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SURGERY

Desmoid Tumor of Anterior Chest Wall: a Case Report

Said Elsagheer[1,2] · Walid Abu Arab[1,3] · Shehab Alhammadi[4]

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Abstract

Desmoid tumors (DTs) are rare non-malignant tumors of mesenchymal origin. They are comprised of benign fibrous tissue with spindle-shaped cells adjacent to collagen. DTs may show aggressive fibroblastic proliferation. They do not metastasize, but may exert local infiltration with a recurrence tendency. DTs are classified into superficial or deep ones. Moreover, they can be of sporadic or familial types. Aggressive fibromatosis usually occurs in the deep soft tissues. It is characterized by proliferation of fibrous tissue and infiltration of the adjacent structures. The clinical management of DTs is very complex and requires a multidisciplinary approach. We present here a 35-year-old male patient with chest wall aggressive fibromatosis, who was treated with surgery followed by adjuvant radiation therapy. The patient is doing well following 12 months of follow-up without local recurrence. Radical surgical resection is the main treatment for DTs. The adjuvant treatment, either chemotherapy or radiotherapy, can be considered when radical resection could not be achieved and if reoperation for a recurrence is not applicable. Close follow-up is essential in such patients.

Keywords Desmoid · Chest wall tumor · Fibromatosis

Introduction

Desmoid tumors (DTs) are rare slowly growing benign neoplasms that arise from fascial or musculoaponeurotic structures. They are composed of benign fibrous tissue with spindle-shaped cells adjacent to collagen. They may show aggressive fibroblastic proliferation. DTs usually do not exhibit signs of metastasis, but they commonly exert local infiltration with a recurrence tendency. The recurrence rate after surgical excision may reach up to 65% [1, 2].They constitute about 3.5% of fibrous tumors, 0.3% of all solid tumors. Furthermore, they are given several names like desmoids fibroma, desmomas, aggressive fibromatosis, desmoplastic fibroma, and low grade sarcoma [3].

DTs are classified according to their location into superficial (as palmar, plantar, penile, infantile digital fibromatosis) or deep (extra-abdominal, abdominal, and intra-abdominal). Moreover, these tumors can be of sporadic type, usually presenting as extra-abdominal/abdominal wall mass, or familial type, mainly intra-abdominal, and often associated with adenomatous polyposis [1].

Although aggressive fibromatosis is one of the nomenclatures of the DTs, it usually describes an aggressive form of it and usually occurs in the deep soft tissues. It is characterized by proliferation of fibrous tissue and infiltration of adjacent structures. It has a greater tendency to recur locally, especially postoperatively, compared to well-circumscribed DTs [1, 2].

The median age at diagnosis of DTs is around 35 years old and the majority of patients are females [2]. Around 15% of the patients with DTs are related to familial adenomatous polyposis (FAP) [2, 4]. The pathological diagnosis of DTs is uneasy. Moreover, the somatic mutation of CTNNB1 in cases unrelated to FAP appears to be important as an important diagnostic marker [2]. Furthermore, the clinical management of DTs is very complex and requires a multidisciplinary approach due to the unpredictable course of the disease, its heterogeneous nature, and the functional consequences related to it [2, 5].

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✉ Walid Abu Arab
walidabuarab@yahoo.com

[1] Thoracic Surgery Department, King Khalid Hospital, Najran, Saudi Arabia

[2] Cardiothoracic Surgery Department, Suez Canal University, Ismailia, Egypt

[3] Cardiothoracic Surgery Department, University of Alexandria, Alexandria, Egypt

[4] Histopathology Department, King Khalid Hospital, Najran, Saudi Arabia

We present a male patient with chest wall desmoids tumor, who was treated with surgical resection followed by adjuvant radiation therapy, following getting his approval for publication.

Case Presentation

A 34-year-old male patient presented with a complaint of pain located at the left upper anterior wall for 3 months. The pain was dull in nature and has no aggravating or relieving factors. No other complaints were present. Patient sought medical advice and orthopedic surgeon described analgesia and asked for X-ray for the left shoulder (Fig. 1) that could not reveal any radiological abnormality. The patient continued to suffer pain, and then he feels swelling at the left upper anterior chest wall. The general surgeon examined the patient and detected a submuscular swelling that is attached to the thoracic cage muscles. CT chest was asked for; and it revealed a well-defined slightly hypodense soft tissue lesion that was located at the anterior chest wall deep to the left pectoralis muscle at the left infra-clavicular region lying on the anterior ends and left sternocostal joint of the first two left ribs. It was measuring 5.5 × 5.7 × 2.5 cm with a clear thin fat plane separating it from the overlying muscles yet no clear separation between it and the underlying intercostal muscles. No other remarkable findings were reported (Fig. 2a). The patient was referred to the thoracic surgery department for consultation. Following the examination of the patient, an MRI chest was done, and it revealed a mass of 5.9 × 2.7 × 2.8 cm in dimensions. The lesion was a well-defined soft tissue mass seen within the left anterior chest wall under the left pectoralis muscle just lateral to the sternum

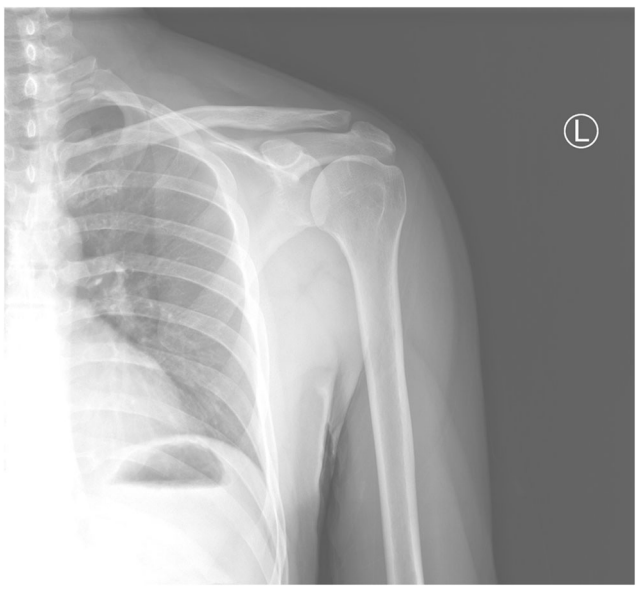

Fig. 1 X-ray of the left shoulder showing no abnormality of the clavicle, scapula, and left shoulder

displaying a hyper-intense signal in the T2-weighted images and iso-intense signal to the muscle in T1-weighted images and shows a homogeneously enhancement. The mass was seen extending between the ribs and abutting the left pleura; however, no obvious enhancement within the pleural membrane appearances is worrisome for a neoplastic process. The mass displaced the pectoral muscle anteriorly with no obvious infiltration to it. There was no evidence of extension of the mass to the underlying rib or lungs or enlarged lymph node within the axilla and in the supraclavicular regions (Fig. 2b).

Following discussion with the patient and his family regarding the available options for getting a biopsy of the lesion; either performing a core needle biopsy or an excisional biopsy with safety margins; they decided to have an excisional biopsy.

Left-sided infra-clavicular skin incision was done. Dissection was taken place up to the level of the left pectoralis muscle. A mass of about 6 × 3 × 3 cm was found infiltrating the pectoralis minor muscle and extending deeply into the intercostals muscles between the first and second ribs. Radical resection of the mass with the first and part of the second rib was decided to be performed. However, the lateral edge of the lesion was approaching the vascular structures and extending beyond the first rib. Consequently, this extension of the lesion has limited the extended resection at the lateral boundary of the lesion. No Chest wall reconstruction was needed. The postoperative period was uneventful, and the patient was discharged home the same day.

The histopathology report revealed an ill-defined lesion composed of proliferating spindle cells forming fascicles interspersed by collagen and thin-walled blood vessels. The cells show elongated, slender, uniform nuclei. No nuclear atypia or necrosis was seen. The proliferating cells infiltrate the adjacent fat, skeletal muscle fibers, and the periosteum of the bone. The cells are positive for SMA, beta catenin, H-caldesmon, and calretinin and negative for CD34 and BCL2. The lesion reaches the inked excision margin at one side (Fig. 3 a and b). The final diagnosis was fibromatosis (desmoid tumor).

The patient was referred to the oncology department, and they advised adjuvant radiotherapy. Follow-up of the patient for the next 12 months show well control with no local recurrence.

Discussion

DTs are rare tumors. Their incidence is around 5–6 per million populations per year. In 8–10% of cases, DTs are located in the chest [6]. When chest involvement is present, they are usually locally aggressive with a high recurrence rate [3]. They are thought to be primary or secondary to trauma, including surgical procedures, familial adenomatous polyposis, and hormonal changes [2, 6]. Our patient had no medical or surgical history and no history of trauma; moreover, he had no

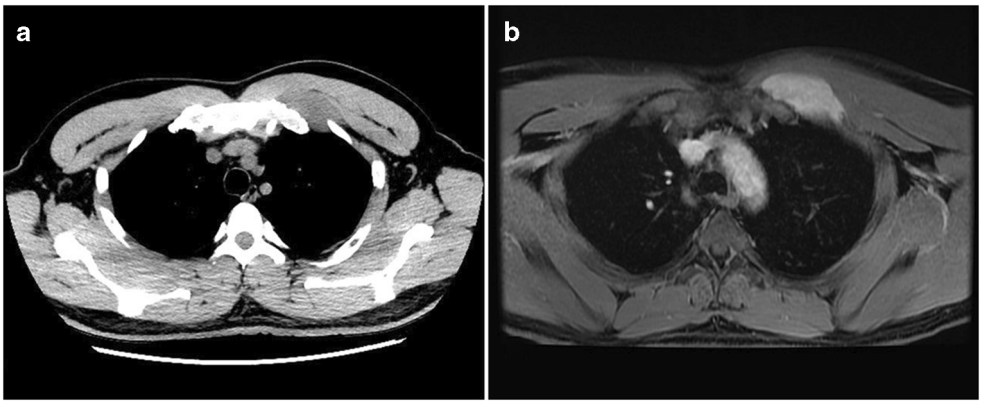

Fig. 2 **a** CT chest (mediastinal window) showing a well-defined slightly hypodense soft tissue lesion (5.5 × 5.7 × 2.5 cm) located at the anterior chest wall deep to the left pectoralis muscle at the left infra-clavicular region lying on the anterior ends and left sternocostal joint of the first two left ribs. **b** MRI chest (T2-weighted image) showing a mass lesion (5.9 × 2.7 × 2.8 cm). The lesion is a well-defined soft tissue mass within the left anterior chest wall under the left pectoralis muscle just lateral to the sternum. The mass seen extending between the ribs and abutting the left pleura

any familial history for similar lesions or familial adenomatous polyposis.

The wide local surgical excision with clear margins is the cornerstone of the treatment [2, 6]. However, it is rather challenging due to the aggressive nature of the tumor. The complete excision might also be difficult due to the critical structures that may be adjacent to the tumor. This limitation was encountered in our patient as the lesion was approaching near the vascular structures at the thoracic outlet at the left side. Although resection margins were macroscopically free, the microscopic margins were proved to be positive on histopathological examination (R1).

In a study published by Brodsky et al. [7], they reported that the positivity or negativity of the resection margins either macroscopically or microscopically can be a determinant factor for the local recurrence. Moreover, the surgical resection should be performed again whenever possible if DTs recur. However, the reoperation is usually more difficult than the first one. Hence, it is desirable to carry out the first resection as extensively as possible and to include an adequate surgical margin in order to avoid the possible recurrence [8].

Some authors have advised for the postoperative radiotherapy for preventing the recurrence. Sherman et al. [9] documented that the local recurrence can be satisfactorily controlled in up to 83% of all cases receiving high-dose postoperative radiotherapy (average 50 Gy). In addition, the effectiveness of radiotherapy in the treatment of the recurrent DTs has also been reported by others [8, 10].

Furthermore, the adjuvant therapy should be considered for positive margins with close follow-up in the view of high recurrence rates [2, 5, 6]. Our patient received adjuvant radiotherapy and followed up for 12 months with no local recurrence. The interesting fact in this patient is that there was no local recurrence during the 12 months of follow-up even with positive resection margin microscopically (R1).

Conclusion

In conclusion, DTs are rare tumors of the chest wall. Radical resection is the main treatment for them. The adjuvant treatment should be considered when radical resection could not be achieved and if reoperation for a recurrence is not applicable. Those patients should be closely followed-up due to the high rate of local recurrence.

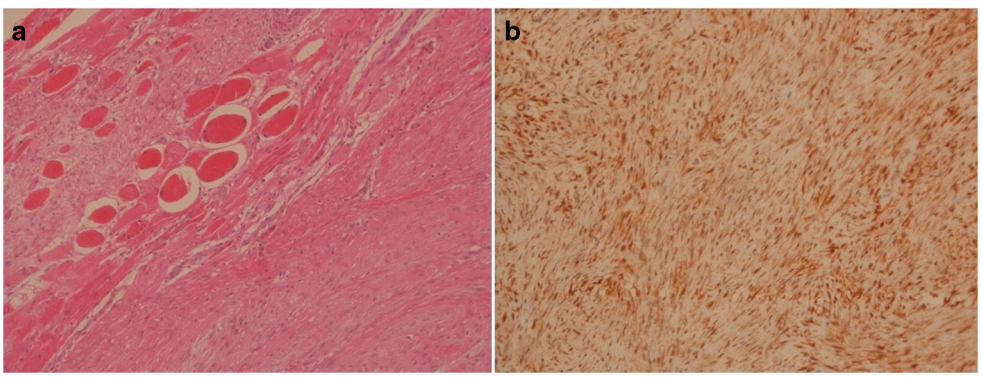

Fig. 3 **a** Hematoxylin and Eosin (H&E) biopsy specimen × 40: showing proliferating spindle cells with oval to elongated nuclei interspersed by collagen and thin-walled blood vessels. No nuclear atypia or necrosis was seen. **b** Immunohistochemistry for beta catenin showed positivity

Authors' Contributions All authors contributed equally in the manuscript.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Code Availability Not applicable.

Ethics Approval Not applicable.

Consent to Participate Not applicable.

Consent for Publication All authors have reviewed the manuscript and have approved submission for publication.

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