RETROPERITONEAL SARCOMA
Local Control

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DISCLOSURE SLIDE

None
Conclusion

The current standard of care for retroperitoneal sarcoma:

• SURGERY

extended en bloc complete resection of the tumour and surrounding viscera (which may be adherent to but not necessarily invaded by the tumour)

  – in an attempt to include a surrounding cuff of normal tissue around the tumour to minimise the marginality of the resection –

  – safely performed in high-volume centres.
Retroperitoneal sarcomas is not a single disease!
  • a group of heterogeneous neoplasms
  • different places/organs/structures.

Biologic behaviour, response to treatment and clinical outcomes vary by histological subtype/grade.

The management plan, including extent of resection and neoadjuvant strategies, should be formulated accordingly.
Anatomy
Anatomy

A = Aorta
IVC = Inferior vena cava
PP = Parietal peritoneum
RMP = Retromesenteric plane
RRS = Retrorenal space
APS = Anterior pararenal space
PRS = Perirenal space
PPS = Posterior pararenal space
TF = Transversalis fascia
PRF = Retrorenal fascia
ARF = Anterior renal fascia
## Retroperitoneal margins

<table>
<thead>
<tr>
<th>RETROPERITONEAL COMPARTMENT MARGIN</th>
<th>DESCRIPTION</th>
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</thead>
<tbody>
<tr>
<td><strong>ANTERIOR</strong></td>
<td>posterior parietal peritoneum, ipsilateral colon and mesocolon, tail of pancreas + spleen or head of pancreas, duodenum</td>
</tr>
<tr>
<td><strong>POSTERIOR</strong></td>
<td>psoas, iliacus, quadratus lumborum, transverses abdominis, muscles, diaphragm</td>
</tr>
<tr>
<td><strong>MEDIAL</strong></td>
<td>IVC (right-sided tumours), duodenum, head of pancreas. Aorta + branches (left-sided tumours), spine, paraspinous muscles</td>
</tr>
<tr>
<td><strong>LATERAL</strong></td>
<td>lateral abdominal musculature, kidney and colon (depending on tumour location)</td>
</tr>
<tr>
<td><strong>SUPERIOR</strong></td>
<td>transverse colon/mesocolon, tail of pancreas or spleen, diaphragm, the right lobe of the liver, the duodenum</td>
</tr>
<tr>
<td><strong>INFERIOR</strong></td>
<td>iliopsoas muscle, femoral nerve, iliac vessels or pelvic sidewall, bladder/rectum</td>
</tr>
</tbody>
</table>
Anatomy
Anatomy
Anatomy
Psoas/posterior abdominal wall sarcoma
Subtypes

Primary retroperitoneal sarcoma

- Liposarcomas (well-diff, dedifferentiated)
- Leiomyosarcoma
- Solitary fibrous tumours
- Malignant PNST
- Synovial sarcoma, Ewing’s sarcoma, Pleomorphic sarcoma, Fibrosarcoma, Spindle cell sarcoma NOS, Pleomorphic rhabdomyosarcoma, Undifferentiated round cell sarcoma, Epithelioid smooth muscle tumour, Desmoplastic small round cell tumour
Variability in Patterns of Recurrence After Resection of Primary Retroperitoneal Sarcoma (RPS)

A Report on 1007 Patients From the Multi-institutional Collaborative RPS Working Group

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Overall survival curve in the whole series

5-year OS = 67%
10-year OS = 46%
Overall survival curve in the whole series

WD LPS 8-yr OS ~ 80%

LeiMS 8-yr OS ~ 40%

DD LPS Gr3 8-yr OS ~ 30%
Cumulative incidence of local relapse

5-year CCI = 26%

10-year CCI = 35%
Cumulative incidence of local relapse

- DD LPS Gr2 ~ 50%
- DD LPS Gr3 ~ 40%
- WD LPS ~ 35%
- LeiMS ~ 10%

Cumulative incidence of distant metastasis

LeiMS $\sim 50\%$

DD LPS Gr3 $\sim 30\%$

DD LPS Gr2 $\sim 6\%$

WD LPS $=0\%$

Surgical extent according to biologic behaviour

Well-differentiated Liposarcoma
Surgical extent according to biologic behaviour

IVC Leiomyosarcoma
Surgical extent according to biologic behaviour

IVC leiomyosarcoma
Initial approach to RP mass

1. Diagnosis?
2. Should it be taken out?
3. Should I be taking it out?
4. What will it involve taking it out?
5. Is it possible/beneficial/risks to resect?
# Role of core needle biopsy

## Retroperitoneal mass

| Biopsy not essential | • imaging reviewed in Sarcoma MDT and is diagnostic and absolutely certain of a resectable retroperitoneal liposarcoma  
|                       | • and no neoadjuvant treatment planned |
| Biopsy                | • lipomatous lesion when radiological uncertainty  
|                       | • include non-lipomatous soft tissue sarcomas, GIST, benign soft tissue tumours, suspected lymphomas, germ-cell tumours or primary/metastatic carcinoma. |
Role of core needle biopsy

Advantage
1. Accurate

Differentiate malignant and benign STT (n = 530)
– Sensitivity = 96.3%
– Specificity = 99.4%
– Positive predictive value = 99.5%
– Negative predictive value = 95.1%

Accuracy = 98%

Strauss DC, Qureshi YA, Hayes AJ, et al JSO 2010
Role of core needle biopsy

Advantage
2. To identify the non-STS malignancies (lymphoma, metastatic carcinoma, germ cell tumours)
Role of core needle biopsy

**Advantage**

3. To differentiate benign from malignant

Schwannoma

Leiomyosarcoma
Role of core needle biopsy

Advantage

4. To subtype and grade soft tissue sarcomas

Preoperative biopsy allows to plan the optimal surgical + neoadjuvant treatment for each patient and to tailor treatment according to the individual sarcoma subtype.
Prognostic factors predicting outcome

EORTC study 62092-22092
STRASS - A phase III randomized study of preoperative radiotherapy plus surgery versus surgery alone for patients with Retroperitoneal sarcoma (RPS)
Role of core needle biopsy

Advantage
To identify chemo-sensitive sarcoma subtypes

Ewing’s sarcoma

- Core needle biopsy:
- Extraskeletal Ewing’s sarcoma

- Preoperative chemotherapy (vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide)

- Resection specimen:
  • marked treatment-related changes and <10% viable tumour.
Chemo-sensitive subtypes

Aug 2012

Nov 2012 – post chemotherapy (4 cycles ifosfamide and doxorubicin)
Chemo-sensitive subtypes

Leiomyosarcoma
04/2013 – 11/2013
6 cycles gemcitabine
Role of core needle biopsy - lipomatous lesions
Role of core needle biopsy

Safe:

- All retroperitoneal sarcomas
  - Local Recurrence (%)
  - Months post-resection of RPS
  - \( p = 0.487 \)
  - CNB
  - No CNB

- All retroperitoneal sarcomas
  - Overall Survival (%)
  - Months post-resection of RPS
  - \( p = 0.275 \)
  - CNB
  - No CNB

- Retroperitoneal liposarcoma only
  - Local Recurrence (%)
  - Months post-resection of RPS
  - \( p = 0.332 \)
  - CNB
  - No CNB

- Retroperitoneal liposarcoma only
  - Overall Survival (%)
  - Months post-resection of RPS
  - \( p = 0.024 \)
  - CNB
  - No CNB
Resectability

Is it beneficial/oncologically sensible to resect the tumour?

Patient benefit v treatment morbidity

“*In the field of surgical oncology: tumour biology* is king, *patient selection* is queen, and *technical manoeuvres* are the prince and princess.*

Occasionally the prince and princess try to overthrow the powerful forces of the King and Queen, sometimes with temporary apparent victories, usually to no long term avail.”
Resectability

Is it beneficial/oncologically sensible to resect the tumour?

- Systemic metastasis
- Incomplete resection of high-grade tumour
- Tumour rupture
- Aggressive tumour biology
Resectability

– Encasement of vessels
Resectability

Spinal involvement
The Royal Marsden

Resectability

Multifocality
Resectability

Multifocality

Local recurrence

- Multifocal
  - 20% 5yr LR free

Local control

5yr LR free survival = 59.3%
Surgical principle of resection

Liberal visceral en bloc resection in an attempt to include an envelope of normal tissue around the tumour to minimise the marginality of the resection - in the hope of improving outcome.
Compartmental resection

The objective is to achieve an envelope of normal tissue along some tumour surfaces by removing adjacent, easily disposable organs while performing what is essentially a marginal excision along critical structures.
Compartmental resection
Compartmental resection
Anterior colon/mesentery
Anterior colon/mesentery
Superior margin - spleen
Superio-medial margin - pancreas
Lateral margin - peritoneal
Lateral margin - peritoneal
Medial margin - aorta
Medial margin - aorta
Posterior margin - psoas muscle

Psoas muscle or psoas aponeurosis?
Posterior margin - psoas muscle
Posterior margin - psoas muscle

High-grade component
Posterior margin - psoas muscle
Inferior vena cava, aorta
Inferior vena cava, aorta
Direct infiltration

Organs or structures should be resected only if directly infiltrated: duodenum, head of pancreas, liver, stomach, diaphragm, major abdominal vessels, bladder/rectum and nerves, bone.
Direct infiltration

Stomach
Direct infiltration

R kidney, liver, diaphragm, abdominal wall and chest wall
Pleomorphic sarcoma

Pelvic exenteration
Extra-abdominal extension

Surgical extent
Extra-abdominal extension
Conclusion

– The retroperitoneum can host a wide spectrum of pathologies, including a variety of rare benign tumours and malignant neoplasms which can be primary or metastatic lesions.

– Other diagnoses must be considered when the radiological appearance is not typical of a retroperitoneal liposarcoma.
Conclusion

– The optimal management of retroperitoneal sarcoma (RPS) is facilitated by pre-treatment diagnosis and staging.

– Image-guided percutaneous core needle biopsy of RPS is strongly recommended.

– A preoperative core needle biopsy is safe and does not affect oncological outcome.

– An open or laparoscopic surgical incision biopsy must be strongly discouraged.
Conclusion

- The best chance of a curative resection is at the time of primary presentation.

- The individual management plan should be determined taking into account both imaging and pathologic findings.
Conclusion

– Biologic behaviour, response to treatment, and clinical outcomes vary by histological subtype of RPS. The management plan, including extent of resection and neoadjuvant strategies, should be formulated accordingly.

– The current standard of care for retroperitoneal sarcoma is extended en bloc complete resection of the tumour and surrounding viscera performed in high-volume centres.
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