Inflammatory myofibroblastic tumours of the lung: a case series

Abstract

Inflammatory myofibroblastic tumours represent a rare group of lesions reported in various organs but with an unclear aetiology. Although so far they are considered benign, some cases of recurrence and invasive behaviour have been noted. We report three consecutive cases of tumours of the lung, with heterogeneous clinical presentation. They were all treated by means of surgery, with good long-term results. Diagnosing an inflammatory myofibroblastic tumour relies mainly on an experienced pathologist.

Keywords

inflammatory myofibroblastic tumour • plasma cell granuloma • fibroxanthoma

Introduction

Inflammatory myofibroblastic tumours are rare, unencapsulated, with some describing them as being quasi-neoplastic tumours derived from the unregulated growth of inflammatory cells. The first people to acknowledge this group of pseudotumours were Umiker and Iverson in 1954 (1). The amount of information about these tumours is scarce, and one of the reasons is the various terms that have been used to describe them; terms based on the particular type of cells are as follows: plasma cell granuloma, plasma cell pseudotumour, inflammatory myofibroblastic proliferation, omental–mesenteric hamartoma, histiocytoma, and fibroxanthoma. All of these terms describe the same group that is inflammatory...
myofibroblastic tumours. At the moment this article was written, it was not yet known whether these lesions represent a primary inflammatory process or if they present a low-grade malignancy with a very strong inflammatory response.

Case series

We report three consecutive cases which had a very heterogeneous clinical and radiological presentation.

Patient 1
A 19-year-old male, nonsmoker, with no other significant pathological problems was admitted after a minimal haemoptysis episode. Standard chest X-ray appeared normal, but after a repeat of the haemoptysis a thorax computed tomography (CT) scan was done which showed complete atelectasis of the left lower lobe caused by a 30-mm well-defined tumour located in the bronchial tree. Bronchoscopy revealed a cherry-like tumour located in the left inferior bronchus which completely obstructed its lumen and bled very easily on touch, which did not make a biopsy possible. At this point, our suspicion was of a carcinoid tumour; therefore, a left lower lobectomy was performed. Abnormal hard bleeding during dissection was noticed, an aspect that will be observed later in other patients also, but at the time it was considered to be the result of long-term atelectasis and inflammation. The postoperative course was uneventful. The histopathological report pointed towards an inflammatory myofibroblastic tumour (Figures 1–3). The images show lung parenchyma with spindle-like cell proliferation, pale eosinophilic cytoplasm, and ill-defined tumour border; mitosis was not encountered, chromatin was finely distributed and nucleoli were small. Organizing pneumonia is a common feature at the edges of the tumour, but it is not mandatory for the diagnosis. The patient had his 8 years of follow-up with no signs of local or systemic recurrence.

Figure 1. Histopathological aspects of inflammatory pseudotumours of the lung.

Figure 2. Histopathological aspects of inflammatory pseudotumours of the lung.

Figure 3. Histopathological aspects of inflammatory pseudotumours of the lung.

Patient 2
A 25-year-old woman presented to the Pneumology Department after a medium haemoptysis episode (250–300 ml). Patient history revealed multiple small haemoptysis episodes in the past month. An emergency bronchoscopy was performed, and it revealed a minor but active bleed from the right inferior lobe bronchial tree, more specific from the posterior basal segment.

As patient history also revealed continuous contact with ovines due to working in agriculture, and the chest X-ray presented a well-defined, round opacity in the inferior part of the right thorax, the initial diagnostic was thought to be a hydatid cyst with a vascular fistula; the CT scan showed a well-defined tumour situated in the inferior right lower lobe, with different semifluid densities throughout, an aspect that is not characteristic of a hydatid cyst (Figures 4 and 5). As the patient was having haemoptysis and the imagistic and bronchoscopic aspects were clear, a right thoracotomy was decided in order to perform a right lower lobectomy to remove the bleeding lesion. During surgery, palpation revealed a solid tumour. The dissection was difficult due to excessive bleeding, aspects similar to the other presented patient. The right lower
CT scan revealed a well-defined tumour in the right inferior lower lobe with mixed semifluid densities, similar to the one in patient 2 (Figures 6–9). Bronchoscopy did not reveal anything significant. This time, with a straight suspicion of an inflammatory myofibroblastic tumour, we decided to do a right lower lobectomy. As in the other cases, hilum and lymph node dissection was difficult because of the excessive bleeding. Histopathological examination confirmed our suspicion of inflammatory myofibroblastic tumours. The patient had his 2 years of follow-up with no signs of local or systemic recurrence.

Figures 4, 5, 6, 7. Thoracic CT scans showing well-defined tumours in the right lower lobe.

Lobectomy was performed with a clean postoperative evolution, and the final diagnosis from pathology was of an inflammatory myofibroblastic tumour. The patient had her 5 years of follow-up with no signs of local or systemic recurrence.

Patient 3
A 76-year-old male, with no respiratory symptoms with a pulmonary opacity discovered during preoperative investigations made for a chronic gallbladder condition. Surgery was postponed, and the patient was referred to a pulmonologist in order to investigate the pulmonary lesion.
The best way of describing this entity is a quasi-neoplastic process determined by an unregulated growth of inflammatory cells. The cause however for this unregulated growth remains unknown. There are several theories regarding it, most of which describe an unchecked immunologic response to a foreign/viral antigen–antibody reaction. These pseudotumours are not limited to the lung and can grow in other organ systems such as the brain or liver (2–10).

Because of their rarity and the limited amount of tissue that can be taken through a biopsy (which finds inflammatory cells only), these pseudotumours remain a diagnostic dilemma in any organ system for both the pathologist and the surgeon. However, it has been noted that, although uncommon, these tumours develop more commonly in the lung or trachea with an increased predilection towards children (9–11).

Inflammatory myofibroblastic tumours are the most common isolated primary tumours of the lung in children under the age of 16 years with more than half of patients being younger than 40 years of age. These lesions represent <1% of all lung tumours, with no discrepancy between gender or race. Many patients are asymptomatic, these tumours usually being discovered incidentally on basic chest X-rays. Some symptoms that may occur are cough, fever, weight loss, fatigue, haemoptysis, dyspnea, clubbing, chest pain, and arthralgia (5,8).

Inflammatory myofibroblastic tumours can be observed on a basic chest X-ray, usually as a solitary nodule or mass of 1–10 cm in diameter, with the lesions usually present in the lower lobes. However, radiographic images and invasive diagnostic procedures such as percutaneous fine-needle aspiration biopsy or bronchoscopy are considered insufficient for a clear diagnosis. Because of this, the best way of reaching a diagnosis and a therapeutic cure remains through surgery (5,8).

Macroscopically, the most common presentation of inflammatory myofibroblastic tumours appears as well-circumscribed, non-encapsulated, firm, usually yellow–white masses containing variable inflammation, haemorrhage, calcification, and rarely cavitations. Most of these lesions are parenchymal with only some located endobronchial, which may cause airway obstruction. Rarely, <5% of cases can present with invasion in the mediastinum and/or chest wall. The cases of local recurrence are attributed to incomplete resection of the lesion. There have been cases of metastasis following the complete resection of the mediastinum or brain. Moreover, rarely there can be active lesions intra- and extrathoracic simultaneous (3,4).

Microscopically, it has been determined that these types of lesions consist of a mixture of fibroblasts and fibrous tissue, granulation tissue, and inflammatory cells (including giant cells, macrophages, eosinophils, lymphocytes, and histiocytes)
and a large number of plasma cells. Further investigation through immunohistochemistry reveals the polyclonal nature of plasma cells with a predominance of immunoglobulin G. The pathological examination on frozen sections at the time of surgery is often indeterminate; the differential diagnosis includes sarcoma, lymphoma, and fibrosis. The pathologist is usually able to eliminate a neoplastic process, however, if the result is uncertain on the frozen section, complete resection, if possible, is recommended (6, 7).

The best course of treatment for inflammatory myofibroblastic tumours remains complete surgical resection which not only achieves cure but also excludes malignancy. Wedge resection may be an adequate treatment if removal is complete and the diagnosis is certain. If it is required for complete resection and if the patient’s pulmonary reserve is adequate, a lobectomy may also be performed. There are several non-surgical treatments such as chemotherapy, radiotherapy, or corticosteroids which may be used in case of incomplete surgical resection, postoperative tumour recurrence, and multifocal disease or if the patient’s lung function does not allow surgical resection. Spontaneous regression may occur sometimes; however, local expansion/invasion may cause significant morbidity and occasional death. Long-term follow-up after complete removal of the mass shows a low incidence of recurrence. In the rare cases of the recurrent disease, the recommended treatment is through a surgical re-resection (2, 3, 6).

Conclusions

Our cases point to the same conclusions as those available in the literature on inflammatory myofibroblastic tumours. The most common symptom found was haemoptysis in two of the patients, the third being asymptomatic. The main course of treatment remains surgical, with anatomical resections being the operative indication if lung function allows it. Important to be mentioned is the uncommon bleeding during surgery.

There were no recurrences in our follow-ups which again confirms surgical resection as being the best approach. Diagnosing inflammatory myofibroblastic tumours relies mainly on an experienced pathologist, seeing as, in the rarity of this type of tumour, and misdiagnosing may be part of it.

Ethics approval and consent to participate

Not applicable.

Consent for publication

Consent was obtained from the patients for publication of this case report and the accompanying images.

Competing interests

The authors declare that they have no competing interests.

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