Desmoid-type chest wall fibromatosis. A six cases series

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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...
Chest wall tumor, chest wall fibromatosis

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 formal 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 microscopy. An immunohistochemical study was performed in all cases. A panel of antibodies was used associating alpha smooth muscle actin, desmin, PS100, CD34, EMA, in all cases. A panel of antibodies was used associating microscopy. An immunohistochemical study was performed in all cases. A panel of antibodies was used associating alpha smooth muscle actin, desmin, PS100, CD34, EMA, 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 mi xoid 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 endopharyngeal 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.
and 8th ribs, atypical resection of the right lower lobe of the lung and a tissue sample of the parietal pleura. Histological analysis confirmed recurrence and pleuropulmonary invasion. Radiotherapy was indicated but the patient was lost-to-follow-up. She consulted 2 years later for a second case of recurrence. This included a 2 cm tumoral nodule of the main axis located on the posterior 5th rib stump extending to the 4th rib and another 1 cm nodule attached to the pulmonary parenchyma. Surgical resection was complete, confirmed microscopically on rib samples.

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

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.

Figure 3  CT Scan of the chest with a mediastinal window and contrast injection (case no. 3): parasternal tumor with chest wall and endo thoracic invasion.

Figure 4  Histology (HE × 400): spindle cell proliferation if parallel bundles intersected with collagen bands. In the lower right image, tumor cells are seen to be positive for alpha smooth muscle actin (IHC × 400).
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 thoracotomy scars or even after simple intramuscular injections. [7,8]. In our series two cases of fibromatosis developed on a surgical thoracotomy 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 Gardner syndrome [7]. Certain authors have identified a deletion of the long arm of chromosome 5 in case of the 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 nerve 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 spicular 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: heterogenous 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 generally 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, carcinoideal 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-inflammatory, and

**Figure 5** Histology (HE ×100): fibromatosis with invasion of bone trabeculae.
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