Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Desmoid Tumor of Anterior Chest Wall: A Case Report

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DOI: 10.36347/sjmcr.2024.v12i07.013 | **Received:** 06.06.2024 | **Accepted:** 10.07.2024 | **Published:** 13.07.2024

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Abstract **Case Report**

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 8-year-old male child 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]. We present a male patient with chest wall desmoids tumor, who was treated with surgical resection followed by adjuvant radiation therapy.

CASE PRESENTATION

An 8-year-old male patient presented with a substantial mass located on the right side of the anterior chest wall. An ultrasound was done, the child's family were informed that the swelling would regress with time. But later, the family noticed that this mass was increasing in size gradually.

Physical examination unveiled a painless, solid mass characterized by hardness, emerging from the right

lower chest. This mass exhibited adherence to its surroundings, with no apparent pathological vasculature. After this examination, a chest MRI was conducted, revealing a well-defined, and heterogeneous parietal solidocystic mass of (30x66x70 mm) in front of the anterior arches of the 7th and 8th right ribs displaying a hyper-intense signal in the T2-weighted images and iso-intense signal to the muscle in T1-weighted images and shows a discrete enhancement. This mass develops intra-abdominally, responsible for scalopping on segment 4 of the libver and arises in intimate contact with the lesser gastric curvature medially, the head of the pancreas posteriorly, and the antropyloric region inferiorly. (fig 1.2)

Subsequently, the decision was made to proceed with surgical intervention. The patient underwent a chest wall mass resection, 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, Hcaldesmon, and calretinin and negative for CD34 and BCL2. 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

Desmoid fibromatosis (DF) is a locally aggressive connective tissue tumor that appears in musculoaponeurotic tissues. It is also known as aggressive fibromatosis, deep fibromatosis, and desmoid tumor [6]. The most typical age range for the onset of DF is between 15 and 60, and females are more likely to experience it [7]. There is a dearth of information about desmoid tumors in children [8]. They account for 0.03% of all childhood neoplasms. Although the cause is uncertain [8], some well-known disorders, such as familial adenomatous polyposis (FAP), can put patients at risk for developing desmoid tumors. Additionally, localized trauma accounts for 25% of known cases in this site [6]. Also, prior surgery and excessive estrogen exposure may be linked to desmoids [9]. These tumors cannot spread by metastasis, they are characterized by slow, gradual growth, local invasion, and local recurrence after surgical excision. Recurrence remains a problem following resection of desmoid tumors with as many as 50% of patients experiencing a recurrence within 5 years, factors associated with recurrence included age, tumor location, and margin status [9].

DF may affect any region but is most frequently found in the extremities, abdominal wall, and abdominal mesentery. There haven't been many specialized studies on tumors of the chest wall. It is a very rare site for these tumors. The presence of a tumor was also found to be independently related to recurrence-free survival. Particularly, the risk of recurrence was increased for extra-abdominal tumors [9].

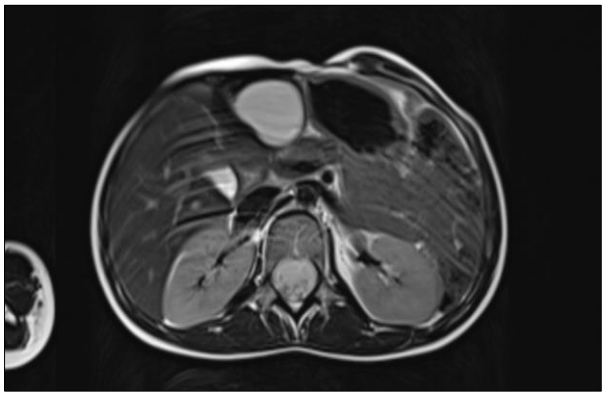

Figure 1: MRI chest (T2-weighted image) showing a mass lesion (3Ox66x70mm). The lesion is a well-defined soft tissue mass within the lower right chest wall with intra abdominal extention.

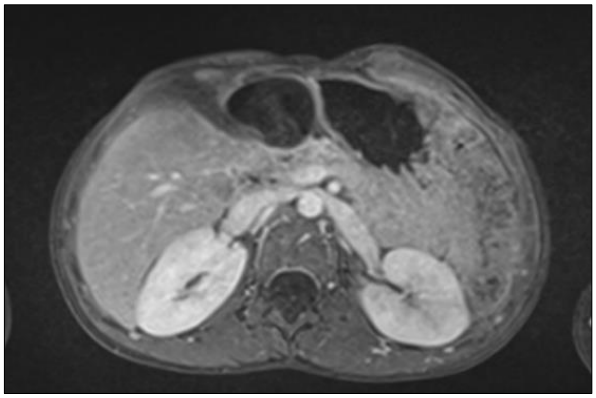

Figure 2: MRI chest (T1 C+ (Gd)) showing a mass lesion (3Ox66x70mm). The lesion is a well-defined soft tissue mass within theower right chest wall discreetly enhanced after gadolinium injection with intra abdominal extention

To diagnose, assess therapy effectiveness, and monitor these tumors, multimodal imaging techniques like ultrasonography, CT, and MR are helpful. A particular diagnosis of benign chest wall lesions typically requires histological specimens, which can be successfully sam- pled under CT or ultrasound guidance.

Management of extra-abdominal desmoid tumors has been treated with surgery, radiation therapy, and chemotherapy. While surgical resection remains central to the management of patients with desmoid tumors, the high rate of recurrence highlights the need for more effective adjuvant therapies [9], The reason for

this is that complete removal of the tumor is uncommon [8].

CONCLUSION

DF should be considered in the differential diagnosis of a chest wall mass and a history of mild trauma in a male- child patient. The goal of its treatment is complete tumor excision and the avoidance of complications. Postoperative chemo- radiotherapy may be replaced with routine postoperative follow- up, and surgical removal of the mass is a wise option if it is possible to do so. This case report aims to shed light on this rare entity and alert physicians not to oversee these tumors in atypical cases.

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