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

Initial Management

Surgery N=15

Medical Management N=2

Watchful Waiting N=27

2 during pregnancy
13 postpartum

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

N=44
Pregnancy-Associated Desmoids

N=44

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

- **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

19 primary
10 recurrent

Response to Pregnancy

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

- **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>Condition</th>
<th>Risk (%)</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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