Desmoid-type chest wall fibromatosis. A six cases series

A. Zehani-Kassar\textsuperscript{a,\ast}, A. Ayadi-Kaddour\textsuperscript{a}, A. Marghli\textsuperscript{b}, I. Ridenec\textsuperscript{c}, H. Daghfous\textsuperscript{d}, T. Kilani\textsuperscript{b}, F. El Meznia\textsuperscript{a}

\textsuperscript{a} Anatomy and Pathologic Cytology Department, Abderrahman Mami Hospital, Ariana, Tunisia
\textsuperscript{b} Thoracic Surgery Department, Abderrahman Mami Hospital, Ariana, Tunisia
\textsuperscript{c} Radiology Department, Abderrahman Mami Hospital, Ariana, Tunisia
\textsuperscript{d} Pulmonary Diseases Department, C Pavilion, Abderrahman Mami Hospital, Ariana, Tunisia

Accepted: 6 September 2010

Summary

\textit{Purpose of the study:} Desmoid-type fibromatosis is a fibroblastic proliferation that develops in the deep soft tissues and is characterized by infiltrative growth and a tendency towards local recurrence but does not metastasize. Fibromatosis of the chest wall is rare, representing only 10 to 20\% of all deep fibromatoses.

\textit{Case report:} We report a six cases series of fibromatosis of the chest wall treated at our institution between 1996 and 2009. There were five men and one woman with a mean age of 42.2. Swelling of the chest wall was the most frequent symptom. Imaging showed an expansive homogeneous mass, invading the rib (four cases) or the sternum (two cases). Surgical resection was performed in all cases. The outcome was favorable (five cases) or marked by local recurrence (one case).

\textit{Discussion:} Although it is considered to be a benign lesion, this tumor can be locally very aggressive, and has a high local recurrence rate depending upon the efficacy of surgical resection.

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Introduction

Desmoid-type fibromatoses of the deep tissue are rare soft tissue tumors characterized by proliferation of fibroblast and myofibroblast-type spindle cells, which infiltrate musculoaponeurotic tissue. They represent 3.5\% of fibrous tumors and 0.03\% of all neoplasms [1]. Although they are considered to be benign tumors, they can be very aggressive locally with a high risk of local recurrence after surgical excision, but do not metastasize [2]. Although the etiology of these tumors is unknown, certain factors play a role in their development and growth. There is no generally accepted protocol for treatment. Nevertheless extremely thorough surgical resection is essential. Radiotherapy can be beneficial in case...
of residual or inoperable tumors. These tumors are usually found in the abdomen. [3]. Fibromatoses of the chest wall are rare and only represent 10 to 20% of all fibromatoses [1,4].

We report a series of six cases of fibromatoses of the chest wall treated by surgical resection and discuss the anatomo-clinical features of this tumor as well as the main differential diagnoses.

Observations

This is a retrospective study of six cases of fibromatosis of the chest wall that were surgically treated at our institution between January 1, 1996 and December 31, 2009 (Table 1). All patients underwent a complete clinical examination, chest X-ray and CT Scan. Magnetic resonance imaging (MRI) was only performed in one patient. Surgical samples were obtained of all tumors. The age and sex of patients, the tumor location and symptoms as well as the radiological features, final histology, treatment and outcome were recorded. The results are presented in Table 1. Samples were fixed in formalin then embedded in paraffin. Tissue samples were cut into four-micron pieces stained with hematoxyline-eosine and examined on conventional optic microscopy. An immunohistochemical study was performed in all cases. A panel of antibodies was used associating alpha smooth muscle actin, desmin, PS100, CD34, EMA, cytokeratin and vimentin. The streptavidin biotin peroxidase complex immunohistochemical technique was used.

Age/gender

The mean age of patients was 42.2 years old with a range of 15 to 67. Half of the patient population was under 40 (three cases). There were five men and one woman (sex-ratio 5).

Medical History

One patient had a history of chest trauma, one year before. Two patients had a history of surgery: coronary artery bypass surgery 7 years before in one patient and a pulmonary hydatid cyst 18 months before the diagnosis of the fibromatosis in the other.

Clinical picture

A progressively expanding swelling in the chest wall was the reason for consulting in all patients. Swelling was isolated in three of these patients. Symptoms of neurovascular compression such as pain and paresthesia were found in one patient. Respiratory symptoms including lower chest pain, coughing and dyspnea were found in one patient. Two patients were in poor general condition. On clinical examination, the swelling in the chest was usually firm, poorly circumscribed, non inflammatory, attached to the deep tissues and sometimes painful. The location was the anterior lower thorax (two cases), parasternal (two cases), pectoral (one case) or dorsal (one case).

Additional tests

Biological tests were normal in all patients. Chest X-ray showed an opaque image eroding the adjacent bone tissue in five cases. It was normal in the 6th case. Chest CT scan showed a homogenous, enlarging mass with lack of enhancement after contrast injection, which was invading the soft tissue but left the pulmonary parenchyma intact. The tumor had invaded the ribs in five cases (Figs. 1 and 2), including one case with invasion of the sternum (Fig. 3). Invasion of the sternum alone was noted in one case. Chest MRI was indicated in one patient to evaluate tumor invasion in neighboring structures, in particular vascular structures. It showed a highly vascularized tumor without vascular invasion. Because of the radiological features showing tumor aggressivity a malignant sarcoma-type tumor was highly suspected. A transparietal biopsy was performed before surgical resection in three cases, suggesting the diagnosis in two cases.

Treatment

All cases were treated surgically. Wide resection with a tumor free margin (> 2 cm) was performed. It was complete in all cases. Resection was extended to include the sternum in two cases and the ribs in five cases. Between one and four ribs were resected. The post-operative course was simple in all cases.

Histological study

Macroscopic examination showed a yellowish or pink tumor, which was often poorly circumscribed, firm and fasciculated when cut. The size varied from 7 to 11.5 cm, the mean size was 8.75 cm. Histological examination showed a tumor with a proliferation of fibroblastic and myofibroblastic-type spindle cells arranged in parallel bundles, with variable cellularity, in the presence of mixoid and edematous areas, coexisting with areas of higher cellular density (Fig. 4). These tumor bundles were intersected by bands of hyalinized collagen. The poorly circumscribed tumor had invaded the soft tissues. On immunohistochemical study the tumor cells were positive for alpha smooth muscle actin and vimentin (Fig. 4). The histological study also confirmed tumoral invasion of the rib or sternum in all cases (Fig. 5). In two cases in which resection was considered to be complete during surgery, microscopic examination revealed tumoral segments or adhesions (cases no. 2 and 5).

Outcome, progression

Patients were followed up for between 8 months and 6 years with a mean follow-up of 3 years. The outcome was favorable in five cases and resulted in recurrence in one case in which resection was shown to be incomplete. In this patient recurrence was diagnosed at 2 years of follow-up revealed by a voluminous tumor of 11 × 7 cm, which reached the 5th and 6th rib and invaded the right lower lobe of the lung. Surgical resection included a tumorectomy, resection of the 5th and 6th ribs as well as of the posterior arches of the 7th rib.
<table>
<thead>
<tr>
<th>Case</th>
<th>Sex/age (years)</th>
<th>Location</th>
<th>History</th>
<th>Clinical</th>
<th>Imaging</th>
<th>Size (cm)</th>
<th>Surgery</th>
<th>Follow-up</th>
<th>Progression</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M/32</td>
<td>7th, 8th, 9th and 10th left ribs</td>
<td>NA</td>
<td>Left anterior lower chest</td>
<td>Well-circumscribed homogenous tumor no contrast enhancement no visceral invasion</td>
<td>10</td>
<td>Complete</td>
<td>5 years</td>
<td>Stable</td>
</tr>
<tr>
<td>2</td>
<td>W/55</td>
<td>8th and 9th right ribs</td>
<td>Chest trauma</td>
<td>Solid attached poorly circumscribed tumor of the right lower chest. Lower chest pain dyspnea cough PGC</td>
<td>Rib lysis with opacity of the external parietal pleura and endo thoracic extension into the soft tissues</td>
<td>10</td>
<td>Complete</td>
<td>6 years</td>
<td>2 cases of recurrence</td>
</tr>
<tr>
<td>3</td>
<td>M/67</td>
<td>Sternum, 2nd and 3rd ribs</td>
<td>Coronary artery bypass</td>
<td>Hard parasternal tumor of the chest attached to the chondrosternal joint of the 2nd rib. Dysphonic-PGC</td>
<td>Aggressive tumor invading the chest wall and the anterior mediastinum, in contact with the ascending aorta</td>
<td>7</td>
<td>Complete</td>
<td>18 months</td>
<td>Stable</td>
</tr>
<tr>
<td>4</td>
<td>M/45</td>
<td>2nd right rib</td>
<td>NA</td>
<td>Tumor in the right pectoral region. Painful-neurovascular symptoms (paresthesia)</td>
<td>Tumor expanding into the ribs with areas of peripheral osteocondensation and cortical narrowing with no signs of muscular or pulmonary invasion</td>
<td>7</td>
<td>Complete</td>
<td>4 years</td>
<td>Stable</td>
</tr>
<tr>
<td>5</td>
<td>M/15</td>
<td>Sternum soft tissues</td>
<td>NA</td>
<td>Tumor of the inferior end of the sternum</td>
<td>Vascularized mass of tissue centered upon the xyphoid apophysis invading the large right muscle of the sternal bone</td>
<td>7</td>
<td>Complete</td>
<td>2 years</td>
<td>Stable</td>
</tr>
<tr>
<td>6</td>
<td>M/39</td>
<td>5th rib</td>
<td>Surgery for a hydatic cyst of the lower left lobe</td>
<td>Dorsal tumor on a scar from a posterior thoracotomy</td>
<td>Deep parietal submuscular tumor across from the surgical site slight peripheral enhancement with contrast medium</td>
<td>11.5</td>
<td>Complete</td>
<td>8 months</td>
<td>Stable</td>
</tr>
</tbody>
</table>

M: man; W: woman; NA: not applicable; PGC: poor general condition.
Figure 1  CT Scan of the chest with a mediastinal window with contrast injection (case no. 1): tumor of the chest wall, well circumscribed, homogenous, lacking enhancement after contrast injection with lysis of the arc of the 7th and 8th left ribs.

Figure 2  CT Scan of the chest with a mediastinal window after contrast injection (case no. 6): posterior chest wall tumor (arrow) well circumscribed, dense tissue, homogenous, with no enhancement, without lysis of the adjacent rib.

Discussion

Desmoid-type tumors, which were previously called deep fibromatoses, were first described in the abdominal wall by Mac Farlane in 1832 [1]. Extra-abdominal types were described later, in particular in the limbs and the scapular belt [5]. Tumors in the chest wall have only been reported in 10 to 20% of cases, while intrathoracic locations are rare. [1,4]. These extra-abdominal types, which are more generally called aggressive fibromatoses, are generally distinguished by their deep location, and slow progressive growth, without metastases but with a high risk of local invasion and post-operative recurrence [5]. These tumors develop on the conjunctive tissue, fascias, aponeuroses or
intramuscular compartments [6]. Although the pathogenic mechanism of these tumors has not been clearly elucidated, different factors seem to play a role. The role of hormones has been suggested, with the influence of estrogens and their antagonists such as tamoxifen, on tumor growth [6, 7].

Certain traumas also seem to play a role in the development of these tumors. Indeed, they can develop in contact with breast implants on the muscles transposed for reconstruction, on surgical thoacotomy scars or even after simple intramuscular injections. [7, 8]. In our series two cases of fibromatosis developed on a surgical thoacotomy scar and in one case it was post-traumatic. The delay between chest surgery and the development of the tumor was 18 months and 7 years respectively. In 15% of cases, fibromatosis was associated with familial polyadenomatosis and was a sign of Garnder syndrome [7]. Certain authors have identified a deletion of the long arm of chromosome 5 in case of association of desmoid tumor-Gardner syndrome [6].

In a series of 53 cases of fibromatosis of the chest wall reported by Abbas et al., the mean age at diagnosis was 39 with a range between 10 and 78. There were slightly more women than men in most studies [9, 10]. Fibromatosis of the chest wall usually presents in the form of a tumor of various sizes but which is often large. In the literature, the size of tumors varies from 5 to 10 cm and is rarely larger than 20 cm. In the series by Kabiri et al., tumor size varied from 2 to 13 cm with a mean of 6 cm [4]. In our series, the average tumor was 8.75 cm. The tumor was attached to deep tissue, firm and painless in most cases. It only becomes symptomatic due to mechanical compression of neighboring organs [9, 11]. In case of invasion of the nervous structures, hyperesthesia, muscular weakness or more rarely pain may develop. In our series neurovascular symptoms such as paresthesia and pain were noted in one case. Chest X-ray shows a parietal tumor that has invaded the soft tissues, the adjacent bone tissue, and in certain cases the nearby periosteum [9]. Ultrasound can reveal abnormal but nonspecific echogenicity [6]. CT Scan can be used to identify the location and size of the tumor. When fibromatosis is in contact with bone, reaction of the adjacent bone varies, but is mainly a spiculated periosteal reaction as well as erosion or even cortical destruction. CT-guided biopsies can sometimes be performed, suggesting a diagnosis in certain cases [12]. Nevertheless, MRI is more sensitive than CT Scan for evaluating bone invasion when the tumor is in contact with bone [5, 12]. It can also be used to monitor lesions after treatment. Classically, the characteristics of fibromatosis on MRI include a poorly circumscribed heterogeneous tumor, with an isosignal on T1 and a high intensity signal on T2 weighted sequences (when the cellular component is predominant) as well as contrast enhancement of internal septa and a low intensity signal of the pseudocapsule in all sequences. Well-circumscribed lesions and low intensity signal T1 and T2 weighted sequences without contrast enhancement have also been described when the fibrous component is predominant. These radiological features are fairly similar to those of soft tissue malignant tumors: heterogeneous signal, poorly circumscribed lesions, T2 weighted high intensity signal, as well as neurovascular or bone invasion. Indeed, low-grade sarcoma is impossible to differentiate from desmoid fibromatosis on imaging and it is therefore the major differential diagnosis [12]. The final diagnosis is histopathological. An initial CT-guided biopsy of the parietal pleura should probably be performed if the lesion is inaccessible in particular to make certain differential diagnoses, which may require primary chemotherapy. On resected tumor samples, the tumor is firm, poorly circumscribed and appears fasciculated when cut. Histologically, there is a proliferation of fibroblast and myofibroblast-like spindle cells arranged in parallel bundles, intersected by more or less thick “cheloid-type” hyalinized collagen bands. Atypical nuclei and mitoses are rare. The poorly circumscribed tumor invades the soft tissues [10]. There is no chondrogenesis or osteogenesis. On immunohistochemistry, the spindle cells express alpha smooth muscle actin and vimentin. They are PS100, CD34 and keratin negative. A differential diagnosis must be made from well-differentiated fibrosarcoma, which can mainly be excluded in the absence of fibroblasts with large hyperchromatic, mitotic nuclei in less abundant stromal collagen. There is no normally accepted treatment protocol. Nevertheless, patient management should be multidisciplinary to prevent the high risk of recurrence [6]. Surgical treatment includes the widest possible resection, which is often complicated. However, carcinological resection without mutilation is often difficult because of the large sized and poorly circumscribed tumor as well as its proximity to vital structures such as the brachial plexus or bone marrow [2, 13]. Local recurrence is frequent and can occur after several operations [5]. Indeed, on the chest wall, besides obtaining a wide tumor free margin, resection of the ribs above and below should be performed with a 2 to 4 cm margin [7, 10]. Another therapeutic alternative includes high doses of radiotherapy associated with surgical treatment. Radiotherapy is beneficial in case of a residual tumor or inoperable locations, and results in recurrence free survival in 77% of cases [14]. Nevertheless there are still potential complications associated with combined radiotherapy and surgery, especially in children, and this treatment not satisfactory [5]. Medical treatment has been attempted when surgery is unsuccessful. Treatment with cytotoxic as well as non-cytotoxic chemotherapies has been reported: hormonal, non-steroid anti-inflammatories, and...
interferon α [5]. Nevertheless, the progression of desmoid fibromatosis is unpredictable. Spontaneous regression has been described in the absence of any treatment or with partial resection, as has tumoral stabilization but also sarcomatous degeneration [6]. Recurrence occurs in 25 to 75% of cases in the chest wall depending on the series [9,15]. Although survival at 5 years is nearly 93%, the probability of recurrence after 5 years is an estimated 29% [11,16]. Even though these tumors do not metastasize, they can result in significant morbidity and death from locoregional invasion [17].

Conflict of interest statement

None.

References