Current therapies for extra-abdominal fibromatosis of the chest wall: a silent giant mass

Göğüs duvarında ekstra-abdominal fibromatozis için güncel yaklaşımlar: Sessiz, dev bir kitle

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Extra-abdominal fibromatosis (desmoid tumor) is a rare and benign lesion of soft tissue which has a strong tendency for local invasion and recurrence. We present a case of a 44-year-old woman with an asymptomatic, intrathoracic but extrapulmonary giant mass requiring wide surgical resection and reconstruction of the chest wall. The tumor which was greater than 10 cm in diameter, was found incidentally on a routine chest radiograph. The patient underwent wide chest wall resection and reconstruction with a sandwich graft. No recurrence has been observed during two years follow-up. Treatment modalities for recurrent and inoperable desmoid tumor include radiotherapy and estrogen antagonists, however, surgical resection with safety margins as for a malignant tumor is the primary treatment choice.

Key words: Chest wall tumor; desmoid tumor; extra-abdominal fibromatosis.

Ekstra-abdominal fibromatozis (dezmoid tümör), lokal invazyonun ve nüksün sık görüldüğü nadir, iyi huylu ve yumuşak bir doku lezyonudur. Bu yazıda, göğüs kafesi içerisinde fakat akciğer dışında, asemptomatik dev kitle nedeni ile göğüs duvarına geniş cerrahi rezeksiyon ve onarım uygulanan 44 yaşında kadın olgu sunuldu. Çapı 10 cm'den büyük olan tümör, düzenli olarak çekilen akciğer grafisinde rastlantısal olarak saptandı. Hastaya sandviç grefti ile geniş göğüs duvar rezeksiyonu ve onarımı yapıldı. İki yıllık izlem süresince herhangi bir nüks saptanmadı. Nüks gelişen ve inoperabl dezmoid tümörlerde radyoterapi ve östrojen antagonistleri gibi tedavi yöntemleri uygulanmak ile birlikte, malign tümörün güvenli cerrahi sınırlar ile çıkarılması ilk tedavi seçeneği olmalıdır.

Anahtar sözcükler: Göğüs duvarı tümörü; dezmoid tümör; ekstra-abdominal fibromatozis.

Extra-abdominal fibromatoses or desmoid tumors are rare soft tissue neoplasms derived from fascial or musculoaponeurotic structures. These aggressive non-metastasizing tumor-like lesions have a strong tendency to local infiltration, with a recurrence rate of about 40%. Patients with these lesions are often asymptomatic and thus commonly present with giant masses. The treatment for these neoplasm remains margin-negative surgical excision, and given the often large size at presentation, may require extensive chest wall resection.

In this case report, we present a patient with an asymptomatic and non-palpable chest wall tumor that was found incidentally on a routine chest radiograph. Current therapy and new trends for the treatment of desmoid tumors are also discussed.

CASE REPORT

A 44-year-old woman was referred to our clinic for evaluation of a chest radiograph abnormality (Fig. 1). She was a smoker with chronic obstructive pulmonary disease (COPD), who had coughing and dyspnea of two years' duration. She had no past medical or surgical history including soft tissue trauma. No family history of fibrous tumors, colonic polyps, or colorectal cancer was present. Results of her physical examination were unremarkable except for her respiratory findings of COPD; results of her blood laboratory tests were within normal limits. Her latest chest radiograph was taken two-years prior on a routine check-up, and no abnormality was seen. A computed tomography scan revealed a large low-density thoracic wall mass

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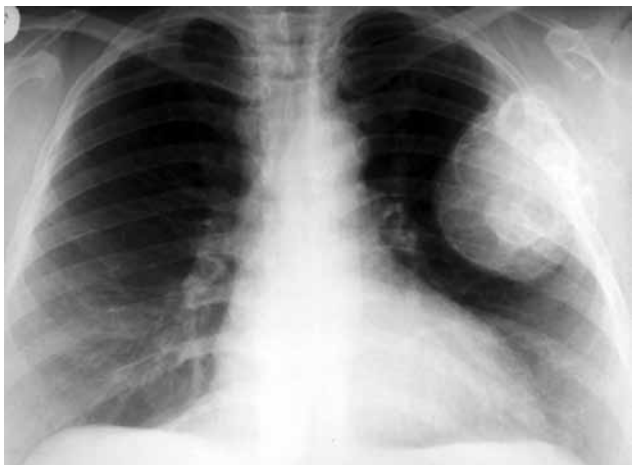

Fig. 1. Chest X-ray is showing a giant mass with lobule contours and calcifications on the left side.

with invasion to the left thoracic wall, protruding into the left pleural cavity (Fig. 2). T1-and T2-weighted magnetic resonance imaging scans demonstrated a high-intensity mass, invading the left thoracic wall. A malignant chest wall tumor was suspected on the basis of these imaging findings. Bone scintigraphy revealed increased uptake on the left 4th and 5th ribs. The patient underwent a left lateral thoracotomy along the 6th intercostal space and resection of the tumor, together with the chest wall, including soft tissue, and the left 3rd through 6th rib with a 2 cm safety margin were performed (Fig. 3). The chest wall deformity of about 15x20 cm was repaired with a prolene mesh-methyl methacrylate sandwich graft. There was no apparent parenchymal invasion. Histopathologic examination revealed the irregular borders of the lesion which infiltrated bone and striated muscle (Fig. 4a). The lesion was hypocellular in broad areas and composed of fibroblastic cells scattered in the collagenous stroma. Fibroblastic cells were arranged in a storiform pattern nearby bone (Fig. 4b, c). There was no necrosis and no marked mitotic activity. There were thick, hyalinized collagen fibers resembling keloid (Fig. 4d). Her post-operative course was uneventful. During 24 months' follow-up, no recurrence has been observed.

DISCUSSION

Desmoids are histologically benign but may behave aggressively at the local level with multiple recurrences being common after complete resection. The shoulder, chest wall, back, and thigh are favorite sites. Chest wall desmoids account for approximately 20% of all patients with desmoid tumors. Trauma, hormones, and heredity have been implicated as etiologic factors. Chest wall desmoids have been described after mastectomy, silicon breast implants, rib fractures, and thoracotomy.[1] Also, desmoid tumors occur with greatly increased frequency in patients with familial adenomatous polyposis. The association of familial polyposis with desmoids and other tissue tumors was originally given the name Gardner's syndrome.[2] In our patient, neither familial polyposis nor trauma history was present as a predisposing etiologic factor.

Intrathoracic desmoids may present an even more difficult diagnostic problem and, depending on location, localized fibrous tumors of the pleura, neurofibromas, ganglioneuromas, fibrosarcomas, fibrous pseudotumors and lung cancers should be considered in the differential diagnosis.[3] Fine needle aspiration may not be sufficient because of the relative hypocellularity of this tumor.[4] Excisional biopsy allows for definitive diagnosis of the chest wall tumors.[5] We did not perform incisional biopsy or needle aspiration biopsy before the operation. There were no distant metastases or additional paranchymal lesion and the tumor was totally resectable.

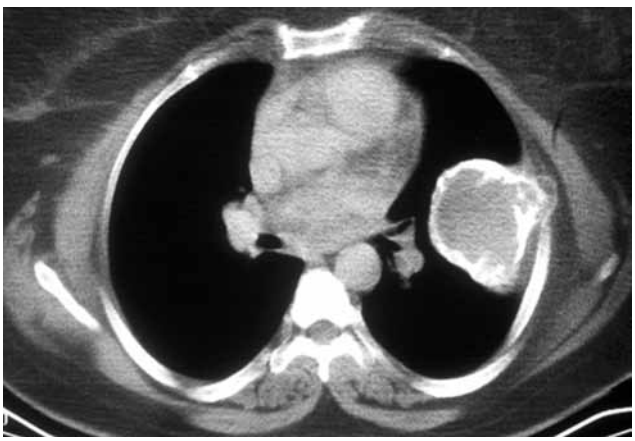

Fig. 2. Thorax computed tomography revealed a chest wall tumor about 11 cm size, protruding into the left pleural cavity without lung invasion.

Fig. 3. Solid gray white variagated cut surface of the mass.

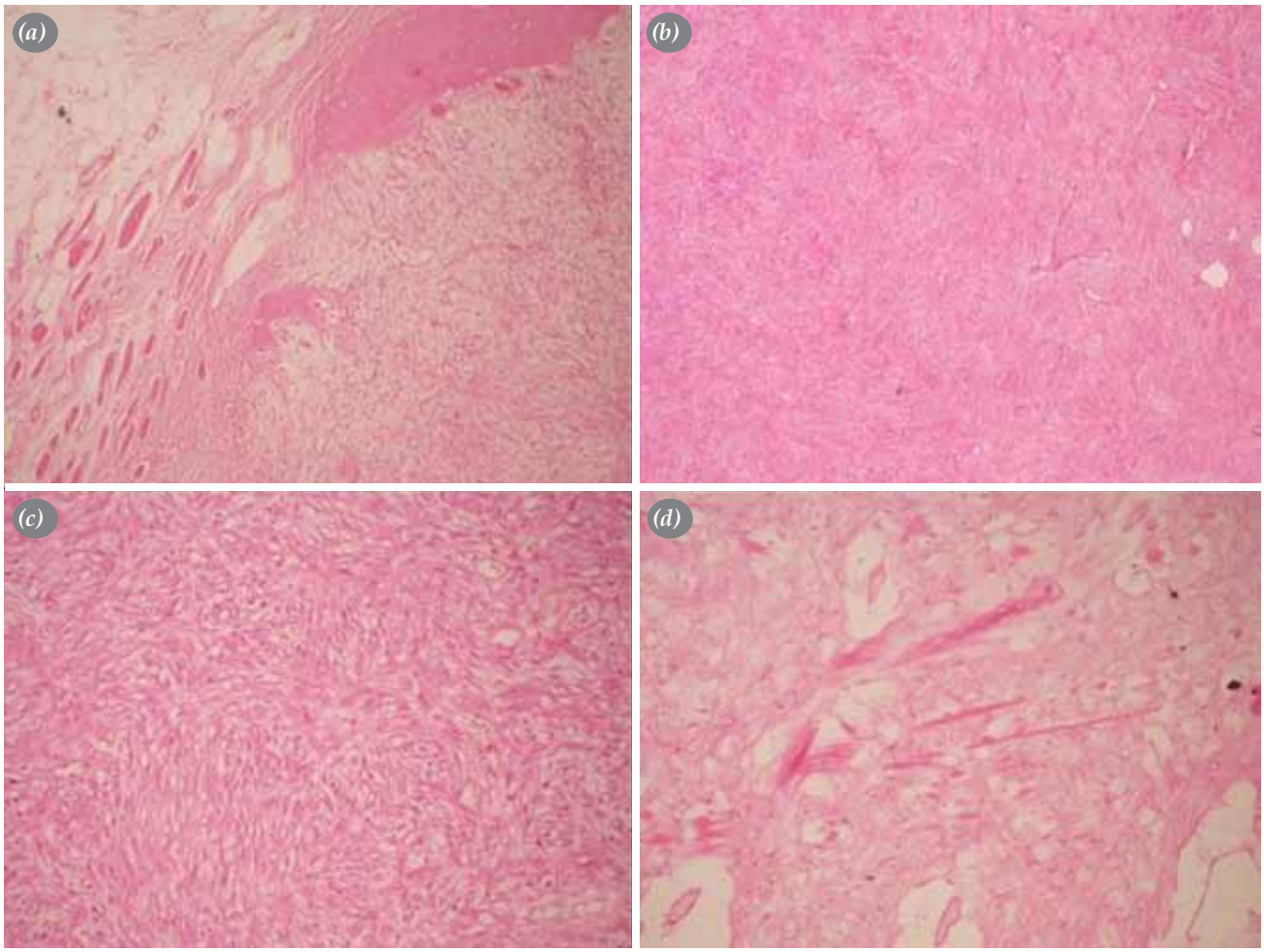

Fig. 4. (a) The lesion was invading bone and striated muscle (H-E x 40), **(b, c)** composed of focal cellular areas with storiform pattern (H-E x 40; H-E x 100 respectively) and **(d)** broad hypocellular areas containing thick hyalinized fibers resembling keloid (H-E x 200).

While it previously has been thought that surgery was the only therapy, nowadays, adjuvant procedures like radiation, hormonal therapy, and chemotherapy are also in practice. Nonsteroidal anti-inflammatory drugs in conjunction with ascorbic acid or tamoxifen have been reported to decrease the growth of some desmoids.[6] In the latest study Deyrup et al.[7] stated that estrogen antagonists may have a role in the treatment of refractory or recurrent extra-abdominal fibromatoses. In young women with recurrent desmoid tumors, tamoxifen has been considered as a choice of treatment.[8] Tamoxifen had also been shown to produce a response in desmoid tumors without estrogen receptors.[9] The mechanism of tamoxifen to desmoid tumors has not been clearly known however these studies showed that tamoxifen has an influence on desmoid tumors.[6-9] Even though these alternative therapies are recommended for unresectable and recurrent desmoids, complete surgical excision still yields the best results.[10] Positive margins at resection, reoperation and postoperative radiation are associated with a high risk of local recurrence.[11] Easter and Halasz[12] claimed that neither adjuvant radiotherapy nor chemotherapy had been shown to reduce the rate of recurrence.

A high rate of recurrence and silent enlargement of these tumors are the main problems for treatment. Further investigations on the relationship between hormonal receptors and desmoid tumor may lead to a new procedure to prevent recurrence and avoid extended surgical interventions. However, wide surgical excision, reconstruction, and differential diagnoses from the other malignancies are essential elements in the current treatment of these rare tumors.

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REFERENCES

1. McKinnon JG, Neifeld JP, Kay S, Parker GA, Foster WC, Lawrence W Jr. Management of desmoid tumors. Surg Gynecol Obstet 1989;169:104-6.
2. Lotfi AM, Dozois RR, Gordon H, Hruska LS, Weiland LH, Carryer PW, et al. Mesenteric fibromatosis complicating familial adenomatous polyposis: predisposing factors and results of treatment. Int J Colorectal Dis 1989;4:30-6.
3. Wilson RW, Gallateau-Salle F, Moran CA. Desmoid tumors of the pleura: a clinicopathologic mimic of localized fibrous tumor. Mod Pathol 1999;12:9-14.
4. Allen PJ, Shriver CD. Desmoid tumors of the chest wall. Semin Thorac Cardiovasc Surg 1999;11:264-9.
5. Pairolero PC. Chest wall tumors. In: Shields TW, LoCicero J 3rd, Ponn RB, editors. General thoracic surgery. 5th ed. Baltimore: Williams & Wilkins; 2000. p. 589-98.
6. Wilcken N, Tattersall MH. Endocrine therapy for desmoid tumors. Cancer 1991;68:1384-8.
7. Deyrup AT, Tretiakova M, Montag AG. Estrogen receptor-beta expression in extraabdominal fibromatoses: an analysis of 40 cases. Cancer 2006;106:208-13.
8. Ohashi T, Shigematsu N, Kameyama K, Kubo A. Tamoxifen for recurrent desmoid tumor of the chest wall. Int J Clin Oncol 2006;11:150-2.
9. Chao AS, Lai CH, Hsueh S, Chen CS, Yang YC, Soong YK. Successful treatment of recurrent pelvic desmoid tumour with tamoxifen: case report. Hum Reprod 2000;15:311-3.
10. Varghese TK Jr, Gupta R, Yeldandi AV, Sundaresan SR. Desmoid tumor of the chest wall with pleural involvement. Ann Thorac Surg 2003;76:937-9.
11. Abbas AE, Deschamps C, Cassivi SD, Nichols FC 3rd, Allen MS, Schleck CD, et al. Chest-wall desmoid tumors: results of surgical intervention. Ann Thorac Surg 2004;78:1219-23.
12. Easter DW, Halasz NA. Recent trends in the management of desmoid tumors. Summary of 19 cases and review of the literature. Ann Surg 1989;210:765-9.