



Case Report

Important Connection Between Pregnancy and Desmoid Tumors

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Abstract

Persistent unexplained pain during pregnancy must be investigated by all healthcare providers as it could relate to growth of desmoid tumors. The importance of this is explained in our case of a 25-year-old woman who developed persistent buttock pain during the first trimester of her pregnancy. She was sent to physical therapy where her pain continued to worsen. The patient then received Magnetic Resonance Imaging (MRI) two years after initial presentation which showed a large soft tissue tumor in the buttock area. Unexplained pain during pregnancy could be linked to desmoid tumors which is important to be aware of for timely diagnoses and early treatment.

Keywords: Desmoid Tumors; Pregnancy; Cryoablation; Unexplained Pain

Introduction

Desmoid tumors, otherwise referred to as desmoid fibromatosis, are histologically bland growths that arise from the deep soft connective tissue and ultimately related to aberrant Wnt/ β -catenin signaling. They are known for being infiltrative and locally invasive. Although they routinely do not have the ability to metastasize, they have a high propensity for local recurrence [1]. This is a rare type of tumor with 2 to 4 cases per million population, but it most commonly occurs in individuals specifically females, between 15 to 60 years of age [2]. In fact, two-thirds of patients who receive this diagnosis are women [3]. The etiopathogenesis of desmoid tumors has been linked to trauma, pregnancy, and oral contraceptives [2]. An explanation for this can be tied to desmoid tumors' growth due to hormonal dependency, as illustrated by in vitro studies revealing that hormone-related proliferation of fibroblasts associated with desmoid tumors will increase during pregnancy and decrease postpartum [4]. This case demonstrates the importance of being aware of the linkage between pregnancy and desmoid tumors, and how primary care providers need to be especially alert when a woman presents with pain in a particular area in the body.

Case Presentation

A 25-year-old woman developed sharp pains in her left buttock during the first trimester of her pregnancy. She had no prior history of injury or infection. However, as her pregnancy progressed, the pain became more constant and enhanced after activity or long periods of sitting down. Postpartum, the patient went to physical therapy, utilized massage therapy, and non-steroidal anti-inflammatory drugs to alleviate the pain, but it still remained. There was no swelling in the leg, palpable mass, or skin discoloration; however, the patient still experienced difficulty moving her toes up and down with gait difficulty.

Differential Diagnosis

During the initial diagnostic assessment of this patient, an X-Ray of the lumbar spine was obtained which appeared normal, while the pelvic X-Ray showed osteitis. However, the patient's symptoms continued, so magnetic resonance imaging (MRI) of the pelvis with and without contrast was completed which revealed a large lobulated enhancing mass measuring 9.6cm x 3.9cm x 4.0cm extending within and anterior to the left gluteus maximus muscle; the mass also extended to the left anterior pelvis (Figure 1). Additionally, the MRI displayed a low signal adjacent area likely representing lobulation measuring 3.1x 7.4cm.

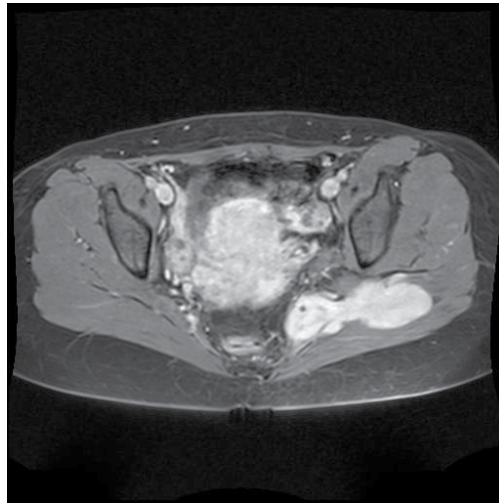


Figure 1: MRI Image of the pelvis with and without contrast. Orange arrow indicates a large lobular mass measuring 9.6cm x 3.9cm x 4.0cm.

An ultrasound-guided biopsy was obtained which was non-diagnostic, but the subsequent computed tomography-guided biopsy showed a cytologically bland spindle cell proliferation arranged in fascicles with admixed thin-walled blood vessels in a collagenous background; by immunohistochemistry, the neoplastic spindle cells exhibited nuclear accumulation of β -catenin (Figure 2). There was no significant cytomorphologic atypia, mitotic activity, or necrosis, and the histological features were diagnostic of desmoid fibromatosis.

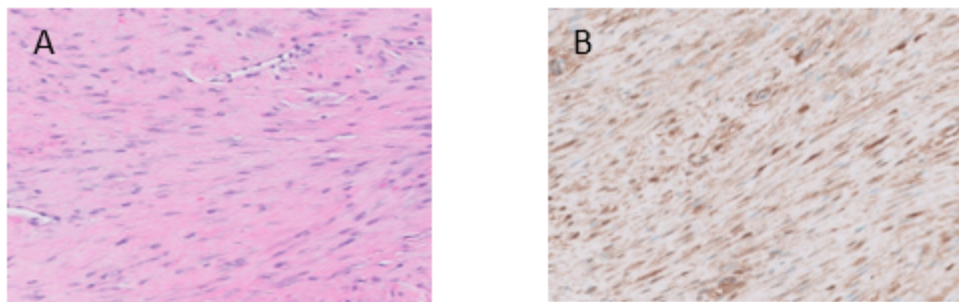


Figure 2: A) Representative photomicrograph of an H&E-stained histologic section of the tumor at 400x magnification. B) Representative photomicrograph of β -catenin immunohistochemistry at 400x magnification.

Results/Treatment

Patient was started on Sorafenib, (Nexavar[®]) which was stopped after 10 days due to Drug Rash with Eosinophilia and Systemic Symptoms (DRESS) syndrome. The tumor was then treated with cryoablation, targeting the lateral portions that were located in the left pelvis. A post-treatment MRI showed decreased overall tumor volume; there was decreased residual viable tumor at the medial border of the treatment cavity in the gluteus maximus muscle extending through the sciatic foramen into the pelvis. However, the patient's symptoms continued to worsen including random increased sharp pain when walking or driving and swelling when she walks too much. Given the growth of the desmoid tumor superficial to the left gluteus maximus muscle, the patient agreed to proceed with a second MRI-guided ablation.

Discussion

The majority of desmoid tumors related to pregnancy are diagnosed within 24 months following delivery, which is consistent with the chronology of this case [4]. Postpartum diagnoses of desmoid tumors were observed to be a result of many key events during

pregnancy such as hormone signaling, mechanical constraints, inherent trauma, and postpartum healing. MRI was key in diagnosing this specific type of tumor, especially in defining the pattern of the tumor and the extent of its involvement, as this form of imaging is known as the primary mode for imaging of desmoid tumors [5].

Planning a course of treatment following the diagnosis of desmoid tumors can be difficult as the progression of these growths can vary greatly. For our patient, surgery was not recommended as there was a high risk of recurrence, potential injury to the sciatic nerve, and increased risk of post-surgical morbidity. Radiation therapy has been shown to be effective in the majority of patients treated, however, secondary malignancies remain a potential risk [5]. The “wait and watch” approach was used after the patient underwent her first cryoablation treatment to see what areas of her tumor grow and how rapidly, in order to decide on the next phase of treatment. The efficacy of cryoablation after the “wait and watch” or surveillance approach was explored in a 30-patient retrospective study [6]. Eleven of 30 patients had partial ablation of the tumor because of proximity to critical structures. The treatment of these 11 patients was similar to our patient who had a tumor in close proximity to the sciatic nerve, superior gluteal nerve, and inferior gluteal nerve. In this retrospective study, eight of these 11 patients had partial regression, while 3 had progression of the untreated tumor. Overall, all patients in the retrospective study had a significant improvement of symptoms, and showed a durable decrease of pain even if complete ablation was not performed. These promising results support the use of secondary cryoablation to address persistent symptoms in our patient.

After the patient underwent cryoablation, surveillance treatment was utilized, and it is important to note that a request for a non-hormonal intrauterine device (IUD) placement instead of hormonal contraceptive was recommended, as birth control increases estrogen and can possibly lead to tumor growth. In fact, antiestrogen medications, such as tamoxifen, have some benefit in desmoid tumor therapy [7]. Similarly, after menopause or bilateral oophorectomies, spontaneous regression of desmoid tumors has been seen, once again indicating desmoid tumors’ reliance on estrogen [4]. Therefore, the dependence of desmoid tumors during pregnancy explains the growth stimulation of these tumors in particular during pregnancy when there is a high influx of this hormone. It is important to utilize effective contraception that do not cause further risk for patients with desmoid tumors where there are safer alternatives.

Conclusion

It is vitally important for health care providers to be aware that persistent unexplained pain during pregnancy may be linked to the growth of desmoid tumors. The knowledge that hormones such as estrogen are linked to desmoid tumor growth and can pose a risk of tumor growth during pregnancy or secondary to hormonal contraceptives used postpartum can affect how providers treat their patients. It is important to be aware of for timely diagnoses and early treatment.

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Statement of Ethics: Stanford has obtained IRB approval to publish retrospective cases.

Conflict of Interest Statement: The authors have no conflicts of interest to declare.

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