A Huge Desmoid Tumor: A Case Report

DEV DESMOİD TÜMÖR: OLGU SUNUMU

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Abstract

A case report of desmoid tumor located within the abdominal wall is presented. A 31 years old woman gravida 5, para 5 admitted to the hospital with a mass, located just under the previous ceaserian section (C/S) scar which she had recognized 6 months after the operation. An exploratory laparotomy has been performed which showed a 15 cm x 6 cm x 6 cm mass beginning subcutanously with the invasion of all abdominal wall except parietal periton. The histological studies confirmed a desmoid tumor.

Key Words: Desmoid tumor, ceaserian section scar

Özet

Karın ön duvarında lokalize bir desmoid tümör olgusu sunulmuştur. Otuz bir yaşında gravida 5, para 5 olan bir kadın hasta eski sezaryen skarı altındaki, operasyondan 6 ay sonra farkettiği bir kitle nedeniyle hastaneye başvurmuştur. Yapılan laparatomi sonucunda cilt altından başlayan, parietal periton harici tüm karın duvarını içine alan 15 x 6 x 6 cm'lik kitle tespit edilmiştir. Histolojik çalışmalar kitlenin desmoid tümör olduğunu göstermiştir.

Anahtar Kelimeler: Desmoid tümör, sezaryen skarı

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Desmoid tumors are histologically benign fibrous neoplasms originating from the musculoaponeurotic structures of the body. Muller first described the term desmoid in 1838, this term is derived from the Greek word desmos, that means tendonlike structure.[1]

Desmoid tumors are reported to account for 0.024-0.035% of all neoplasms.[2,3] When present in patients with familial polyposis of the colon, the prevalence of desmoid tumors are as high as 13%.[4] The biggest desmoid tumors are usually seen with Gardner Syndrome which are measuring up to 10 cm in diameter.[5]

Desmoid tumors are usually called as aggressive fibromatosis because they have a great tendency to infiltrate the neighbouring tissues and they are often adherent to surrounding structures. The aggressive form of the fibromatosis defines marked cellularity and aggressive local behavior. The tendency to the recurrence makes the treatment of these relatively rare fibrous tumors challenging. The patient with desmoid tumor are mostly 20 to 40 years old. They have usually 3 important localizations: The first one is the abdominal wall muscles especially during or after pregnancy. The second place is the shoulder, chest wall, back and hip muscles. Thirdly, it is seen as retroperitoneal neoplasms inside the abdominal cavity at the patients with Gardner syndrome.

Here presented is a huge desmoid tumor located on abdominal wall.

Case Report

A 31 years old woman, gravida 5, para 5, was referred to our hospital for suspected mass at the abdominal wall under the previous C/S scar. She had a past history of C/S operation due to the twin pregnancy in january 2002. She had no complaints about the mass but had a disturbing feeling of

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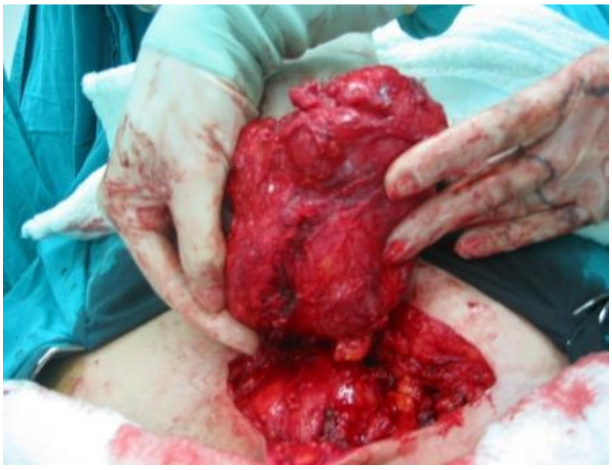

Figure 1. A total 15 cm x 6 cm x 6 cm mass was completely excised with free margins.

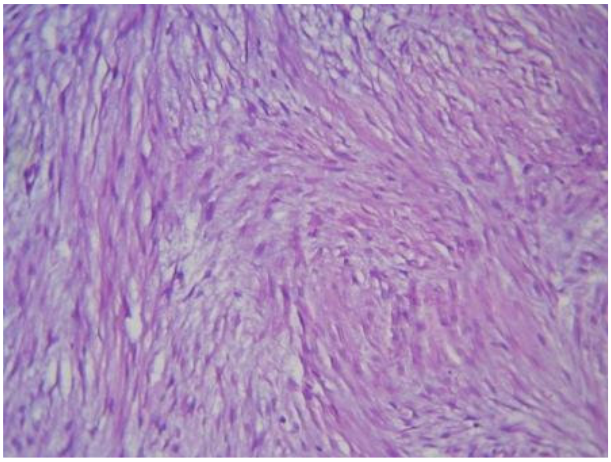

Figure 2. The histopathology of the specimen HxEx100.

bloating. The tumor markers were in the normal ranges, hemoglobin level was 12 g/dL and the haematocrite was 35%. All the biochemical parameters were in the normal limits. At the pelvic examination, we found a mass measuring nearly 15 cm x 5 cm x 6 cm in diameter just beneath the skin including the hypogastrium and both inguinal regions. The mass was strongly fixed to the neighbouring tissues.

The uterus was in normal size and consistency, both of the ovaries were normal. The transabdominal ultrasound examination showed a solid mass occupying the all abdominal wall structures, measuring 15 cm x 5 cm x 6 cm. At the exploratory laparotomy, there was a mass beginning just under the skin to the parietal periton. After confirmation of desmoid tumor by histopathology, we have extirpated the tumor mass totally from the anterior abdominal wall (Figure 1, 2). The defect in the abdominal wall was reconstricted by a Prolen-Mesh®. The specimen was sent for histopathological examination. Postoperatively, the patient underwent colonoscopic examination for the elimination of the familial poliposis. The findings were normal. The patient had an uneventful postoperative course and was discharged from the hospital on the 4th day.

Discussion

Desmoid tumors are benign, deep and infiltrating fibromatous growth originating from fascia and muscular aponeurosis. They have a tendency to infiltrate adherent tissues. The desmoid tumor is often seen in female gender after surgical trauma.[6,7] Our case had a history of a C/S 2 years ago. It is believed that the occurence of the desmoid tumor may appear about 4 years after the surgical trauma. The desmoid tumors may be seen sporadically or may be associated with familial adenomatosous polyposis. Clinically, the desmoid tumors seen with familial poliposis are more agressive than the other types of the tumor. The sporadic cases are usually seen in the abdominal wall and mostly in women.[3] Gansar et al reported that the prevalance of desmoid tumors are higher in parous women as we mentioned in our patient.[8] Desmoid tumors of the abdominal wall are mostly seen after a pregnancy.[3] It may be due to high estrogen level and progesterone withdrawal. During the fertile period, the growth of the tumor is fast and goes parallel with the estrogen levels. At the menopause, they may undergo to spontaneous regression.[5] We know that the biology of this disease is related to the endogenous hormonal environment and that estrogen receptors have been shown in desmoid tumors itself. For this reason, Kiel et al recommend that endocrine treatment may be employed in inoperable desmoid tumors.[9]

The treatment of desmoid tumors consist of surgical resection, radiotherapy, chemotherapy, anti-inflammatory treatment and hormone-therapy. All

these treatments have different indications. Among those, surgical excision is the first choice of treatment. Surgery always aims a radical tumor resection with free margins. For this reason, we must secure a good reconstruction with synthetic or autolog material in the surgical field. In our case, the large tumor resection with tumor free margins and reconstruction of the abdominal wall was performed with Prolen-Mesh®.

The other treatment modalities are generally prefered in incomplete tumor resection, recurrence of the tumor or morbide conditions of the patients.

Desmoid tumors are locally aggressive tumors, with no metastatic potential for far organ systems, that generally are amenable to treatment modelities, such as surgery and radiation therapy. Systemic therapy is just considered for selected cases that are not able to be extirpated by surgical approach. Desmoid tumors in adults have a limited response to chemotherapy and such treatment should only be considered before embarking on radical treatment to avoid obvious negative consequences and delayed complications.

Radiotherapy is also recommended in patients with extended local disease having the possibility of significant morbidity. After incomplete resection of the tumor, the patients should be followed carefully for the tumor recurrence. If the recurrence occurs, the immediate radiotherapy should be the first choice.

Until now, the most effective treatment of accessible and smaller desmoid tumors is the resection with negative margins, although it may not guarantee local recurrence of the disease. It has been reported that the recurrence rate of the tumor after surgery is 40-45%. The factors affecting recurrence rate after resection are the completeness of surgical margins, age of the patient, the tumors size, the existance of trisomy 8 in the lesion, family history and time past after surgery.[10]

In conclusion, although the surgical resection is the first choice, the treatment of desmoid tumors remains uncertain. Radical resection with free margins remains the principal determinant of outcome with the risk of local recurrence. Non-surgical treatment can only be used in large and unresectable masses, without definite success.

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