Desmoid tumour of the chest wall

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Case report

The authors report the case of a 35-year-old female patient, without particular antecedents and a non-smoker, who consulted for chest pain that had been evolving for several weeks, without any change in her general condition. The pain was parietal, opposite the second left intercostal space, irradiating in the arm and reproduced on palpation. The rest of the physical examination did not reveal anything specific. The chest X-ray (Fig. 1) revealed well-defined, ovoid opacity, in projection of the second left intercostal space. The computed tomography (CT) (Fig. 2) confirmed the existence of a homogenous left parietal tissue mass, centred on the second intercostal space with invasion of the second and third ribs. This lesion appeared well defined, spontaneously isodense to the muscles, without enhancement after intravenous injection of the contrast agent. A left lateral thoracotomy was then performed, enabling the healthy macroscopic excision of a hard tumour, invading the middle arch of the second and third ribs, thrust out on the pulmonary parenchyma without adherence. The anatomopathological examination of the surgical piece (Fig. 3) revealed a lesion with poor cellularity consisting of bundles of fibroblasts without nuclear atypia, with a conjunctive type stroma between the cells with collagen fibres, indicating a diagnosis of desmoid tumour. The index of proliferation by Ki67 was low, under 10% (Fig. 4). Unfortunately, the resection was not in a healthy area. In view of the incomplete nature of the excision, complementary radiotherapy was discussed during the multidisciplinary meeting. After a free interval of several months, the patient again complained of parietal pain and the CT confirmed the local recurrence, requiring further surgery.

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Discussion

Desmoid tumours are benign lesions corresponding to slowly evolving, infiltrating, fibroblast proliferations. Although non-metastatic, their infiltrating and recurrent nature renders their morbidity-mortality significant, so that the term “aggressive fibromatosis” is often used to characterise it. These tumours account for 0.03% of all neoplasia with an annual incidence estimated at 2–4 per 1,000,000. They are often found in familial adenomatous polyposis (FAP) and Gardner’s syndrome [1]. Other factors of risk such as trauma, surgery, pregnancy or even the use of oral contraceptives have been reported [2]. Women are more likely to have desmoid tumours (sex ratio: 2/1) with a mean age ranging from 20 to 40 years [3]. Although the aetiology is multifactorial, certain genetic factors have been identified in patients with desmoid tumours. Mutations of the adenomatous polyposis of the colon (APC) gene have been found in patients presenting a Gardner’s syndrome or a FAP. In the sporadic forms, the anomalies may involve the gene coding for betacatenine (CTNNB1). These mutations result in an increase in the intracellular betacatenine concentration that induces an increase in fibroblast proliferation due to the interaction with transcription factors [1]. From a histological point of view, desmoid tumours macroscopically come in the form of invading fibrous masses adhering to the neighbouring structures. The colour is pale since they are not highly vascularised. They correspond to a clonal proliferation of well-differentiated fibroblasts, arranged in strips, within a matrix of collagen fibres. The mitotic index is low and there is no nuclear atypia. There are no haemorrhagic and necrotic rearrangements. Calcifications are rare and myxoid degeneration variable. Although the appearance is often well defined in imaging, there is no peripheral capsule and their outlines are poorly defined [4].

Desmoid tumours are often intra-abdominal (up to 69% according to the series), involving the chest wall or...
Desmoid event tumour

In the chest wall, the desmoid tumour is the second or third left ribs. Desmoid tumour, in particular in cases of FAP or Gardner’s syndrome [2]. Impairment of the chest wall accounts for 20% of the extra-abdominal locations. The starting point of the tumour is most often the intercostal muscles. The symptomatology remains non-specific and the most common signs are dyspnoea, chest pain, palpation of a mass, coughing or even vourse of the chest wall [5]. Finally, multiple locations are found in 15% of the cases, in particular in case of impairment of the extremities [1].

Imaging is very important in the care of desmoid tumours. The ultrasound examination is used to explore the superficial locations revealing hypoechochogenic and homogenous lesions with variable vascularisation with Doppler examination [2]. In the CT, they appear well-defined, isodense to the muscles in spontaneous contrast, as illustrated in our case report. After the intravenous injection of the idoine contrast agent, they may or may not be enhanced. In our case, the lack of enhancement may be due to the little cellular nature of the lesion brought to the histological analysis. In case of contrast enhancement, the intensity remains variable. Desmoid tumours are rather homogenous, without haemorrhagic or necrotic rearrangement [6]. The magnetic resonance imaging (MRI) is the choice examination for the exploration of desmoid tumours. The excellent resolution in contrast in the analysis of soft tissue renders the study of the extent of the lesions and their relationship with the neighbouring structures precise. This helps direct the therapeutic attitude. In T1, desmoid tumours are rather isosignal compared with the muscles. Their signal in T2 is variable and usually intermediate. In case of major cellularity or myxoid component, the T2 signal is higher. However, if the collagen contingent is predominant, the signal is reduced. Therefore, the presence of bands in hyposignal T1 and T2 related to the collagen fibres is classically reported in desmoid tumours, although not specific. Finally, the contrast enhancement after injection of gadolinium chelates is also variable, more intense when the lesion presents high cell proliferation. The MRI is also useful in patient monitoring. In fact,
recurrent lesions present a high cellularity giving them a high T2 signal and a more intense contrast enhancement [2,7,8]. The role of position emission tomography (PET) with 18-FDG has not been clearly established. It may be interesting in the detection of recurrences as well as in an evaluation of the therapeutic response of patients receiving imatinib [1]. Although the histological characteristics may account for the appearance of desmoid tumours in imaging, the diagnosis requires anatomopathological proof. Radioguided biopsies are useful in the establishment of the diagnosis. The operator will usually find a hard lesion, resistant to the placement of biopsy material [2]. The main differential diagnosis of the desmoid tumour of the chest wall is fibrosarcoma, the most common primary malignant lesion in the adult of the soft tissue of the chest wall. The differentiation remains very difficult in imaging, in particular for low-grade sarcoma. For the high-grade lesions, the presence of an area of intraloesional necrosis may help in the diagnosis [9].

A multidisciplinary approach is required to obtain optimum care for the patients. The asymptomatic lesions may be monitored, especially since cases of spontaneous cure have been described. Full surgical excision is the choice treatment for symptomatic desmoid tumours [1]. It is often difficult to obtain healthy margins due to the infiltrating nature of the lesions or local anatomic conditions, as in our case. In fact, the anatomical relationships of the chest wall often render full excision difficult. In our case, there is a major costal invasion, illustrating the locally aggressive and infiltrative nature of these lesions. In addition, a pleural extension may be feared due to the location. The local recurrence of desmoid tumours is frequent and up to 50% for the extra-abdominal locations [2]. It is estimated at up to 63% according to the series for impairment of the chest wall [10]. Adjuvant radiotherapy decreases the rate of local recurrence, especially if the excision is incomplete. For the non-operative forms, an association of radio and chemotherapy is recommended. Hormone therapy, treatment with non-steroid anti-inflammatories, alpha interferon or even imatinib (a tyrosine kinase inhibitor) is also discussed [1]. Although the prognosis for survival of patients is rarely involved, the treatment of desmoid tumours remains difficult and sometimes mutilating, especially when the extremities are involved.

**Conclusion**

Desmoid tumours are benign tumours. However, they have a high locoregional invasive potential rendering their ablation often incomplete with a non-negligible risk of local recurrence. Their appearance in imaging remains variable, directly correlated to their histological characteristics. The diagnosis is raised in view of any mass of soft tissue of well-defined appearance, invading the neighbouring structures, especially when there are predisposing factors such as familial adenomatous polyposis or Gardner’s syndrome.

**References**