Desmoid tumour. The risk of recurrent or new disease with subsequent pregnancy: a case report

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Desmoid tumours are rare, benign tumours arising from fibrous tissue in muscle fascia or aponeurosis. They are most common in women of child-bearing age and most often appear during or after pregnancy in this age group. The recommended treatment is wide surgical excision, if possible, but unresectable tumours may be treated with radiotherapy, anticancer drugs, nonsteroidal anti-inflammatory agents or antiestrogenic compounds. The recurrence rate is high and seems to be related to the achievement of resection margins free of tumour. The literature is not specific about how to counsel woment who have had a desmoid tumour and subsequently wish to have a child. Patients should be advised that these tumours may be estrogen sensitive but subsequent pregnancy is *not necessarily* a risk factor for recurrence or development of new disease.

Les tumeurs desmoïdes sont des tumeurs bénignes rares qui émanent de tissus fibreux de l'aponévrose musculaire. Elles sont plus fréquentes chez les femmes en âge de procréer et font le plus souvent leur apparition pendant ou après une grossesse dans cette catégorie d'âge. On recommande comme traitement une excision chirurgicale large si possible, mais les tumeurs irrésécables peuvent être traitées au moyen de la radiothérapie, de médicaments anticancéreux, d'agents anti-inflammatoires non stéroïdiens ou de composés anti-œstrogéniques. Le taux de récidive est élevé et semble lié à la résection de marges non atteintes par la tumeur. Les écrits ne sont pas précis sur la façon de conseiller les femmes qui ont eu une tumeur desmoïde et veulent par la suite avoir un enfant. Il faudrait préciser aux patientes que ces tumeurs peuvent être sensibles aux œstrogènes mais qu'une grossesse subséquente *ne constitue pas nécessairement* un facteur de risque de récidive ou d'apparition d'une nouvelle maladie.

Desmoid tumours are rare, benign tumours occurring most commonly in the anterior wall of the rectus abdominis muscle. They arise from fibrous tissue in the fascia or aponeurosis. These tumours are confounding because histologically they are benign and nonmetastasizing yet they infiltrate extensively and have high recurrence rates.[1–3]

Hormonal sensitivity of desmoid tumours has been established indirectly. Although these tumours occur in men and children, previous pregnancies and hormonal constitution have been cited in the literature as the most likely factors influencing disease onset.[1,4–6] Genetic factors may also play a role.[1]

Given that the hormonal or genetic environment may have an influence on desmoid tumour onset, counselling patients about what they may expect about tumour recurrence or new tumour growth with subsequent pregnancies becomes a case-management issue. We describe 2 patients who presented with desmoid tumours shortly after pregnancies and present the results of follow-up examinations after subsequent pregnancies.

Case reports

Case 1

A 28-year-old woman who had an abdominal mass that had been present for about 12 months was referred by her family physician. The patient first noted the tumour immediately after the delivery of her second child, approximately 3 years after her first preg-

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nancy. She had no history of abdominal surgery or blunt trauma. On physical examination the lump was 4 × 5 cm in dimension, tender, firm and relatively immobile when the abdominal muscles were contracted. Computed tomography showed that the mass was confined to the abdominal wall in the region of the left rectus muscle with no obvious extension into the abdominal cavity. It measured 3.0 to 4.0 cm in diameter and was 2.5 cm deep in its thickest portion.

Biopsy showed a firm nodule of yellow and white tissue. Microscopic examination showed a proliferative lesion comprising well-differentiated fibroblasts and fibrocytic cells. The tumour was seen to be infiltrating the adjacent skeletal muscle. The collagen was abundant between the proliferating cells, but there were no cytologic features of malignancy and there was a virtual absence of mitotic activity.

A desmoid tumour was diagnosed and was widely excised 2 months later. The anterior rectus sheath was incised about 2 cm on either side of the mass and carried out and down, through and including rectus muscle. The tumour extended to but did not invade the peritoneal cavity. As a result the posterior rectus sheath was excised to include a 2-cm margin of healthy tissue. Pathology reports of frozen sections from the abdomen showed no evidence of tumour, and the incision was closed. Given that hormonal factors are suspected in the etiology of desmoid tumours the patient was advised to refrain from taking birth control pills or becoming pregnant for a couple of years.

Seven months postoperatively a dermatofibroma developed in the patient's left breast. It was excised under local anesthesia. Follow-up CT of the abdomen 1 year after the abdominal surgery showed no recurrence of the desmoid tumour. The patient wished to become pregnant and, given the possible hormonal influence on the etiology of the disease, wanted to know whether a subsequent pregnancy would affect her chances of recurrence or the development of new disease. A search of literature did not outline any specific recommendations in this regard. Follow-up examinations and CT showed no evidence of recurrence.

Fifteen months postoperatively became pregnant but miscarried. A subsequent pregnancy, about 24 months postoperatively, resulted in a vaginal delivery without complication. A therapeutic abortion was performed 39 months after the original surgery. There was no evident recurrence or new tumour growth at follow-up 60 months after the desmoid tumour was removed.

Case 2

This 28-year-old woman presented with a lump, measuring 1 to 2 cm, in the left upper rectus muscle just below the costal margin. The patient had noticed the mass approximately 6 months earlier. She had been delivered of a healthy child approximately 12 months before first noticing the mass. There was no history of abdominal surgery or blunt trauma. CT showed a very smooth soft-tissue mass measuring about 1.3 × 1.2 cm. No extension of the lesion into the peritoneum or overlying subcutaneous tissues was apparent.

A biopsy specimen of the tumour showed a fibromatosis infiltrating between muscle bundles. Desmoid tumour was diagnosed and was removed by wide excision 3 weeks later. En bloc full-thickness resection was carried out. Microscopic examination showed tumour margins that were unaffected and the tumour was negative for estrogen and progestin receptors. This patient became pregnant 13 months after tumour excision and was delivered without complication. Follow-up physical examination and abdominal CT show no evidence of recurrent or new disease 46 months postoperatively.

Discussion

Desmoid tumours are generally classified as abdominal, extra-abdominal or mesenteric, with abdominal tumours occurring most frequently in women of child-bearing age.[1] They are quite rare,[1] so the general surgeon or pathologist seldom sees them. Morphologically, desmoid tumours are nonencapsulated, rubbery, greyish-white lesions consisting of collagenous fibroblasts and fibrocytes.[1,6] The absence of marked pleomorphism, cellularity and mitotic activity distinguishes desmoid tumours from low-grade fibrosarcomas.[6,7] Despite nonmetastasizing behaviour, desmoid tumours are aggressive and infiltrating, making them difficult to completely excise. Generally, their growth is slow yet progressive,[2,3,6,8] but it can be quite rapid in fertile females.[1] Desmoid tumours can become very large if left untreated,[6] but they are largely asymptomatic[9,10] unless they begin to encroach on or invade adjacent structures such as muscles, tendons, organs, nerves or blood vessels.[2]

Etiology

The most commonly cited etiologic factors for the onset of desmoid tumours are surgical or blunt trauma, pregnancy or, possibly, genetic factors.[1,4,6] The predominance of tumours in the anterior abdominal wall of patients with previous pregnancy may suggest that trauma sustained during pregnancy and childbirth is an etiologic factor in these cases.[9]

Pregnancy and related hormonal

factors appear to play a more important role in the pathogenesis of the tumour.[1,6,9] Lim and associates[11] reported finding estrogen and antiestrogen binding sites in 33% and 80%, respectively, of desmoid tumours, and a number of studies have shown some success in treating unresectable desmoid tumours with antiestrogenic compounds.[5,12–18]

Treatment

Desmoid tumours should be widely excised unless the surgical procedure would result in unacceptable disfigurement or dysfunction.[19–21] Some large intra-abdominal or extra-abdominal tumours cannot be totally resected, and some studies have shown success with adjuvant radiotherapy in the control or cure of the disease.[20–23]

Anticancer drugs,[24,25] non-steroidal anti-inflammatory agents[26] and antiestrogenic compounds[5,12–19] have been used to control desmoid tumors.

Prognosis

The recurrence rate for excised desmoid tumours is estimated to be 40%,[1,2] and recurrence generally occurs within 1 to 2 years of the initial treatment.[1,16,20,21] Abdominal tumours have the best rate of cure after primary treatment, with a recurrence rate of 25%. [1] This seems to reflect the greater ease with which these tumours can be excised with unaffected margins. If recurrence does occur, then re-excision of residual or recurrent disease is considered appropriate because secondary recurrence rates have been found to be equal to or lower than primary rates.[27]

Because desmoid tumours are most common in women predominantly during and after pregnancy, those who have a desmoid tumour after pregnancy are often anxious to know whether they have a higher risk of a new or recurrent tumour developing with subsequent pregnancies. Unfortunately little has been published to indicate what the risk factors might be. Caldwell[10] described a woman with an abdominal desmoid tumour that was detected 6 weeks after her fourth pregnancy. Because of its size the tumour was not resected and over time it spontaneously regressed. A little over a year later the patient became pregnant yet the tumour continued to regress in size.

In our first patient a desmoid tumour developed immediately after delivery of her second child. She has had 3 subsequent pregnancies (1 carried to term) without recurrence or development of a new tumour for 60 months after initial excision. The second patient had a desmoid tumour excised and became pregnant 13 months later. Again there was no evidence of disease for 46 months after tumour excision. Women should be counselled about the etiologic factors regarding this disease. They should know that there is a 25% chance of recurrence, but it would seem from these reports that subsequent pregnancy does not necessarily mean that the disease will return or a new will develop.

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