Case report

The evolution of a pleural nodule into a giant fibrous tumor associated with hypoglycemic coma

Antonio D’Andrilli*, Claudio Andreetti, Mohsen Ibrahim, Erino A. Rendina

Department of Thoracic Surgery, University of Rome “La Sapienza”, “Sant’Andrea” Hospital, Rome, Italy

Received 7 November 2006; received in revised form 12 January 2007; accepted 23 January 2007; Available online 20 February 2007

Abstract

We hereby present the exceptional case of hypoglycemic coma associated with a giant benign localized fibrous tumor of the pleura (LFTP). A 79-year-old woman was found to have a small peripheral nodular lesion in the right hemithorax 7 years ago. A fine needle aspiration biopsy provided a diagnosis of LFTP, but the patient refused surgery. Six years later hypoglycemic coma and respiratory insufficiency appeared in association with a giant mass completely filling the right chest and shifting the mediastinum contralaterally. The tumor was completely resected, and complete relief of the hypoglycemic and respiratory symptoms was achieved. The patient is in good metabolic and respiratory conditions and disease-free 14 months after the operation.

© 2007 European Association for Cardio-Thoracic Surgery. Published by Elsevier B.V. All rights reserved.

Keywords: Localized fibrous tumor of the pleura; Pleural neoplasm; Hypoglycemia

1. Introduction

Benign localized fibrous tumors of the pleura (LFTP) are rare, usually slow-growing neoplasms. They present with greatly variable size, ranging from small nodules to large masses occupying the entire hemithorax. Their association with respiratory insufficiency depends on the size, and the concomitant occurrence of hypoglycemia is exceptional [1]. Hereby, we report the exceptional case of hypoglycemic coma and respiratory insufficiency associated with a giant benign LFTP.

2. Case report

Seven years ago a 79-year-old woman was incidentally found to have a nodular 2.5 cm lesion of the right parietal pleura by means of chest-X-ray and subsequent CT scan of the thorax (Fig. 1a). Fine needle aspiration biopsy (FNAB) provided a diagnosis of benign LFTP. The patient refused surgery due to her advanced age and to the diagnosis of benign lesion (Fig. 2).

Two years later the woman repeated a chest-X-ray that documented minimal size variation of the pleural lesion. No further radiological control was performed since that time.

Fourteen months ago the woman required an urgent hospital admission with the diagnosis of hypoglycemic coma and respiratory insufficiency. Glycemic level was 20 mg/dl. The related neurological symptoms relieved after prompt glucose administration. The CT scan of the chest showed a giant mass filling the entire right hemithorax and shifting contralaterally the mediastinum (Fig. 1b and c). The patient was transferred to our Division for surgical treatment. The patient presented persistent hypotension and edema at the lower limbs. Arterial blood gases showed severe hypoxemia and hypercapnia (PaO2: 46 mmHg; PaCO2: 69 mmHg). Recurrent hypoglycemic episodes requiring glucose administration were registered. Pulmonary function tests documented severe restrictive and obstructive disease (FEV1: 27.8% of the predicted value). The diagnosis of benign LFTP was confirmed by FNAB. Flexible bronchoscopy showed extrinsic compression of the right main bronchus with displacement of the carina contralaterally without neoplastic infiltration. The origin of all the right segmental bronchi was markedly restricted.

The patient underwent a right posterolateral thoracotomy. Several thin-wall and easily bleeding vascular adhesions were found between the neoplasm and the adjacent parietal and visceral pleura. The base of attachment was identified on the parietal pleura in the upper posterior region. The neoplasm entirely filled the hemithorax and the lung was collapsed at the hilum. A complete removal of the tumor was performed after adequate mobilization. Intensive Care Unit admission was required in the first 24 h after surgery for hemodynamic monitoring. The tumor size was 22 cm ×
16 cm × 13 cm and it weighed 2250 g. Macroscopic examination showed a smooth-surfaces gray tumor with a thin membranous capsule. On cut section the neoplasm was composed of dense fibrous tissue with cystlike partially hemorrhagic structures. Spindle-shaped CD 34 positive cells, partially organized in storiform pattern, were identified. No nuclear pleomorphism was found. Mitotic count was 2 mitoses/10 high-power-fields (HPF).

No major intraoperative or postoperative complication occurred. The only minor complication in the postoperative course was a persistent air leak that stopped 7 days after surgery. Respiratory insufficiency and recurrent hypoglycemia completely regressed after resection. Postoperative in-hospital stay was 9 days. The patient is in good respiratory and metabolic conditions and disease free 14 months after surgery. The CT scan of the chest performed 1 year after surgery shows the complete right lung re-expansion.

3. Discussion

LFTP are benign in over 80% of the cases [1,2]. Most of these tumors are found incidentally on routine chest radiograph, since related symptoms are not present in over 50% of the cases [3], especially when the lesion is small sized. Symptoms tend to be more common in larger lesions and may include local symptoms such as dyspnea, chest pain and chronic cough as well as systemic symptoms including fever, weight loss, hypertrophic pulmonary osteo-arthropathy with or without digital clubbing, and more rarely hypoglycemia [1,3—7]. Sporadic cases of hemoptysis and galactorrhea have been reported [1]. Complete regression of all symptoms is systematically observed after radical surgical treatment [1—7].

Severe hypoglycemia has been reported in association with large localized pleural tumors in 2—4% of the patients. Tumor production of insulin like active substances and insulin like growth factor have been recently indicated as the most likely mechanisms responsible for hypoglycemic events [1,3,6,7]. The hypothesis of an increased glucose consumption by the tumor has been also substantiated as one of the possible explanation of the hypoglycemic phenomenon [1].
The presentation of a LFTP with hypoglycemic coma associated with respiratory insufficiency is exceptional and no similar case has been reported in literature [1].

The sudden appearance of a severe respiratory insufficiency a few days before surgery, demonstrates that also symptoms related to a giant mass may occur very late, when a mediastinal shifting is already present.

The clinical behavior of benign LFTP may be variable. Although most of these tumors generally show a slow growth, cases with rapid enlargement or sudden growth increasing have been observed. In our experience, the small lesion identified 6 years before surgical intervention remained nearly unmodified for 2 years, but it was found with a ten times greater diameter after the following 4 years. To the best of our knowledge only one case of resected LFTP larger than this has been reported in the English literature [4].

A malignant transformation of benign LFTP has been sometimes reported and malignant features in recurrences of originally benign tumors have been described [2,5,7]. Based on this evidence, surgical resection should always be recommended for patients with acceptable operative risk. Long-term follow-up should be the rule after surgery and should also be recommended to patients excluded from surgical therapy, as a means to control the evolution of the disease and the possible related risks.

Almost all patients with benign LFTP are cured by complete excision of the lesion [1,6,8]. Local recurrence rates are generally reported under 2% [2,6], although recurrence rates as high as 10% have been observed in some series [4]. However re-resection is usually possible. Compleness of the resection and limited size of the neoplasm represent the principal favorable prognostic factors [3,4,6].

References