

Management of a large abdominal wall desmoid tumor during pregnancy.

Case report



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Desmoid tumors, characterized by aggressive local infiltration of surrounding tissues, are uncommon benign neoplasms with no metastatic potential, that occasionally may attain large size.

We report a case of a 37-year-old woman with an abdominal wall desmoid tumor that appeared and grew rapidly during her pregnancy, diagnosed by trucut core biopsy.

Complete surgical excision of a 20x16cm in size tumor and immediate reconstruction with mesh was performed in the postpartum period. She had no postoperative complications and no recurrence at 2-year follow-up.

Optimal management of large abdominal wall desmoids during pregnancy has to be individualized, with wide surgical excision remaining the treatment of choice.

KEY WORDS: Abdominal wall; Aggressive fibromatosis; Desmoid tumor.

Background

Desmoid tumors account for only 0.03% of all neoplasms. These rare tumors are slow growing and histologically benign, exhibiting fibroblastic proliferation that arises from fascial or musculoaponeurotic structures. Despite their benign histological appearance they are diffusely infiltrative, but without metastatic potential^{1,2}. Treatment of these tumours is whenever possible complete resection with a tumour free margin³, as there is a high risk of local recurrence ranging from 20 to 90%⁴. These tumours can appear anywhere in the body but the most common site of predilection is the anterior abdominal wall, with an incidence of 50%^{5,6}. Desmoid tumors during pregnancy are uncommon and optimal management of this tumor has yet to be defined.

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Case Report

A 37 year-old woman, 16 weeks pregnant, presented with a painless swelling in the right anterior abdominal wall at the level of umbilicus. She had noticed the lump of about 3x2cm size in the beginning of her pregnancy and reported that this gradually doubled in size over the last 5 weeks to 6x4cm. The patient had one previous pregnancy and an elective caesarean section 2 years earlier, through a pfannenstiel incision.

Ultrasonography showed a well defined solid mass arising from the right rectus muscle, while trucut core biopsy of the lesion confirmed features consistent with a desmoid tumour.

The mass grew rapidly during the next weeks with the progress of her pregnancy remaining uneventful. Therefore she underwent a second elective caesarean section at 38 weeks' gestation.

Computed tomography (CT) scan in the post partum period revealed a solid mass 20x 16cm extending from the right hypochondrium superiorly to the pubic bones inferiorly (Fig. 1, 2).

Patient was treated by wide local excision with at least 1cm of surrounding macroscopic normal tissue (Fig. 3).



Fig. 1: View of the anterior abdominal wall.



Fig. 2: CT scan showing a solid mass in the right rectus muscle.

Reconstruction was performed with a large coated polypropylene mesh that was sutured to the excision edge of the anterior abdominal wall musculature.

Histologically, it was a typical abdominal desmoid tumour. The patient's postoperative recovery was uneventful and she remained well at 2-year follow up with no evidence of tumour recurrence or development of incisional hernia.

Discussion

Aggressive fibromatoses (AF) or desmoid tumors are fibroblastic lesions with aggressive, infiltrative and destructive growth, which frequently recur if not widely resected⁷.

Desmoid tumors occur in three main anatomical positions: limb or limb girdle, abdominal wall and retroperitoneally. Limb or limb girdle and intra-abdominal tumours have a high rate of recurrence, whereas this is uncommon with adequately treated abdominal wall disease². The management of extra-abdominal AF is challenging because this disease does not respect the usual surgical rules relating to resection and recurrence⁸.

While most cases of desmoid tumors are sporadic, some are associated with familial adenomatous polyposis (FAP), and these are most often intra-abdominal. There are also cases of familial desmoid tumors, often involving one extremity, in patients without FAP. In both FAP and familial non-FAP tumours, mutations of the adenomatous polyposis gene on the long arm of chromosome 5 have been incriminated⁴. The resultant loss of ability to degrade beta-catenin and elevated beta-catenin levels promotes fibroblastic proliferation⁹. The prevalence of desmoid tumor in FAP is 10-25%¹⁰⁻¹².

The aetiology of desmoids has not been well defined. An antecedent history of trauma to the site of the tumor, often surgical in nature, may be elicited in approximately 25% of cases¹³.

Desmoid tumors are more common in women¹ with a peak incidence between 25 and 35 years of age¹⁴. It has been suggested that the growth of desmoid tumors is stimulated by estrogens, based on a higher incidence of their occurrence in women of childbearing age and their association with pregnancy^{8,15,16}.

Moreover, Clark and Phillips in their review reported an apparent tendency for desmoids to develop, especially in the abdominal wall, during or soon after pregnancy or while taking combined oral contraceptives, as well as regression after menopause and oophorectomy¹⁶.

It has also been shown that oestrogen treatment may induce, both in experimental animals¹⁷⁻²⁰ and in humans²¹, the appearance of desmoids, which regress after suspension of the treatment or with the use of progesterone^{18,19}.

Estrogen binding sites have been identified in 25-75% of desmoids²²⁻²⁴, but the absence of estrogen receptors (ER) in such tumours has also been reported²⁵⁻²⁷.



Fig. 3: Intraoperative view of the desmoid tumor measuring 20cm in its maximum diameter.

Progesterone receptors (PR) have been reported to be present in desmoid tumors by several investigators^{22,25}. Ishizuka et al²⁸ in their study assessed the immunohistochemical expression of oestrogen, progesterone and androgen receptors in desmoids. (ER)a and (ER)b were both detected in 7.4% of desmoid tumors, (PR)-A and (PR)-B were detected in 25.9% and 33.3% respectively, and androgen receptors (AR) in 52.9%.

The evaluation of hormonal receptors in aggressive fibromatosis could be predictive for a clinical response to hormonal agents and may be of prognostic significance in the natural history of these tumours²³.

Diagnosis can be confirmed by Trucut core biopsy with an accuracy of greater than 90%. Open incision biopsy has been reported to have complication rate of 17%³. Whenever possible the tumor should be excised widely, attempting to achieve negative histological margins, but this should not be at the expense of loss of function⁸. After wide excision and mesh reconstruction, recurrence of abdominal wall lesions is unusual². Bertani et al²⁹ suggest for desmoid tumors of the anterior abdominal wall wide surgical excision and immediate plastic reconstruction with mesh, after intraoperative confirmation by frozen sections of disease-free margins of >1cm.

Radiation therapy is used in patients with unresectable tumors or tumours that would require extensive resection including amputation or major chest or abdominal wall resection³⁰. Nuyttens et al³¹ showed that the addition of radiotherapy to surgical resection with positive margins significantly improved local control of disease when compared to surgery alone (75% versus 41%). Therefore radiotherapy should be considered after a non-radical tumor resection and should be given preferably in an adjuvant setting. Also Baumert et al⁴ in their study showed that postoperative radiotherapy significantly improved the progression-free survival (PFS) compared to surgery alone and suggested that it should always be considered after a non-radical tumor resection.

The medical treatment of desmoid tumors is based on three main classes of drugs: hormonal agents, anti-inflammatory agents and cytotoxic agents. Because of their rarity, there are no randomised studies conclusively proving that AF responds to endocrine manipulation. However, several single-arm trials and case reports document both stabilization and regression of desmoid tumors to hormones alone or in combination with NSAIDs. One of the most commonly used antiestrogens in AF is tamoxifen. Other hormonal agents tested include toremifene, progesterone, medroxyprogesterone acetate, prednisolone, testolactone and gozerealone³².

Azzarelli et al³³ reported promising results with cytotoxic drugs in a study of 30 patients with advanced inoperable desmoid tumors who were treated with low-dose methotrexate and vinblastine, documenting disease stability in 60% of patients and partial response in the remaining 40%. A number of reports suggest that the response of AF to chemotherapy could be slow, and

therefore treatment may have to be administered for prolonged periods. This could be attributed to the unique pathological features of tumours, which is characterized by the presence of abundant collagen tissue, scanty malignant cells, especially in the center of the tumors and rare mitoses³².

Conclusively the use of such systemic treatments remains experimental or applicable to situations in which the more conventional modalities have already been tried. Adjuvant radiation has been suggested to diminish local recurrence and should be used selectively, especially in margin-positive tumors. Surgery remains the treatment of choice.

Conflict of interest statement

The authors have no financial or personal associations that might pose a conflict of interest in connection with the submitted article.

Riassunto

I tumori desmoidi che sono caratterizzati da infiltrazione locale aggressiva dei tessuti circostanti, sono rari neoplasmi benigni, senza potenziale metastatico che occasionalmente possono raggiungere grandi dimensioni.

Noi riportiamo il caso di una donna di 37 anni con un tumore desmoide della parete addominale che era comparso e cresciuto rapidamente durante la gravidanza, ed è stato diagnosticato con biopsia core trucut.

È stata effettuata una resezione chirurgica completa del tumore, di dimensioni 20x16cm, e ricostruzione immediata con rete nel periodo post-parto. La paziente non ha avuto complicazioni postoperative o ricorrenza dopo 2 anni di follow up.

Il trattamento ottimale dei grandi tumori della parete addominale deve essere individualizzato, con la resezione estesa, di rimanere il trattamento di scelta.

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