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Case Report
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An Aggressive Malignant form of Fibrous Solitary Tumor of the Pleura: A Case Report and Literature Review

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Abstract

Background: The solitary malignant fibrous tumor of the pleura is rare histological entity with aggressive potential.

Case Presentation: The authors report the case of a 55 years old male who presented with left chest pain. The diagnosis of a solitary malignant fibrous tumor of the pleura was evoked on the basis of CT scan chest and confirmed by histopathological analysis of a tissue biopsy obtained by anterior mediastinotomy.

Conclusions: If surgery is effective for benign forms, chemotherapy alone or coupled with radiotherapy remains the only therapeutic option for the malignant forms of these tumors.

Keywords: Tumor of the pleura; Fibrous solitary tumor; Malignant tumor; Chemotherapy

Introduction

The solitary fibrous tumor of the pleura is a rare fibroblastic entity, most often localized and benign, accounting for less than 5% of all pleural tumors. Less than 10% of these tumors are labelled malignant with life-threatening metastasis [1]. Clinical manifestations may be respiratory or extra-thoracic with arthralgia, clubbing of the fingers and hypoglycaemic seizures. We report the observation of a patient with chest pain with dyspnea in whom the diagnosis of solitary malignant fibrous tumor of the pleura was evoked on the CT scan and then confirmed by the pathological examination of a tissue biopsy obtained by mediastinotomy.

Case Presentation

A 55-year-old male with a history of smoking, operated 3 times (in 2005, 2007 and 2009) for recurrent Dermato fibrosarcoma protuberans (Darier-Ferrand sarcoma, DFSP) of the right anterior thoracic wall. He had since 4 months pain in the left chest wall radiating to the left upper limb, dyspnea on exertion and weight loss of 6 kg in 4 months. The symptomatology was aggravated one week before presentation by the accentuation of the dyspnea. On admission, he had a general condition preserved with a blood pressure 150/80 mmHg and tachypnoea at 34 cycles/min associated with edema of the lower limbs. The chest examination was in favour of a consolidating lesion affecting the middle part of the left hemithorax. The chest X-ray showed opacity of the left hemithorax with mediastinal deviation to the right side (Figure 1). On CT chest with contrast, a mass, massively necrotic, occupied the left hemithorax, having close contact with the pericardium and the main pulmonary artery which was encircled by more than 180°. This mass engages the left stem bronchus and the left lower lobar bronchus and was laminated (Figure 2). It was associated with an invasion of the diaphragm with the presence of Para tracheal mediastinal adenopathy. There were no extra-thoracic lesions. The histopathological study of a large biopsy core obtained by anterior mediastinotomy under general anaesthesia was in favour of a solitary malignant fibrous tumor of the pleura (Figures 3 and 4). In fact, the patient was operated before for Darrier-Ferandsarcoma of the right chest wall. Actually we initially thought of a metastasis but after discussion with pathologists and oncologists we concluded a solitary malignant fibrous tumor of the pleura. After a discussion at the multidisciplinary consultation meeting, the decision of a radio-chemotherapy was made. The patient is currently followed in oncology department in 2nd chemotherapy cure.

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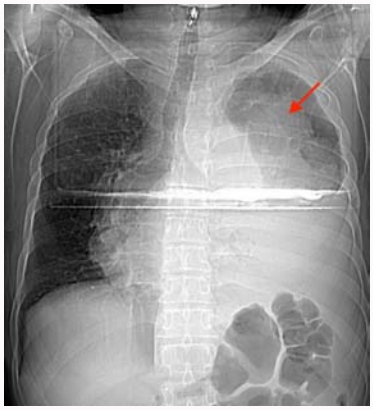

Figure 1: Chest X-ray showing opacity of left thoracic hemi champ with mediastinal deviation to right side.

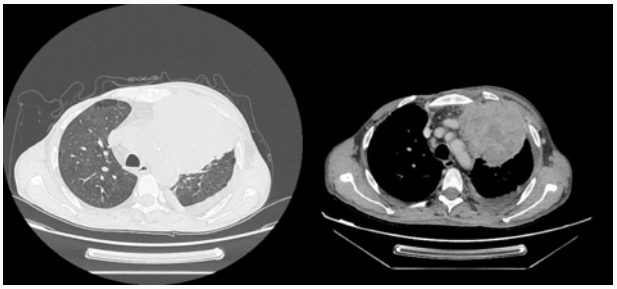

Figure 2: Thoracic tomodensitometry with injection of contrast medium, which targets a tissue mass, massively necrotic, having contact with the left ventricle and the pulmonary artery it encompasses.

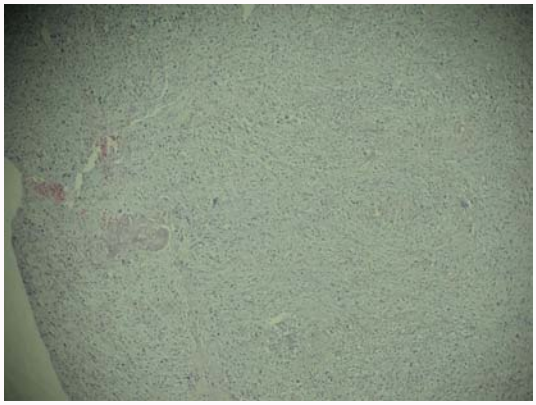

Figure 3: Malignant solitary fibrous tumor of fusocellular architecture, cells display cytonuclearatypieslabeled with mitosis figs (x20).

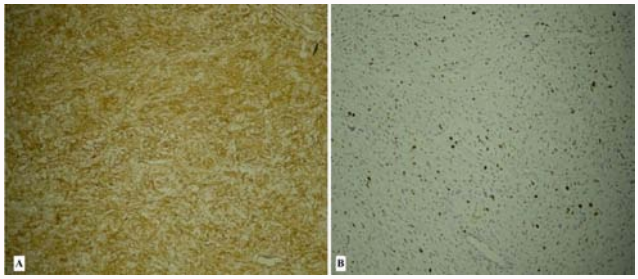

Figure 4: Malignant solitary fibroid tumor. **A**: Diffuse and intense cytoplasmic labeling by CD34 (x 20). **B**: Nuclear labeling of mitotic cells by Ki67 (x 20).

Discussion

The solitary fibrous tumors of the pleura represent a rare histological entity belonging to the pleural tumor groups of sub-mesothelial origin with variable clinical expression. This condition can occur at any age, but predominates in patients over 50 years with a balanced sex ratio in the majority of reported cases [2]. Clinically, these tumors may manifest as chest pain, associated or not with dyspnea or cough. In some cases, haemoptysis or obstructive pneumonia may be in the clinical presentation [3,4]. The intensity of these symptoms is often correlated with the aggressiveness and size of the tumor by the compression effect it exerts not only on the respiratory system but also on the cardiac chambers responsible for dyspnea at rest as the case of our patient. Some solitary fibrous tumors may be associated with certain paraneoplastic syndromes such as clubbing of the fingers, a hypertrophic pneumonic osteoarthropathy of Pierre Marie, but especially hypoglycaemia in about 5% of cases, which is secondary to the secretion by the tumor of an insulin- Like substances [2,5,6].

Chest x-ray is a diagnostic step, usually showing a rounded, homogeneous and well-defined opacity. The diagnosis of malignancy can be evoked before a speculated opacity associated mainly with pleural effusion [4].

The thoracic computed tomography will complement the assessment by confirming the pleural origin and tissue nature of the tumor, as well as its localization and its relationship with the pleura and adjacent organs [7]. The tumor often appears very limited and homogeneous, but a heterogeneous aspect can be observed. As is the case in our patient, the malignant nature of a solitary fibrous tumor of the pleura is strongly evoked, whenever there are necrotic-haemorrhagic rearrangements on the CT scan [8]. The thoracic computed tomography allows us to study the loco-regional extension of the tumor, an essential element of the therapeutic decision.

Some authors have noted the value of magnetic resonance imaging (MRI), which makes it possible to determine the fibrous nature of the tumor, especially in the T1 sequence, which distinguishes it from other mediastinopulmonary, parietal and diaphragmatic structures [8,9]. At this stage, the histological diagnosis by biopsy of the tumor mass, with histological analysis should be proposed [10,11].

For our patient, we obtained biopsy by anterior mediastinotomy. This technique allowed us, under general anaesthesia, to have large biopsy fragments necessary for a consistent histological analysis thus avoiding the misdiagnosis. From the histopathological point of view, malignant and benign solitary fibrous tumors are defined. The malignant forms, more rare, are defined by numerous mitoses, areas of necrosis, and a nuclear pleomorphism [10]. Immunohistochemical analysis, using antibodies (Ac Bcl2, CD99, CD34, vimentin, keratin ...), makes it possible to affirm the positive diagnosis and to eliminate differential diagnoses such as sarcomas, Mesotheliomas [10].

The reference treatment is a complete surgical resection of the tumor at its implantation base, which can sometimes extend to the pulmonary parenchyma or to the chest wall in the event of invasion [2,12]. In the aggressive forms of solitary fibrous tumors of the pleura, surgical resection may be followed by chemotherapy associated or not with radiotherapy, the real interest of which remains to be demonstrated. In the specific case of our observation, in view of the invasiveness of the tumor radiologically and the results of the pathological examination including histology and

immunohistochemistry, the decision of a radio-chemotherapy was retained. Indeed, ifosfamide and adriamycin chemotherapy is of interest in malignant inoperable forms at the outset, or for the significant reduction in the size of an operable tumor [13]. This chemotherapy could be justified in neo-adjuvant, in large tumors or in postoperative, in case of incomplete resection [13]. Radiotherapy may also be indicated after surgical resection, in case of histological signs of malignancy, especially if this resection has been incomplete. Because of an evolutionary profile that is difficult to evaluate, remote radio clinic surveillance is recommended by some authors in search of metastasis [14,15].

Conclusion

The diagnosis of solitary fibrous tumors of the pleura is histological, facilitated by the complement of immunohistochemistry. The management is surgical excision without delay. However, for aggressive and/or metastatic malignancies, chemotherapy alone or in combination with radiotherapy remains the only therapeutic option that can improve prognosis in terms of survival.

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