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
Well-defined mass, iso-intense compared with muscle.
Heterogeneously hyperintense mass, swirled appearance in the lesion

From BIDMC PACS
The lesion extending into the pelvis. It is 3.4 x 2 x 8.7 cm.
MR T2 FS

Signal of fat has been suppressed.

There is no obvious fatty tissue within the mass because the high signal is not suppressed.

From BIDMC PACS
MR T1 post Gd

Fat suppression

Vessels are enhanced.

This is a hypervascular lesion.

From BIDMC PACS
MR T2 Local Invasion

Local infiltration of levator ani muscle

From BIDMC PACS
An evidence of tumor infiltration: levator ani muscle is abnormally enhanced.

MR T1 post Gd

From BIDMC PACS
Discussion

• Aggressive angiomyxoma (AAM)
• Differential diagnosis
Aggressive angiomyxoma

- 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

From
http://www.geocities.com/sampyroy2000/AgA1.jpg
AAM Image findings

- **CT**
  - Well-defined mass within the pelvis or perineum that is iso- or hypoattenuated 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
Well-defined mass, Iso-intense compared with muscle

From BIDMC PACS
Heterogeneously hyperintense mass, swirled appearance in the lesion.
MR T1 post Gd

Fat suppression

Vessels are enhanced.

This is a hypervascular lesion.

From BIDMC PACS
AAM Pathology

• Spindle cells separated by a myxoid stroma
• Variably sized vessels.
• Mitotic activity has been shown to be rare in most cases
MR T2WI showing hyperintensity caused by fluid-like myxoid stroma

From Sarah T. Stewart, MD et al., *Radiology* 2004;233:697-700 (Right)

AAM Pathology

MR T1 post Gd, showing heterogeneously enhanced by hypervascularity

From Sarah T. Stewart, MD et al., *Radiology* 2004;233:697-700 (Right)

& http://www.telepathology.mds.qmw.ac.uk/ (left)
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

From http://www.wheelessonline.com/image3/i1/lipm1.jpg
Myxoid liposarcoma

Hyperintense fatty tissue at T1

Low signal intensity at FS

From Mark J. Kransdorf, MD, et al., Radiology 2002;224:99-104
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](http://erl.pathology.iupui.edu/C603/IMAGES/5AF10.JPG)
Myxoid lipoma

Infiltrating angiolipoma

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

Histology of Infiltrating angiolipoma, showing adipose tissue

From http://www.sapporo-dermpath.com/angilipo0204622/angiolipo.html
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></th>
<th>High Fat Content</th>
<th>Hypervascularity</th>
<th>Common Location</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Aggressive Angiomyxoma</strong></td>
<td>X</td>
<td>O</td>
<td>Pelvis, Perineum</td>
<td>Whorled-appearance, Large and infiltrating</td>
</tr>
<tr>
<td><strong>Myxoid liposarcoma</strong></td>
<td>O</td>
<td>O</td>
<td>Lower ext., Intramuscular</td>
<td>Var-defined</td>
</tr>
<tr>
<td><strong>Myxoid lipoma</strong></td>
<td>O</td>
<td>X</td>
<td>Intramuscular</td>
<td>Well-defined</td>
</tr>
<tr>
<td><strong>Infiltrating angiolipoma</strong></td>
<td>O</td>
<td>O</td>
<td>Intramuscular</td>
<td>Well-defined</td>
</tr>
<tr>
<td><strong>Myxoma</strong></td>
<td>X</td>
<td>X</td>
<td>Intramuscular</td>
<td>Well-defined</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

- Sarah T. Stewart, MD and Shirley M. McCarthy, MD, PhD, Case 77: Aggressive Angiomyxoma, Radiology 2004;233:697-700.
- Antonio Aversa do SoutoI; Flavio S. DominguesI; Leila ChimellillI; Armando M. LemosI 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 !!