Desmoid Tumors and Pregnancy

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History

• Johannes Muller coined the term “desmoid” in 1838

• In 1832, John MacFarlane reported an “organized sarcomatous tumor between the layers of the abdominal muscles” in women who had children

Muller (1838), *Ueber den feinern Bau und die Formen der krankhaften Geschwulste*
MacFarlane (1832), *Clinical Reports of the Surgical Practice of the Glasgow Royal Infirmary*
Background

• Desmoid fibromatosis is partly modulated by hormonal signaling

• Role of specific signaling pathways, such as those mediated by estrogen, is unclear
Rationale

• Desmoids are often diagnosed in young women during or after pregnancy

• This raises concern about potential progression during the current pregnancy or recurrence with a subsequent one

• Limited existing data to guide women with desmoids and their treating clinicians regarding future pregnancy
Desmoid-Type Fibromatosis and Pregnancy
A Multi-institutional Analysis of Recurrence and Obstetric Risk

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Objectives

• Evaluate women with desmoids in different clinical scenarios
  • Desmoids arising during or shortly after pregnancy
  • Unresected desmoids who later got pregnant
  • Resected desmoids who later got pregnant

• Identify disease-related risk
  • Risk of progression with watchful waiting
  • Risk of recurrence after resection
  • Risk of recurrence during subsequent pregnancy

• Identify obstetric risk
Study Cohort (N=92)

- Data from 4 high volume centers in 4 countries
  - Istituto Nazionale dei Tumori (Milan, Italy)
  - Institut Gustave Roussy (Villejuif, France)
  - Princess Margaret Hospital and Mount Sinai Hospital (Toronto, Ontario, Canada)
  - Brigham and Women’s Hospital and Dana-Farber Cancer Institute (Boston, Massachusetts, USA)

- Included
  - Women with desmoids and pregnancy before, during, or after desmoid diagnosis

- Excluded
  - FAP-related desmoids
  - Infantile fibromatosis
  - Palmar/plantar fibromatosis
Pregnancy-Associated Desmoids

N=44

**Initial Management**
- Surgery N=15
  - 2 during pregnancy
  - 13 postpartum
- Medical Management N=2
- Watchful Waiting N=27

**During or After Pregnancy**
- Progression N=17
  - Surgery N=6
  - Medical Management N=6
  - Watchful Waiting N=5
  - Watchful Waiting N=10

**No Progression** N=10
Pregnancy-Associated Desmoids

Definitive Management

- Surgery N=21
- Medical Management N=8
- Watchful Waiting N=15

N=44

- After initial management
  - 17/27 (63%) women offered watchful waiting as initial management progressed
  - 6/29 (21%) women offered non-operative therapy as initial management required surgery for definitive management
  - 23/44 (52%) women treated non-operatively

- After definitive management
  - 4/44 (9%) women progressed
  - 2/15 (13%) women offered surgery as initial management progressed
  - 5/44 (11%) women experienced spontaneous regression
Case Example

• Young woman diagnosed with 3 cm abdominal wall desmoid after first pregnancy
  • Treatment – watchful waiting, with no growth

• During second pregnancy, tumor grew
  • Treatment – watchful waiting postpartum, with no further growth

• During third pregnancy, tumor grew
  • Treatment – surgery postpartum

• Desmoid only grew during pregnancy
Pregnancy With Unresected Desmoids

- **N=29**
  - **Surgery N=6**
  - **Medical Management N=2**
  - **Watchful Waiting N=8**
  - **Progression N=16**
  - **No Progression N=13**

**Response to Pregnancy**

- **16/29 (55%)** women with unresected desmoids progressed during pregnancy
- **0/8** women managed with watchful waiting progressed
Pregnancy After Resected Desmoid

N=19

- Progression: N=4
  - Surgery: N=2
  - Medical Management: N=1
  - Watchful Waiting: N=1
- No Progression: N=15

- 4/19 (21%) women with previously resected desmoids progressed during pregnancy
Pregnancy and FAP-Associated Desmoid

- Very limited data

- 47 women with FAP
  - 22 who had never been pregnant
  - 25 who had been pregnant at least once

- Desmoids in women who had been pregnant had a significantly more indolent course
  - Trend towards pregnant women requiring less treatment for desmoid

Church and McGannon (2000), *Dis Colon Rectum*
Obstetric Concerns

• Pregnancy-associated desmoids often arise in abdominal wall muscle (67% in our series)
  • Trauma? Stretching?

• Presence of desmoid did not lead to fetal loss
  • Mesenteric desmoid could make the pregnancy high risk

• Impact on course of pregnancy
  • 1 woman with a vaginal desmoid had to have a C-section
Subsequent Pregnancies

N=15

- Surgery N=3
- Medical Management N=1
- Progression N=4
- No Progression N=11

- 4/15 (27%) women who had a subsequent pregnancy after management of a desmoid progressed during that pregnancy
  - All had been managed after prior pregnancy with watchful waiting
Abdominal Wall Mesh

- Standard synthetic surgical mesh has very little elasticity
  - Great for providing a strong abdominal wall closure
  - NOT so great when the abdomen is stretching during pregnancy

- Biologic mesh, an alternative if considering subsequent pregnancy, has more elasticity
  - Laxity in abdominal wall can mimic a hernia
  - May stretch more during pregnancy
  - Derived from pig, cow, or human cadaver, which may conflict with religious beliefs

- Obstetrician should be notified about the type abdominal wall reconstruction
  - May wish to have a surgeon available if considering a C-section, depending on the level of the mesh
Conclusions

• Having a desmoid does NOT preclude pregnancy
  • Women should NOT be counseled to avoid getting pregnant
  • Women should NOT be advised to have an abortion if pregnant

• Desmoids arising during pregnancy or already present before pregnancy may grow during the course of the pregnancy
  • ~50% can successfully avoid surgery
  • 13% undergoing surgery recur

• Desmoids resected prior to pregnancy recur infrequently during pregnancy (21%)
Conclusions

• Ultimately,
  • 32% required surgery
  • 54% managed with watchful waiting
  • 14% spontaneous regression

• Women can successfully carry subsequent pregnancies after initial desmoid
  • Progression is usually only in women managed with watchful waiting

• The presence of a desmoid does not increase obstetric risk
  • Synthetic mesh used for reconstruction after resection of an abdominal wall desmoid is not as flexible as native tissue
## Summary for Counseling

### TABLE 4. Available Data for Counseling in Women Affected by Sporadic DF

<table>
<thead>
<tr>
<th>Description</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>New diagnosis of DF during or shortly after pregnancy</td>
<td></td>
</tr>
<tr>
<td>Risk of relapse after complete resection</td>
<td>13%</td>
</tr>
<tr>
<td>Risk of progression with watchful waiting</td>
<td>63%</td>
</tr>
<tr>
<td>Spontaneous regression</td>
<td>11%</td>
</tr>
<tr>
<td>Risk of failure after any first active treatment (initial or delayed until the time of progression)</td>
<td>10%</td>
</tr>
<tr>
<td>Overall managed without resection</td>
<td>52%</td>
</tr>
<tr>
<td>Pregnancy after previous diagnosis of DF</td>
<td></td>
</tr>
<tr>
<td>Risk of DF recurrence/progression</td>
<td>42%</td>
</tr>
<tr>
<td>DF recurrence/progression safely managed with either active treatment or watchful waiting</td>
<td>94%</td>
</tr>
<tr>
<td>Multiple lines of active treatments needed for progression</td>
<td>6%</td>
</tr>
<tr>
<td>Spontaneous regression was described after progression as well</td>
<td>7%</td>
</tr>
<tr>
<td>Obstetric risk</td>
<td></td>
</tr>
<tr>
<td>Obstetric complications related to DF in both mother and fetus</td>
<td>0%</td>
</tr>
<tr>
<td>Intra-abdominal/pelvic DF should be anyway considered at higher risk (few data available)</td>
<td></td>
</tr>
<tr>
<td>Cesarean delivery to be considered in case of macroscopic DF in particular anatomic sites</td>
<td></td>
</tr>
<tr>
<td>Postpartum incisional hernia after previous abdominal wall full-thickness mesh repair is an issue</td>
<td></td>
</tr>
</tbody>
</table>

Fiore et al (2014), *Ann Surg*
Thank you

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