Case Presentation and Mini-review

A Case with Infiltrating Perineal Mass

Lin, Kuei-yu (Paul),
National Taiwan University, School of Medicine Year VI
Gillian Lieberman, M.D.
Beth Israel Deaconess Medical Center

History

• 43 year old woman, asymptomatic
• 1998: Bartholin’s gland enlargement, grape size
• Aug 2000: obvious growth noted → surgical excision + pathology exam: aggressive angiomyxoma
• Sep 2000: palpable spongy vulvar mass → re-excision → MR F/U
• Dec 2001: post-operative scarring
• Jun 2004: tumor recurrence, observe
• Mar 2005: MR F/U

MR-T1WI

Well-defined mass, iso-intense compared with muscle
From BIDMC PACS

MR T2WI

Heterogeneously hyperintense mass, swirled appearance in the lesion
From BIDMC PACS

MR T2

The lesion extending into the pelvis. It is 3.4 x 2 x 8.7 cm
From BIDMC PACS

MR T2 FS

Signal of fat has been suppressed.
There is no fatty tissue within the mass because the high signal is not suppressed.
From BIDMC PACS
Fat suppression. Vessels are enhanced. This is a hypervascular lesion.

Local infiltration of levator ani muscle.

An evidence of tumor infiltration. Levator ani muscle is abnormally enhanced.

Aggressive angiomyxoma (AAM)
- Rare benign soft-tissue tumor with myxoid and vascular components
- Involving mainly the pelvis—particularly the perineum—in women of childbearing age
- Propensity to grow to large sizes
- Locally infiltrative, No metastases
- Propensity to recur locally

CT
- Well-defined mass within the pelvis or perineum that is isodense or hypointense relative to muscle.
- Moderately enhanced

AAM Image findings

- T1-weighted MR images:
  - Isointense when compared with muscle
  - Moderately enhanced by Gd
  - Lack of high fat content
- T2-weighted MR images
  - High signal intensity
  - Distinctive internal architecture composed of a swirled or layered appearance

AAM Pathology

- Spindle cells separated by a myxoid stroma
- Variably sized vessels.
- Mitotic activity has been shown to be rare in most cases
**AAM Pathology**

**Differential diagnosis**

- Myxoid Liposarcoma
- Myxoma
- Infiltrating angiolipoma
- Myxoid lipoma

**Myxoid liposarcoma**

- Malignant tumor
- Most commonly occurs in lower extremities within the intramuscular fat
- Post Gd: Homogenous enhancement
- Some has high fat content

**Myxoid liposarcoma**

Histology of Myxoid liposarcoma, showing plenty of adipose tissue


**Myxoid lipoma**

- Morphologic variant of lipoma
- Benign counterpart to liposarcoma
- Predominantly myxoid in nature
- Mature adipose tissue

Myxoid lipoma

Histology of Myxoid lipoma, showing plenty of adipose tissue

From http://erl.pathology.iupui.edu/C603/IMAGES/5AF10.JPG

Infiltrating angiolipoma

• Benign soft-tissue mass
• Regional infiltration; No metastasis
• Hypervascular lesion similar to AAM
• Usually found in the thigh
• Composed of mature lipocytes

From Mark J. Kransdorf, MD, et al. Imaging of Fatty Tumors, Radiology 2002;224:99-104

Myxoma

• Benign mesenchymal neoplasm
• Lacks the vascular component
• Mainly intramuscular

MR T1 post Gd, showing not enhanced centrally due to lack of vascular component


Differential diagnosis

<table>
<thead>
<tr>
<th>Common location</th>
<th>High Fat Content</th>
<th>Hypervascularity</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aggressive Angiomyxoma</td>
<td>X</td>
<td>O</td>
<td>Pelvis, Perineum</td>
</tr>
<tr>
<td>Myxoid liposarcoma</td>
<td>O</td>
<td>O</td>
<td>Lower ext., Intramuscular</td>
</tr>
<tr>
<td>Myxoid lipoma</td>
<td>O</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Infiltrating angiolipoma</td>
<td>O</td>
<td>O</td>
<td>Intramuscular</td>
</tr>
<tr>
<td>Myxoma</td>
<td>X</td>
<td>X</td>
<td>Intramuscular</td>
</tr>
</tbody>
</table>
AAM Treatment

• Primary treatment: Surgical excision
• Recurrence rates of 36%–72%
• Radiation therapy or chemotherapy is not helpful

Conclusion

• MR imaging shows the angiomatous and myxomatous natures of AAM well
• MR images is valuable in diagnosis of this tumor, evaluation of tumor extent, and planning of surgery
• Histology and radiology
• Make use of different MR sequences

Reference

• Antonio Aversa do Souto; Flavio S. DominguesI; Leila ChimelliII; Armando M. LemosI Souto Salvador, Bahia - Brazil, September 20 to 23, 2005

Acknowledgements

• Gillian Lieberman, MD
• Pamela Lepkowski
• Larry Barbaras
• Ivan Pedrosa, MD
• Jesse Wei, MD
• Avneesh Gupta, MD

Thank you for your attention!!