

# Chest Wall Desmoid Tumor Mimicking a Pancoast – Tobias Tumor

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### Abstract

Desmoid tumors of the chest are rare and invasive tumors. We report the case of a 66-year-old patient operated through a Cormier Dartevelle Grunewald incision. After resection of the first two ribs to improve exposition, we proceeded to a careful control of the proximal and distal extremities of the sub-clavian artery that was invaded by the tumor. A termino-terminal anastomosis was performed after removal of the invaded segment. One-piece resection was not possible because of the size of the mass. The resection of the intra-foraminal extension was performed with an intra-thoracic approach by a neurosurgeon. The postoperative course was uneventful.

Keywords: Desmoid tumor; Chest wall; Sub-clavian artery; Surgery

## Introduction

Desmoid tumors of the chest are rare and represent borderline tumors. They are characterized by a tendency to local invasion and a high rate of recurrence. Wide resection is the best therapeutic option, and obtaining negative margins is a challenge in some complex presentations.

We report the case of an operated desmoid tumor of the chest wall localized in the left apex that clinically mimics a Pancoast-Tobias tumor.

#### **Case Report**

A 66-year-old man with the history of a road accident 13 years ago causing a left leg fracture without a thoracic trauma, complained from left cervical and shoulder pain since 7 months. He first consulted an orthopedist and it was believed that the pain was related to cervical radiculopathy, but there was no improvement under symptomatic treatment. In the meanwhile, the man developed a ptosis and a myosis of the left eye (Figure 1). By the second medical consultation, a chest roentgenogram showed a left apical mass.

The computed tomography scan showed a well limited mass on the postero-superior mediastinum measuring 12 cm  $\times$  8.5 cm  $\times$  7.5 cm that compress the trachea, the esophagus and the other mediastinal components. There was osteolysis of the posterior arc of the first and



Figure 1: Left Ptosis and myosis.

the second left ribs and of the transvers processes of D2. The subclavian vessels seemed to be invaded by the tumor (Figure 2).

Magnetic resonance imaging of the neck and thorax revealed a soft tissue capsulated lesion in the para-vertebral zone that invades the intervertebral foramen.

A trans-parietal biopsy by a fine needle was performed, and concluded to a desmoid tumor. The decision to make a surgical resection



Figure 2: CT showing the tumor involving the left sub-clavian artery (arrow).

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was collective and it was encouraged by the good performance state of the patient.

The intervention was performed on collaboration with neurosurgeons and vascular surgeons. The patient was intubated with a double lumen tube, and was positioned supine with the neck hyperextended and the head turned away from the tumor. We decided to proceed via a Cormier Dartevelle Grunewald incision. We first resected the first two ribs to improve exposition. The brachial plexus was identified and reclined away from the tumor. Then we proceeded to a careful control of the proximal and distal extremities of the subclavian artery, which was just sheathed by the tumor.

A termino-terminal anastomosis was necessary to perform because of a located invasion of the vessel (Figure 3). One-piece resection was not possible due to the size of the mass. So, the resection of the intraforaminal extension was done with an intra-thoracic approach by the neurosurgeon. At the end of the operation, the incision was closed respecting the anatomical layers.

The evolution was uneventful. There were no shoulder instability or muscle weakness. The patient was seen 30 days after surgery and report a slight improvement of the ptosis (Figure 4).

An adjuvant radiotherapy was indicated to prevent a local recurrence.

## Discussion

Desmoid tumors are neoformations that belong to the group of soft tissue sarcomas. They developed from fasciae, aponeuroses and striate muscles, and they are known with the name of "sarcoma with low grade malignancy" [1,2]. Macroscopically, they are firm and rubbery with a tendency to local invasion [3]. They are rare and represent 0.3% to 0.5% of all solid tumors.

Often found on the abdomen, the chest wall localization is



Figure 3: Per operative identification and control of different structures.



Figure 4: Post-operative scar.

exceptional and represent 8% to 10% of all cases. Defects in connective tissue represent the etiological basis in the development of this kind of tumors [4]. Mechanisms remain unclear and different hypothesis were proposed to be involved like traumatisms, genetic mutations and hormonal effects. In effects, approximately 2% of all desmoid tumors are associated with familial adenomatous polyposis. This association is a result of germ-line mutations of the adenomatous polyposis coli gene [4]. A stabilization of b-catenin protein is found in sporadic cases, and it represents the result of somatic mutation in either b-catenin or APC gene [4]. The history of traumatism is found in about 25% of all cited cases, and sex hormone seems to take a part in the development of these tumors.

Desmoid tumors can be found in all ages, and cases were reported from 11 to 74 years old patients. These tumors can reach big sizes, up to 210 mm [4], and symptoms are mainly the result of compressing neighboring organs like trachea, esophagus, big vessels of the mediastinum, and other structures. Radiological exploration by a computed tomography or magnetic resonance imaging allows to define limits of the mass, and to judge the possibility of surgical resection.

Pre-operative biopsies are performed when the tumor invade vital structures and the proposed procedure is enough complex to attempt reconstruction after surgical resection [1,3]. Wide radical resection of the mass and involved structures is the mainstream treatment to achieve R0 surgical limits and surgical margins of 4 cm are optimal.

Recurrence rate is high when resection is incomplete and it ranges from 25% to 75% of cases [2-4]. In the work published by Abbas et al. [3], the positive margins were a significant factor of local recurrence (p<0.0001) as well as reoperation (p=0.0199) and the absence of postoperative radiation therapy (p=0.0027). Abrao et al. [1] found in his work that the mean disease-free interval was 41 months (8 to 101 months with a standard deviation of 43.09 months).

One-bloc resection is a challenge for the surgeon especially for big tumors, those that involve different structures and those whose access is difficult [3], like in our case.

The choice of the best surgical approach is important to ensure the most radical resection. In our case, the exposition was good through the Cormier Dartevelle Grunewald incision, and the control of the brachial plexus was easy to do.

The post-operative course was uneventful especially for the shoulder because the clavicle and its articulation with the manubrium were conserved. Similar case was described in the literature, and surgeons used the Dartevelle approach [2]. On the post-operative course, they noted a weakness of the considered arm and physiotherapy with an arm sling were necessary for recovery.

The role of adjuvant therapy is controversial. Radiotherapy is an option proposed to reduce the incidence of local recurrence when surgical margins are positive, and even with negative ones [3]. A total dose of 60 Gy over six weeks seems to be well tolerated with good results [2]. Radiation might be indicated also in patient with unresectable tumors, in local recurrences or as a palliative treatment [3,4].

In addition to radiation, others options might be used as an adjuvant treatment like anti-estrogen medications, colchicine, non-steroidal anti-inflammatory agents, and conventional tumor chemotherapy [3]. Tamoxifen, a selective estrogen receptor modulator, has yielded the widest therapeutic experience. Response was noticed in 51% of cases in a meta-analysis done by Serpell et al. [5]. Indometacin and sulindac are the non-steroidal anti-inflammatory agents of choice in the pharmacological treatment of desmoids [4].

An association of tamoxifen with non-steroidal anti-inflammatory

drugs was associated with 70% of response as it was reported by Waddel et al. [6]. Combination of high dose of tamoxifen (120 mg/day) with sulindac (300 mg/day) was described too as an adjuvant treatment for desmoid tumors [4].

Concerning chemotherapy, unsatisfactory results were reported by different authors and the hypocellular feature of this type of tumors and the low mitotic indices can be an explication. Imatinib mesylate has been reported to be active in desmoid tumor [4]. All these adjuvant options are still controversial.

#### Conclusion

Desmoid tumors of the chest wall are rare. Wide resection is a challenge to achieve in some complex cases. Positive margins and reoperation are associated with high rate of recurrence. Adjuvant treatment, especially radiation therapy, is often indicated to reduce this risk.

#### References

- Abrao FC, Waisberg DR, Fernandez A, Bernardo WM, Pêgo-Fernandes PM, et al. (2011) Desmoid tumors of the chest wall: surgical challenges and possible risk factors. Clinics (Sao Paulo) 66: 705-708.
- Franco S, Andrea DA, Asadi N, Nicola C, Guido Ca, et al. (2013) Desmoid Tumour of the Thoracic Outlet in a 70 Year-old Man Successfully Removed Through Cervico-thoracic Dartevelle Approach. Heart Lung Circ 22: 224-228.
- Abbas AE, Deschamps C, Cassivi SD, Nichols FC, Allen MS, et al. (2004) Chest-wall desmoid tumors: results of surgical intervention. Ann Thorac Surg 78: 1219-1223.
- Mátrai Z, Tóth L, Szentirmay Z, Vámos FR, Klepetko W, et al. (2011) Sporadic desmoid tumors of the chest: long-term follow-up of 28 multimodally treated patients. Eur J Cardiothorac Surg 40: 1170-1176.
- Serpell JW, Paddle-Ledinek JE, Johnson WR (1996) Modification of growth of desmoid tumours in tissue culture by anti-oestrogenic substance: a preliminary report. Aust N Z J Surg 66: 457-463.
- Waddel WR, Gerner RE, Reich MP (1983) Nonsteroid antiinflammatory drugs and tamoxifen for desmoid tumors and carcinoma of the stomach. J Surg Oncol 22: 197-211.