33 year old woman with enlarging finger mass

Los Angeles Society of Pathologists
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November 13, 2007
Clinical History

- 33 y/o pregnant female with a slow-growing mass on the left middle finger.
  - Seen in clinic on 3/07 while five months pregnant.
  - States mass has been enlarging rapidly since pregnancy.

- Rapid growth over five-month period (3/07 → 8/07)

- PMH: None
- PSH: C-section x 2
- All: NKDA
- Meds: None
March 2007

August 2007
Physical exam:

- No Lymphadenopathy of Axilla or Epicondylar regions
- **Mass on Distal Phalanx of L MF, fungating, involves nail**
  - Circumference: 12 cm
  - Length: 4.5 cm
- Mild Fluctuance on Ulnar aspect
- +TTP, Erythematous base
- Sensation intact RDN/UDN
Pertinent Labs:

WBC 7.5, Hgb 14.4, Hct 43.8, Plt 267
BMP NL
Alk Phos: 90, AST 36, ALT 56, T. Protein 8.6,
Albumin 4.9
UA: + Protein 30 mg/dL
PT/PTT: 12.2(0.92)/31.6
β-HCG: Negative
CRP 5.8
Cx Negative
PPD: 8x10 mm
Gross Findings (Left Middle Finger)

- 15.3 x 2.5 x 0.2 amputated finger
- Large necrotic ulcerating hemorrhagic mass (7.5 x 5.5 x 3.2 cm)
  - Hemorrhagic friable tan yellow tissue
- Proximal resection margin free of tumor
Differential???
Differential Diagnosis

- Clear cell sarcoma
- Epithelioid sarcoma
- Synovial sarcoma
- PNET/Ewing sarcoma
- Rhabdomyosarcoma
- Hemangiopericytoma
- Leiomyosarcoma
- Glomangiosarcoma
- Melanoma
- Adnexal Tumors
- Primary Carcinoma
- Primary Carcinoma
- Adnexal Tumors
- Epithelioid Sarcoma

- Clear Cell Sarcoma
- Melanoma
- Rhabdomyosarcoma
- Leiomyosarcoma
- Hemangiopericytoma

MSA
MyoD1
SMA
Desmin
- PNET/Ewing Sarcoma
- Synovial Sarcoma
Immunohistochemistry

**Positive (+):**
- Vimentin
- Muscle-specific Actin
- Smooth muscle Actin
- CD34
- bcl-2

**Negative (-):**
- Cytokeratin
- EMA
- CK5/6
- Desmin
- MyoD1
- S-100
- Mel-MART
- Chromogranin
- CD31
- vWF
- FLI-1
- CD45
- CD99
Differential Diagnosis

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- Clear cell sarcoma
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- Glomangiosarcoma
- Hemangiopericytoma
Final Diagnosis

Malignant Glomus Tumor
(Glomangiosarcoma)
History
Glomangiosarcoma

- Lumley & Stansfeld (1972) → 24 yr old pt
  - Malignant glomus tumor(?)

Aiba et al. (1988): Glomangiosarcoma arising in benign glomus tumor

Aiba, M et al. Glomangiosarcoma in a glomus tumor. An immunohistochemical and ultrastructural study. 1988 Cancer; 61: 1467-1471. (Figures 1,2)


(Figures 1, 2, 3, 6)
Microscopic Findings

**H&E:**
- Glomus Cells: Round ± Spindle cells (GABG), Mitotic figures, Nuclear atypia, Hypercellularity
- Blood vessels
- + Stroma- Hyalinized, myxoid

**IHC:**
- Similar staining pattern to benign:
  - Positive: MSA & SMA (Weaker), Vimentin (More Intense)
  - Negative: Keratin, Neural, Endocrine, Vascular, Desmin
  - Basement membrane: Collagen IV, Reticulin, or PAS-D
- Malignant: also bcl2, p53, inc. Ki67 (MIB-1) index*

**EM:**
- Fewer microfilaments, pinocytotic vesicles, Basal Lamina (incomplete)
**Location**

**Glomus Tumors:**
- Hand (>50%)
  - Distal Phalanx, Palm, Wrist
- Soft Tissue & Skin (Other sites)

**Other:**
- Lung/Mediastinum:
  - Inc. Trachea
- GI Tract:
  - Stomach, Small & Large Bowel
- GU Tract:
  - Kidney, Vagina, Uterus
- Sellar region
- Bone, Nerve, Orbit
- Head & Neck:
  - Oral & Nasal Mucosa
- Coccyx

**Glomangiosarcoma:**
- Lower Extremity (75%)
- Soft Tissue & Skin (Other sites)

**Other:**
- Lungs/Mediastinum:
  - Inc. Trachea
- GI Tract
- GU Tract:
  - Bladder
- Sellar region
- Bone
  - Hand: 9 cases
Clinical Features

<table>
<thead>
<tr>
<th>Frequency (%)</th>
<th>SXS</th>
<th>Duration</th>
<th>Location</th>
<th>Size Cm</th>
<th>Gender Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign (Hand)</td>
<td>Pain</td>
<td>7 years (40 yr)</td>
<td>Dermal/subQ</td>
<td>&lt;1 (3-4)</td>
<td>3:1 F:M 30-50 yr</td>
</tr>
<tr>
<td>Benign (Other)</td>
<td>Pain/Mass effect/Other</td>
<td>&gt;7 years (20 yr)</td>
<td>Dermal/subQ/Internal organs</td>
<td>Skin&lt;1-3 Deep &gt;1</td>
<td>M&gt;F 40-70 yr (27-82 yr)</td>
</tr>
<tr>
<td>Multiple (Benign)</td>
<td>± Pain</td>
<td>Variable</td>
<td>Dermal/subQ</td>
<td>&lt;1-3 (9.0)</td>
<td>M&gt;F Children</td>
</tr>
<tr>
<td>Malignant (Other)</td>
<td>Pain/Mass effect/Other</td>
<td>Monthly-Years (9 yr)</td>
<td>Dermal/subQ/Internal organs</td>
<td>Skin&lt;1-3 Deep &gt;1</td>
<td>M&gt;F 40-70 yr (20-84 yr)</td>
</tr>
<tr>
<td>Malignant (Hand)</td>
<td>Pain</td>
<td>Monthly-Years (18 yr)</td>
<td>Dermal/subQ</td>
<td>&lt;1-3 (7.5)</td>
<td>8:1 F:M 30-77 yr</td>
</tr>
</tbody>
</table>
Gould et al. (1990) → 3-way classification system

- Locally infiltrating “benign” glomus tumor (LIGT)
  - Lumley & Stansfeld’s case

- Glomangiosarcoma arising in benign glomus tumor (GABG)
  - Enzinger & Weiss, Aiba et al’s case

- Glomangiosarcoma arising de novo (GADN)
  - Braithwaite et al’s case
  - Our Case (?)
<table>
<thead>
<tr>
<th>Hand (n=9)</th>
<th>SXS</th>
<th>Duration</th>
<th>Location</th>
<th>Size</th>
<th>Sex</th>
<th>Age</th>
<th>Treatment, Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>+ Benign component (GABG)</td>
<td>Pure Sarcoma-like (GADN)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wetherington et al. (1997)</td>
<td>Pain</td>
<td>1 Year</td>
<td>Thumb</td>
<td>0.5 cm</td>
<td>F</td>
<td>30 yr</td>
<td>Excision/Reexcision Alive, no recurrence</td>
</tr>
<tr>
<td>Fuentes et al. (2005)</td>
<td>Pain</td>
<td>1 Year</td>
<td>Thenar</td>
<td>0.5 cm</td>
<td>F</td>
<td>36 yr</td>
<td>Excision/Reexcision Alive, no recurrence</td>
</tr>
<tr>
<td>Gould et al. (1990)</td>
<td>Pain</td>
<td>1 Year</td>
<td>Bet. 2\textsuperscript{nd} &amp; 3\textsuperscript{rd} metacarpals</td>
<td>3 cm</td>
<td>F</td>
<td>17 yr</td>
<td>Excision Alive, no recurrence</td>
</tr>
<tr>
<td>Khoury et al. (2005)</td>
<td>None</td>
<td>5 Months</td>
<td>Base of Thumb</td>
<td>3 cm</td>
<td>F</td>
<td>48 yr</td>
<td>Excision/Reexcision + Chemotherapy Mets to Lung (Alive)</td>
</tr>
<tr>
<td>Park et al. (2003)</td>
<td>Pain</td>
<td>5 Months</td>
<td>Palm</td>
<td>1.5 cm</td>
<td>F</td>
<td>74 yr</td>
<td>Excision Alive, no recurrence</td>
</tr>
<tr>
<td>?Our Case (2007)</td>
<td>Pain</td>
<td>18 years</td>
<td>Third finger (Distal tip)</td>
<td>7.5 cm</td>
<td>F</td>
<td>33 yr</td>
<td>Ray Amputation Alive</td>
</tr>
<tr>
<td>Folpe et al. (2001)</td>
<td>??</td>
<td>??</td>
<td>Finger (2) Wrist (1)</td>
<td>0.5-3 cm</td>
<td>2 F, 1 M</td>
<td>38-77 yr</td>
<td>Excision Alive, no recurrence</td>
</tr>
</tbody>
</table>
Khoury et al. Malignant Glomus Tumor: A case report and review of literature, focusing on its clinicopathologic features and immunohistochemical profile. 2005 Am J Dermatopathol; 27(5): 428-431. (Figure 4)
DE NOVO GLOMANGIOSARCOMA:
- Total 14 cases, 3 recurrences, **5 metastases, 5 deaths**
  - Kreutz et al. M 33 Thigh (10 cm) Met. to Maxilla (Alive)
  - Braithwaite et al. M 63 Nose (2 cm) Distant mets (Died)
  - Watanabe et al. F 47 Left hip (1 cm) Distant mets (Died)
  - Gaertner et al. M 69 Lung (9.5 cm) Distant mets (Died)
  - Shim et al. F 57 Bladder (6.5 cm) Distant mets (Died)
  - Choi et al. F 78 Mediastinum (4.5 cm) Local invasion (Died)
  - Gould et al. F 72 Cheek (5 cm) Local recurrence (Alive)
  - Gould et al. F 26 Back/Axilla (15 cm) (Alive)
  - Hiruta et al. M 44 Right thigh (7 cm) (Alive)
  - Matsumoto et al. F 16 Brachial muscle (5 cm) (Alive)

GLOMANGIOSARCOMA ARISING IN BENIGN TUMOR:
- Total 12 cases, 4 recurrences, **1 metastases**
  - Yu et al. M 47 Trachea (Size?) +Distant mets (Status?)

LOCALLY INFILTRATIVE GLOMUS TUMOR:
- Total 5 cases, 2 recurrence, no metastases or deaths
Atypical Glomus Tumors

Folpe et al. (2001): New Criteria (n=52)

- **Malignant:**
  - Deep seated + Large size (>2 cm) or Atypical Mitoses or High nuclear grade + >5 mitoses/50 hpf

- **Uncertain malignant potential:**
  - Superficial + >5 mitoses/50 hpf or Deep seated only or Large size only

- **Symplastic:**
  - High nuclear grade only
  - No other malignant features

- **Glomangiomatosis**
Treatment & Prognosis

- **Surgical:**
  - Complete excision
- **Adjuvant:**
  - Chemotherapy and/or Radiotherapy
- **Long-term follow-up:**
  - LIGT, GABG: Local recurrence*
  - GADN & Folpe: Local recurrence, Metastasis, Death

*1 metastasis reported in the GABG group
Summary-Glomangiosarcoma

- Larger, Deep-seated
- Older Age, Males
  - Hands: Still Females
- Indolent Course but....
  - Local Recurrence & Metastasis
  - Classification questionable
- Difficult to diagnose
  - Round cell tumors
  - Spindle cell tumors
- Clinical features non-specific
Follow-Up

- Pt doing well.
- No recurrence or metastatic disease.
- No adjuvant therapy.
Acknowledgements

- Dr. Paul Dinh, Orthopedic Hand Surgery
- USC-LA County Hospital
Glomangiosarcoma 
Hand (n=9)

**Benign component (GABG):**
- Wetherington et al. (1997) ➔ Thumb
- Fuentes et al. (2005): Hypothenar eminence
- Gould et al. (1990) ➔ Between 2\textsuperscript{nd} & 3\textsuperscript{rd} Metacarpals

**Pure Sarcoma-like (GADN):**
- Khoury et al. (2005) ➔ Base of thumb
  - Metastatic glomangiosarcoma
- Park et al. (2003) ➔ Palm

**Other:** Folpe et al. (2001) ➔ 2 Finger, 1 Wrist