Case report

An unusual cause of haemoptysis in a young male

N Barbetakis*, A Efstathiou, T Xenikakis, H Konstantinidis and I Fessatidis

Address: Cardiothoracic Surgery Department, Geniki Kliniki – Euromedica, Thessaloniki, Greece
Email: N Barbetakis* - nibarb@otenet.gr; A Efstathiou - akhna@otenet.gr; T Xenikakis - txenikakis@yahoo.gr;
H Konstantinidis - harikonsta@yahoo.gr; I Fessatidis - fessatidis@genikikliniki.gr
* Corresponding author

Abstract

Inflammatory myofibroblastic tumour (IMT) are reported to occur in a variety of sites, including the head and neck, abdominal organs, central nervous system and urinary tract. They only rarely occur in the lung.

We report a case of a 25-year-old male admitted with haemoptysis. His chest radiograph showed a peripheral right lung opacity and computed tomography revealed a right lower lobe soft tissue density mass. Bronchoscopy and fine needle aspiration were unhelpful, a diagnosis of pulmonary carcinoma was made, and the patient underwent a right lower lobectomy. On pathology, the tumor was found to be an inflammatory pseudotumor. These lesions are extremely rare, constituting less than 1% of pulmonary malignancies, but are known to occur in young patients. We believe clinicians need to retain an index of suspicion for the presence of this disease in young patients, which can masquerade as more common malignancies.

Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare disease that usually occurs in the lung. It is also known as plasma cell granuloma, inflammatory pseudotumor, xanthogranuloma and fibrous histiocytoma [1]. The notion of IMT being a reactive lesion or a neoplasm is controversial [2]. Because of its rarity, its biologic nature, natural history and response to treatment have yet to be completely defined. A case of a 25-year-old male who was admitted with hemoptysis, right lung mass and underwent a right lower lobectomy is presented. Biopsy of the resected specimen confirmed the diagnosis of an IMT.

Case presentation

A 25 year-old-male presented with a history of cough with bloody mucoid sputum for the previous two months. He also had anorexia and a history of intermittent upper respiratory tract infections for two years. Clinical examination revealed diminished breath sounds at the right lung base. Laboratory investigation reported normochromic – normocytic anaemia, with low iron serum level and elevated erythrocyte sedimentation rate (76 mm/first hour).

His chest radiograph revealed a peripheral right lung opacity, and computed tomography (CT) demonstrated a right lower lobe soft tissue density mass [Figures 1, 2]. Enlarged mediastinal lymph nodes were not identified. Bronchoscopy was negative. Transthoracic fine-needle aspiration under CT guidance was not diagnostic. A complete study of bone, brain and abdomen was also performed to assess the stage of a presumed bronchogenic carcinoma and no extrapulmonary involvement was noted. Surgery was carried out to obtain diagnosis and achieve cure. The patient underwent a right lower lobectomy and complete lymph node dissection through a right posterolateral thoracotomy. Macroscopically, the
mass appeared well-circumscribed and yellowish in colour on cut section. Microscopically the mass was composed of fibroblasts, collagen and inflammatory cells mainly of lymphocytes and plasma cells (Figure 3). There was no mitotic activity. No microorganisms were detected even on special stains. The overall features suggested an inflammatory pseudotumor. All lymph nodes were negative for malignancy. The postoperative period was uneventful and the patient discharged home on postoperative day 10. The patient remained well and asymptomatic two years later.

Discussion

Inflammatory myofibroblastic tumor of the lung is rare and its incidence is reported to be 0.04–1% of all pulmonary tumours [1]. Although these lesions can grow at a wide variety of other sites, they usually arise within the lung [3]. Pulmonary IMT is the most common lung tumor in patients younger than 16 years, and there does not appear to be a predilection for sex [4,5].

Common symptoms include cough, dyspnoea, fever, pleuritic pain and haemoptysis. A significant proportion of cases (30–70%) remain asymptomatic [4]. A history of upper respiratory tract infections or pneumonia is reported in approximately 30% of cases [6]. In our case, anorexia and hemoptysis were the main symptoms and a history of previous respiratory infection was also reported. Occasional cases of inflammatory pseudotumors of the lung are reported in which bacteria and fungi are isolated [7].

Previous studies have reported anaemia, elevated erythrocyte sedimentation rate, thrombocytosis and polyclonal hypergammaglobulinemia [8,9]. The first two findings were detected in our patient.

The radiologic features of inflammatory pseudotumors of the lung have been analyzed by Agrons et al [10]. Computed tomographic scan shows a nodule or a mass in approximately 90% of patients and multiple nodules in 5%. Secondary infiltration of hilum, mediastinum and airways occurs rarely. Calcification or cavitation is also reported but it is very infrequent [11]. Generally, computed tomography is not able to identify any specific features of inflammatory pseudotumors and all patients are eligible for surgery with suspected lung cancer as the diagnosis.

Needle biopsy has been suggested as a feasible approach to the diagnosis [12]. Fine needle aspiration shows a mixture of inflammatory cells, including plasma cells, fibroblasts and pneumocytes. Such findings are non-specific, given that inflammatory lesions of different origin can present the same picture. Moreover, inflammation and fibrosis can sometimes represent a reaction around a malignant tumor. Cerfolio et al proposed that these preoperative procedures are unnecessary and recommended complete resection for both diagnosis and treatment [1]. Attempted fine-needle aspiration cytology in our case also failed to establish diagnosis.

Gross pathology demonstrates that pulmonary inflammatory pseudotumors typically form a well-defined, firm, lobulated parenchymal nodule or mass with a whorled and often heterogeneous appearance on cut section. Histologically, IMT is composed of a variable inflammatory and mesenchymal cellular mixture including plasma cells,
During a stringent and prolonged follow up.

years later the patient is in an excellent condition under-

ous regression of IMT has been reported only infre-

chemotherapy or steroids is uncertain [1,16]. Spontane-

bouring anatomic structures (chest wall, diaphragm)also

In conclusion, IMT is a rare disease with similar character-
istics to those of a true tumor. clinicians should retain an
index of suspicion for the presence of this disease in
young patients with symptoms or chest radiographs
which suggest the presence of a malignancy. Surgical
resection when possible, is recommended as the treat-
ment of choice with an excellent outcome. Long-term fol-
low up is imperative to detect recurrence.

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histochemical and ultrastructural features. The mass was composed of fibroblasts, collagen and inflam-

histiocytes, lymphocytes and spindle cells. Therefore,
depending on the predominant cellular components,
many synonyms for this disease have been described. Pet-
tinato et al referred to this entity as IMT because the bulk
of the lesion invariably consisted of not specific inflam-
matory cells but proliferative myofibroblasts and fibro-
blasts [13]. Most of the spindle cells are myofibroblasts,
which show immunohistochemical staining for vimentin
and smooth muscle actin and consistent ultrastructural
features. The spindle cells commonly have low cellular
atypia and no mitotic activity. The differential diagnosis
of IMT is multifarious because of its variable cellular
admixture. It includes malignant lymphoma, lymphoid
hyperplasia, pseudolymphoma, plasmacytoma, malignant
fibrous histiocytoma, sarcomatoid carcinoma, sclero-
osing haemangioma, sarcoma and chronic nodular
pneumonitis [14]. These lesions can be differentiated by
careful attention to cellular atypia, necrosis, mitotic
activity, immunoreactivity or clonality [1,2].

The treatment of choice of inflammatory pseudotumor of
the lung is surgery [15]. Wedge resection, if radical is suit-
able for curative purposes. When it is not technically feas-
able, the lesion is removed with major resection
(lobectomy or pneumonectomy). In some cases, neigh-
bouriing anatomic structures (chest wall, diaphragm)also
need to be excised. The effectiveness of radiotherapy,
chemotherapy or steroids is uncertain [1,16]. Spontane-
ous regression of IMT has been reported only infre-
quently. In our case, a right lower lobectomy was per-
formed with radical lymph node dissection and two
years later the patient is in an excellent condition under-
going a stringent and prolonged follow up.

In conclusion, IMT is a rare disease with similar character-
istics to those of a true tumor. clinicians should retain an
index of suspicion for the presence of this disease in
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Figure 3
The mass was composed of fibroblasts, collagen and inflam-
matory cells mainly of lymphocytes and plasma cells (Hema-
toxylin-Eosin × 100).