Case report

Inflammatory pseudotumor: a controversial entity

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Abstract

Inflammatory pseudotumor of the lung is considered to be a rare, benign, neoplastic lesion, consisting mainly of spindle mesenchymal cells, sometimes in such a way that its histological appearance mimics that of a spindle cell sarcoma, fibrous histiocytoma or fibrosarcoma. On the occasion of a case managed in our department, the literature is reviewed, in an attempt to clarify some issues concerning this tumor. Emphasis is given to complete resection of the tumor for both diagnostic and therapeutic purposes. Malignant behavior may occur and recurrence is possible.

Keywords: Plasma cell granulomas; Histiocytomas of the lung; Spindle cell sarcoma; Solitude nodule; Coin lesion; Complete resection; Recurrence; Malignancy

1. Introduction

Plasma cell granulomas (PCG) or histiocytomas of the lung (also widely known as inflammatory pseudotumors) are usually considered to be benign tumors principally occurring in younger patients. Bahadori and Liebow reported a series of 40 cases in 1973 [1] describing the tumor in its typical appearance as the most common isolated primary tumor of the lung in children younger than 16 years of age. Twenty-five of the 40 patients were females, ranging from 13 to 68 years of age, whereas in 24 patients the lesion was discovered on a routine chest X-ray without previous history or related disease. Another large series of 24 patients has been recently reviewed in a retrospective study [2], whereas sporadic cases with unusual location or behavior have been reported.

2. Case report

A 50-year-old non-smoking female was submitted to diagnostic evaluation for recurrent pulmonary infections which revealed, on chest X-ray, a nodule of 3.5 × 3 cm in diameter (Fig. 1), presented as coin lesion and located in the posterior basal segment of the right lower lobe. The remaining other lung fields as well as the mediastinum, the hilum bilaterally and the chest wall were normal. From her medical history noteworthy pathology was not recorded. A CT scan of the thorax confirmed the chest radiography finding, whereas CT scans of the abdomen and of the brain were negative for any abnormality. Fine needle aspiration (FNA) under CT was performed and cytological examination revealed no malignant disease.

The patient was submitted to right posterolateral thoracotomy. From the frozen section of the tumor mass, the existence of malignant disease could not be excluded. Thus, and because of the age of the patient and her normal pulmonary function, a lower lobectomy was performed. Macroscopically, there was a solid solitary mass, with a homogeneous gray color, circumscribed, located within the lung parenchyma, 2.5 cm from the surgical margins (Table 1).

Microscopically, the mass consisted of spindle mesenchymal and inflammatory cells. There were no mitotic activity or atypia of the cells. Numerous non-neoplastic plasma cells with sparse Russel bodies, enough lymphocytes, scattered histiocytes and two small foci of calcification constitute inflammation. The histological characteristics were compatible with an inflammatory pseudotumor (plasma cell hypotype) (Fig. 2).

The postoperative course was uneventful, the patient was
discharged from the hospital one week postoperatively and three years after the surgical resection the patient remains free of disease.

3. Discussion

Plasma cell granulomas are lesions considered in the past to be not truly neoplastic, but composed of a variety of inflammatory cells, predominantly plasma cells. Such lesions account for less than 1% of all lung tumors with no sex preponderance. No specific findings on physical and laboratory examinations exist. Many cases are diagnosed by chance on radiological examination, whereas persistent fever, cough, dyspnea, pulmonary infection, bronchitis [3] or hemoptysis [4] are prominent in some cases. Weight loss, fever and fatigue have been also described [2].

Radiological examination demonstrates solitary peripheral nodule or mass, while in a few cases extraparenchymal involvement – including hilar, mediastinal and airway invasion, may be detected. According to Agrons et al. the lesions are usually of heterogeneous attenuation on CT and of intermediate signal intensity on T1-weighted magnetic resonance imaging [5]. In any case, radiographic images and invasive diagnostic procedures – including bronchoscopy and trans-thoracic fine needle biopsy – may be not sufficient for histological diagnosis. Consequently, surgery is crucial for both diagnostic and therapeutic reasons [2-4].

Macroscopically, PCG appears as a solitary yellowish-white, well-circumscribed mass containing variable inflammation, hemorrhage, calcification and rarely cavitation.

Local invasiveness is frequently detected in pathological examination. This finding is considered to be more often associated with symptoms than the non-invasive type [2]. Local recurrence is attributed to incomplete resection of the primary lesion in cases of invasive PCG. Metastasis of the tumor in mediastinum or the brain even many years after complete resection has been also described [2,3]. Rarely, simultaneous intra- and extrathoracic locations may occur. Association with other malignancies in sporadic cases has been reported [3].

Malignant lymphoma and low-grade sarcoma may have areas resembling PCG [3]. Differential diagnosis includes lymphoma, sarcoma, malignant fibrous histiocytoma,

<table>
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<th>Right lower lobe (cm)</th>
<th>Tumor</th>
<th>Symptoms</th>
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<tr>
<td>Maximal diameter (cm)</td>
<td>Distance from surgical margins (cm)</td>
<td>Cough, fever, recurrent pulmonary infections</td>
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<tr>
<td>13 × 10 × 3.5</td>
<td>3.1</td>
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malignant plasmacytoma and fibrosis [2,3]. Histopathological identification can be sometimes very difficult. In this context, PCG has not to be considered as a strictly distinct histological entity, but incorporated into a wide spectrum of pathologies ranging from benign fibrohistiocytoma or inflammatory pseudotumor, fibrohistiocytic type, to fibrohistiocytic lesions of borderline malignant appearance, to unequivocal malignant fibrous histiocytoma [6].

Special interest has developed in the etiology and the nature of the lesion, which were both considered unknown for some time. Our attitude about the histological identity of PCG has been changed: PCG has been accepted to be benign neoplastic tissue rather than inflammatory or reactive lesion [7]. This concept has been recently challenged from clinical demonstration of recurrence and cytogenetic evidence of acquired clonal chromosomal abnormality [7,8]. In recent years, PCG has been increasingly recognized in other sites such as liver, viscera, pelvis and soft tissue.

Pathogenesis of the lesion has been attributed to an aberrant or exaggerated response to tissue injury without an established cause.

Fig. 2. Pathological specimen. (a) The spindle mesenchymal cells (presumably fibroblasts) are very prominent, suggesting the possibility of a connective tissue tumor. There are no mitosis or atypia. Some plasma cells are present. HE, × 10. (b) Plasma cells are the most numerous cells, with only scattered histiocytes. HE, × 20.
The treatment of choice is unequivocally surgical resection, though steroid administration has been reported to be efficacious. Spontaneous regression has been also described. Radiotherapy should be considered in patients who had incomplete surgical resection or postoperative recurrences and in patients with non-resectable disease due to associated medical conditions. The role and effectiveness of radiotherapy remains controversial. Application of chemotherapy seems to be without benefit. Prognosis of these rare tumors appear to be excellent for pulmonary PCG after complete surgical excision, while more aggressive behavior has been described in extrapulmonary PCG, which are supposed to be of a higher malignant potential.

Consequently, PCG is a rare benign neoplasm mimicking cancer in clinical and radiological presentation. As preoperative investigation is not diagnostic, excision of the mass is imperative in order to exclude malignancy. Frozen section may be inconclusive. In any case, complete removal of the tumor is the treatment of choice. Complete resection remains the key to prevent recurrence. If recurrence occurs, re-resection is needed.

References