Case report

Solitary fibrous tumor of the pleura: A giant finding, a benign entity?

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ABSTRACT

Solitary fibrous tumor of the pleura (SFTP) is a rare tumor. The prognosis is relatively good, but up to 20% of the cases are malignant. We presented a case of a large mass of the thorax, showing malignant poor prognosis features.

A 73-year-old male, former smoker, presented with a history of dry cough. Chest CT revealed a left bronchopulmonary mass (11 cm × 14 cm) extending from the costal pleura to the left hilum. Histopathologic examination of the transbronchial lung biopsy was consistent with SFTP. A left pneumonectomy was performed due to invasive nature of the lesion.

According to features associated to poor prognosis (lung tumor ≥ 10 cm, hypercellularity, abundant intratumor blood vessels and areas of necrosis), and in spite of proliferation index (Ki67+ < 1%), the case was discussed in a multidisciplinary meeting, and was assumed to be malignant. At the present time, the patient is his fifth month after surgery, in tight follow up.

This case highlights a rare SFTP presenting bad prognosis features, that although completely resected should have long-term follow-up due to the high risk of recurrence.

1. Introduction

Solitary fibrous tumors of the pleura (SFTP) are rare, originated from the mesenchymal cells, representing less than 5% of all pleural tumors [1]. SFTP are usually asymptomatic in early stages, mostly detected incidentally by chest imaging [2]. The prognosis is relatively good because of the slow-growing and low metastatic potential, but up to 20% of the cases are malignant [3,4]. Although, it is important to determine whether the tumor is benign or malignant before deciding on treatment options and estimating prognosis, this can sometimes be difficult. We present a case of a large mass of the thorax, showing poor prognosis features.

2. Case description

A 73-year-old male, former smoker, with past history of chronic obstructive pulmonary disease, was referred to the pulmonology outpatient clinic due to dry cough associated with a thoracic lesion on chest X-ray. On physical observation, he had an ECOG performance status of 1, and the chest auscultation revealed diminished breath sounds on the middle and lower left hemithorax. A chest X-ray showed a large homogenous opacity in the middle-to-lower left lung field, which was confirmed by a chest computed tomography (CT) describing bronchopulmonary mass, measuring 11 × 14 cm, extending from the costal pleura to the hilum. Histopathologic examination of the transbronchial lung biopsy was consistent with SFTP. A left pneumonectomy was performed due to the invasive nature of the lesion, location and size. The post-operative period was uneventful.

Upon macroscopic examination of the resected specimen, an heterogeneous mass with 15 × 15 × 8cm was observed. Microscopically, irregular fascicles of spindle cells, with hypercellularity, numerous blood vessels and areas of necrosis, and in spite of proliferation index (Ki67+ < 1%), the case was discussed in a multidisciplinary meeting, and was assumed to be malignant. At the present time, the patient is his fifth month after surgery, in tight follow up.

With these findings a benign SFTP was suspected but a left pneumonectomy was performed due to the invasive nature of the lesion, location and size. The post-operative period was uneventful.

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vessels with sclerosing walls, focal hemorrhage, several areas of necrosis and mitotic figures (3 mitoses per 10 high power fields) were present. Resection margins were tumor free. Immunohistochemistry showed positivity for STAT-6, bcl-2, CD 34 and CD 99 (Fig. 3 A–D). According to features associated to poor prognosis (lung tumor ≥ 10 cm, hypercellularity, abundant intratumor blood vessels and areas of necrosis), and in spite of proliferation index (Ki67+) <1%, the case was discussed in multidisciplinary team meeting and the patient was proposed for close follow-up.

At the present time, the patient is the patient is his fifth month after surgery, in tight follow up, without any sign of focal recurrence or metastization.

3. Discussion

SFTP are uncommon tumors originated from the mesenchymal cells of the pleura. Although it represents less than 5% of all pleural tumors, it has been increasingly recognized over the past few years [2,3]. The peak of incidence is between the 5th and 7th decade of life although it is seen in all ages [5]. There is a lack of association with environmental exposure (radiation, tobacco, asbestos or other toxicants), and inherited risk factors are unknown.

Fig. 1. A - Chest radiography showed a homogenous opacity in the middle-to-lower left lung field. B, C - Computed tomography of the chest showed a pulmonary mass, measuring 11 × 14 cm, extending from the costal pleura to the hilum.
At presentation, approximately 40–60% of patients have nonspecific pulmonary symptoms, typically cough, shortness of breath, or chest pain [6]. Rarely, hemoptysis and obstructive pneumonitis may occur as a result of airway obstruction. In other cases, an intrathoracic mass is incidentally detected on a routine chest X-ray in asymptomatic individuals [2].

Making the differential diagnosis between benign and malignant SFTP is usually problematic for predicting the disease prognosis. According to some case series, a malignant behaviour of SFTP is suspected in presence of the following histologic features: hypercellularity, pleomorphism, tumor necrosis, more than 4 mitoses per ten high-power fields and infiltrative margins [4]. Immunohistochemistry may be useful in differentiating the SFTP from mesotheliomas and intrapleural sarcomas. Usually SFTP is vimentin, CD 34, CD 99, bcl-2 and STAT6 positive and cytokeratin negative.

Therefore, the present case showed some poor prognosis features, namely lung tumour ≥10 cm, hypercellularity, abundant intratumor blood vessels and several areas of necrosis. A tight clinical follow-up is recommended to detect the recurrence [7].

Complete surgical resection is the gold-standard treatment and the most important prognostic factor [8]. The choice of surgical approach (video-assisted thoracoscopic surgery (VATS) and standard thoracotomy) is essentially based on the tumor size, the difficulty of removal and surgical team expertise. As in the described case, large tumors could be more difficult to resect due to the invasive behaviour and an extended resection may be necessary [5]. The role of adjuvant radio and chemotherapy in malignant SFTP remains unclear since there is no systematic assessment due to the rarity of the tumor [8].

Recurrence rates for completely resected malignant SFTP range from 14 to 63% in pedunculated and sessile tumors, respectively, and occur mainly in the first 24 months after surgery. Despite a complete resection, malignant SFTP still have a poor prognosis with a 5-year rate survival of 45.5%. A long and systematic follow-up is mandatory because of the risk of recurrence of SFTP regardless of the pathological type obtained after surgical resection. Recurrent tumors should be resected. Combined chemotherapy with temozolomide plus bevacizumab can be considered in locally advanced, recurrent or unresectable malignant SFTP [9].

4. Conclusions

This case highlights a rare SFTP presenting malignant features, that although completely resected and potentially with good prognosis, should have long-term follow-up due to the high risk of local and metastatic recurrence.

Statement of ethics

The ethics committee of Hospital Garcia de Orta in Almada, approved this study.

Written informed consent was obtained from patient.

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Author contributions

All the authors had full access to all the data of the patient and contributed to the conception, drafting and critical revision of the case report.

Declaration of competing interest

The authors have no conflicts of interest to declare.
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Fig. 3. A – The resected specimen disclosed mesenchymal neoplasm with moderate cellularity, with oval and fusiform nucleated cells. B – Fusiform nucleated cells with hemangiopericytic vessels. C – Immunohistochemical strong and diffuse nuclear expression of STAT6.