Desmoid Tumor of the Anterior Abdominal Wall: Case Report

Anterior Karın Duvarı Desmoid Tümörü

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ABSTRACT Desmoid tumors are histologically benign lesions arising from musculoaponeurotic structures. They are commonly seen in fertile women between 25-40 years of age. Here in we present a case with desmoid tumor located in the anterior abdominal wall. A 68 years-old female patient presented with the complaint of pain in the upper-left quadrant of the abdomen. Radiologic examination showed a well-circumscribed solid mass in size of 7x5 cm located between abdominal wall muscles. Surgical excision was performed within 2-3 cm clear surgical margins. The patient was discharged on the second postoperative day. Histopathological investigation was reported to be desmoid tumor. Desmoid tumors are mesenchymal origin tumors which have tendency to grow slowly. Surgical resection in which sufficient amount of normal tissue is removed together with tumoral mass provides an efficient therapy. Since high recurrence is observed, long-term follow-up should be performed after surgery.

Key Words: Desmoid disease, hereditary; abdominal wall; surgery

ÖZET Desmoid tümörler müsküloaponörotik dokulardan gelişen histolojik olarak benign lezyonlardır. Sıklıkla 25-40 yaş arası fertil bayanlarda görülürler. Bu yazıda karın ön duvarına yerleşmiş desmoid tümörlü bir olgunun sunulması amaçlandı. 68 yaşında kadın hasta karın sol üst kadranda ağrı şikayeti ile başvurdu. Radyolojik incelemede karın duvarı kasları arasında 7x5 cm büyüklüğünde düzgün sınırlı solid kitle tespit edildi. 2-3 cm temiz cerrahi sınırlarla eksizyon yapıldı. Hasta ameliyat sonrası 2. günde taburcu edildi. Histopatolojik inceleme sonucu karın ön duvarı desmoid tümör olarak rapor edildi. Desmoid tümörler mezenşimal kökenli ve yavaş büyüme eğiliminde olan tümörlerdir. Yeterli miktarda sağlam dokunun da tümöral kitle ile birlikte çıkarıldığı cerrahi rezeksiyon, etkin bir tedavi sağlamaktadır. Yüksek rekürrens gözlendiğinden ameliyat sonrası uzun dönem takip mutlaka yapılmalıdır.

Anahtar Kelimeler: Dezmoid hastalık, kalıtsal; karın duvarı; cerrahi

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Desmoid tumors are histologically benign lesions arising from musculoaponeurotic structures. Desmoid tumors account for 0.03% of all neoplasms and 3% of all soft tissue tumors. These are commonly seen in fertile women between 25-40 years of age.[1,2] They occur most frequently in the abdominal wall at a rate of 50%.[3,4] The etiology is not known exactly. They are distinguished from fibrosarcomas by not metastasizing to the other organs. Ultrasonography (USG), computed tomography (CT) and magnetic resonance imaging (MRI) are used in the diagnosis of the disease.

In this article, it was aimed to present a 68 years-old of case with desmoid tumor located in the anterior abdominal wall.

CASE REPORT

A 68 years-old female patient presented to our polyclinic with the complaint of pain and swelling in the upper-left quadrant of the abdomen lasting approximately for 3 months. There was no history of smoking, drug use and previous surgery. Her family history was non-contributory. A solid mass was palpated in the left-upper quadrant of the abdomen during the physical examination. Biochemical parameters were within normal ranges and tumor markers were negative in the laboratory investigation. A mass with a sharp margin consisting of solid and cystic areas in the left-upper quadrant of the abdomen was determined by the USG. A well-circumscribed solid mass in size of 7x5 cm consisting small cystic components in patches located between abdominal wall muscles in the lower neighborhood of left arcus costarium was determined by CT (Figure 1). Pathological lymphadenopathy was not encountered. Surgical excision was performed within 2-3 cm clear surgical margins by receiving informed consent from the patient (Figure 2). The defect was covered with 10x15 cm polypropylene mesh. The patient had an uneventful postoperative period and she was discharged on the second postoperative day. Histopathological investigation was reported to be desmoid tumor located in the anterior abdominal wall (Figure 3). Recurrence was not encountered and incisional hernia was not developed in 12th month of her follow-up.

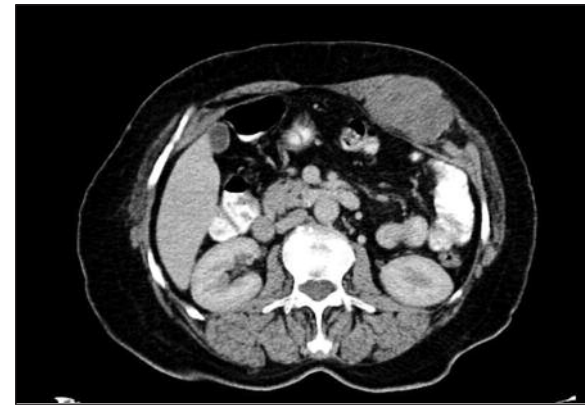

FIGURE 1: CT examination revealed desmoid tumour located between abdominal wall muscles.

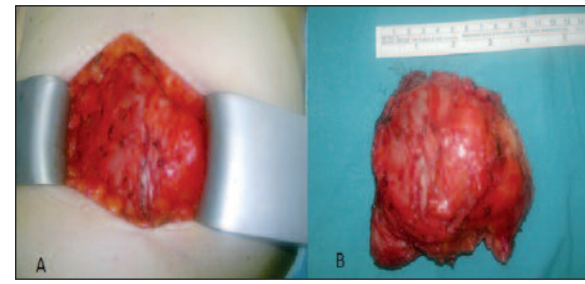

FIGURE 2: Intraoperative pictures of surgery for abdominal wall desmoid tumor. **A)** Abdominal wall with tumor. **B)** Macroscopic view of the tumor.

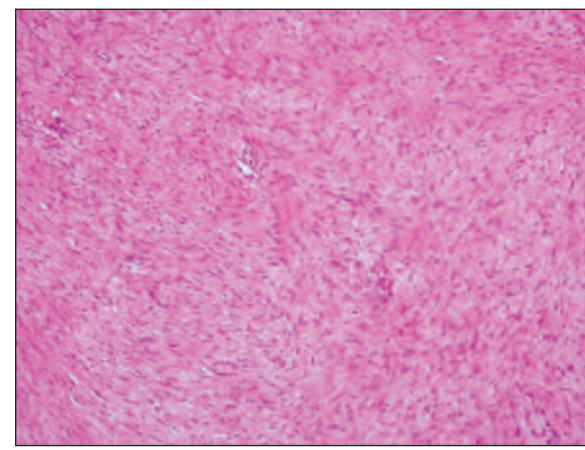

FIGURE 3: Microscopic view of the excised rectus desmoid tumor showing fascicles of fibroblastic spindle cells with abundant intercellular collagen. (Hematoxylin and eosin stain; original magnification × 100.)

DISCUSSION

Desmoid tumors are rarely seen mesenchymal tumors. They are also called as aggressive fibromatosis and musculoaponeurotic fibromatosis. A tumor occurring in the abdominal wall of a woman after delivery was described by McFarlane for the first time in 1832. However, description of desmoid tumor was used by Mueller in 1838.[5,6] Its incidence was 2-4 per million. It is commonly seen in young females. Male/Female ratio is 1/2. Although they are known to be benign tumors, they have a high trend towards local invasion and recurrence. Desmoid tumors have no tendency to metastasize. The etiology

is not known exactly, but multifactorial pathogenesis is a matter of discussion. A close correlation was determined between the area in which tumor occurrence was seen and the previous trauma or surgical intervention occurred in that area. In addition to this, a close correlation was determined between desmoid tumor and pregnancy, external estrogen intake, Gardner's syndrome and Familial adenomatous polyposis (FAP) syndrome.[1,7] Gardner's syndrome, which is a variant of familial adenomatous polyposis (FAP) syndromes, arises from mutations in the adenomatous polyposis coli (APC) gene located on the long arm of chromosome 5 and it is an autosomal dominant disease. It is characterized by multiple colorectal polyps, a great number of osteomas and presence of mesenchymal tumor in the soft tissue. In these patients, association with desmoid tumor was shown a rate of 10-15%.[8] Affection of the women in reproductive age much more by the tumor suggested its relationship with estrogen. There are some studies showing that tumor frequency increases in the individuals using oral contraceptives and tumor regresses during menopause of tamoxifen usage.[9] Ranitamo et al. demonstrated the correlation between growth rate of tumor and estrogen levels. According to this, the slowest growth was determined to be during maidenhood. A marked increase in menarche and a decrease in menopause were determined.[10] Growth rate in male patients is independent from the age it is as low as in the elderly females.

Desmoid tumors have four distinct types according to the age they are seen. These are: juvenile (in young ladies and generally located extra-abdominally), fertile (during reproductive age and generally located intra-abdominally), middle-age (seen on an equal basis in both genders and predominantly located intra-abdominally) and advanced-age (seen on an equal basis in both genders and may show location intra-abdominally or extra-abdominally) groups.[10] Our case was a 68 years-old aged patient without any specific past medical history and desmoid tumor was located in the anterior abdominal wall. Extra-abdominal desmoid tumors may occur mainly in the extremities, neck, trunk and the various parts of the body. Abdominal desmoid tumors often show trend to be located in the abdominal wall, intestinal wall and mesentery.[11,12] Desmoid tumors of anterior abdominal wall originate from the musculoaponeurotic structures of abdominal wall especially like rectus abdominus and internal oblique muscle and less commonly external oblique muscle and transversus abdominus muscle and their fascias.

Histologically, desmoid tumours accounts for fusiform myofibroblastic cells with ovoid or circular vesicular nuclei that ordinarly comprise paralel bundle. Cells are arranged in a linear order and separated from each other by collagen.[13]

USG, CT and MRI can be used in the diagnosis of the disease and for determination of relationship with the surrounding structures and recurrence. On USG, desmoid tumor is seen as a lesion with smooth but without sharply circumscribed margins and in varying echogenicity. On CT, desmoid tumor appears as an isodense-hyperdense mass rather than well-circumscribed homogeneous muscle tissue. On MRI imaging; heterogeneity with irregular margin is seen and hypointensity (low signal intensity) on T1-weighted images and heterogeneity (variable signal intensity) on T2-weighted images are seen. Hypointensity on T2-weighted images shows that collagen deposits are in high concentrations.[14]

Definitive diagnosis of the disease is made by histopathological investigation.[11]

Although the treatment of choice is surgical excision; radiotherapy, chemotherapy, hormonal and anti-inflammatory treatment can also be combined with surgical treatment. Basis of surgical treatment is radical local excision. Desmoid tumors located in the anterior abdominal wall can be excised easily by maintaining safe surgical margins of 2-3 cm generally. Defect occurring after excision is generally repaired with mesh reconstruction. We also excised the mass present in the anterior abdominal wall of our patient by maintaining approximately a safe margin of 2 cm. Then the defect occurring in the anterior sheath of rectus was repaired by using a 15x10 cm prolene mesh.

Recurrence in desmoid tumors following surgery is seen at a rate of 20-70% even though it is dependent on the localization of the tumor and the extent of the surgery. Desmoid tumors in the abdominal wall have lower recurrence at a rate of 20-30% and recurrence is frequently seen in the first 6 months after surgery. Radiotherapy is an alternative treatment option either in the patients unresectable or surgical resection is associated with severe morbidity rate and possibility of loss of organ function or in the patients with a positive surgical margin.[15] We determined the excision margins in our case as the surgical margin would be negative and pass near to approximately 2 cm away from the mass. We performed the resection and sent the specimen for histopathological investigation. In the histopathological investigation, it was seen that the spindle-shaped cells in uniform shape and size were predominant in the specimen. It was determined that these spindle-shaped cells formed clusters and showed elongations to the surrounding tissue in patches. There was not nuclear hyperchromasia in the cells and these cells were staining immunohistochemically positive for vimentin. All the surgical margins were determined to be negative. Therefore, no adjuvant therapy was needed in addition to surgical treatment during postoperative period.

In conclusion, desmoid tumors are mesenchymal origin tumors which have tendency to grow slowly. Despite these tumors have no tendency to recur; they may cause pressure effect on the surrounding organs and accordingly loss of organ function by locally progressing. Surgical resection in which sufficient amount of normal tissue is removed together with tumoral mass provides an efficient therapy. Since high recurrence is observed, long-term follow-up should be performed after surgery. MRI is an imaging method which can be used safely during follow-up.

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