Desmopastik Fibroblastoma'nın Seyrek Lokalizasyonu: Göğüs Duvarı

A Rare Localization of Desmoplastic Fibroblastoma: Chest Wall

Desmoplastik Fibroblastoma / Desmoplastic Fibroblastoma

Hüseyin Çakmak[1], Arif Osman Tokat[2], Sezgin Karasu[2], Rüçhan Aygün[3]
[1]Göğüs Cerrahisi Kliniği, Sağlık Bakanlığı Ankara Onkoloji Eğitim ve Araştırma Hastanesi, Ankara, Türkiye.
[2]Göğüs Cerrahisi Kliniği, Sağlık Bakanlığı, Ankara Eğitim ve Araştırma Hastanesi, Ankara, Türkiye.
[3] Patoloji Kliniği, Sağlık Bakanlığı Ankara Onkoloji Eğitim ve Araştırma Hastanesi, Ankara, Türkiye.

Özet

Desmoplastic fibroblastoma is a rare, benign, soft tissue tumor. It predominantly occurs within the subcutaneous tissue or muscle. Desmoplastic fibroblastoma usually appears in various locations, including the upper and lower extremities, posterior neck, upper back, abdominal wall and hip. Herein, we present a 17-year-old male patient who had a desmoplastic fibroblastoma on the chest wall. The diagnosis is histologically verified and differentiated from other soft tissue tumors. The rare localization of this lesion in the chest wall is particularly interesting.

Anahtar Kelimeler

Dezmoplastik Fibroblastoma; Göğüs Duvarı; Yumuşak Doku Tümörü

Abstract

Dezmoplastik Fibroblastoma seyrek görülen, benin bir yumuşak doku tümörüdür. Yaygın olarak subkutan dokuda ve kas içinden kaynaklanır. Dezmoplastik Fibroblastoma genellikle üst ve alt ekstremitelerde, boyun posteriorunda, sırtın üst kısmında, karın duvarında ve kalaçada görülür. Biz burada göğüs duvarında Dezmoplastik Fibroblastoma saptanan 17 yaşında erkek hastayı sunmaktayız. Hastanın tanısı histolojik olarak konmuş ve diğer yumuşak doku tümörlerinden ayrılmıştır. Lezyonun nadir bir lokalizasyonu olan göğüs duvarı yerleşimi özellikle ilginçtir.

Keywords

Desmoplastic Fibroblastoma; Chest Wall; Soft Tissue Tumors

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Corresponding Author: Arif Osman Tokat: Sağlık Bakanlığı, Ankara Eğitim ve Araştırma Hastanesi, Göğüs Cerrahisi Kliniği, Ulucanlar 06340 Ankara, Turkey.
Phone : +903125952281 GSM: +905323160994 Fax: +903123633396 E-mail:aostokat@hotmail.com

Introduction

Desmoplastic fibroblastoma is a rare benign soft tissue tumor. It was first described in 1995 by Evans.[1] The name "collagenous fibroma" was also used in many papers in recent years, but "desmoplastic fibroblastoma" was adopted in World Health Organization classification of soft tissue and bone tumors in 2002.[2]

We present, herein, a case of Desmoplastic fibroblastoma which arises as a mass in the left side of the chest wall extending to the hemithorax via intercostal space.

Case Report

A 17-year-old woman admitted to our clinic with painless and gradually enlarged mass in the left side of the chest wall. The mass was first observed 4 months earlier. No traumatic episode or inciting event was reported by the patient. Her past medical history was unremarkable. Physical examination revealed a 5,5 x 5,5 x 11 cm firm mass located in the left side of chest wall at the anterior axillary line and repose on the 5th and 6th rib. There were no pathological findings of the overlying skin. A computed tomography scan revealed a soft tissue dense mass containing small foci of calcification extending to the intercostal space with no periosteal reaction or bony erosion. The mass density was measured as approximately equal to that of fat tissue (Fig. 1).

In the operative procedure the mass was found in the subcutaneous layer extending between the muscular layers through the intercostal space and settling on the surface of the costal bones. The tumor was totally removed by surgical excision. The resected mass was well-circumscribed and disc-shaped. On cross-section, a pearl gray color and homogenous firm consistency mass without hemorrhage or necrosis was observed. Postoperative course was uneventful and the patient is doing well at postoperative six months.

Histologically, the tumor was well-circumscribed from the muscle tissue, hypocellular and characterized by a proliferation of large, stellate and spindle cells set in a densely collagenous and myxocollagenous stroma. Neither cellular atypia nor mitotic figures were observed. Inflammatory cells such as lymphocytes, plasma cells and foamy histiocytes were absent. There was no tumor necrosis. Perivascular hyalinization was observed in the lesion. Tumor cells stained for CD68 (PGM-1) and vimentin, there was no immunoreactivity for S100, actin, desmin (Fig. 2).

Discussion

Desmoplastic fibroblastoma is a recently defined benign fibroblastic/myofibroblastic tumor.[1,2] Miettien and Flesch published the largest series of 63 cases collagenous fibroma which defined the clinicopathological characteristics of the tumor. It has male dominance of 1:4, and occur between the ages of 16 and 83 years. It can appear in deep sections of the subcutis, in fascia, aponeurotic tissue or skeletal muscles. Desmoplastic fibroblastoma commonly presents as a firm, well circumscribed, painless, slow-growing mobile masses. It usually appears in various locations, including the upper extremities, posterior neck, upper back, lower extremities, abdominal wall and hip. The tumor ranges in size from 1 to 20 cm in maximum dimension.[3,4] Entrapment of the adjacent muscle and fat is common, but aggressive growth is not a feature of desmoplastic fibroblastoma.[1,3,5]

On gross examination the tumor appears oval, disc-shaped or fusiform with a firm, homogenous pearl-gray color consistency. Microscopically, it is composed of medium-sized to large "reactive appearing" spindled to stellate fibroblasts sparsely distributed in a fibromyxoid to densely fibrous background; mitotic figures is very rare or absent, tumor necrosis is not seen, and vascularity is low.[1] Evans, Nielsen and et al. designated the condition as a 'neoplasm' because no inciting event was clinically mentioned, and no specific cause of a reactive fibrous proliferation was microscopically identified.[1,2]

The differential diagnosis for desmoplastic fibroblastoma includes benign, locally aggressive soft tissue tumors, such as desmoid fibromatosis, nodular fasciitis, fibroma of tendon sheath, neurofibroma and myxoma. For cases with skeletal muscle involvement, low-grade fibromyxoid sarcoma and fibromatosis might be entertained.[1-4] The treatment of desmoplastic fibroblastoma is a total surgical excision and no tumor recurrence has been reported.[2]

In our case, all the features of desmoplastic fibroblastoma such as age, gender, symptoms and tumor behavior are similar with the literature, but the localization of the tumor is novel. Less than 100 cases of desmoplastic fibroblastoma have been reported in the English literature. However, there is only one case of desmoplastic fibroblastoma that has reported on the chest wall.[6]

Surgical resection of chest wall tumors, whether primary or

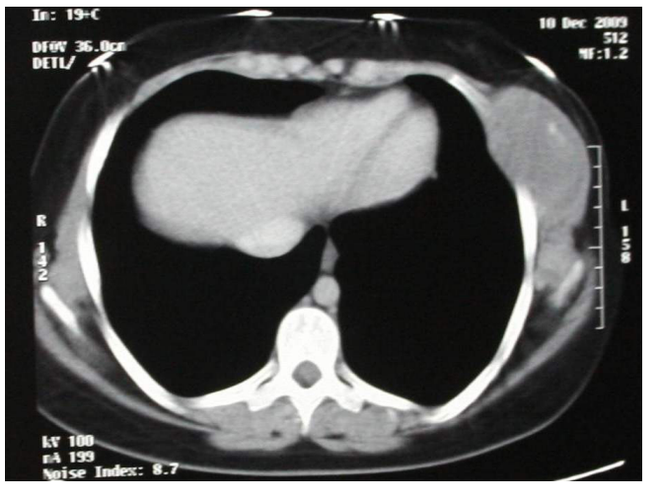


Figure 1. At thorax CT, mass containing small foci of calcification is extending to the intercostal space with no periosteal reaction or bony.

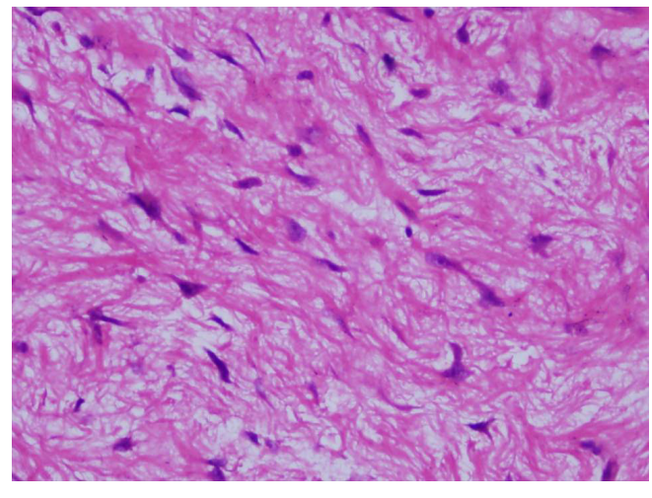


Figure 2: Photomicrogram of the case. Hypocellular and characterized by a proliferation of large, stellate and spindle cells set in a densely collagenous and myxocollagenous stroma. (HE x100)

metastatic, should be considered when the tumor is well demarcated. The general approach is to perform a wide excision of all involved structures, regardless of size. For a primary tumor of the rib, also the ribs immediately above and below, the adjacent muscles and underlying pleura, all should be excised. For tumors of chest wall, to determine the best treatment plan, it is essential to choose a correct diagnostic procedure for the individual patient: fine-needle aspiration, incisional or excisional biopsy, or immediate chest wall resection. In our case, we preferred immediate chest wall resection. In cases of a prior malignancy with a current clinically and radiologically malignant mass, a needle biopsy is enough to confirm the diagnosis of recurrence or metastasis. In general, however, excisional biopsy is preferable to needle or incisional biopsy.[7] In our case; no any findings of distant organ involment at whole body scanning and rapid enlargement of the tumor made us think, it was a primary malign chest wall tumor. Surgical resection was performed without biopsy.
Although a rare entity, desmoplastic fibroblastoma in younger patients may exist with chest wall presentation. During operation, surgeon and pathologist should consider collagenous fibroma as a possible diagnosis: Thereby, inappropriately aggressive surgery is avoided.

Competing interests

The authors declare that they have no competing interests.

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