

# Giant Desmoid Tumor in the Pelvis: A Case Report

Davut GÜVEN<sup>1</sup>, İsmail KILICO<sup>1</sup>, Yıldırım DURAK<sup>1</sup>, İdris KOÇAK<sup>1</sup>

Cazip ÜSTÜN<sup>1</sup>, Oğuz AYDIN<sup>2</sup>

Samsun, Turkey

A 30 years old patient presented with complaint of abdominal discomfort and distension. The patient was gravida 1 and para 1. Ultrasonographic examination revealed a 90x83 mm sized mixed echoic pelvic mass located at the posterior of uterus. With an exception of high CA125 level of 63 IU/ml (cut-off is 35), the other serum laboratory findings were in normal ranges. She had not any history of familial or chronic diseases and she had a C-section six years ago. A laparotomy was performed and uterus, bilateral tuba uterina and ovaries observed as normal. A 20x25 mm sized, creamy coffee colored, solid natured, lobulated, hard mass was located at the paraumbilical region, bound to anterior abdominal via a thin pedicle.

**Key Words:** Desmoid tumor, Pelvic mass

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## Introduction

Desmoid tumor is a monoclonal, fibroblastic proliferation arising in musculoaponeurotic structures. This connective tissue hyperplasia infiltrates locally, recurs frequently after resection but does not metastasize. Abdominal desmoid occurs sporadically, in association with some familial syndromes and often represents a clinical dilemma for surgeons. The enigmatic biology and anatomical location of abdominal desmoids make treatment recommendations difficult. This distinct pathological entity is reviewed with a specific focus on a etiology and management. Desmoid tumor is a rare lesion representing < 3% of all soft tissue tumors with an estimated incidence of 2-4 new cases per million per year. An association with familial adenomatous polyposis of the colon (FAP) and Gardner's syndrome has been well documented. Abdominal and extra-abdominal desmoids occur more frequently in FAP patients, with an incidence of 3.5%-32%. In the original Gardner kindred the incidence was 29%.<sup>1</sup>

## Case Report

A 30 years old patient presented with complaint of ab-

dominal discomfort and distension. The patient was gravida 1 and para 1. Ultrasonographic examination revealed a 90x83 mm sized mixed echoic pelvic mass located at the posterior of uterus (Figure 1). With an exception of high CA125 level of 63 IU/ml (cut-off is 35), the other serum laboratory findings were in normal ranges. She had not any history of familial or chronic diseases and she had a C-section six years ago. A laparotomy was performed and uterus, bilateral tuba uterina and ovaries observed as normal. A 20x25 cm sized, creamy coffee colored, solid natured, lobulated, hard mass was located at the paraumbilical region, bound to anterior abdominal via a thin pedicle. The pedicle was clamped and mass was removed for pathological evaluation. Patological examination was reported as abdominal desmoid tumor (Figure 2). One year follow-up of the patient after the surgery was completely normal.



Figure 1: USG image.

<sup>1</sup>Departman of Obstetrics and Gynecology and <sup>2</sup>Pathology Ondokuz Mayıs University School of Medicine, Samsun

Address of Correspondence: Davut Güven  
Cumhuriyet mamallesi Atatürk bulvarı  
11. sokak No:1 Bahadır apartmanı D:6  
Atakum, Samsun  
dguven@omu.edu.tr,  
davut\_guven@yahoo.com

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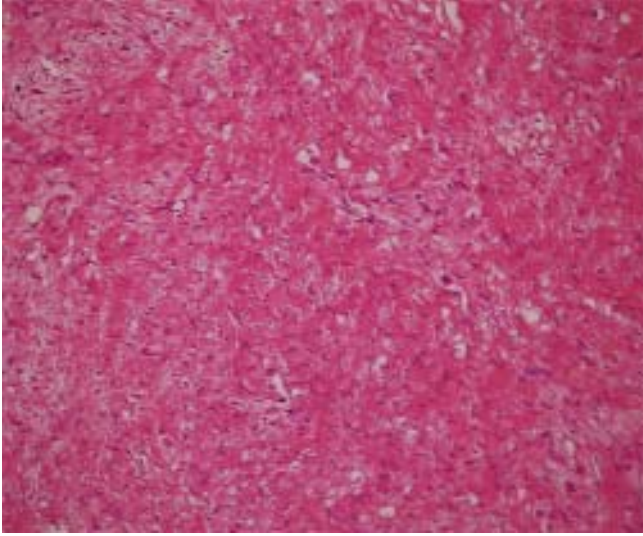


Figure 2: Tumoral lesion with atypical mesenchymal cells that have fusiform nuclei and eosinophilic cytoplasm (H&E, x200).

## Discussion

Desmoid tumors are benign deep fibromatoses, originating from fascia and muscle aponeurosis with an infiltrating growth. Only sporadic cases localized within the thorax wall or retroperitoneally are described. Desmoid tumor is often associated with female gender and occasionally with surgical trauma. It has a higher prevalence in women who experienced pregnancy. Our case had a history of a C/S 6 years ago. Depending on tumor size, therapy and negative resection margins, recurrence occurs in up to 45%.<sup>2</sup>

Therefore, a differentiation from other solid tumors is impossible using morphological criteria. Histology is the only evidentiary method which demonstrates long fascicles of spindle cells of variable cell-density with few mitoses and absence of atypical nucleus-separations. Characteristically, there is a diffuse cell infiltration of adjacent tissue structures. Immunohistochemical response for actin can be partially positive and immunohistochemical muscle cell markers delimit desmoid tumors from fibrosarcoma.<sup>3</sup>

Surgery always aims at radical tumor resection with free margins, which, depending on localisation of surgery, may leave major soft tissue defects behind. Although abdominal wall integrity after full-thickness surgery can be restored with direct sutures, reconstruction with synthetic materials is a common technique in major abdominal wall defects. Albeit in our study the big tumor resection and reconstruction of the abdominal wall was performed with direct sutures.<sup>4</sup>

Although hard data on abdominal wall desmoid tumors are

difficult to amass, given their uncommon nature, many other published series contend that recurrence is a major issue and that some molecular determinants are probably involved in its development.<sup>5</sup>

In conclusion, the treatment of desmoid tumors remains enigmatic. Non-surgical treatment resulted in diverse and unpredictable outcome and is considered to be an opportunity in patients with unresectable lesions or for adjuvant therapy. Radical resection with clear margins remains the principal determinant of outcome with the risk of local recurrence.

## Pelvisde Dev Bir Desmoid Tümör:

### Vaka Sunumu

30 yaşındaki hastanın bir yıldır karın ağrısı ve şişkinlik şikayeti vardı. Gravida 1, parite 1 olan hastaya yapılan USG'de Uterus posteriorunda 90x83 mm boyutlarında mixeikoik kitle görüldü. Hastanın CA125 değerinin 63 U/ml (Üst sınır 35 U/ml) olması dışında tüm laboratuvar verileri normaldi. Ailesel ya da kronik bir hastalığı olmayan, 6 yıl önce sezaryen operasyonu geçiren hastaya yapılan laparotomide uterus, tüpler ve overler normal görünümdeydi. Karın ön duvarına ince bir sapla bağlanmış, umblikusa yakın, 20x25cm boyutlarında sütlü kahve renginde solüt, lobüle, sert bir kitle görüldü.

**Anahtar Kelimeler:** Desmoid tümör, Pelvik kitle

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