CANCER EDUCATION DAY

Management of Non-Extremity Sarcoma

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Presenter Disclosure

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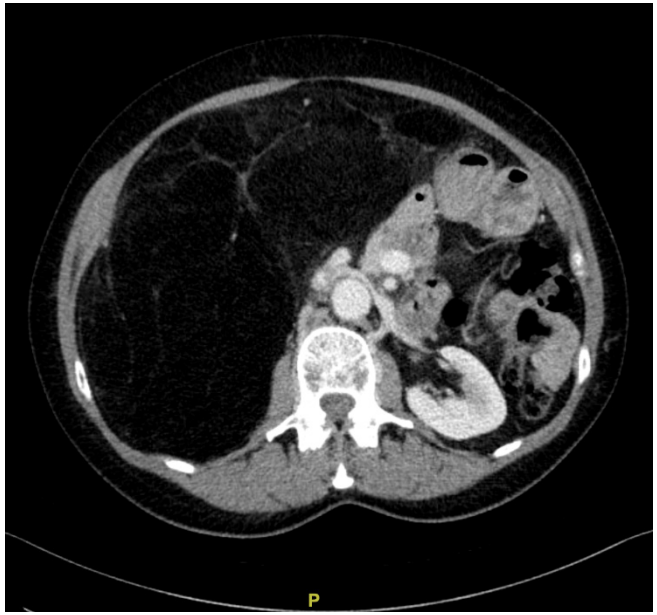
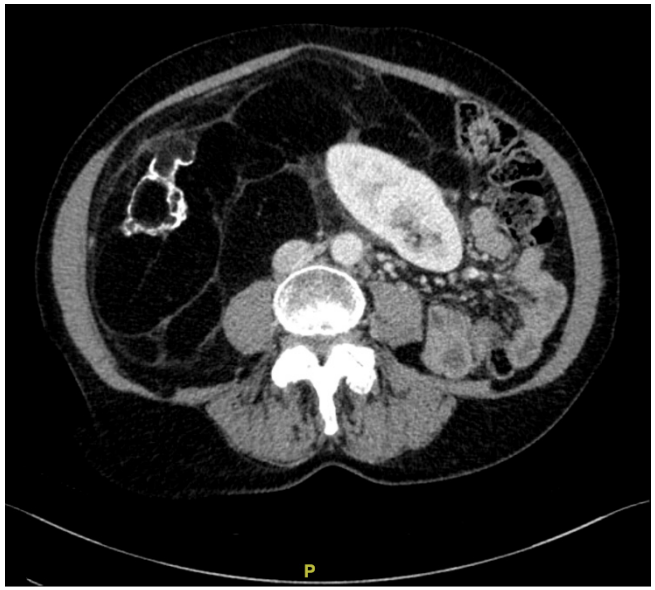
Overview

- Common presentations, work up, management
- Importance of multidisciplinary management
  - Retroperitoneal sarcoma
  - Abdominal wall masses/sarcoma
  - Inguinal / groin sarcoma

Case 1

- 65 yo F presented to ER with symptoms of chest tightness
  - CT PE, negative
  - Incidental finding of right retroperitoneal lipomatous mass
  - CT AP done following day
- Complex lipomatous mass 23.1 x 19.5 x 29.6 cm, leftward displacement of kidney, small and large bowel





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Retroperitoneal Sarcoma

- Work up:
  - History and physical: in retrospect noted increase in abdominal girth and clothing fitting differently
  - Core biopsy: well differentiated liposarcoma, MDM2 +
  - Split renal scan (nuclear medicine): left kidney – 88 %, right kidney 12 % of total renal function
  - Review at Sarcoma MCC:
    - Large but resectable, encasement of right kidney requires resection
    - Discussion regarding preoperative radiation – large field, not recommended

Retroperitoneal Sarcoma

TABLE 1 Histologic subtypes of primary retroperitoneal sarcoma

Histologic subtype	MSKCC, 2016 [$n = 675$]	TARPSWG, 2020 [$n = 1942$]	Total [$n = 2617$]
Well-differentiated liposarcoma	186 (28%)	446 (23%)	632 (24%)
Dedifferentiated liposarcoma	213 (32%)	829 (43%)	1042 (40%)
Leiomyosarcoma	140 (23%)	352 (18%)	492 (20%)
Solitary fibrous tumor	33 (5%)	105 (5%)	138 (5%)
Malignant peripheral nerve sheath tumor	23 (3%)	54 (3%)	77 (3%)
Other	69 (9%)	156 (8%)	164 (8%)
Unclassified/undifferentiated pleomorphic sarcoma (UPS)	–	61 (3%)	
Sarcoma NOS	35 (5%)	21 (1%)	
Translocation associated and other	34 (5%)	74 (4%)	

Data are expressed as *n* (%)

MSKCCC Memorial Sloan-Kettering Cancer Center,[101] *TARPSWG* Transatlantic Australasian Retroperitoneal Sarcoma Working Group,[102] *NOS* not otherwise specified

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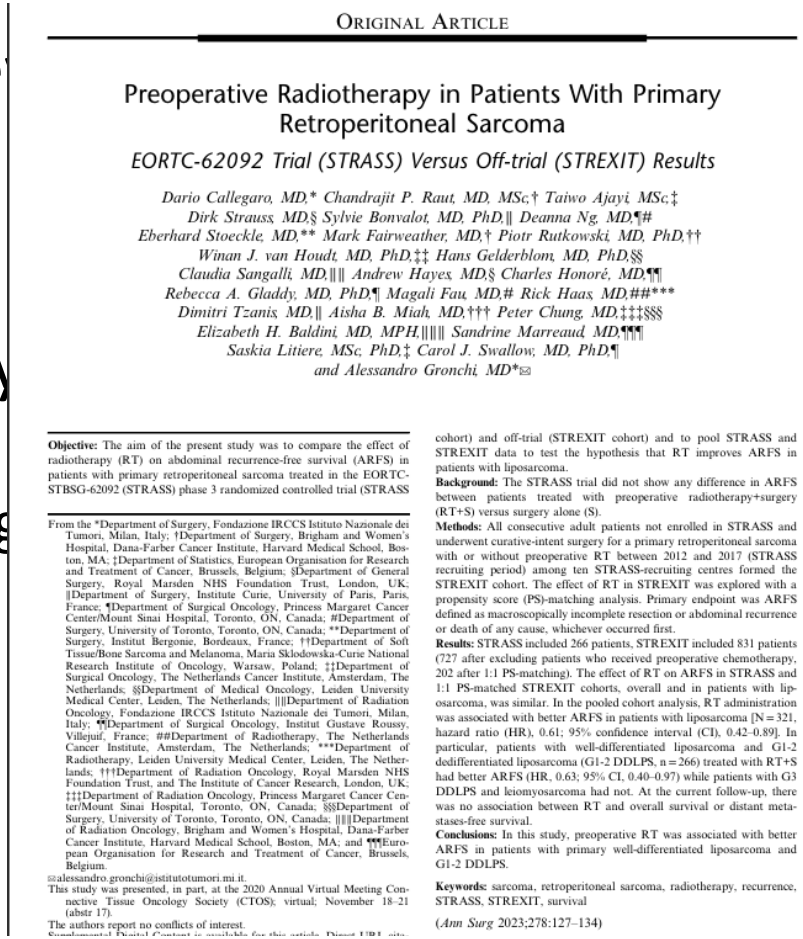
Swallow et al., 2021

Principles of Management

- Establish diagnosis, re
- Goal is complete (R0),
- Preservation of critica
  reconstruction case by
  - Use of radiation thera
    anticipated close marg



ORIGINAL ARTICLE

Preoperative Radiotherapy in Patients With Primary Retroperitoneal Sarcoma

EORTC-62092 Trial (STRASS) Versus Off-trial (STREXIT) Results

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Objective: The aim of the present study was to compare the effect of radiotherapy (RT) on abdominal recurrence-free survival (ARFS) in patients with primary retroperitoneal sarcoma treated in the EORTC-STBSG-62092 (STRASS) phase 3 randomized controlled trial (STRASS cohort) and off-trial (STREXIT cohort) and to pool STRASS and STREXIT data to test the hypothesis that RT improves ARFS in patients with liposarcoma.

Background: The STRASS trial did not show any difference in ARFS between patients treated with preoperative radiotherapy+surgery (RT+S) versus surgery alone (S).

Methods: All consecutive adult patients not enrolled in STRASS and underwent curative-intent surgery for a primary retroperitoneal sarcoma with or without preoperative RT between 2012 and 2017 (STRASS recruiting period) among ten STRASS-recruiting centres formed the STREXIT cohort. The effect of RT in STREXIT was explored with a propensity score (PS)-matching analysis. Primary endpoint was ARFS defined as macroscopically incomplete resection or abdominal recurrence or death of any cause, whichever occurred first.

Results: STRASS included 266 patients, STREXIT included 831 patients (727 after excluding patients who received preoperative chemotherapy, 202 after 1:1 PS-matching). The effect of RT on ARFS in STRASS and 1:1 PS-matched STREXIT cohorts, overall and in patients with liposarcoma, was similar. In the pooled cohort analysis, RT administration was associated with better ARFS in patients with liposarcoma [N = 321, hazard ratio (HR), 0.61; 95% confidence interval (CI), 0.42–0.89]. In particular, patients with well-differentiated liposarcoma and G1-2 dedifferentiated liposarcoma (G1-2 DDLPS, n = 266) treated with RT+S had better ARFS (HR, 0.63; 95% CI, 0.40–0.97) while patients with G3 DDLPS and leiomyosarcoma had not. At the current follow-up, there was no association between RT and overall survival or distant metastases-free survival.

Conclusions: In this study, preoperative RT was associated with better ARFS in patients with primary well-differentiated liposarcoma and G1-2 DDLPS.

Keywords: sarcoma, retroperitoneal sarcoma, radiotherapy, recurrence, STRASS, STREXIT, survival

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This study was presented, in part, at the 2020 Annual Virtual Meeting Connective Tissue Oncology Society (CTOS); virtual; November 18–21 (abstr 17).

The authors report no conflicts of interest.

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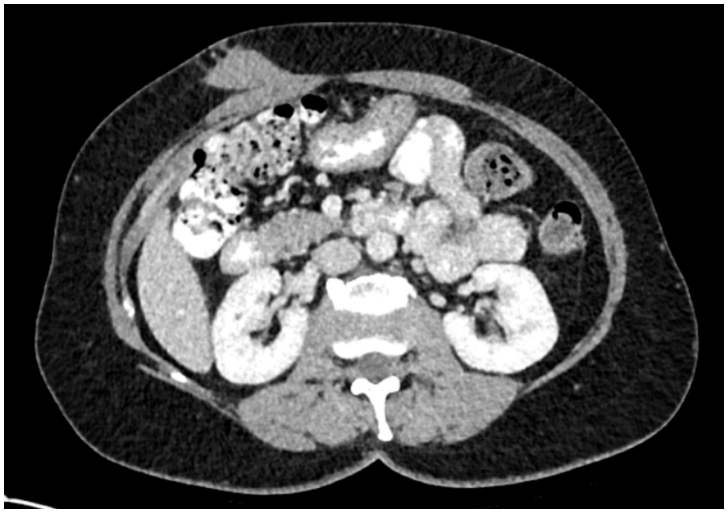
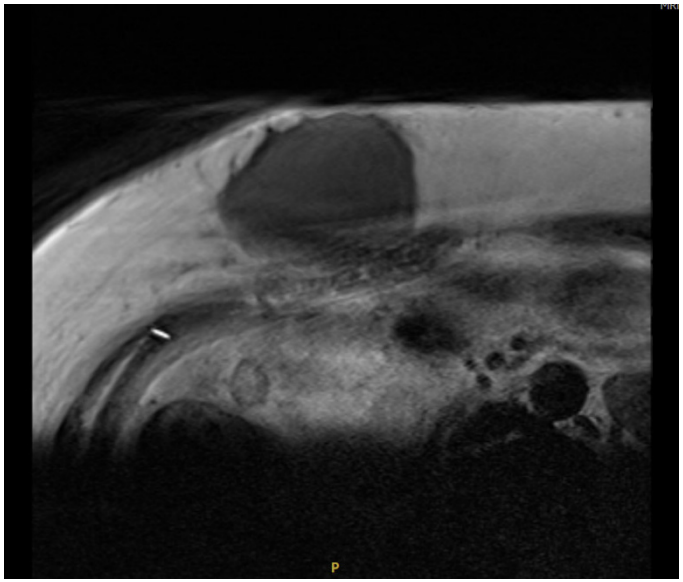
Case 2

- 61 yo F
- Presented to Family MD with grape-sized lump in RUQ abdomen
  - Growing steadily, never drained
- Had US, then CT
- Core Biopsy:
  - Bland fibroblastic proliferation, favour fibromatosis, desmoid type





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Abdominal Wall Sarcoma

- Fibromatosis – desmoid type
  - Evaluation for symptoms – patient was minimally symptomatic, only with direct pressure on area
  - Plan: saw medical oncology, NSAIDs were recommended

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Annals of
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ORIGINAL ARTICLE – BONE AND SOFT TISSUE SARCOMAS

Spontaneous Regression of Primary Abdominal Wall Desmoid Tumors: More Common than Previously Thought

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ABSTRACT

Purpose. The relevance of the initial observational approach for desmoid tumors (DTs) remains unclear. We investigated a new conservative management treatment for primary abdominal wall DTs.

Methods. Data were collected from 147 patients between 1993 and 2012. The initial therapeutic approaches were categorized as front-line surgery [surgery group (SG), $n = 41$, 28 %] and initial observation or medical treatment [nonsurgery group (NSG), $n = 106$, 72 %]. The cumulative incidence of the last strategy modification was estimated using competing risk methods with variable censoring times.

Results. Of the 147 patients, 143 were female (97 %). In the SG, 27 patients (66 %) required full-thickness abdominal wall mesh repair. In the NSG, 102 patients (96 %) underwent initial observation and four received medical treatment. In the NSG, the 1- and 3-year incidences of changing to medical treatment (no further changes during the follow-up) were 19 % [95 % confidence interval (CI) 11–28] and 25 % (95 % CI 17–35), respectively, and the 1- and 3-year incidences of a final switch to surgery were 14 % (95 % CI 8–22) and 16 % (95 % CI 9–24), respectively. An initial tumor size of >7 cm was associated with a higher strategy modification risk ($p = 0.004$). Of the 102 patients initially observed, 29 experienced spontaneous regression over a median follow-up period of 32 months. All second-intent resections were macroscopically completed, with R0 resections achieved in 82 % of patients.

Conclusions. This study supports an initial nonsurgical approach to abdominal wall DTs ≤7 cm, followed by surgery based on tumor growth in select cases.

Surgery has historically been the primary treatment for patients with resectable desmoid tumors (DTs).[1,2] Recently, the 2012 Guidelines for soft tissue sarcoma included observation as an option for selected patients with resectable DTs.[3,4] These modifications were made on the basis of recent retrospective analyses, some of them including children.[5–11] However, the criteria for selecting the patients for whom watchful waiting is most beneficial require clarification.[12]

Conflicting results from recent retrospective studies investigating the impact of surgical margins illustrate the heterogeneity of rare disease forms, including tumors causing both indolent and more aggressive disease, requiring different treatment options, as well as the unknown host/tumor factors influencing tumor progression.[1,2,5,13,14]

There is consensus that abdominal wall tumors demon-

voked DVT in distant past

up – 6 months

-during the “wait and see” period, 1/3 of patients remained stable, 1/3 spontaneously regressed

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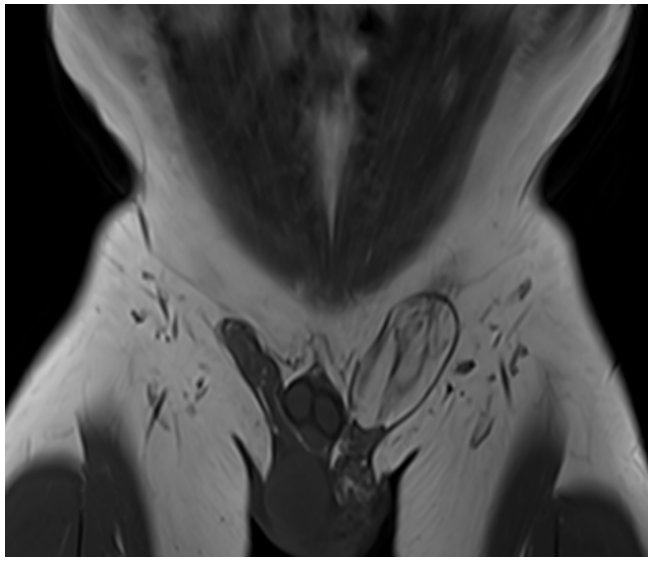
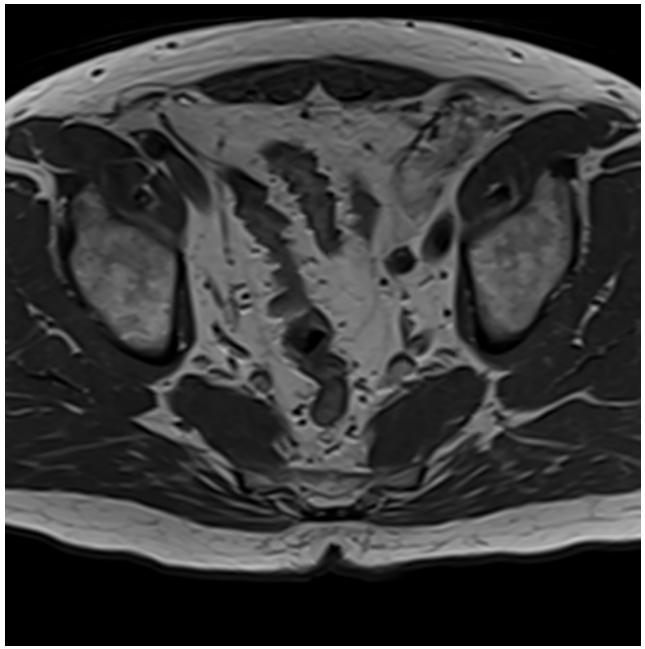
Principles of Management

- Clinic visit at 6 months:
  - Repeat Ultrasound – growth of mass by ~2 cm, more symptomatic and painful
  - Plan: surgical resection with abdominal wall resection, mesh reconstruction
    - R0 resection

Case 3

- 65 M presented to a general surgeon with an inguinal lump, large and non-reducible
- Seen by a general surgeon for possible inguinal hernia repair
- No imaging pre-operatively
- Open inguinal hernia repair attempted
  - Large fatty mass encountered, peeled from spermatic cord/contents
  - Pathology consistent with well-differentiated liposarcoma, MDM2 +





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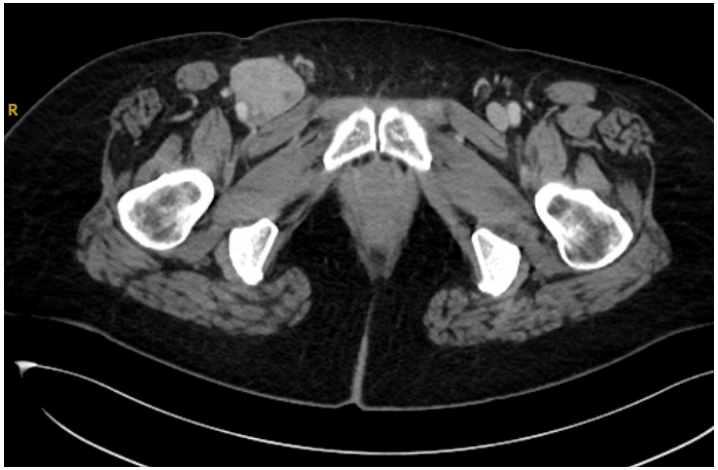
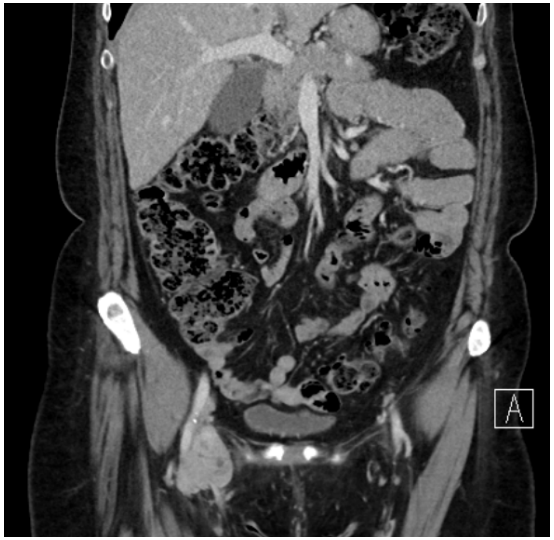
Spermatic cord sarcoma

- Review of case at Sarcoma MCC
- Imaging consistent with residual well-differentiated liposarcoma, no de-differentiated sarcoma noted
- Surgical resection planned to include: spermatic cord/abdominal wall, left testicle, retroperitoneal dissection of cephalad extension
- Discussion and consult with radiation oncology: negative margin resection feasible, complex reconstruction (abdominal wall) would be complicated by pre-operative radiation

Inguinal sarcoma

- 67 yo F
- Self-palpated lump in right inguinal region, firm and solid
- Not consistent with hernia
- Imaging: US – possible enlarged lymph node
  - Core biopsy: low grade leiomyosarcoma





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Inguinal sarcoma

- CT / MRI imaging:
  - Occlusion of common femoral vein, sarcoma likely originating from wall
  - No lymphedema or leg symptoms
  - Abutment of common femoral artery, 180^{o}
- Sarcoma MCC discussion:
  - Given abutment of femoral artery and interest in preservation without resection, preoperative radiation therapy recommended

Principles of management

- For uncharacteristic physical exam findings, cross-sectional imaging is critical
  - Inguinal imaging should include pelvis
  - CT as initial imaging is sufficient
- MCC discussion prior to final treatment planning is critical
  - Risks/benefits of radiation

Question & Answer