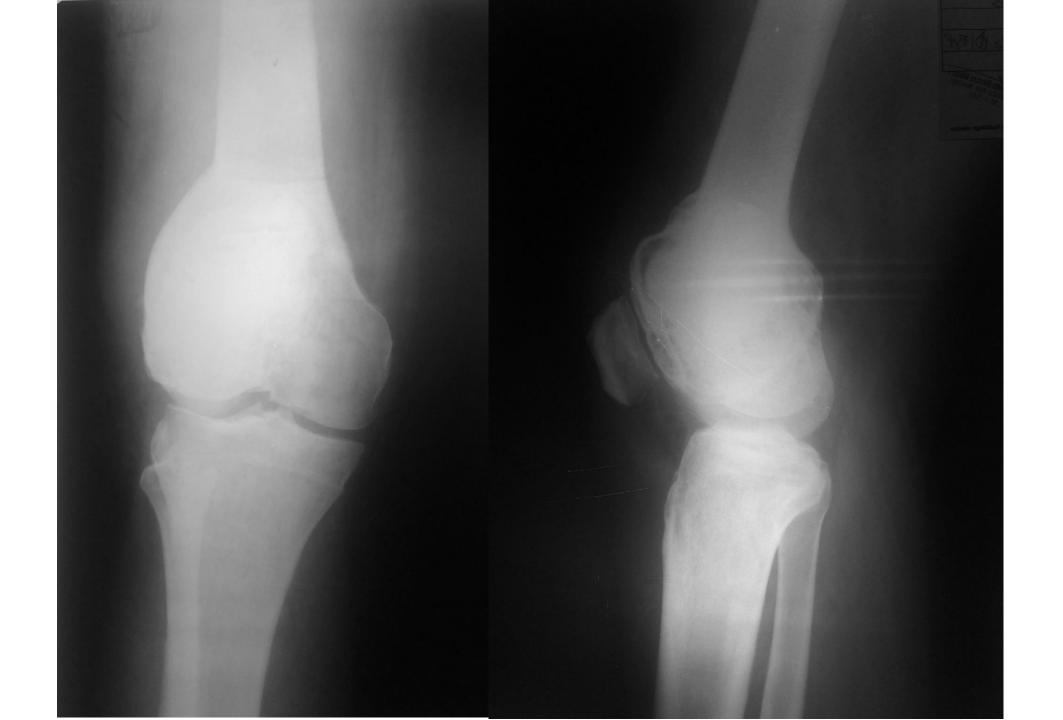






MICROSCOPIA: Neoplasia de estirpe mesenquimal constituida por una proliferación de células fusadas, son de nucleos pequeños y medianos, elongados, con escasas figuras de mitosis y citoplasma eosinófilo que se disponen en haces entrelazados con escaso estroma interpuesto. Se identifican además focos de material osteolde, hisiocitos, vasos sanguíneos de paredes engrosadas, sufusiones hemorrágicas y leve infiltrado inflamatorio mononuclear. No se observan células gigantes.

La piel está libre de lesión. FD.-RA.-GA.-AS.-RF.-



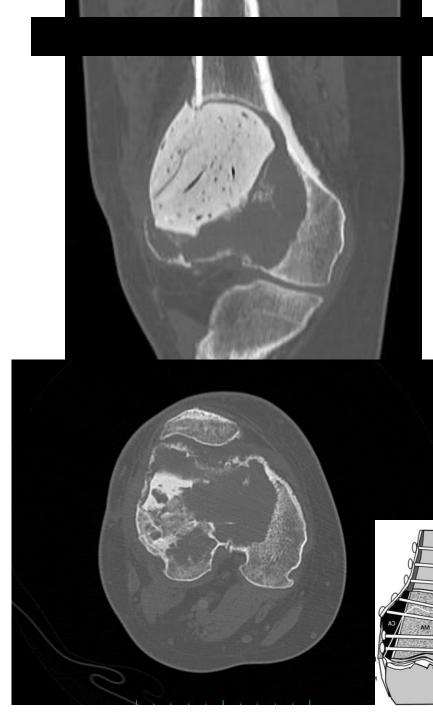


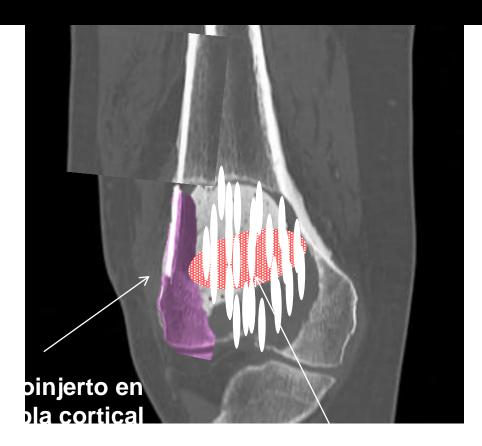
- 36 yo male

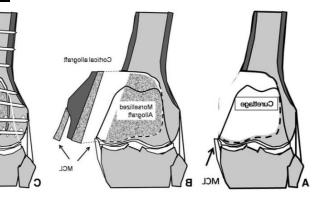
ONCOLOGICAL SUMMARY:

• August 2018: local recurrence

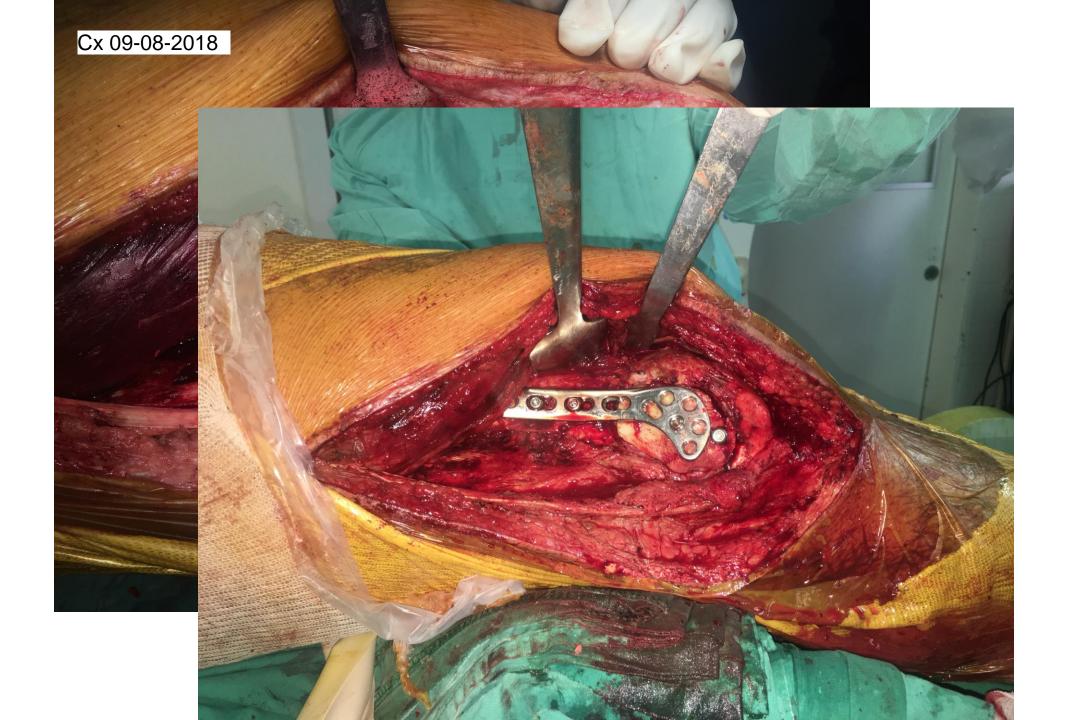








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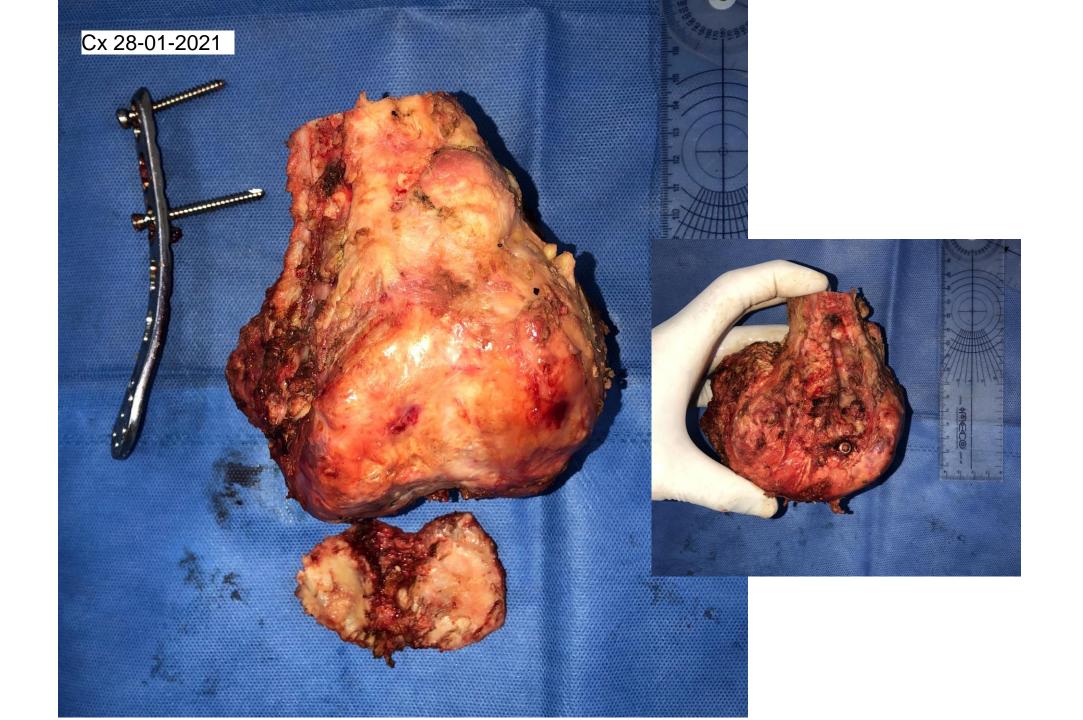




- 38 yo male

- May 2020: 2nd local recurrence
- Treatment decision:
 - En block resection
 - Distal femur endoprosthetic reconstruction

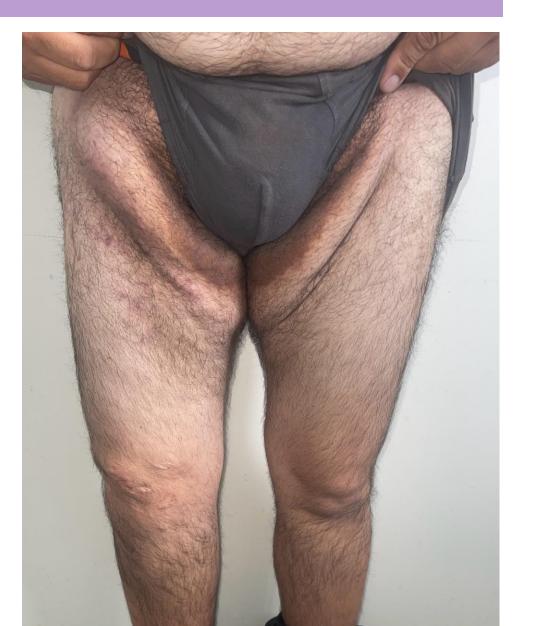




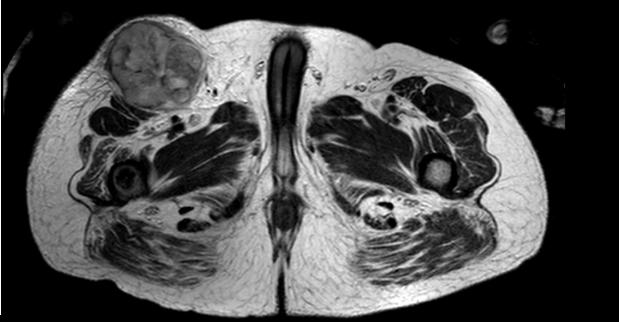




- 42 yo male
- Last control February 2024 (3 ys postop)
- Good functional outcome
- Rigth groin mass





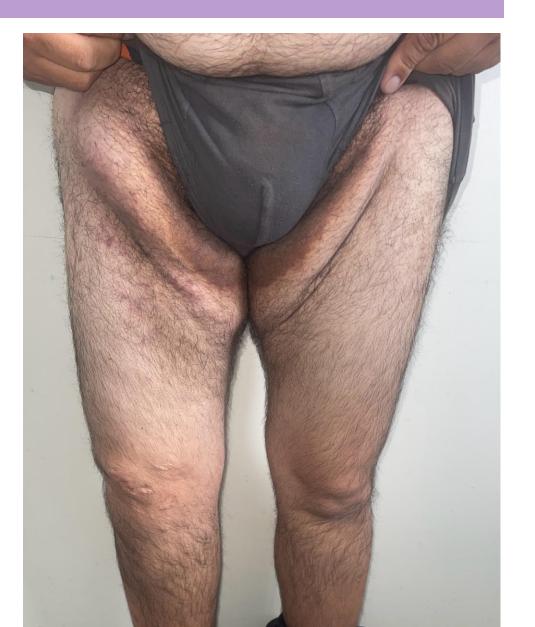


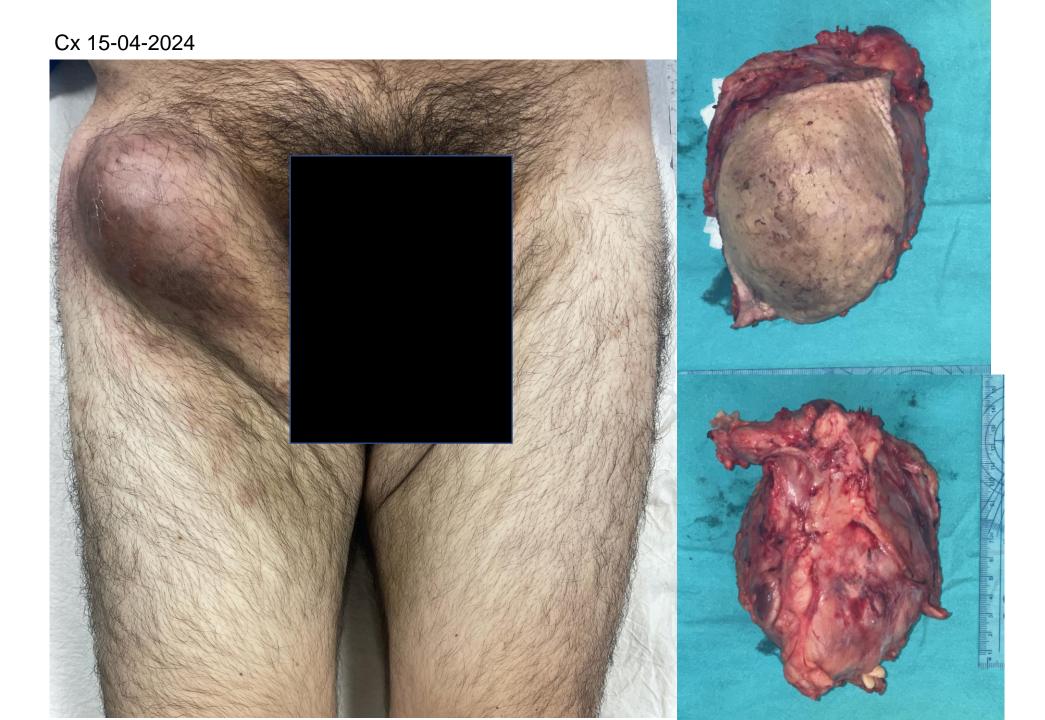


5 mm



- 42 yo male
- Last control February 2024 (3 ys postop)
- Good functional outcome
- Treatment decision:
 - Excisional biopsy







- 42 yo male
- No comorbidities -

ONCOLOGICAL SUMMARY:

• AP report:

INFORME

Biopsias previas nº135929, 142165, 150086, 151202 con diag. de Tumor de ANTECEDENTE: células gigantes. Tratado con Denosumab.

MATERIAL: 1.-Tumor de partes blandas de región inguinal. 2.-Ganglio locorregional.

MACROSCOPIA: 1.-Se recibe pieza quirúrgica que llega sin señalizar, mide 18x14x10 cm y pesa 805 g. Incluye piel de 16x10 cm y partes blandas adheridas. La superficie externa es irregular, lobulada. Al corte se identifica un tumor que mide 15x10,5x9,5 cm, sólido, heterogéneo, de coloración amarillenta, con áreas blanquecinas y otras hemorrágicas. Es de consistencia blanda, en sectores friable. En la periferia se observa un nódulo de 2,5 cm, blanquecino-grisáceo. 2.-Ganglio linfático que mide 3,5x1,5 cm.

1.-El tumor descripto está constituido por una proliferación de células MICROSCOPIA: fusadas con núcleos medianos y grandes, irregulares, hipercromáticos, en sectores vesiculosos con nucleolo prominente, con figuras de mitosis (8 en 10 CGA) y citoplasma eosinofilo mal definido que se disponen en haces cortos, con extensas áreas de necrosis tumoral. El estroma es fibronconectivo y exhibe vasocongestion, sufusiones hemorragicas y leve infiltrado inflamatorio mononuclear.

El nódulo descripto en la periferia corresponde a un ganglio linfático con metástasis tumoral.

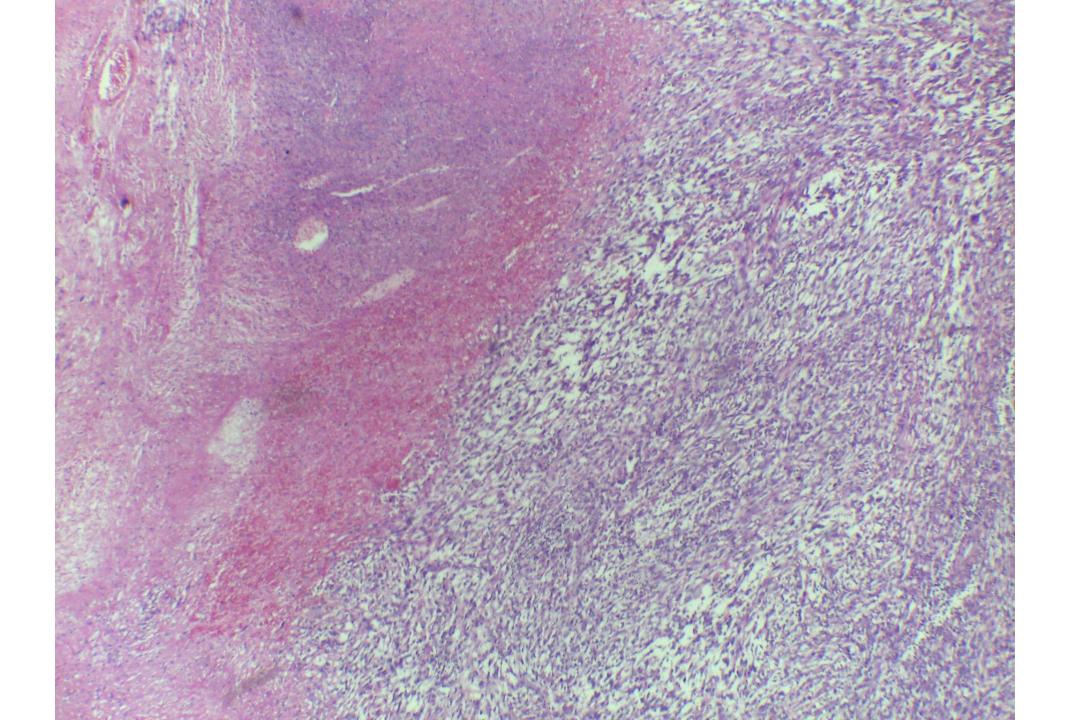
Los márgenes quirúrgicos están a menos de 1 mm del tumor. 2.-Ganglio linfático libre de tumor. LM.-LF.-GA.-

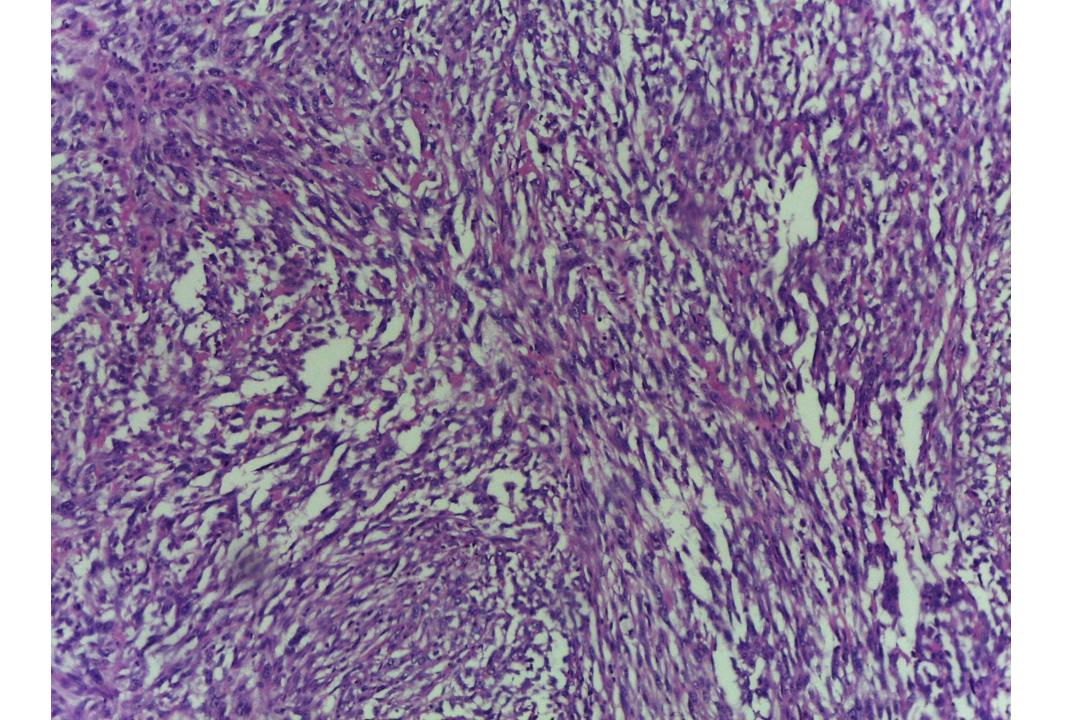
Diagnóstico Tumor fusocelular maligno Metástasis de Sarcoma

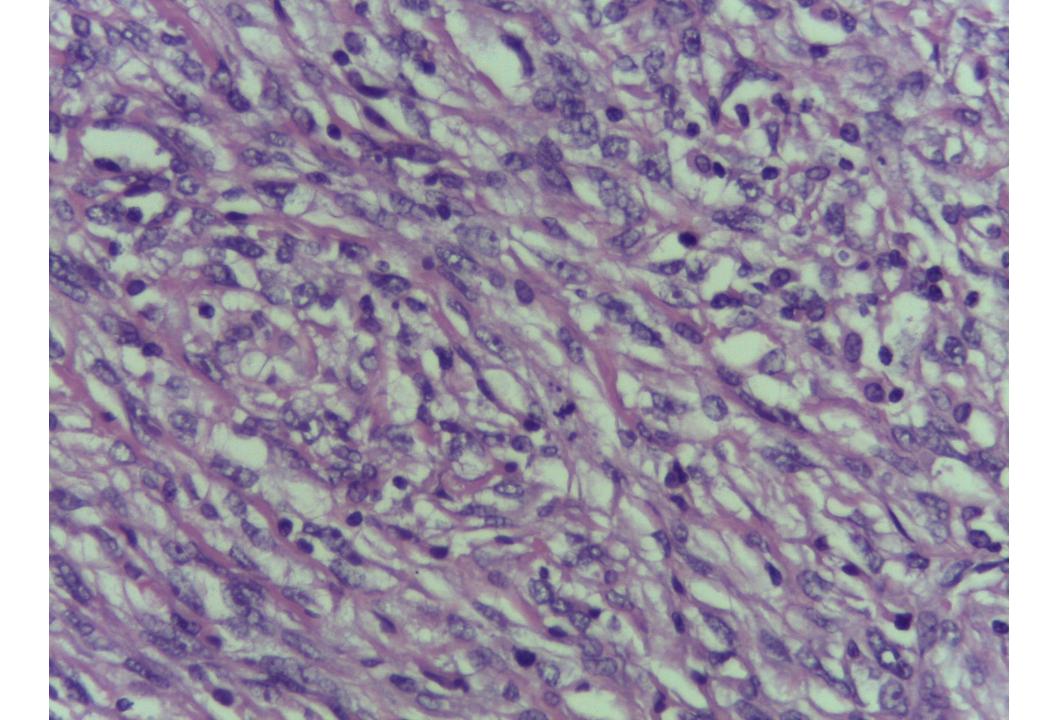
NOTA

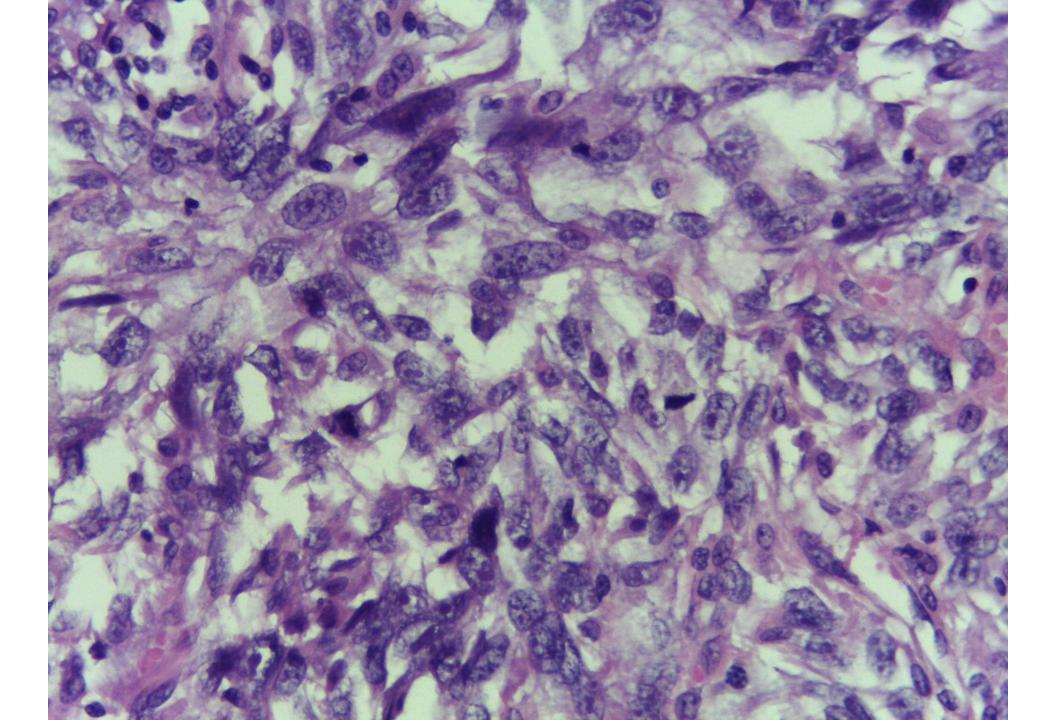
Hallazgos compatibles con Tumor de células gigantes maligno secundario.

Nº Protocolo: . .163.109 Página: 1











- 42 yo male
- No comorbidities

- Torax/Abdomen/ Pelvis CT Scan May 2024:
 - Torax ok
 - Abdomen ok
 - Intrapelvic mass





DISCUSSION

Unusual presentation of a GCT of bone. No visceral metastasis. Distant lesions (groin and pelvis)

More studies? (Pelvic MRI in process / PET CT?) How would you plan the treatment of this patient? (Denosumab again? / Other sistemic strategies / soft tissue pelvic mass resection?