Principes of surgery of sarcomas

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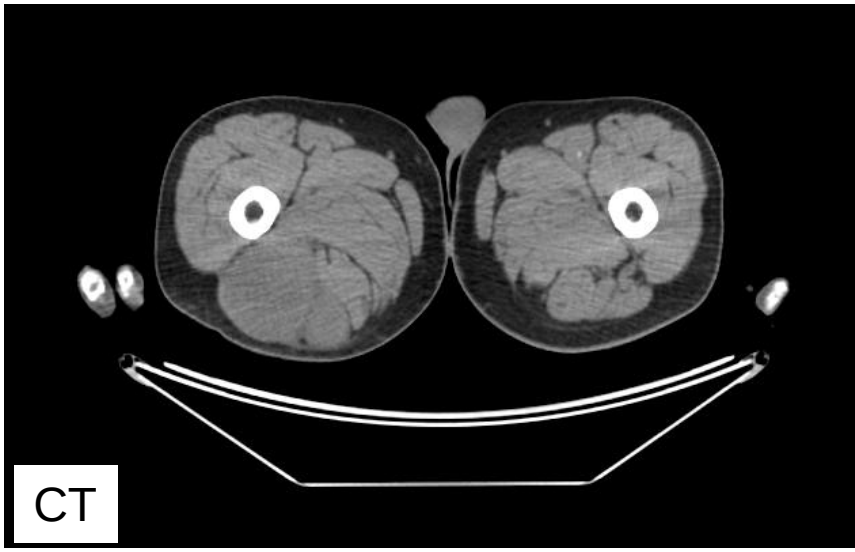
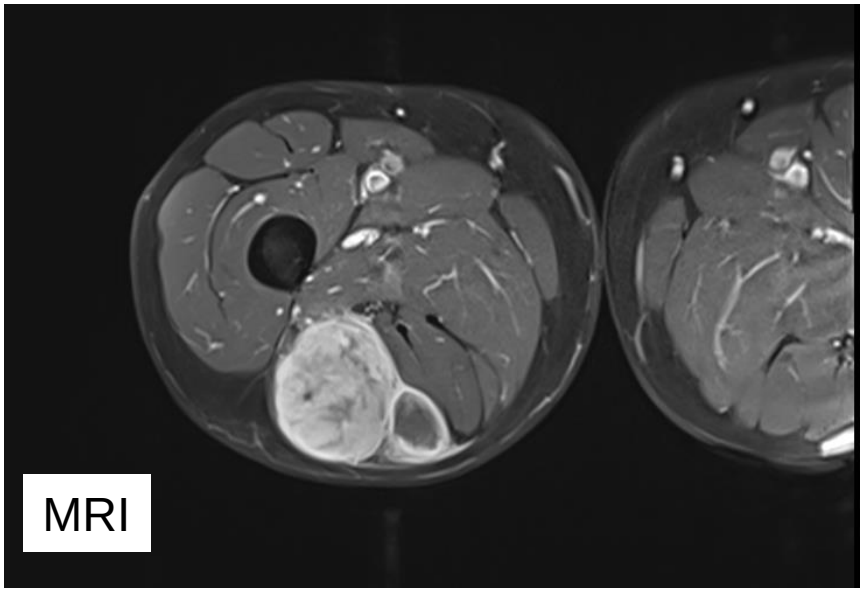
LIMBS

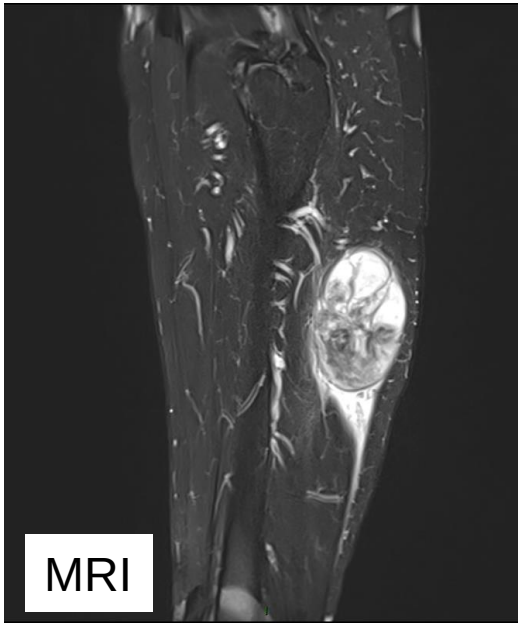
Diagnosis: imaging

- **MRI** is the main imaging modality in the extremities

- Standard radiographs and CT may be useful:
  to rule out a bone tumour,
  to detect bone erosion
  to show calcifications
  to rule out a myositis ossificans

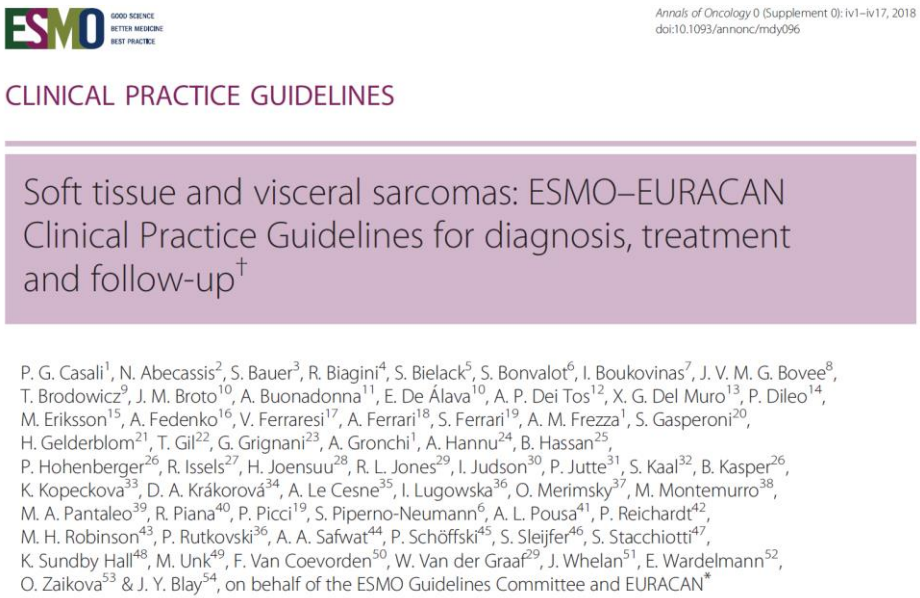






First step: tayloring surgery to histology

Surgery is the standard treatment of all patients with an adult type, localized STS



ESMO GOOD SCIENCE BETTER MEDICINE BEST PRACTICE

Annals of Oncology 0 (Supplement 0): iv1–iv17, 2018
doi:10.1093/annonc/mdy096

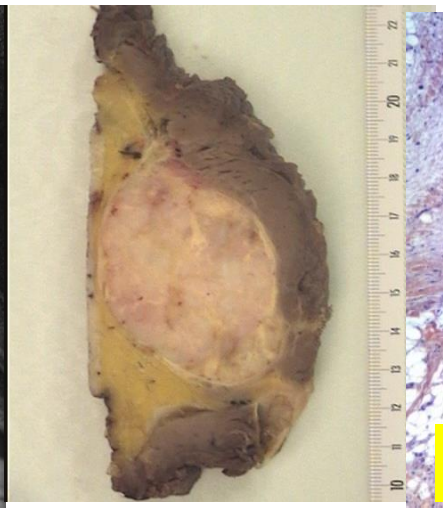
CLINICAL PRACTICE GUIDELINES

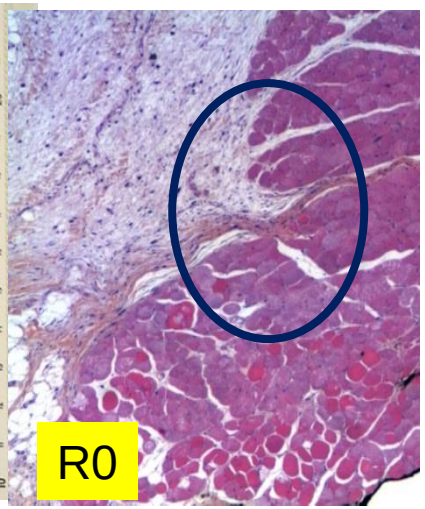
Soft tissue and visceral sarcomas: ESMO–EURACAN Clinical Practice Guidelines for diagnosis, treatment and follow-up†

P. G. Casali[1], N. Abecassis[2], S. Bauer[3], R. Biagini[4], S. Bielack[5], S. Bonvalot[6], I. Boukovinas[7], J. V. M. G. Bovee[8], T. Brodowicz[9], J. M. Broto[10], A. Buonadonna[11], E. De Álava[10], A. P. Dei Tos[12], X. G. Del Muro[13], P. Dileo[14], M. Eriksson[15], A. Fedenko[16], V. Ferraresi[17], A. Ferrari[18], S. Ferrari[19], A. M. Frezza[1], S. Gasperoni[20], H. Gelderblom[21], T. Gil[22], G. Grignani[23], A. Gronchi[1], A. Hannu[24], B. Hassan[25], P. Hohenberger[26], R. Issels[27], H. Joensuu[28], R. L. Jones[29], I. Judson[30], P. Jutte[31], S. Kaal[32], B. Kasper[26], K. Kopeckova[33], D. A. Krákorová[34], A. Le Cesne[35], I. Lugowska[36], O. Merimsky[37], M. Montemurro[38], M. A. Pantaleo[39], R. Piana[40], P. Picci[19], S. Piperno-Neumann[6], A. L. Pousa[41], P. Reichardt[42], M. H. Robinson[43], P. Rutkovski[36], A. A. Safwat[44], P. Schöffski[45], S. Sleijfer[46], S. Stacchiotti[47], K. Sundby Hall[48], M. Unk[49], F. Van Coevorden[50], W. Van der Graaf[29], J. Whelan[51], E. Wardelmann[52], O. Zaikova[53] & J. Y. Blay[54], on behalf of the ESMO Guidelines Committee and EURACAN*

QUALITY OF SURGERY: MARGINS

- The standard surgical procedure is a one block wide excision with negative margins (R0)
- → This implies removing the tumour with a rim of normal tissue around it → the surgeon must know that this is a sarcoma +++
- There is no need to resect a rim of normal tissue if the tumor is benign! → by definition unplanned surgery is marginal
- Imaging is not specific of sarcoma → Diagnosis must be done BEFORE by **pre op biopsy** ++
- Anticipation of reconstructions
- It must be carried out by a surgeon specifically trained in sarcoma





- Biopsy: UPS
- MDT: Pre op RX
- T 11 cm, R0: smallest margin = 6 mm muscle
- 20% viable cells

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ESMO
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DIAGNOSIS: BIOPSY

- The standard approach to diagnosis consists of multiple core needle biopsies, by using ≥14–16 G needles
- A pathological expert review/validation is required in all cases when the original diagnosis was made outside a reference centre
- An excisional biopsy may be the most practical option for <3 cm superficial lesions
- A frozen-section technique for immediate diagnosis is discouraged

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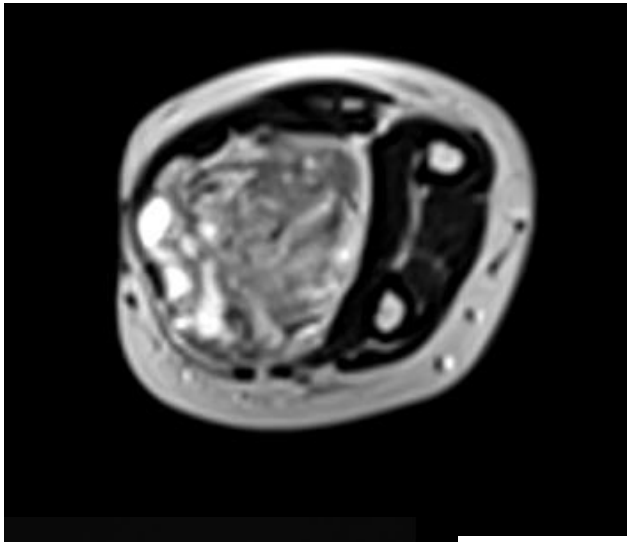
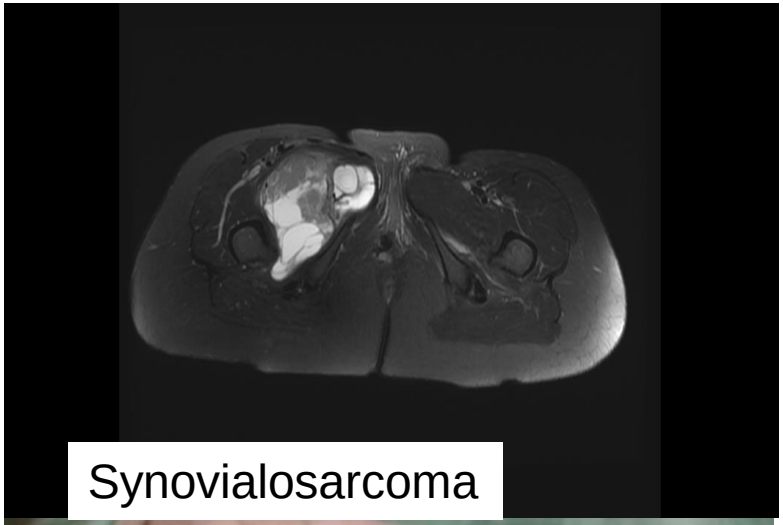
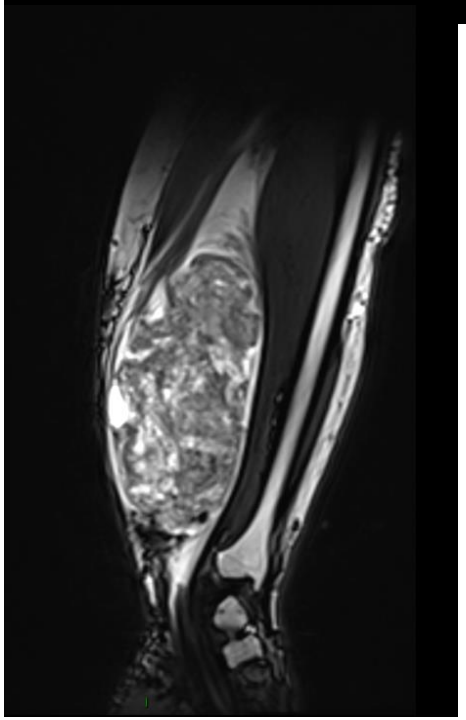
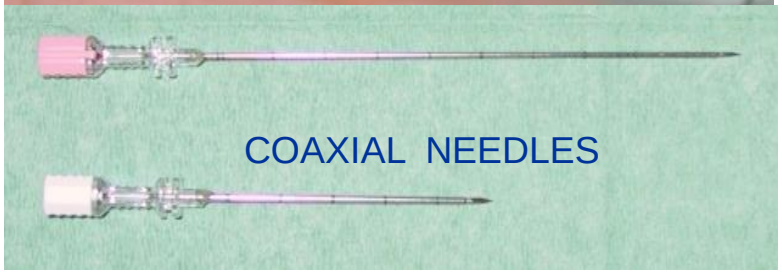




Synovialosarcoma





Fusion transcript: c11orf 95/MKL2
described in chondroïdes Lipoma

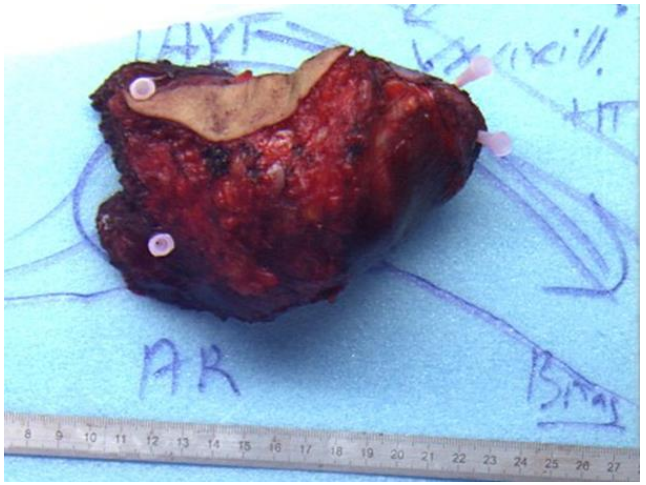
Surgery and Margins

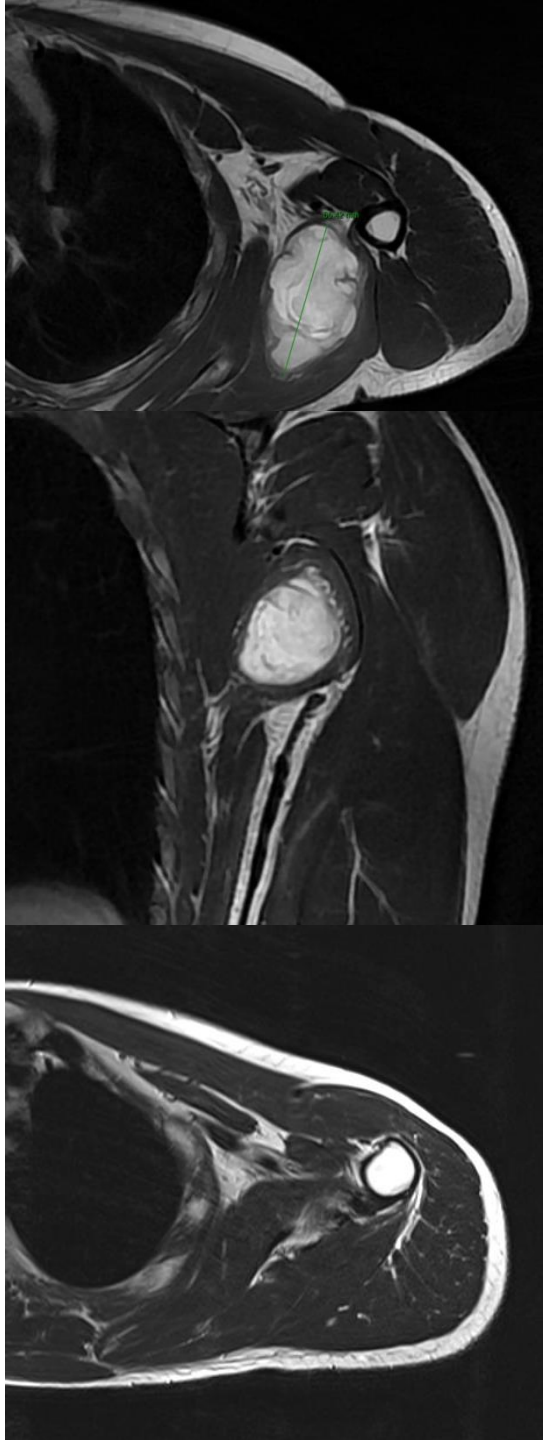
Surgery:

- After percutaneous biopsy
- To remove the tumor without seeing it with a rim of normal tissue

Anatomopathology:

- Pleiomorphic rhabdomyosarcoma
- Grade histopronostique III
- Size 90 mm
- R0 muscle
- Anterior margin 6 mm
- Posterior margin 25 mm
- Down margin 5 mm
- Top margin 20 mm





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STRATEGY: MULTI DISCIPLINARY TUMOR BOARD (MDT)

- ASSESMENT OF EXTENSION (CHEST ABDOMINAL CT)
- SURGERY/MARGINS
- RADIOTHERAPY
- NEO ADJ CHEMOTHERAPY
- ILP

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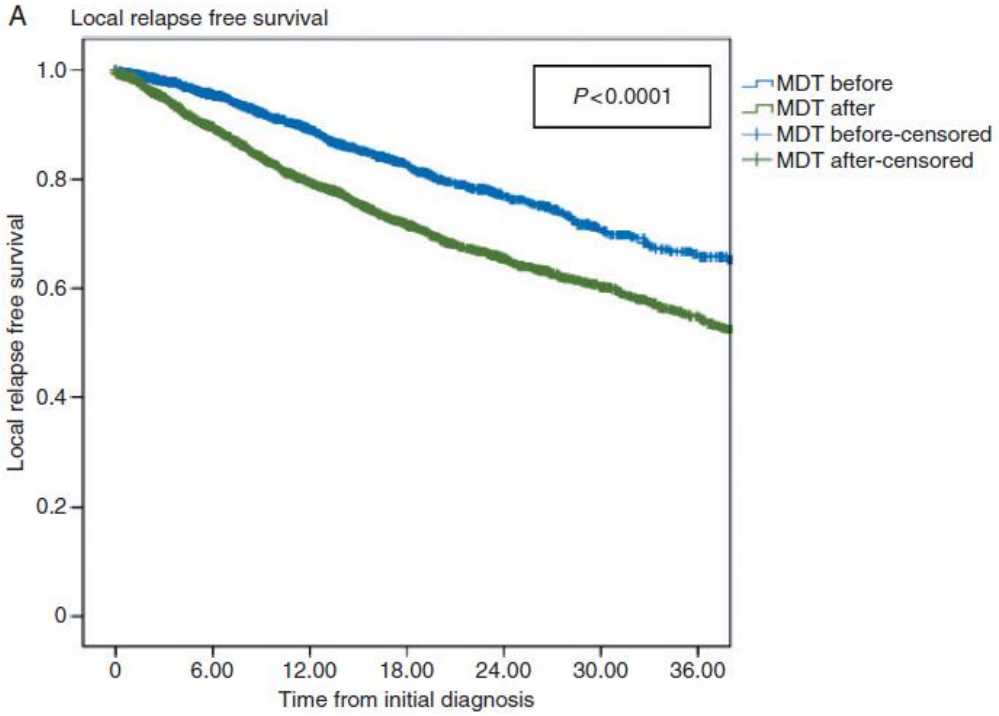
ESMO GOOD SCIENCE BETTER MEDICINE BEST PRACTICE

Annals of Oncology 28: 2852–2859, 2017
doi:10.1093/annonc/mdx484
Published online 28 August 2017

ORIGINAL ARTICLE

Improved survival using specialized multidisciplinary board in sarcoma patients

J.-Y. Blay[1,2*], P. Soibinet[3], N. Penel[4], E. Bompas[5], F. Duffaud[6], E. Stoeckle[7], O. Mir[8], J. Adam[8], C. Chevreau[9], S. Bonvalot[8,10], M. Rios[11], P. Kerbrat[12], D. Cupissol[13], P. Anract[14], F. Gouin[15], J.-E. Kurtz[16], C. Lebbe[17], N. Isambert[18], F. Bertucci[19], M. Toumonde[7], A. Thyss[20], S. Piperno-Neumann[10], P. Dubray-Longeras[21], P. Meeus[1,2], F. Ducimetière[1,2], A. Giraud[7], J.-M. Coindre[7], I. Ray-Coquard[1,2], A. Italiano[7†] & A. Le Cesne[8†], on behalf of the NETSARC/RREPS and French Sarcoma Group–Groupe d'Etude des Tumeurs Osseuses (GSF-GETO) networks[‡]



12528 patients

BIOPSY = SARCOMA (with NO METASTASIS)

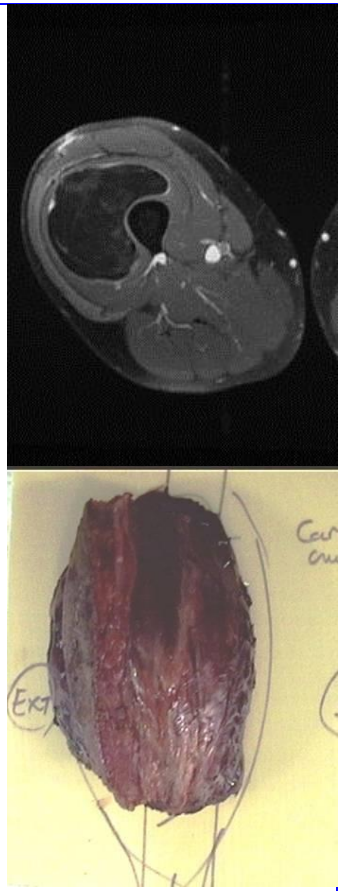
- High grade
- Deep
- Locally advanced (Mutilating surgery)
- Anatomical constraints
- Recurrence

Discussion of neo adjuvant treatment in MTB

According to presentation

- Low grade
- Superficial
- Small tumor
- Non mutilating surgery
- No anatomical constraints

Surgery at first



WDLPS

After excisional biopsy → Re excision (Radiotherapy does not palliate inadaquate surgery)

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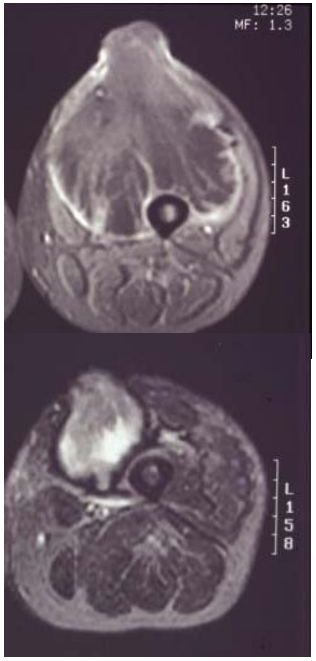
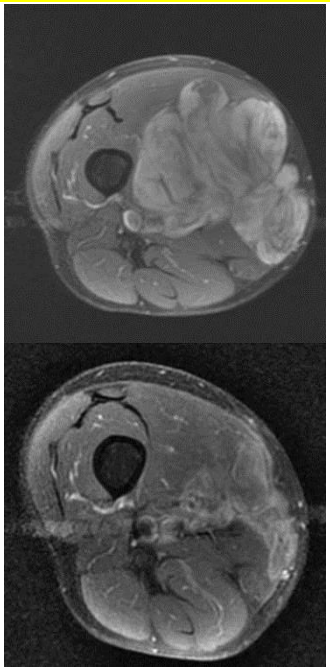
Neo adjuvant strategies in sarcoma

2nd recurrence MPNST
In pre irradiated field ILP



UPS G3 Post
« whoops »
Pre op CT



Myxoid LPS with no round cells
Pre op RT

ESMO GOOD SCIENCE BETTER MEDICINE BEST PRACTICE

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doi:10.1093/annonc/mdy096

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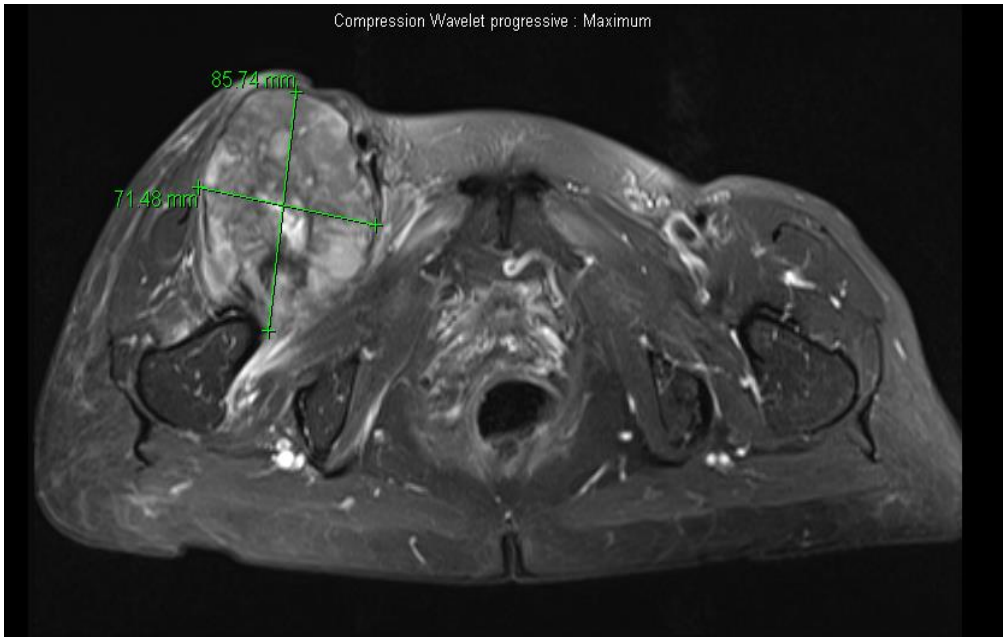
Limbs: reconstructions ++

Reconstructive surgery after pre op RT



October 2012: Pre op RT
MRI Post RT 50.4 Gy

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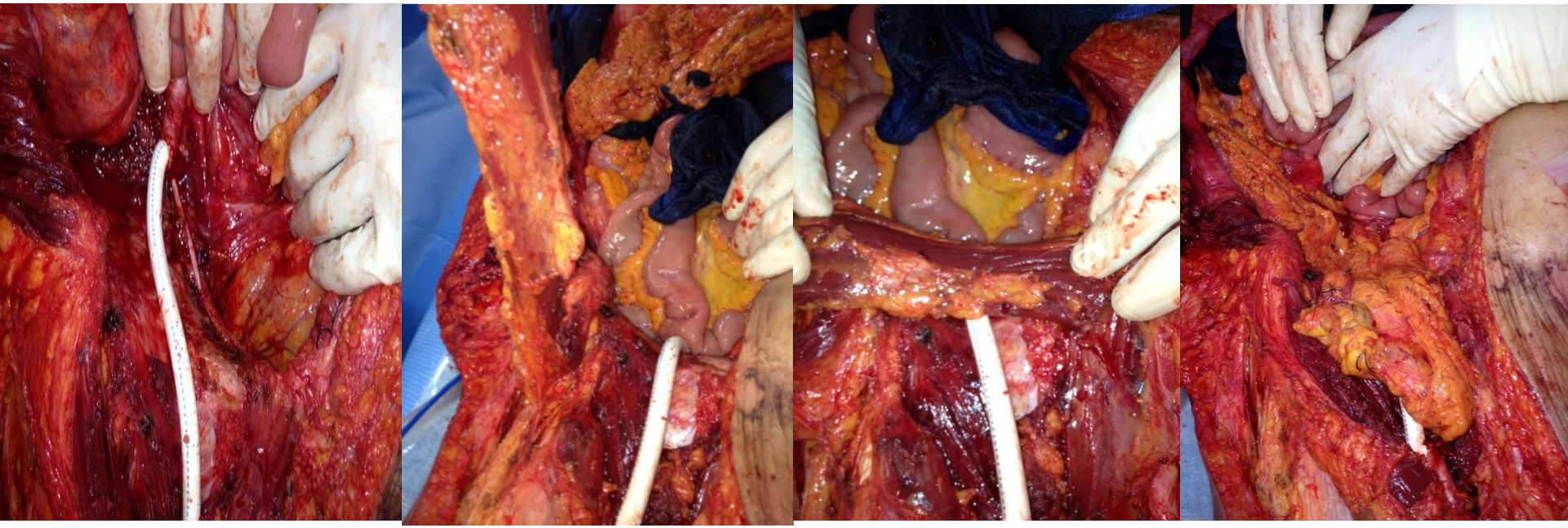
Surgery oct 2012: Anticipation resection/reconstructions

- Vascular graft
- Sartorius flap
- Pedicled rectus abdominis flap
- Omentoplasty



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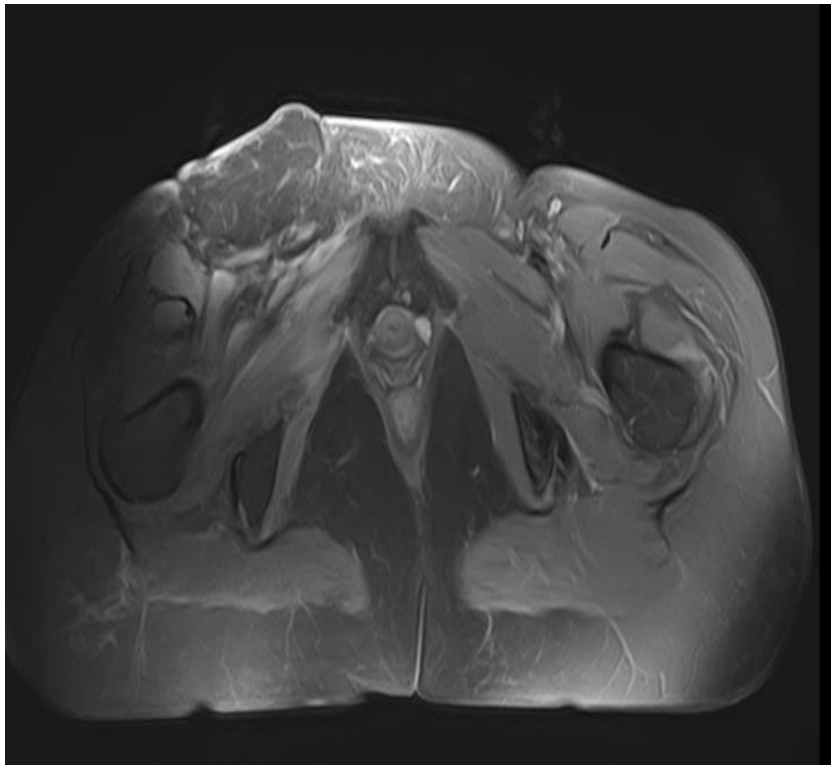

- T 15 X 7 cm, R0 1 mm, Vessels partially included, 8% viable cells
- December 2018: MRI and Chest CT: normal

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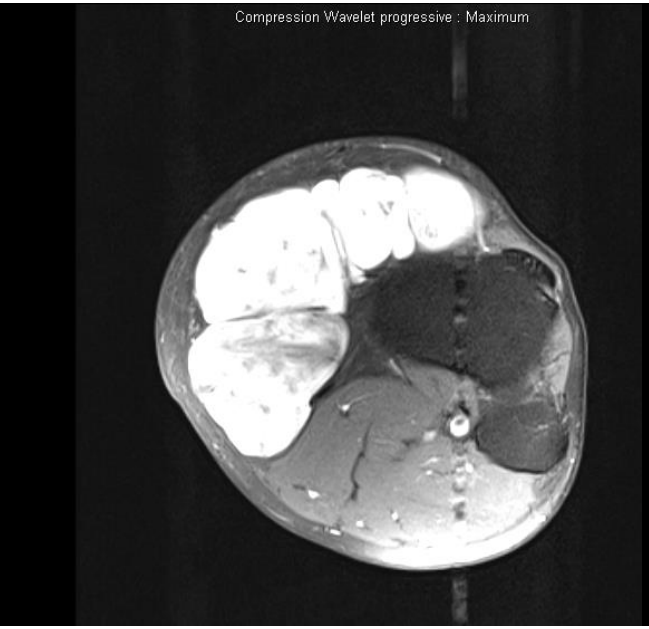
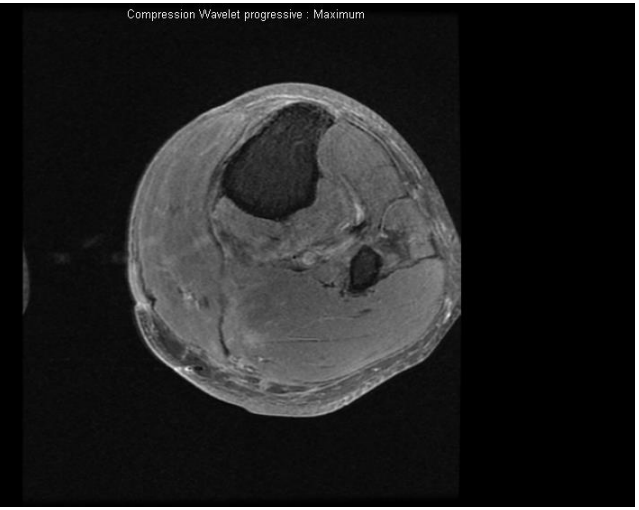
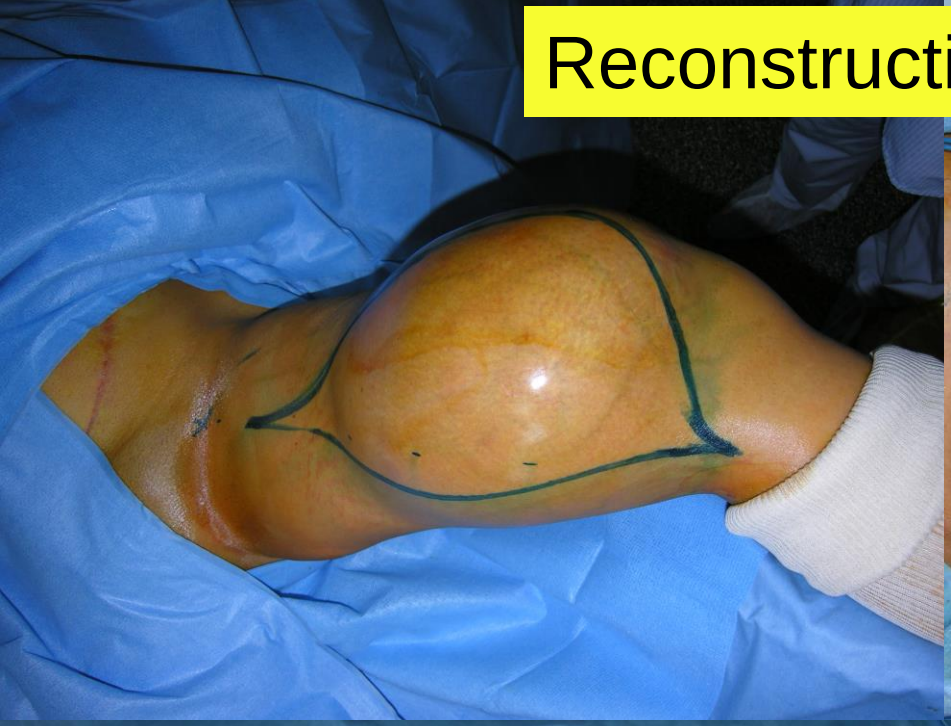
Reconstructive surgery

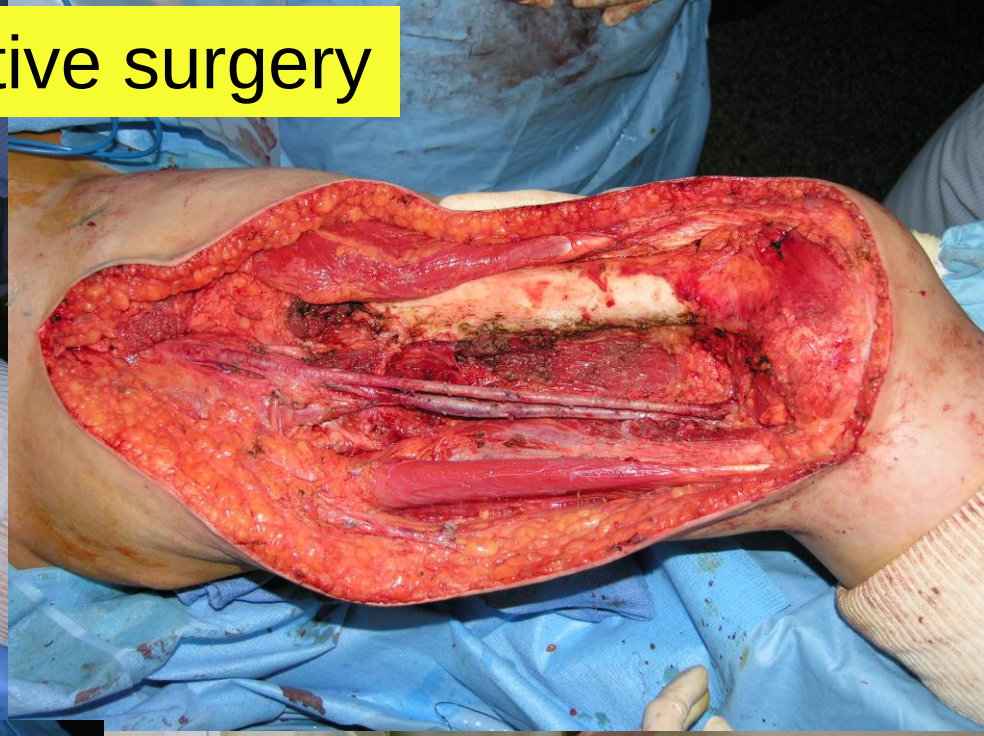


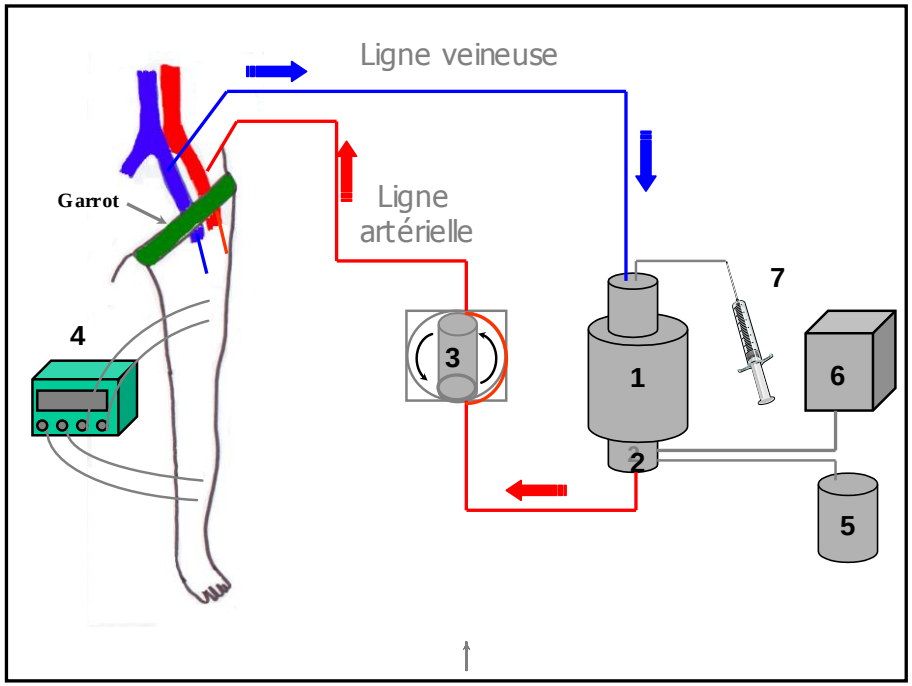


Myxoid LPS
Neo adj. RT
Flap + skin graft

Reconstructive surgery







Isolated Limb Perfusion
ILP

Ann Surg Oncol (2017) 24:194–201
DOI 10.1245/s10434-016-5462-2

Annals of
SURGICAL ONCOLOGY
OFFICIAL JOURNAL OF THE SOCIETY OF SURGICAL ONCOLOGY

ORIGINAL ARTICLE – BONE AND SOFT TISSUE SARCOMAS

Primary Extremity Soft Tissue Sarcomas: Does Local Control Impact Survival?

Sylvie Bonvalot, MD, PhD[1], Antonin Levy, MD[2,3,4], Philippe Terrier, MD[5], Dimitri Tzanis, MD, PhD[1], Sara Bellefqih, MD[2], Axel Le Cesne, MD[4,6], and Cécile Le Péchoux, MD[2]

TABLE 5 Literature overview

Study	Time period	No. P/R	FU (med years)	AR %	5-year LR %
Coindre et al.[25] 1996 (FSG)	1980–1989	546/0	5	4	29
Trovik et al.[26] 2000 (SSG)[c]	1986–1991	559/0	6.3	NA	20
Zagars et al.[27] 2003 (MDACC)[c]	1960–1999	678/170	9.5	NA	15
Weitz et al.[28] 2003 (MSKCC)[c]	1982–2001	1072/189	4.6	7	21
Khanfir et al.[29] 2003 (GR)	1975–1996	133/0	10	NI	22
Eilber et al.[30] 2003 (UCLA)	1975–1997	607/146[b]	7.3	5	10±1[a]
Gronchi et al.[31] 2005 (INT)	1980–2000	642/269[b]	8.9	5	14
Stoeckle et al.[32] 2006 (BI)[c]	1996–2002	205/0	4.4	0	13
Bonvalot et al. (GR)	1993–2012	531/0	7	2	8

ESMO GOOD SCIENCE BETTER MEDICINE BEST PRACTICE

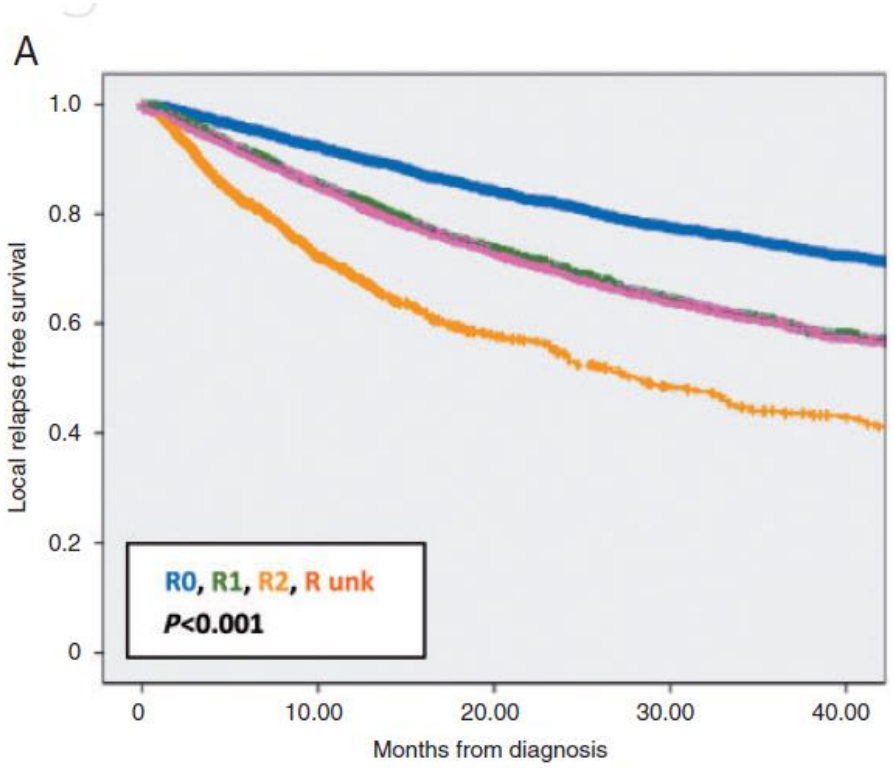
Annals of Oncology 0: 1–11, 2019
doi:10.1093/annonc/mdz124
Published online 9 April 2019

ORIGINAL ARTICLE

Surgery in reference centers improves survival of sarcoma patients: a nationwide study

J.-Y. Blay[1,2,3*], C. Honoré[4], E. Stoeckle[5], P. Meeus[1,2,3], M. Jafari[6,7], F. Gouin[1,2,3,8,9], P. Anract[10], G. Ferron[11], A. Rochwerger[12], M. Ropars[13,14], S. Carrere[15], F. Marchal[16], F. Sirveaux[16], A. Di Marco[17], L. R. Le Nail[18], J. Guiramand[19], G. Vaz[1,2,3], J.-C. Machiavello[20], O. Marco[21], S. Causeret[22], P. Gimbergues[23], F. Fiorenza[24], L. Chaigneau[25], F. Guillemin[26], J.-M. Guilloit[27], F. Dujardin[28], J.-P. Spano[29], J.-C. Ruzic[30], A. Michot[4], P. Soibinet[26], E. Bompas[8,9], C. Chevreau[11], F. Duffaud[12], M. Rios[13,14], C. Perrin[13,14], N. Firmin[15], F. Bertucci[19], C. Le Pechoux[4], F. Le Loarer[4], O. Collard[1,2,3], M. Karanian-Philippe[1,2,3], M. Brahmi[1,2,3], A. Dufresne[1,2,3], A. Dupré[1,2,3], F. Ducimetière[1,2,3], A. Giraud[10], D. Pérol[1,2,3], M. Toulmonde[5], I. Ray-Coquard[1,2,3], A. Italiano[5], A. Le Cesne[4], N. Penel[6,7] & S. Bonvalot[31], on behalf of the NETSARC/REPPS/RESOS and French Sarcoma Group–Groupe d'Etude des Tumeurs Osseuses (GSF-GETO) Networks[†]

35794 patients



R0

R1

R2

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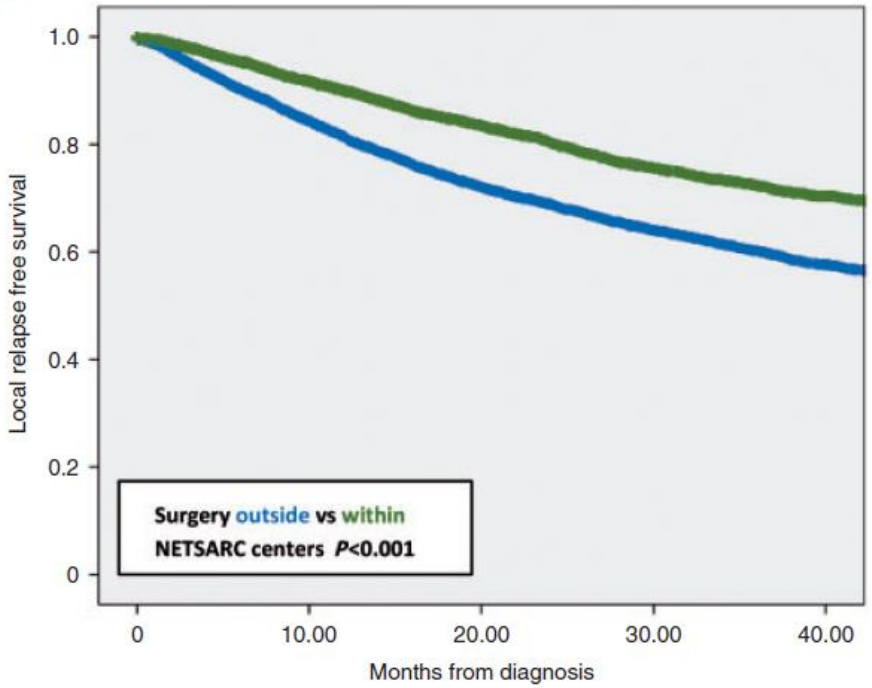
35794 patients

F



Trunk STS

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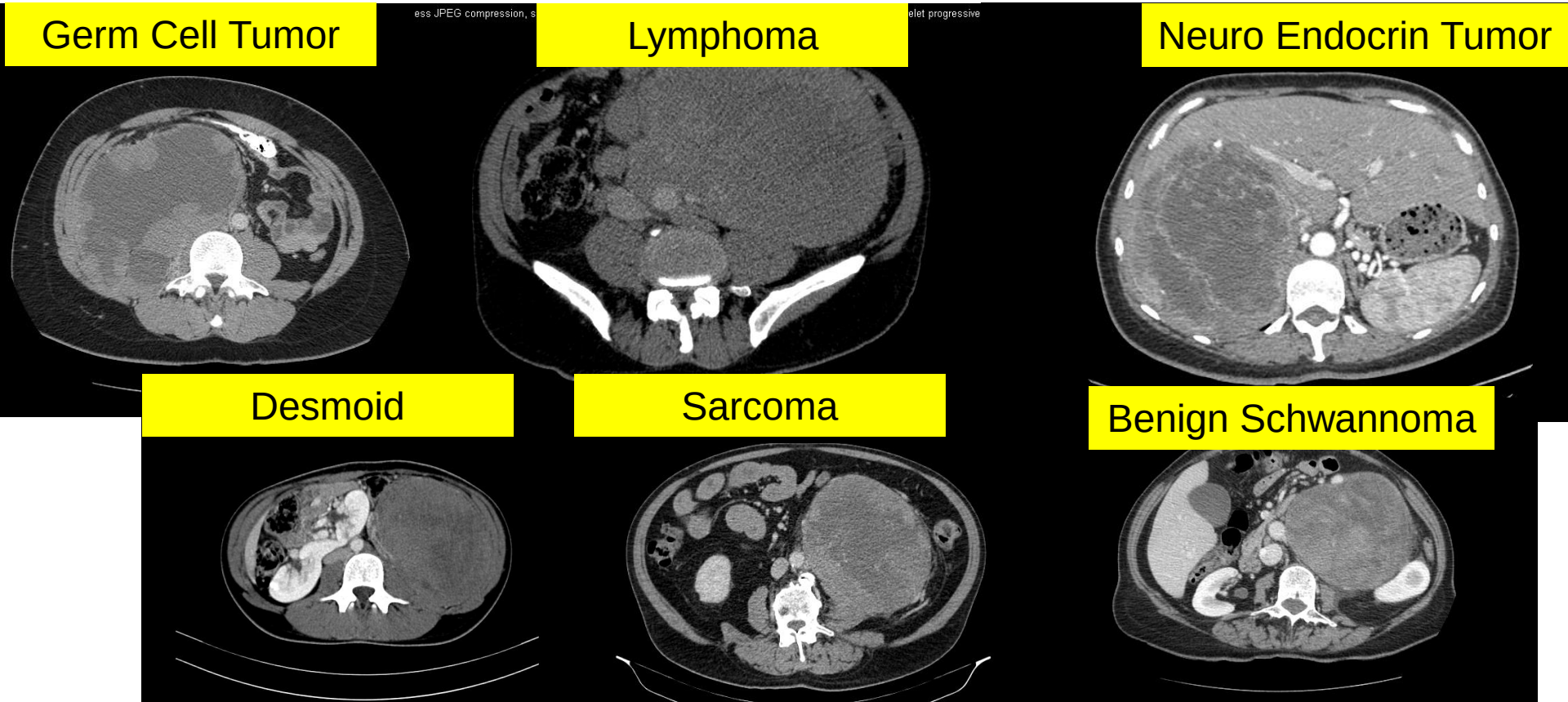
Where is the sarcoma?

Where is the sarcoma?

- Serum tumor markers
- Percutaneous biopsy
- ESMO Guidelines
  (Ann Oncol 2018)



Ann Surg Oncol
DOI 10.1245/s10434-014-3965-2

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SURGICAL ONCOLOGY
OFFICIAL JOURNAL OF THE SOCIETY OF SURGICAL ONCOLOGY

ORIGINAL ARTICLE – BONE AND SOFT TISSUE SARCOMAS

Management of Primary Retroperitoneal Sarcoma (RPS) in the Adult: A Consensus Approach From the Trans-Atlantic RPS Working Group

Trans-Atlantic RPS Working Group

ABSTRACT

Background. Retroperitoneal soft tissue sarcomas (RPS) are rare tumors that include several well-defined histologic subtypes. Although surgery is the mainstay of curative therapy, no universally accepted recommendations concerning the best management have been developed to date. Optimization of the initial approach is critical for maximizing patient outcomes.

Methods. An RPS Trans-Atlantic Working Group was established in 2013. The primary aim was to evaluate the current evidence critically and to develop a consensus document on the approach to this difficult disease. The outcome applies to primary RPS that is nonvisceral in origin. The evaluation included sarcomas of major veins (inferior vena cava, renal vein, ovarian/testicular vein), undifferentiated pleomorphic sarcoma of the psoas, and ureteric leiomyosarcoma (LMS). It excluded desmoid, lipoma and angiomyolipoma, gastrointestinal stromal tumors, visceral sarcomas such as those arising from the gut or its mesentery, uterine LMS, prostatic sarcoma, paratesticular/spermatic cord sarcoma, Ewing's sarcoma, alveolar/embryonal rhabdomyosarcoma, primitive peripheral neuro-ectodermal tumor, sarcoma arising from teratoma, carcinosarcoma, sarcomatoid carcinoma, clear cell sarcoma, radiation-induced sarcoma, paraganglioma, and malignant pheochromocytoma.

Results. Management of RPS was evaluated from diagnosis to follow-up, and a level of evidence was attributed to each statement. This rare and complex malignancy is best managed by an experienced multidisciplinary team in a specialized referral center. The best chance of cure is at the time of primary presentation, and an individualized management plan should be made based on the statements included in this article.

Conclusions. International collaboration is critical for adding to the current knowledge. A prospective registry will be set up.

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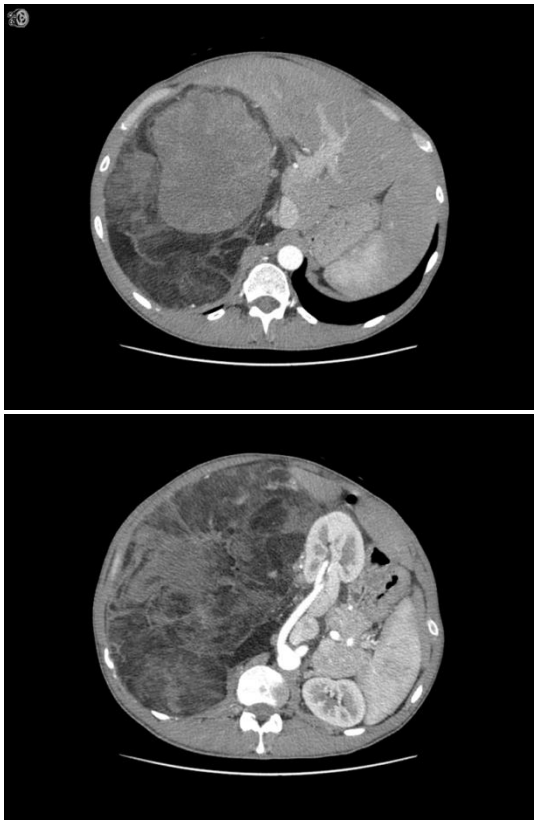
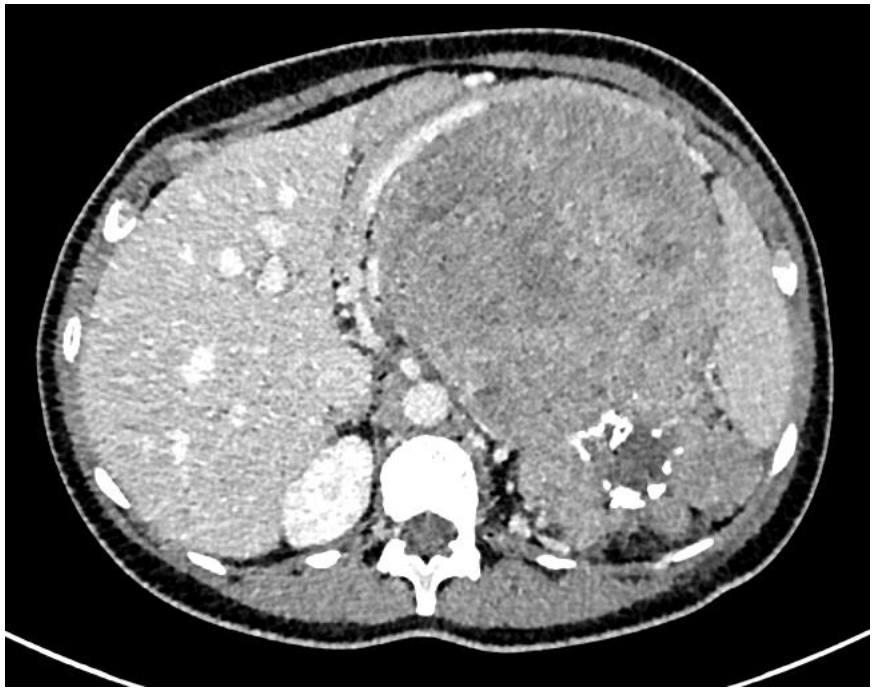
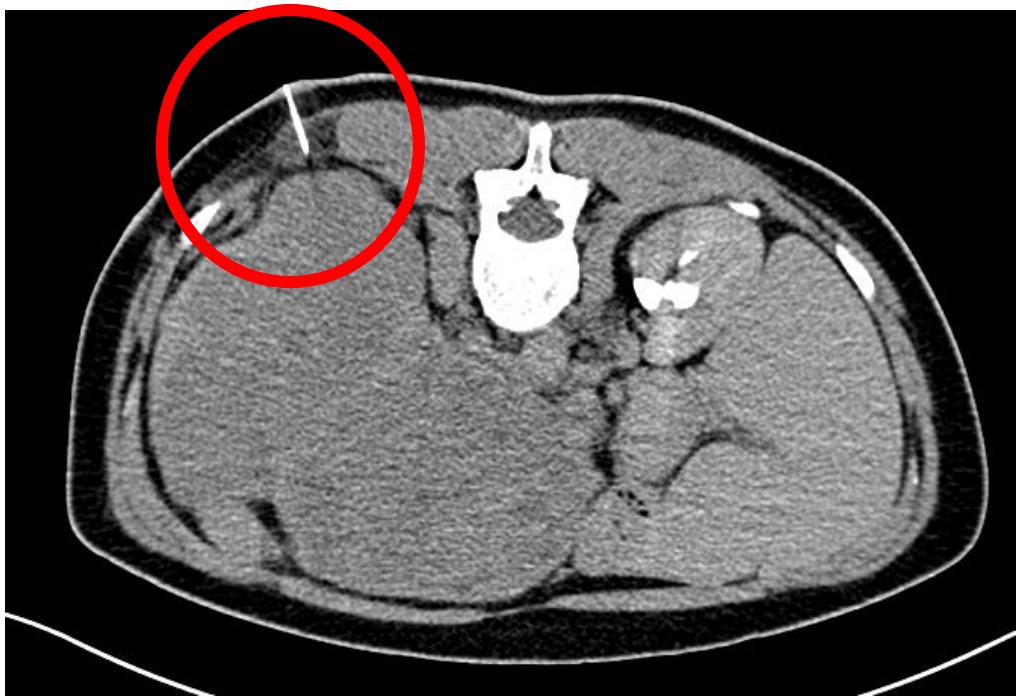
3. Image-guided percutaneous coaxial core needle biopsy (14 or 16 gauge) is strongly recommended (Fig. 1) unless the imaging is pathognomonic (e.g., heterogeneous dedifferentiated/well-differentiated liposarcoma) and no preoperative treatment is planned (IVA).[25–28]

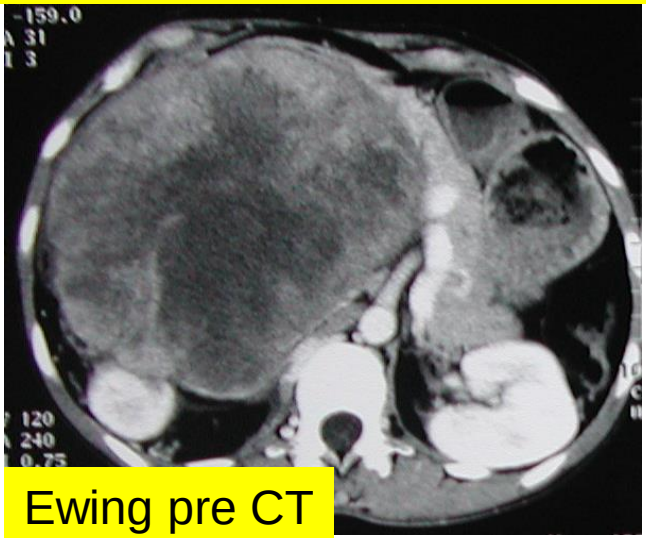


Percutaneous biopsy via a retro peritoneal route

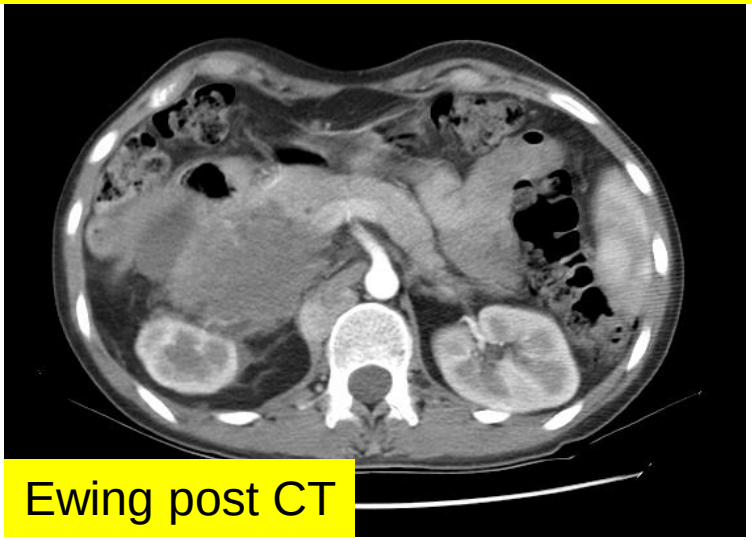




Multidisciplinary sarcoma team to discuss the best strategy



Ewing pre CT



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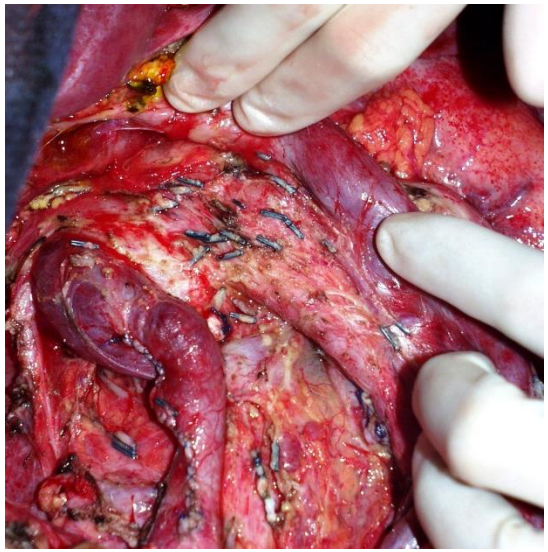

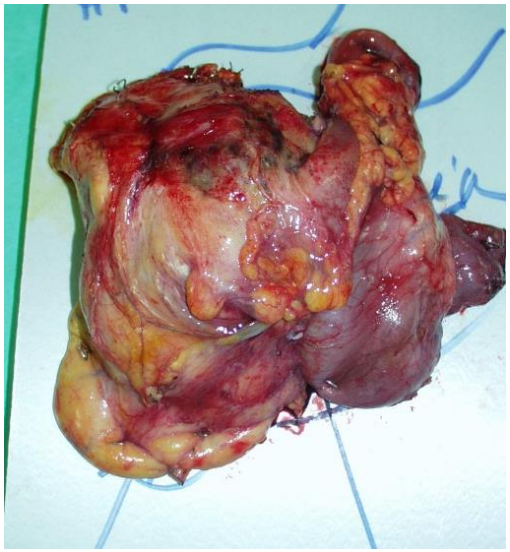

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Second step: recognize the entire tumor

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the Journal of Cancer Surgery

EJSO 43 (2017) 1191–1198

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Review

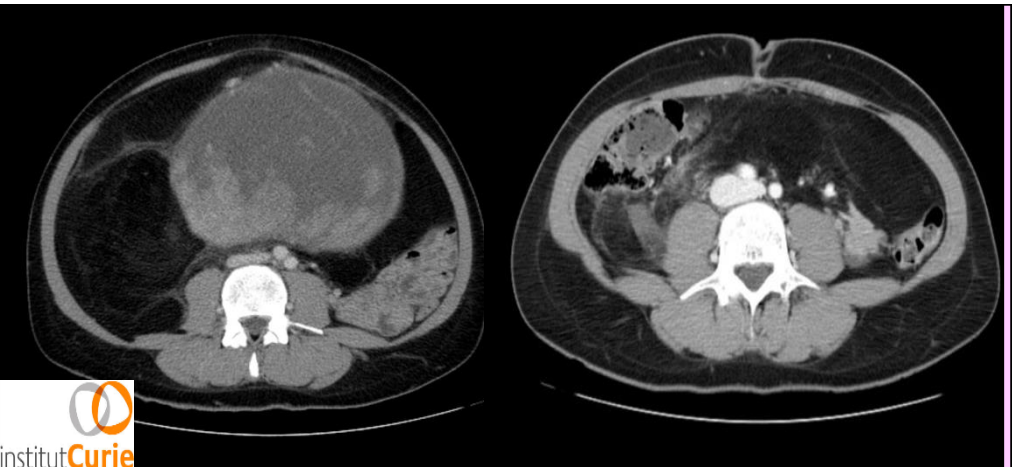
Primary retroperitoneal soft tissue sarcoma: Imaging appearances, pitfalls and diagnostic algorithm

C. Messiou [a,*], E. Moskovic [a], D. Vanel [b], C. Morosi [c], R. Benchimol [d], D. Strauss [e], A. Miah [f], H. Douis [g], W. van Houdt [e], S. Bonvalot [h]

Risk of under estimation of well differenciated part → R2

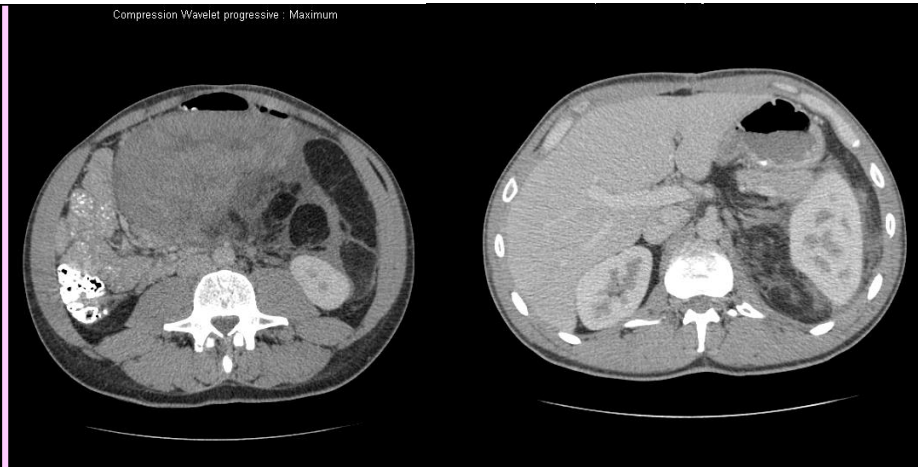
Pre op

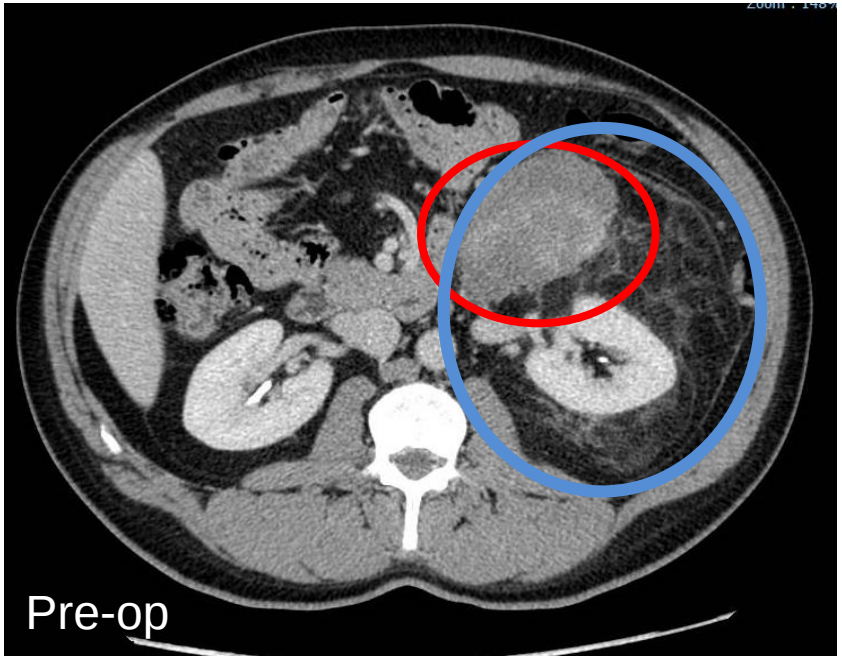
Post op

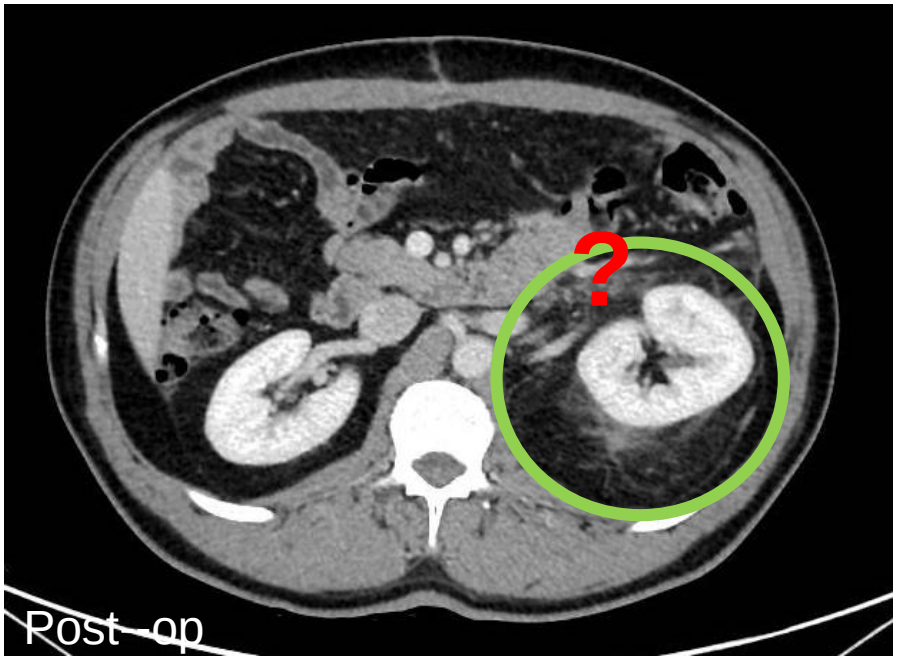


Pre op

Post op




Pre-op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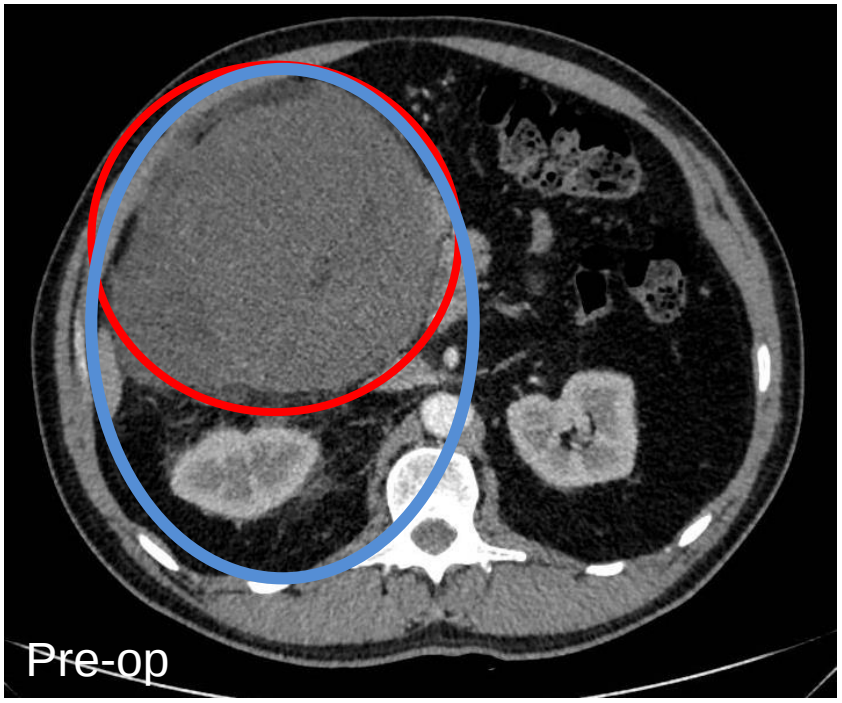
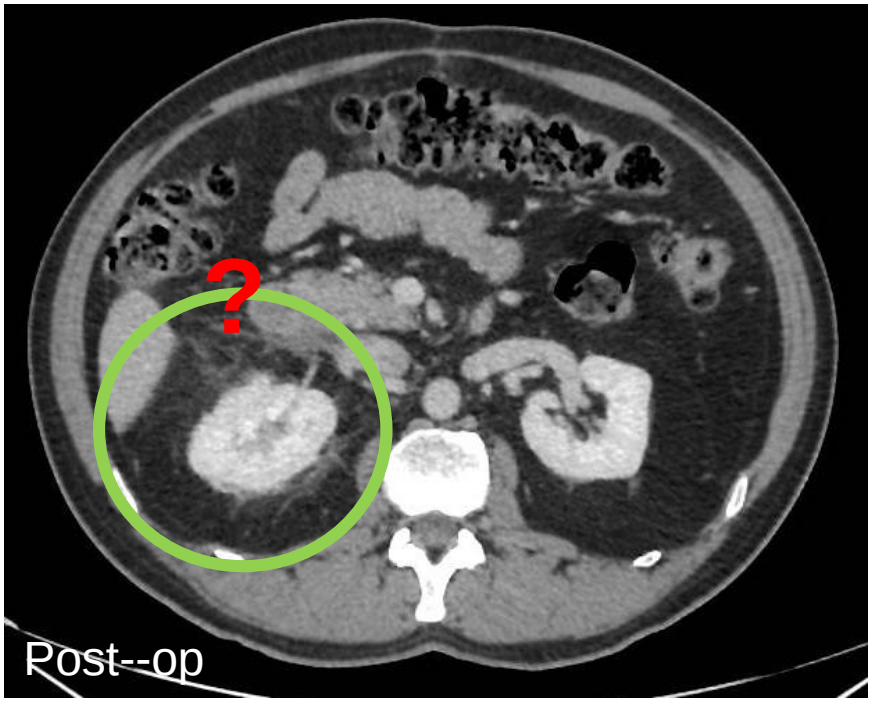
Post--op


Pre-op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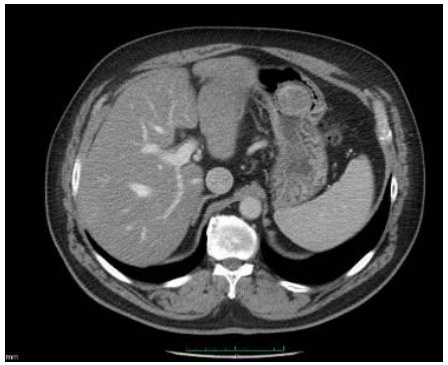
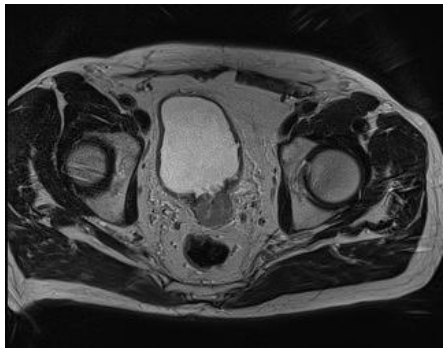
Post--op

Third step: extent of resection

Extent of resection

Visceral sarcoma





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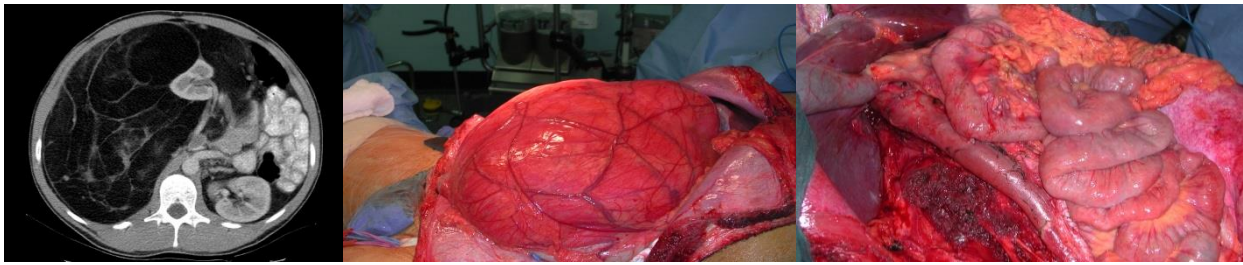
Soft tissue Sarcoma

Limbs and retroperitoneum = similar philosophia



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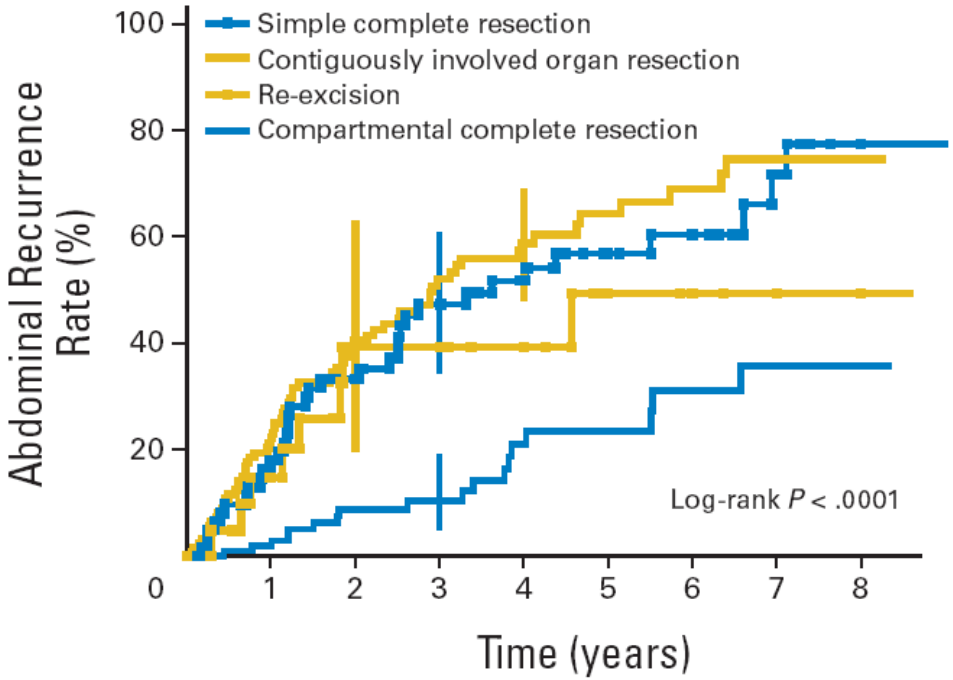
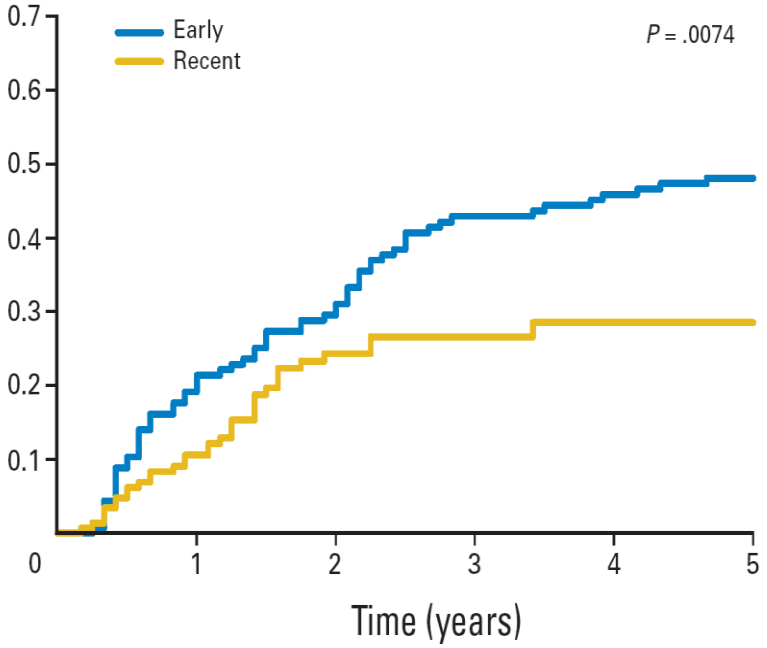
ORIGINAL REPORT



Bonvalot et al. JCO 2009



Gronchi et al. JCO 2009

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ORIGINAL ARTICLE – BONE AND SOFT TISSUE SARCOMAS

Surgical Management of Primary Retroperitoneal Sarcomas: Rationale for Selective Organ Resection

Mark Fairweather, MD[1,2], Jiping Wang, MD, PhD[1,2,3], Vickie Y. Jo, MD[2,3,4], Elizabeth H. Baldini, MD, MPH[2,3,5], Monica M. Bertagnolli, MD[1,2,3], and Chandrajit P. Raut, MD, MSc[1,2,3]

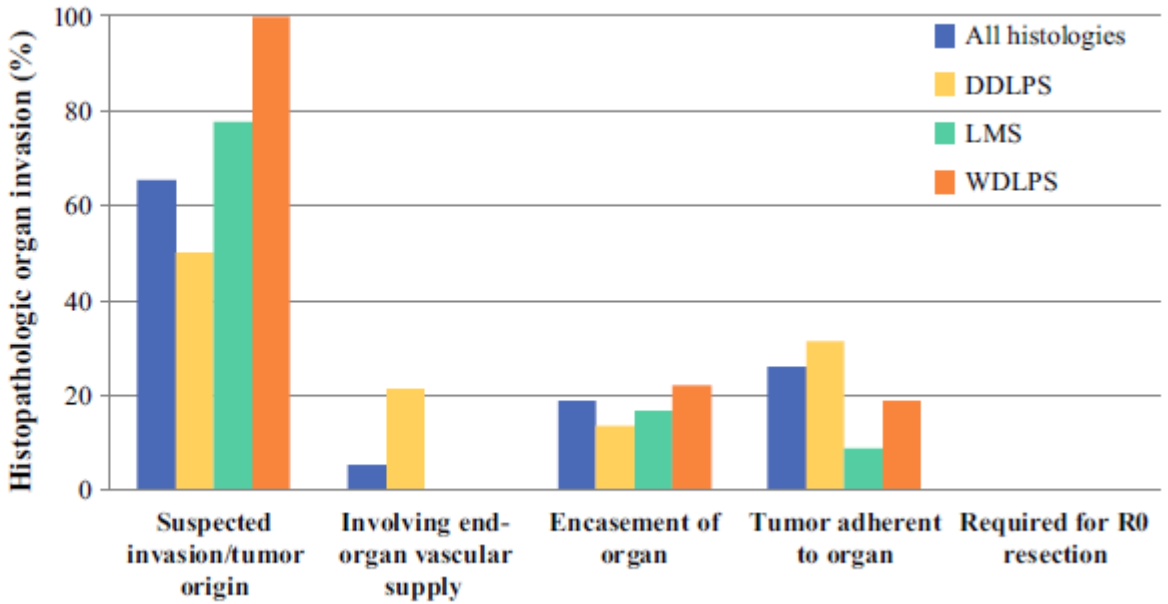

FIG. 3 Rate of histopathologic organ invasion based on the rationale for organ resection for all histologies and by histologic subtype ($n = 302$)

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ORIGINAL ARTICLE – BONE AND SOFT TISSUE SARCOMAS

Technical Considerations in Surgery for Retroperitoneal Sarcomas: Position Paper from E-Surge, a Master Class in Sarcoma Surgery, and EORTC–STBSG

Syvie Bonvalot, MD, PhD[1], Chandrajit P. Raut, MD, MSc[2], Raphael E. Pollock, MD, PhD[3], Piotr Rutkowski, MD[4], Dirk C. Strauss, MD[5], Andrew J. Hayes, MD[5], Frits Van Coevorden, MD[6], Marco Fiore, MD[7], Eberhard Stoeckle, MD[8], Peter Hohenberger, MD, PhD[9], and Alessandro Gronchi, MD[7]

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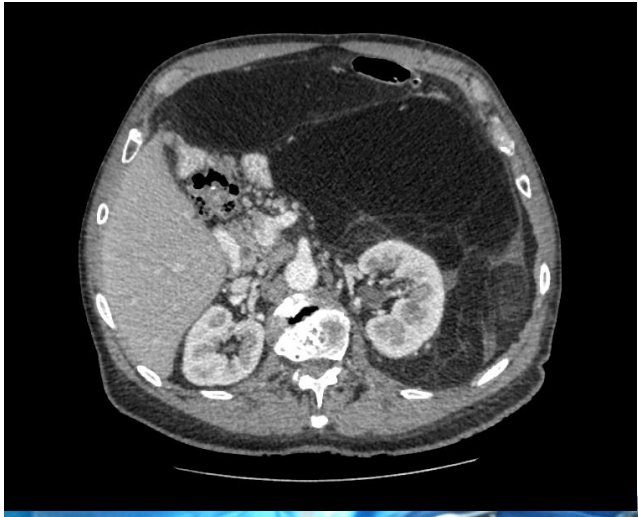

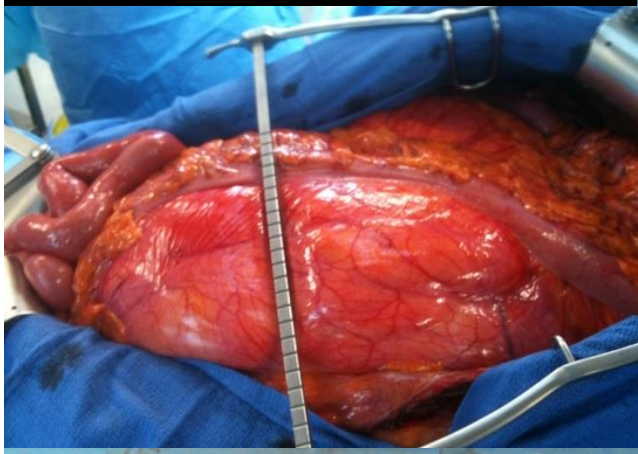

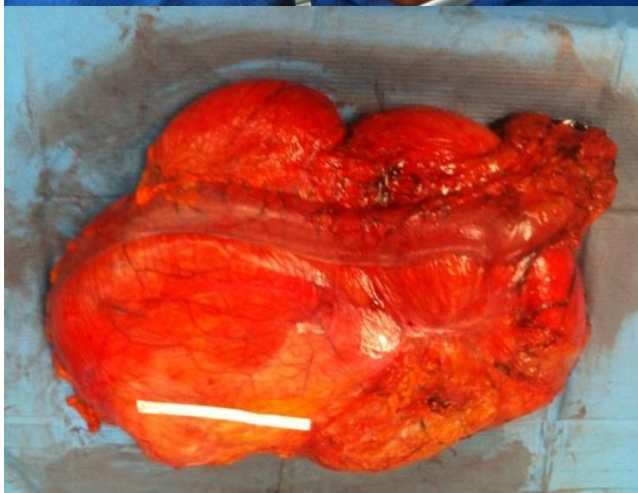

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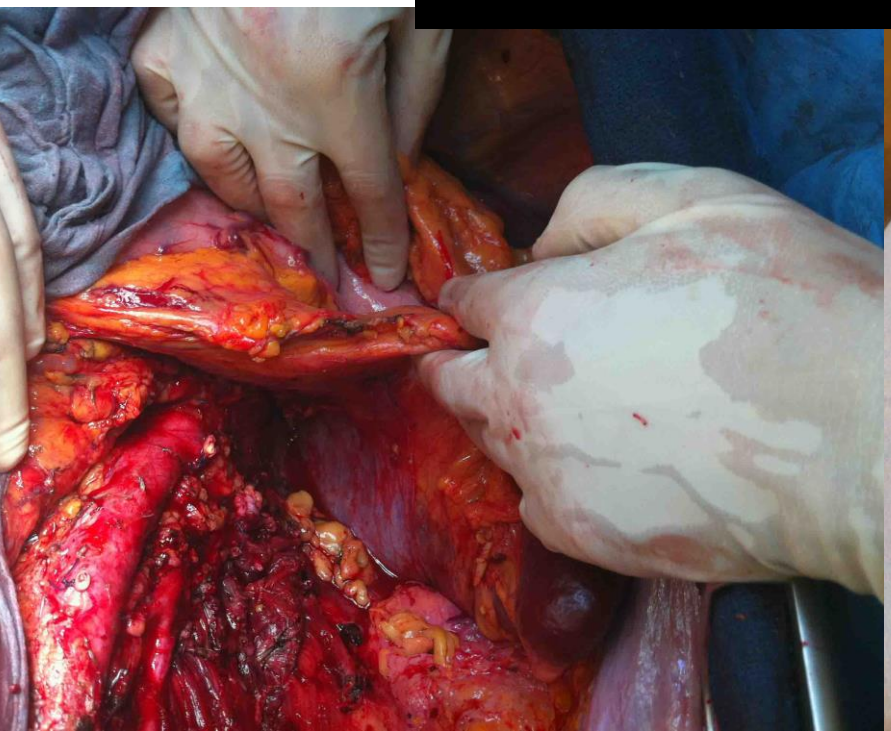
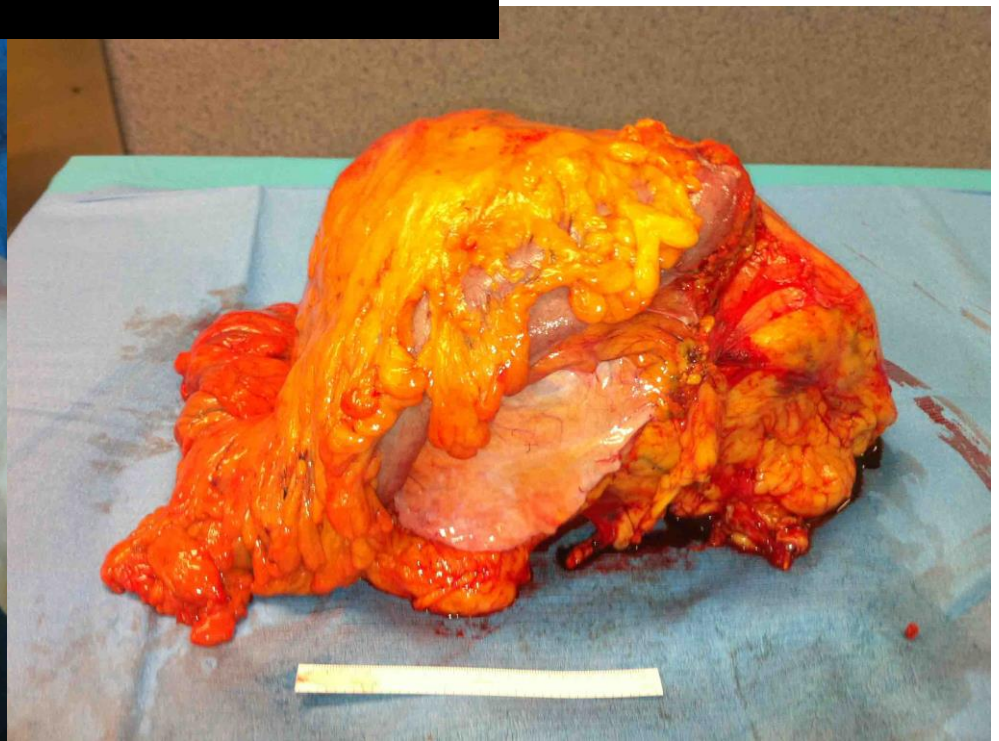
Compression Wavelet progressive : Maximum





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ORIGINAL ARTICLE – BONE AND SOFT TISSUE SARCOMAS

Management of Primary Retroperitoneal Sarcoma (RPS) in the Adult: A Consensus Approach From the Trans-Atlantic RPS Working Group

Trans-Atlantic RPS Working Group

ABSTRACT

Background. Retroperitoneal soft tissue sarcomas (RPS) are rare tumors that include several well-defined histologic subtypes. Although surgery is the mainstay of curative therapy, no universally accepted recommendations concerning the best management have been developed to date. Optimization of the initial approach is critical for maximizing patient outcomes.

Methods. An RPS Trans-Atlantic Working Group was established in 2013. The primary aim was to evaluate the current evidence critically and to develop a consensus document on the approach to this difficult disease. The outcome applies to primary RPS that is nonvisceral in origin. The evaluation included sarcomas of major veins (inferior vena cava, renal vein, ovarian/testicular vein), undifferentiated pleomorphic sarcoma of the psoas, and ureteric leiomyosarcoma (LMS). It excluded desmoid, lipoma and angiomyolipoma, gastrointestinal stromal tumors, visceral sarcomas such as those arising from the gut or its mesentery, uterine LMS, prostatic sarcoma, paratesticular/spermatic cord sarcoma, Ewing's sarcoma, alveolar/embryonal rhabdomyosarcoma, primitive peripheral neuro-ectodermal tumor, sarcoma arising from teratoma, carcinosarcoma, sarcomatoid carcinoma, clear cell sarcoma, radiation-induced sarcoma, paraganglioma, and malignant pheochromocytoma.

Results. Management of RPS was evaluated from diagnosis to follow-up, and a level of evidence was attributed to each statement. This rare and complex malignancy is managed by an experienced multidisciplinary team specialized referral center. The best chance of cure is a time of primary presentation, and an individualized m agement plan should be made based on the statem included in this article.

Conclusions. International collaboration is critical adding to the current knowledge. A prospective reg will be set up.

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Trans-Atlantic RPS Working Group

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12a. In the case of primary RPS, surgery should be aimed at achieving macroscopically complete resection, with a single specimen encompassing the tumor and involved contiguous organs, and at minimizing microscopically positive margins. This is best achieved by resecting the tumor en bloc with adherent structures even if not overtly infiltrated (IIIA)[32] (Fig. 2).

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ORIGINAL ARTICLE – SARCOMA

Survival Benefit of the Surgical Management of Retroperitoneal Sarcoma in a Reference Center: A Nationwide Study of the French Sarcoma Group from the NetSarc Database

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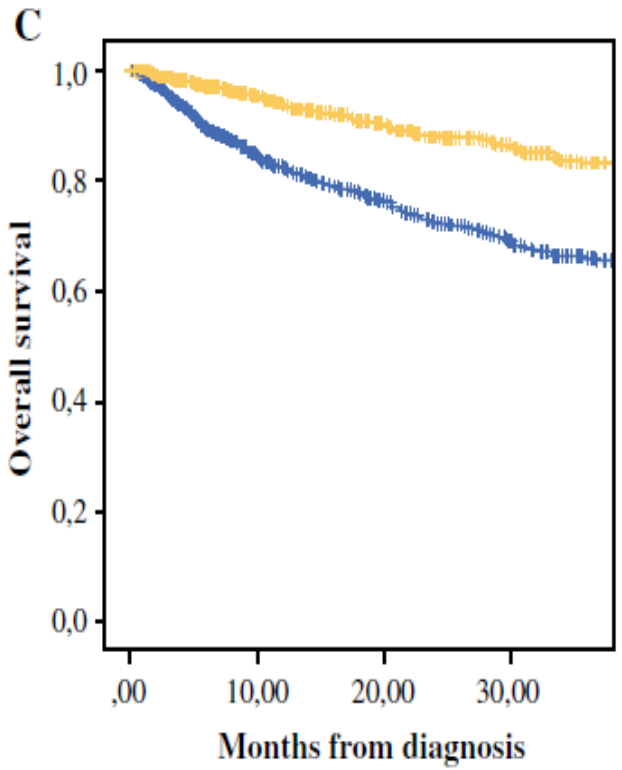


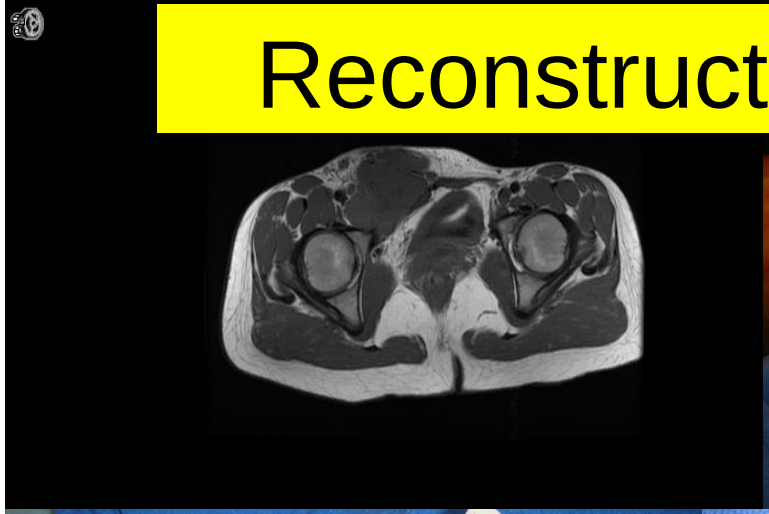
2945 primary RPS
2010-2017

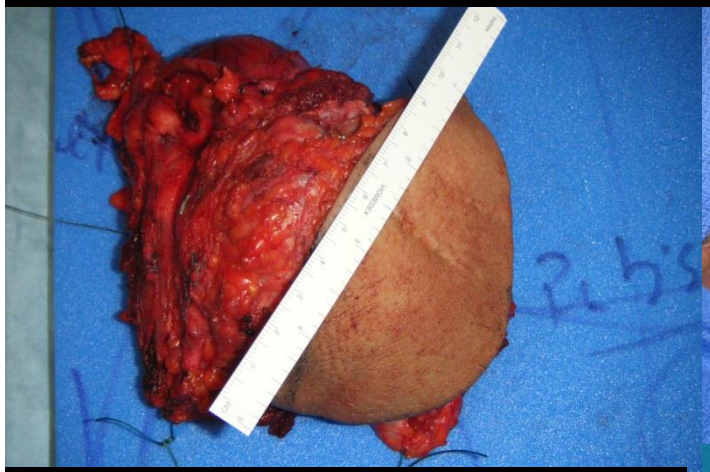
Surgery in a sarcoma center

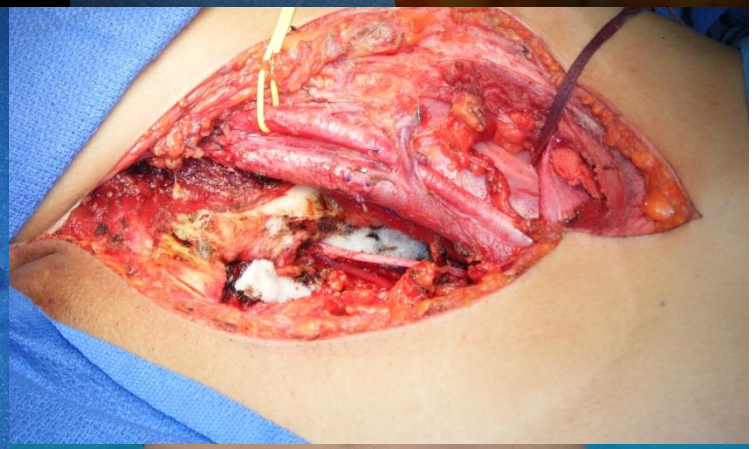
Surgery outside a sarcoma center

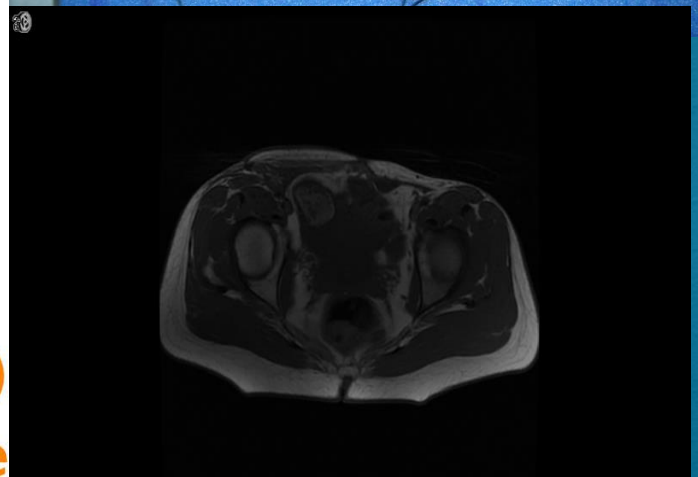
ASO mai 2019

Reconstructive surgery









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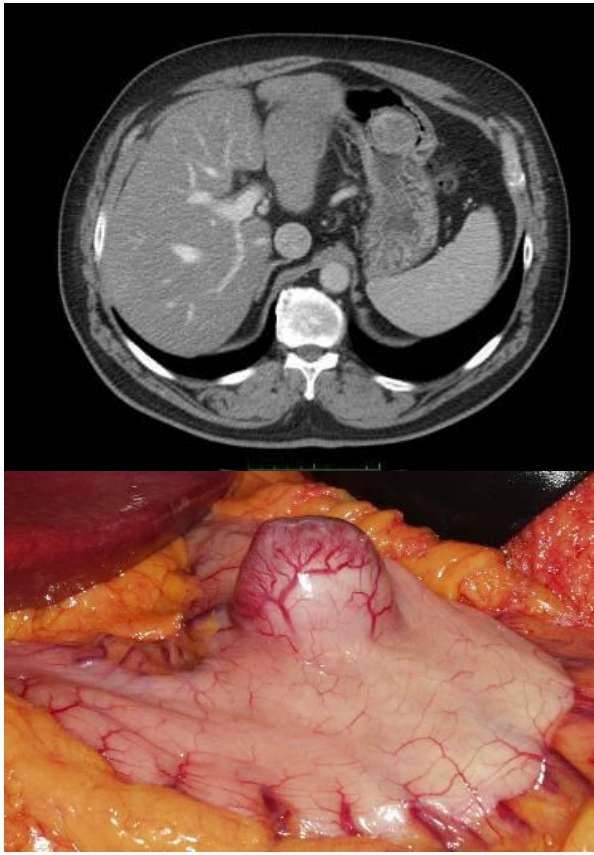
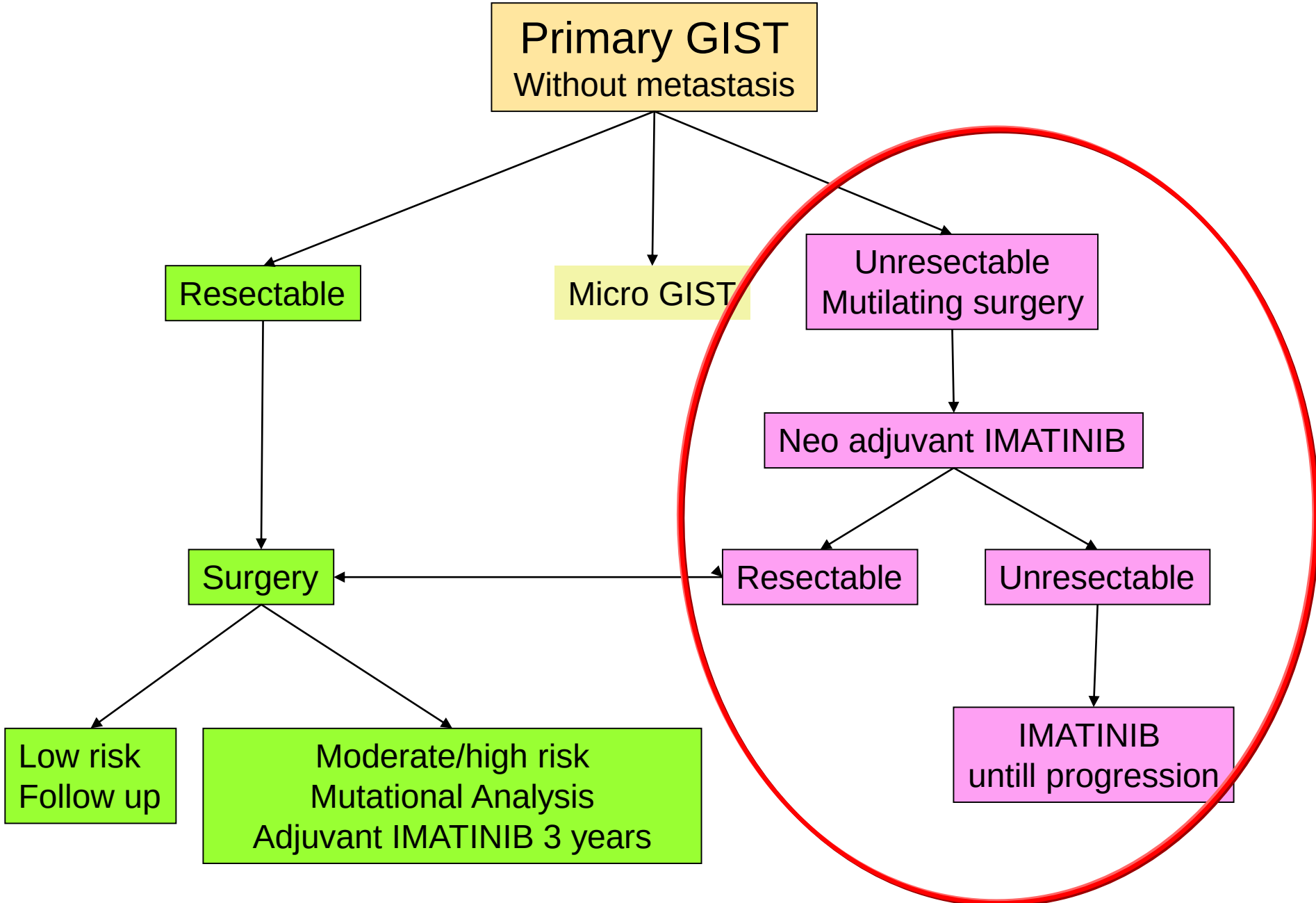
VISCERAL SARCOMA AND GISTs

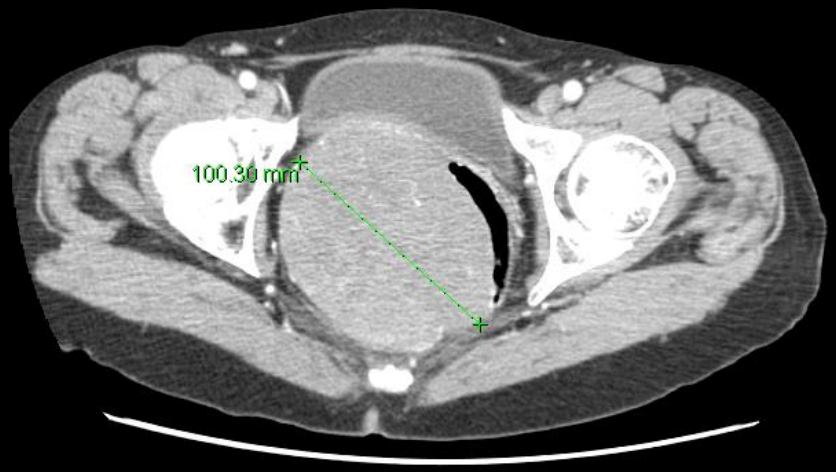
Surgery in Primary Disease

- Complete macroscopic resection without rupture
- With microscopically negative margins over the organ of origin (R0 resection)
- Given the rarity of lymphatic invasion, systematic lymph node dissection is not recommended
- Laparoscopy is not recommended if there is a risk of tumor rupture



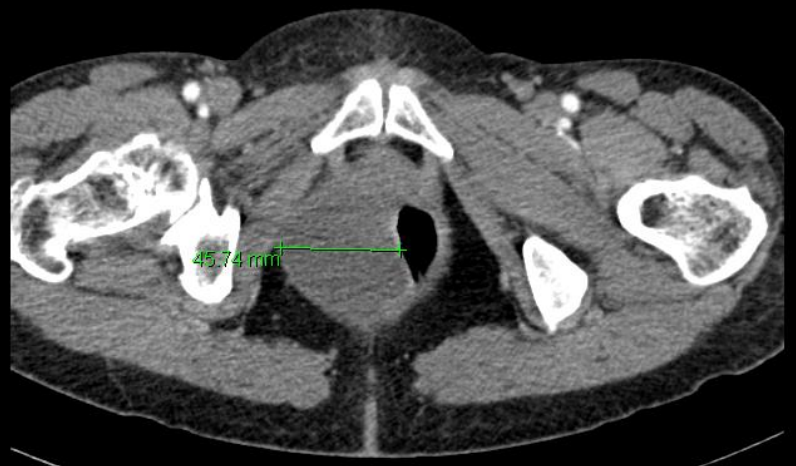
Global strategy





Initial CT

CT 9 months treatment



Conclusions

- Surgery must be done with the knowledge that it is a sarcoma
- Per cutaneous biopsy must be done before the surgery
- Best strategy is decided in multidisciplinary tumor board
- Objective of surgery is to obtain clear margins = a rim of normal tissue around the tumor
- Surgery should be done in a sarcoma center

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