

Re-resection of Giant Aggressive Fibromatosis of the Chest Wall

Göğüs Duvarının Dev Agresif Fibromatozis'inde İkincil Rezeksiyon

Çağatay Tezel, Altan Kır, Cemal Asım Kutlu

Department of Thoracic Surgery, Süreyyapaşa Chest Diseases and Thoracic Surgery Teaching Hospital, İstanbul, Turkey

ABSTRACT

The synonym of Desmoid tumor called "Aggressive Fibromatosis" develops from the muscle or aponeurosis, but rarely in the chest wall. We report a 35 year old patient who underwent complete (R0) resection of a fibromatosis tumor in the chest wall 4 years previously. However, the tumor recurred and re-resection of the previous chest wall reconstruction with almost the half of the upper part of the hemithorax was performed. Adjuvant radiotherapy in the negative margin is undetermined but growing case studies show reduced recurrence in patients who underwent radiotherapy. (*Tur Toraks Der 2012; 13: 71-3*)

Key words: Desmoid tumor, chest wall tumor, aggressive fibromatosis

Received: 03.05.2010

Accepted: 28.06.2011

ÖZET

Desmoid tümör olarak adlandırılan "Agresif Fibromatozis" kas veya aponörozlardan gelişir, nadiren göğüs duvarı kaynaklıdır. Olgumuz 35 yaşındaki hasta, 4 yıl önce göğüs duvarı kaynaklı fibromatosis nedeniyle R0 rezeksiyon uygulanmış bir olgu olup, tümör nüksü nedeniyle hemitoraksın yarısına yakın kısmı rezeze edilmiş ve göğüs duvarı rekonstrüksiyonu uygulanmıştır. Halen cerrahi sınır negatif hastalarda adjuvan radyoterapinin etkinliği kanıtlanmamış olsa da radyoterapi uygulanmış artan olgularda nüks azalmaktadır. (*Tur Toraks Der 2012; 13: 71-3*)

Anahtar sözcükler: Desmoid tümör, göğüs duvarı tümörü, agresif fibromatozis

Geliş Tarihi: 03.05.2010

Kabul Tarihi: 28.06.2011

INTRODUCTION

Desmoid tumor or aggressive fibromatosis often appears as infiltrative, usually well-differentiated, dense fibrous tissue, with a locally aggressive nature [1]. Those tumors are known for their tendency to recur. Extensive resection with a wide surgical margin is essential where the vital structures permit [2,3]. Although the adjuvant radiotherapy is not common after curative resection, the possibility of the results of false positive surgical margin due to the size of the tumor caused us to adopt the notion of adjuvant radiotherapy.

CASE

A 35 year-old man with a chest wall mass was referred for surgical intervention. Previously, he underwent chest wall resection of three ribs including the first rib on the right hemithorax 4 years earlier. At that time, radiological images of the tumor including chest X-ray and thorax magnetic resonance image showed the location of the tumor invading the chest wall (Figure 1). Pathological examination revealed a desmoid tumor of the chest wall that was reconstructed with prolene mesh with negative surgical margins. No additional treatment modality was preferred after the complete excision of the tumor.

From that time, follow-up was incomplete until the patient suffered from pain due the enlarging right mass on the operated side. Chest X-ray revealed a well defined mass on the right hemithorax. Chest tomography scan detected the 11x7x5 cm diameter tumor located on the anterior lower part of the right hemithorax with central necrosis. Additional radio nuclear imaging was done and Positron Emission Tomography disclosed a low grade differentiate tumor with the standard uptake value of 5.8 (Figure 2).

Re-resection of the tumor was planned. Preoperative tests were done. The patient underwent right thoracotomy beginning from the previous thoracotomy scar to the sub-mamarian fold. It was not possible to reach the tumor without cutting the reconstruction material previously placed. The mesh was removed. A near *hemi-thoracotomy* beginning from the rest segment of the 3rd rib to the 7th rib posteriorly, and on the anterior side, down to the 8th costo-sternal junction chest wall was resected enblock. A well-defined capsule, except for some areas of invasion to the anterior segment of the upper lobe, was also stapled with the tumor. A gross defect was reconstructed with polytetrafluoroethylene (PTFE) mesh and methyl metacrylate with the sandwich technique. Before closure of the thoracic cavity, two chest tubes in



Figure 1. A) Computed tomography and magnetic resonance images, B) showing chest wall tumor before the first operation

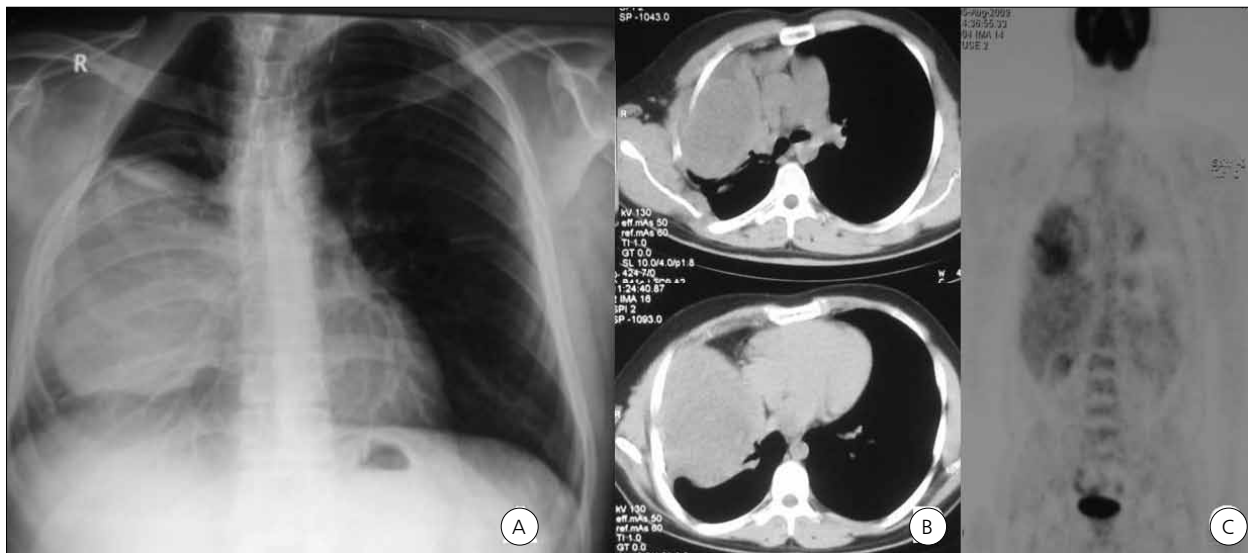


Figure 2. A) Chest X-ray and computed tomography, B) shows tumor recurrence, PET/CT, C) illustrates a probability of a low grade differentiated tumor

the thorax and two hemovac drains between the prosthesis and the muscles were placed to avoid seroma. Prosthetic material showed good performance without complications. With the application of reconstructive techniques the cosmetic result is satisfactory.

The pathological examination of specimen showed a glistening white, coarsely trabeculated cut surface. Histopathologically, the tumor was typically poorly circumscribed with infiltration of the surrounding soft tissue structures. Proliferation of elongated spindle like cells of uniform appearance was common. The cells were lacking in atypia or hyperchromasia. The cells strongly expressed vimentine. Other immunohistochemical results including CD34 were negative, Ki67 proliferation index was 3% and beta katherinin was positive. Those findings revealed the diagnosis of fibromatosis. Most of the cells have the features of fibroblasts. As revealed in the first operation, surgical margins were negative.

The postoperative period was unremarkable. He was discharged on postoperative day 5. Although the mar-

gins were negative, the patient was referred to radiation therapy.

DISCUSSION

Desmoid tumor, or the synonym aggressive fibromatosis, is a rare soft tissue tumor that is a grossly circumscribed lesion of solid fibrous and uniform nature arising from the connective tissue sheaths of striated muscle. Desmoid tumors can arise in any skeletal muscle but mostly develop in the anterior abdominal wall and shoulder girdle [1,2]. The incidence is 0.03% of all neoplasms [4].

Even though the etiology is not clear, predisposing factors are previous trauma and even previous thoracic surgery could be the initiative for tumor development and genetic predispositions like familial adenomatous polyposis and Gardner's syndrome [5]. This tendency for recurrence make the treatment challenging, as described in the present case. Even though the tumors are benign the reported risk of local recurrences is as high as 40-75% [6].

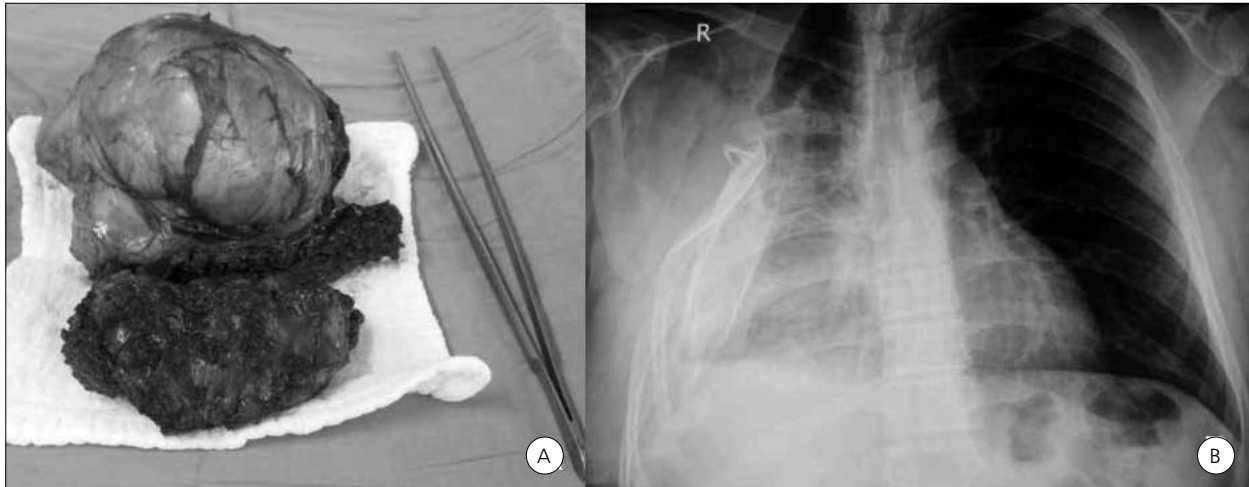


Figure 3. A) Resected specimen with adequate surgical margin, B) Postoperative chest X-ray shows right hemithoracotomy restored with prosthetic material

Radiologically, computed tomography and magnetic resonance imaging are used for the diagnosis and follow-up of desmoid tumors. During follow-up, MRI is superior to CT scan in defining the pattern and extent of involvement as well as in determining whether recurrence has occurred after surgery [7].

The diagnostic test is biopsy of the tumor. On electron microscopic examination, the spindle cells of desmoid tumors appear to be myofibroblasts. This finding is thought to represent an abnormal proliferation of myofibroblasts, which normally disappear gradually during the later stages of wound healing. Immunostaining with vimentin, alpha smooth muscle actin, muscle actin, and desmin are helpful in distinguishing the tumors in the differential diagnosis. As performed during the course of diagnosis, immunohistochemical results of the case including CD34 were negative, Ki67 proliferation index was 3%, and beta katherin was positive.

The prognosis is unexpected. Rock et al. [8] reported that of 194 cases of extra-abdominal desmoid tumors, 132 (68%) recurred an average of 1.4 years after surgery. Although distant spread has not been documented in long-term follow-up studies, these tumors have a strong propensity to recur locally after resection. Although Sherman et al. [9] reported that local recurrence could be controlled satisfactorily in 75-83% of all cases receiving high-dose (50 Gy on average) postoperative radiotherapy, the role of radiotherapy is unclear. Another study referring to the usefulness of radiotherapy from Abbas et al. [6] showed that almost a third of the patients who did not have postoperative radiation therapy had recurrent tumor. The overall 5-year survival rate was 93%, the 5-year local recurrence rate despite aggressive surgical intervention was 29%. The principal of oncological surgery is resection with as wide a surgical margin as adjacent structures such as the brachial plexus, great vessels, and spine allow [3].

A surgical resection should be performed whenever possible if desmoid tumors recur. Reoperation is usually more difficult than the first operation; extensive resection as possible is sometimes troublesome as it may include first operation reconstruction material. However, it is desirable to achieve an adequate surgical margin in order to avoid recurrence. Other treatment options have included external beam radiation therapy, antiestrogen medications, colchicine, nonsteroidal antiinflammatory agents, and conventional tumor chemotherapy [6].

The ideal treatment has not been established yet, and for the present, surgery remains the cornerstone of an effective management. Although adjuvant radiotherapy is still contravened in Ro tumors, reports show radiotherapy is beneficial.

REFERENCES

1. Allen PJ, Shriver CD. Desmoid tumors of the chest wall. *Semin Thorac Cardiovasc Surg* 1999;11:264-9.
2. Shields CJ, Winter DC, Kirwan WO, Redmond HP. Desmoid tumors. *Eur J Surg Oncol* 2001;27:701-6.
3. Pairolero PC, Arnold PG. Chest wall tumors. Experience with 100 consecutive patients. *J Thorac Cardiovasc Surg* 1985;90:367-72.
4. Nuyttens JJ, Rust PF, Thomas CR Jr, Turrisi AT 3rd. Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors: A comparative review of 22 articles. *Cancer* 2000;88:1517-23.
5. Shimizu J, Kawaura Y, Tatsuzaw Y, et al. Desmoid tumor of the chest wall following chest surgery: Report of a case. *Surg Today* 1999;29:945-7.
6. Abbas AE, Deschamps C, Cassivi SD, et al. Chest wall desmoid tumors: results of surgical intervention. *Ann Thorac Surg* 2004;78:1219-23.
7. Casillas J, Sais GJ, Greve JL, et al. Imaging of intra and extra abdominal desmoid tumors. *Radiographics* 1991;11:959-68.
8. Rock MG, Pritchard DJ, Reiman HM, et al. Extra-abdominal desmoid tumors. *J Bone Joint Surg Am* 1984;66:1369-74.
9. Sherman NE, Romsdahl M, Evans H, et al. Desmoid tumors. A 20-year radiotherapy experience. *Int J Radiat Oncol Biol Phys* 1990;19:37-40.