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Evaluation of Desmoid-type Chest Wall Tumors: A Single Center Experience

Göğüs Duvarının Desmoid Tip Tümörlerin İncelenmesi: Tek Merkez Deneyimi

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Evaluation of Desmoid-type Chest Wall Tumors: A Single Center Experience

ABSTRACT

Objective: While chest wall desmoid-type tumors are rare, they do pose significant challenges in their management due to their high rate of recurrence. The purpose of this study is to evaluate our clinical experience of these tumors while reviewing the relevant literature.

Material and Methods: A retrospective study was conducted of patients diagnosed with desmoid-type chest wall tumors who underwent surgery at Gaziosmanpasa University Faculty of Medicine between 2007 and 2022. While all patients underwent surgical management, two patients received radiotherapy after their second recurrence. Demographic findings, clinical features, operative methods, local recurrences, and follow-up criteria were recorded as part of the study. Gastroscopies and colonoscopies were performed on all patients to exclude Gardner syndrome.

Results: Five of the patients in our study were female and one was male, which represented 16.6% of the group. The age range of the study group was between 14 and 76 years old. The recurrence rate was 33.3%, with two patients experiencing three tumor recurrences who received radiotherapy between the second and third recurrences. One patient, who still had a tumor, died due to a myocardial infarction. We did not observe any major complications in the other patients. All of the patients under follow-up are still alive.

Conclusion: While the primary goal of the treatment should be to perform as wide a resection as possible, this is insufficient for the prediction or prevention of recurrences, and close follow-up is necessary for the management of locally invasive tumors.

Keywords: Aggressive fibromatosis, desmoid tumors, epidemiology, soft-tissue tumor, thoracic surgery

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ÖZET

Amaç: Göğüs duvarı desmoid tipi tümörleri nadir görülür ve yüksek nüks oranları nedeniyle yönetimlerinde önemli zorluklar oluşturur. Bu çalışmanın amacı, bu tümörlerle ilgili klinik deneyimlerimizi aktarmak ve ilgili literatürü gözden geçirmektir.

Gereç ve Yöntem: Gaziosmanpaşa Üniversitesi Tıp Fakültesi' nde 2007-2022 yılları arasında ameliyat edilen ve histopatolojik olarak desmoid tipi göğüs duvarı tümörü tanısı almış hastalar retrospektif olarak incelendi. Tedavide tüm hastalara cerrahi uygulandı ve iki hastaya ikinci nüksünden sonra radyoterapi verildi. Demografik bulgular, klinik özellikler, cerrahi yöntemler, lokal nüksler ve takip kriterleri kaydedildi. Gardner sendromunu dışlamak için tüm hastalara gastroskopi ve kolonoskopi yapıldı.

Bulgular: Çalışmamızdaki hastaların beşi kadın ve biri erkekti. Çalışma grubunun yaş aralığı 14 ile 76 yaş aralığındaydı. Nüks oranı %33,3 olarak hesaplandı. İki hastada üçer kez tümör nüksü görüldü. Bu hastalara ikinci ve üçüncü nüksleri arasında radyoterapi uygulandı. Bir hasta, tedavi ve takip sürecinde miyokard enfarktüsü nedeniyle hayatını kaybetti. Diğer hastalarda herhangi bir major komplikasyon izlenmedi, tüm hastalar sağ olarak halen takiptedir.

Sonuç: Tedavideki birincil hedef mümkün olduğunca geniş bir rezeksiyon yapılması olmalıdır. Ancak bu tek başına nüksleri öngörmek veya önlemek için yeterli değildir. Desmoid tip tümörlerin yönetimi için hastaların yakın takibi önemlidir.

Anahtar Sözcükler: Agresif fibromatozis, desmoid tümörler, epidemiyoloji, torasik cerrahi, yumuşak doku tümörü.

Introduction

Desmoid tumors (DTs), which are soft-tissue tumors characterized by a proliferation of fibroblasts and myofibroblast-type spindle cells, infiltrate musculoaponeurotic tissue. They are rare pathological entities, representing 3.5% of fibrous tumors and 0.03% of all neoplasms (1). DTs are low-grade malignancies belonging to the sarcoma group and have microscopic features similar to, or even indistinguishable from, fibromas or fibrosarcomas (1, 2). Histopathologically, DTs are benign, locally aggressive tumors that do not tend to metastasize. DTs of the chest wall are uncommon and their etiology remains uncertain, although it is thought that trauma, hormonal factors, and genetic causes may all play a role in their development (2, 3). A common approach to primary treatment for DTs is wide surgical resection and RT or a combination of surgical resection and RT, although adjuvant treatments are also recommended in the postoperative period to reduce the risk of recurrence (2, 4). Recent alternative adjuvant therapies include tyrosine kinase inhibitors, gamma-secretase inhibitors, chemotherapy agents, estrogen receptor inhibitors, and cryoablation. Nirogacestat, a gamma-secretase inhibitor, is the preferred systemic therapy option. Tyrosine kinase inhibitors are also reasonable alternatives. Cytotoxic chemotherapy may be preferred for patients who require a more rapid response. Cryoablation, a percutaneous ablation technique, has recently been described in a few prospective reports as a newer treatment option. (5, 6). The unpredictable course of tumor requires as close and as long-term follow-up as possible. As the possibility of long-term recurrences must always be considered, the importance of long-term and close follow-up is emphasized. The primary treatment for DTs is wide surgical resection, although adjuvant treatments are also recommended in the postoperative period to reduce the risk of recurrence (2, 4). The unpredictable course of tumor requires as close and as long-term follow-up as possible. As the possibility of long-term recurrences must always be considered, the importance of long-term and close follow-up is emphasized. The purpose of this study is to evaluate our clinical experience with these tumors while reviewing the relevant literature.

Material and Methods

This study was approved by the local ethics committee (Approval No. 22-Kaek-096) and was conducted in accordance with the Declaration of Helsinki. This cross-sectional descriptive study included six patients who were diagnosed with chest wall DTs and underwent surgery between 2007 and 2022 at the Faculty of Medicine, Gaziosmanpasa University. Demographic data, clinical features, operative methods, local recurrences, and follow-up criteria were collected. All patients had gastroscopies and colonoscopies to rule out Gardner syndrome. Preoperatively, patients were evaluated with conventional radiological imaging using Computed Tomography (CT) and Magnetic Resonance Imaging (MRI). During surgery, all patients underwent a wide resection, including skin and musculature structures, at least 2 cm away from the tumor lesion, as well as rib resections at least one rib above and below the tumor site. Frozen sections and surgical margins were studied in all patients whenever possible. Chest wall defects greater than 5 cm were closed by grafting, especially when defects occurred in the anterior region. None of the patients had a preoperative needle biopsy and electromyography was used to evaluate any neurological disorder. Recurrences were detected using computed tomography and magnetic resonance imaging. Two patients received radiotherapy after the second recurrence, and postoperative follow-ups were conducted using CT and MRI every six months for the first five years, then followed by annual check-ups.

Results

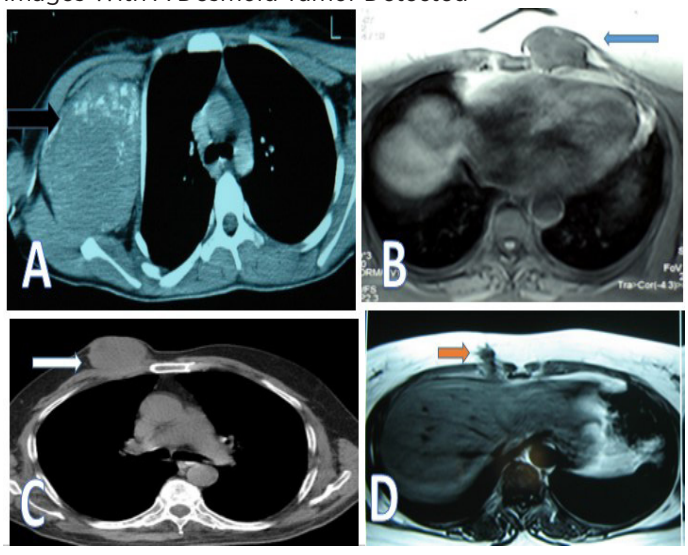
Our study included six patients, five of whom were female and one male, representing 16.6% of the group. The ages of the patients ranged from 14 to 76 years. The patients were evaluated preoperatively using conventional radiological imaging using Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) (Figure I A, B, C, D). During the surgery, all of the patients underwent a wide resection. This included skin and musculature structures at least 2 cm away from the tumor lesion, as well as rib resections at least one rib above and below the tumor site (Figure II A, B, C). All patients underwent surgical removal of the tumor, along with the surrounding tissue, and

Table I. The Clinical Features Of Patients With Desmoid-Type Tumors

Case	Age/Gender	Clinical Presentation	Tumor size	Surgery –surgical margins	Follow-up-recurrences	Progression
1	14/Female	Right axillo-thoracic	12 cm	Complete Inappropriate for surgical margins	Three recurrences 24+12+RT+36 months Still annual check-ups	Free of tumor
2	54/Female	Left parasternal 2.-3. costal cartilage	3 cm	Complete Negative surgical margins	24 months Out of follow-up	Free of tumor
3	36/Female	Inframammarial 6.-7. costal cartilage	2 cm	Complete Negative surgical margins	36 months Still annual check-ups	Free of tumor
4	76/Male	Right anterior chest wall right 3.-5. rib	7 cm	Complete Negative surgical margins	Three recurrences 36 months 12+18+RT-6 months	(Transition to sarcomatoid form. Died due to myocardial infarction with tumor)
5	53/Female	Right parasternal 8.-9. costal cartilage	3 cm	Complete Negative surgical margins	36 months Still annual check-ups	Free of tumor
6	41/Female	Left midaxillary 6.-7. Rib	3 cm	Complete Negative surgical margins	36 months Still annual check-ups	Free of tumor

the median follow-up was 36 months. Two patients, Case 1 and Case 4, had three tumor recurrences and received radiotherapy between the second and third recurrences, making the recurrence rate 33.3%. (Figure III A, B) Despite radiotherapy, the third recurrences occurred in both patients at the sixth month and in three years. Right-sided intermediate ulnar paresis, due to brachial plexus involvement, was detected in Case 1 before surgery, and the condition persisted after the surgery. Pathological analyses confirmed desmoid-type histologic features in all patients, and the Case 4 showed sarcomatoid transition in his recurrent tumor (Figure IV A, B). While preparing for surgery, this patient died in the hospital due to a myocardial infarction. One patient, Case 2, whose surgical margins were positive on permanent section surgery, was offered re-operative resection, but this was refused by the patient, and the patient also opted out of future follow-ups after the second year (Table I). At the time of writing, four patients are still attending annual check-up examinations and have had no signs of recurrent tumors.

Figure I. Computed Tomography and Magnetic Resonance Images With A Desmoid Tumor Detected



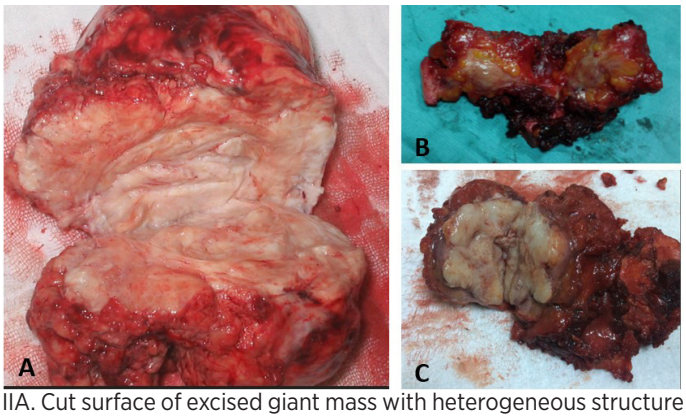
IA. Axial computed tomography image (black arrow shows calcifications), which demonstrates the huge tumor dwelling in the right axillo-thoracic area (Case 1)

IB. T1-weighted axial magnetic resonance image of the desmoid tumor with heterogeneous enhancement, low signal bands on the left side of the sternum (blue arrow) (Case 2)

IC. Computed tomography imaging of well-bounded desmoid tumor (white arrow) with subcutaneous placement just left side of the sternum (Case 4)

ID. Desmoid tumor formation with an irregular contour that penetrates the chest wall (orange arrow) (Case 3)

Figure II. Images of Desmoid Tumor Removed Through Surgery (Case 1)



IIA. Cut surface of excised giant mass with heterogeneous structure (white and fibrous appearance) (Case 1)
IIB. Excised chest wall desmoid tumor with intercostal muscle and costal cartilage (Case 3)
IIC. Excised chest wall desmoid tumor with skin and surrounding musculature structure (Case 4)

Figure III. Images of recurrences of Desmoid Tumor Cases

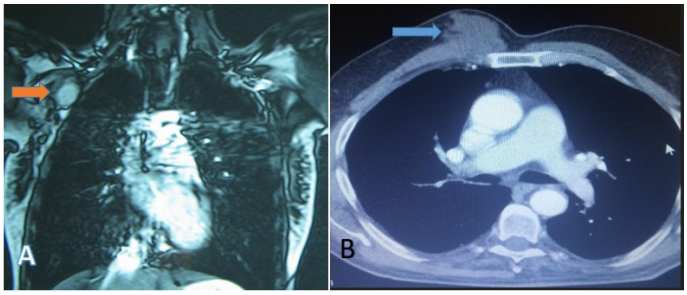


Fig. IIIA Case 1 Coronal Magnetic Resonance image showing well-circumscribed nodular lesion in the axillary fossa adjacent to the axillary artery (red arrow)
Fig. IIIB Case 4 Computed Tomography image showing a mass lesion with irregular borders and soft tissue density located subcutaneously in the right sterno-costal area in the mid-thoracic section image (blue arrow)

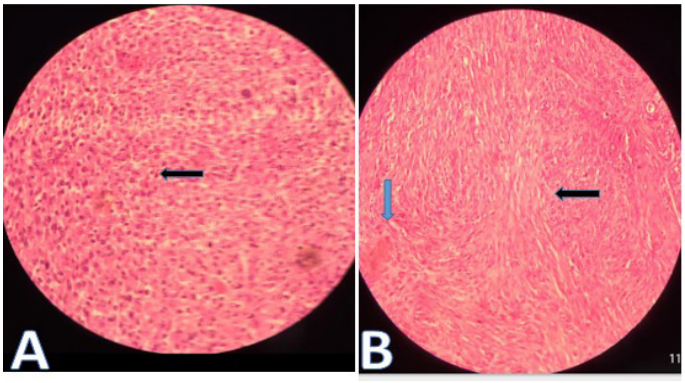
Discussion

DTs were first described by John MacFarlane in 1832 as abdominal wall tumors, with extra-abdominal types being later identified, particularly in the shoulder girdle and lower limbs (1). 10%–20% of tumors are located on the chest wall and an intrathoracic tumors is extremely rare (2). DTs are sometimes referred to as low-grade sarcomas or aggressive fibromatosis, as these synonyms more accurately describe their natural behavior. Chest wall DTs are rare and, due to their high recurrence rates, pose significant challenges in their management, surgical treatment, and follow-up. The exact pathological mechanisms behind these tumors are not yet fully understood, although there have been suggestions that hormonal factors, particularly estrogen, trauma to the chest wall, and previous thoracic operations, including breast implants, muscular transplants, and even intramuscular injections, may all play a role in their etiopathogenesis (2). None of the patients in our study had a history of trauma or a previous operation. Genetic predisposition, carried by the Y chromosome, or the long arm of the fifth chromosome in Gardner syndrome, which is frequently associated with women, results in 15% of patients possibly developing DTs. Our findings were consistent with previous reports, with women being slightly more susceptible to DTs than men (3, 4).

The majority of patients complain of chest pain and firm masses on their chest wall, which they have detected themselves. The sizes of tumors are typically large upon hospital examination due to their asymptomatic nature (3). Symptomatology varies due to tumor involvement of adjacent structures, and the involvement of brachial plexus, as with Case 1, has also been previously reported (7). As DTs in the mammary region may be misdiagnosed as breast tumors, meticulous screening methods are required to obtain an accurate differential diagnosis (8). CT and MRI studies are helpful for diagnosing and delineating tumor margins in preparation for surgery, and MRI is preferable in cases where bone involvement is suspected. Radiologic features of DTs are similar to those of soft tissue malignant tumors, with well-circumscribed masses that are predominantly homogeneous or heterogeneous. A preoperative incisional biopsy or needle biopsy may suggest certain diagnoses and provide helpful information regarding surgical margins and the determination of the tumor differentiation, which may be needed before surgery. However, tumor spillage or contamination is also possible (2, 3). The use of preoperative biopsies was not preferred. During surgery, the primary objective is to achieve a wide resection 2–4 cm from the tumor and rib resections above and below the tumor site with negative surgical margins. Chest defects can be closed with a muscle flap, autologous bone grafts, polytetrafluoroethylene (PTFE), or other similar synthetic tissues. Histological examinations have

demonstrated long fascicles of spindle cells of variable cell-density with few mitoses and the absence of atypical nuclear separations. Characteristically, there is a diffuse cell infiltration of adjacent tissue structures. Immunohistochemically, the spindle cells are positive for vimentin, smooth muscle actin, and muscle-specific actin, which reflects a fibroblastic-myofibroblastic differentiation. The possibility of transition to a sarcomatoid form of tumor during the follow-up period must be considered (8).

Figure IV. Histological Features of Operational Materials from Case 4



IVA. Spindle fibroblastic cells (black arrow) with elongated nuclei in a collagen-rich stroma

IVB. Microhemorrhages (blue arrow) and slender, spindle-shaped tumor cells arranged with ill-defined parallel fascicles (black arrow) that are loculated by abundant interstitial collagen without increased mitotic activity. (Hematoxylin-eosin, ×20). The same patient with a sarcomatoid transition.

(Permission was obtained from the pathology department for histopathological images)

Recurrence rate after surgery for DTs is high and has been reported to range between 25% and 75% in different series (3). The recurrence rate of this study is 33.3%. Some of the predictive parameters for recurrence suggested by authors include age, sex, incomplete resection, positive surgical margins, multiple recurrences, and the absence of radiotherapy (2). While recurrences after surgery are unpredictable, it is essential to aim for a wide resection with negative surgical margins during the first surgery (10). However, obtaining negative surgical margins may not be the only predictive key for recurrences. Furthermore, providing negative surgical margins may be difficult, particularly in the axillary and cervicothoracic regions. As occurred with Case 2, histopathologically negative frozen sections may become positive in permanent sections (3, 10). There is no difference in overall recurrences between microscopic negative and positive margins. In some cases, the main objective of management may be to preserve vital organs and a patient's quality of life (9, 11). Although the efficacy of radiotherapy after surgery in preventing recurrences is uncertain, it may be beneficial in delaying the onset of recurrences (12, 13). Radiotherapy is most suitable for unresectable or partially resectable residual tumors or in positive surgical margins, although some authors may prefer radiotherapy despite negative surgical margins (14, 15). A complete response after radiotherapy is difficult, but radiotherapy is recommended if the tumor is in the upper thorax and neck adjacent to the airway and major vessels (16). After the primary surgery, we preferred surgical resection for the first recurrence. Two patients, Case 1 and Case 4, received radiotherapy after second recurrences. However, tumor recurrences were detected again six months and three years after radiotherapy. Sarcomatoid transition, as with Case 4, and also spontaneous remission, have been reported in cases of DTs with no therapy or after a partial resection (1, 8). A protracted follow-up period is recommended because late recurrences have been reported (17). DTs have a five-year survival rate of nearly 93%, and since they rarely metastasize, death due to DTs has seldom been reported (16, 18). For unresectable cases, therapeutic agents such as estrogen receptor blockers, antifibrinolytic agents, nonsteroidal anti-inflammatory drugs, chemotherapy, tyrosine kinase inhibitors, or a combination of the above, have been applied with some promising outcomes (19). However, despite these successfully sporadic case reports, extensive success has not been reported (3).

Conclusion

Based on the collective data, it appears that standardized management still seems to be deficient, likely due to the varying development of these cases. The number of patients in our study group is low. Our own limited experience has taught us that each patient must be assessed on an individual basis, and if possible, a comprehensive resection with

negative surgical margins remains the mainstay of the treatment. Furthermore, a thorough and prolonged follow-up period may aid in the management of these locally aggressive tumors.

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