Well-differentiated giant "lipoma-like" liposarcoma of the posterior mediastinum: a case report

MASSIMILIANO PACI, SALVATORE DE FRANCO, ALBERTO CAVAZZA*, VALERIO ANNESSEI, GUGLIELMO FERRARI, FRANCESCO FALCO**, GIORGIO SGARBI

Division of Thoracic Surgery
*Department of Pathology
**Division of Respiratory Disease
Santa Maria Nuova Hospital - Reggio Emilia

Riassunto
Il liposarcoma è la neoplasia mesenchimale maligna più frequente nell'adulto ma la sua localizzazione mediastinica è rara. Nell'articolo descriviamo il caso di una donna di 74 anni con una massa del mediastino posteriore con impegno a clessidra di entrambi gli emitoraci, probabilmente già presente ad una radiografia di 4 anni prima. Alla TC del torace la massa appariva disomogenea ed a prevalente contenuto adiposo. Una biopsia trans-toracica eco-guidata con tru-cut, eseguita preoperatoriamente, aveva dimostrato la presenza di tessuto adiposo con aspetti citologici moderatamente atipici. La massa è stata pertanto completamente asportata per via toracotomica destra. Il decorso postoperatorio è stato regolare, la paziente è viva e sta bene, senza evidenza di recidiva, a 6 mesi dall'intervento. In questo report rivediamo la storia naturale, la patologia e la prognosi di queste lesioni. E ne discutiamo i metodi di diagnosi e gestione chirurgica.

Parole chiave: tumori del mediastino, liposarcoma

Summary
Well-differentiated giant "lipoma-like" liposarcoma of the posterior mediastinum: a case report. M. Paci, S. De Franco, A. Cavazza, V. Annessi, G. Ferrari, F. Falco, G. Sgarbi

While liposarcoma is the most common malignant mesenchymal neoplasm in adults, a mediastinal position is rare. We describe here the case of a 74-year-old female with an hourglass-shaped mass in the posterior mediastinum, which was probably present on a roentgenogram 4 years earlier. On a CT scan of the chest, the mass appeared non-homogeneous with a mainly adipose content. A preoperative ultrasonically guided tru-cut transthoracic biopsy revealed the presence of adipose tissue with mildly atypical cytological features. The mass was therefore completely excised by means of a right thoracotomy. The postoperative course was uneventful and the patient is alive and well and has had no recurrence in the 6 months since surgery. We review here the natural history, pathology, and prognosis of the disease, and discuss methods of diagnosis and management of such lesions.

Key words: mediastinal neoplasms, liposarcoma

Chir Ital 2003; 55, 1: 101-104

Correspondence to: Dr. Giorgio Sgarbi - 1st Department of Surgery, Division of Thoracic Surgery - Santa Maria Nuova Hospital - Viale Risorgimento, 80 - 42100 Reggio Emilia, Italy.
Introduction

Liposarcoma is a malignant mesenchymal neoplasm that rarely involves the mediastinum. It strikes both men and women, with no difference in incidence rates, at an average age of 45. While most patients are asymptomatic, symptomatic patients present primarily respiratory symptoms, general chest pain, and, rarely, signs and symptoms of superior vena cava obstruction. Dysphagia is sometimes present for liquids and solids, especially in the posterior sites. In 25% of cases there is significant weight loss. Radiographically, liposarcoma presents as a lobulated mass with ill-defined borders that compresses and may infiltrate adjacent structures. Its density is mostly similar to that of a benign lipoma. We describe the case of a 74-year-old female with a malignant mesenchymal tumour of the posterior mediastinum presenting the histological characteristics of a well-differentiated "lipoma-like" liposarcoma. We comment on the natural history, pathology, prognosis, diagnostic methods and surgical management of the disease.

Case report

A 74-year-old non-smoking Caucasian woman presented to the Emergency Department with dyspnoea and dry cough which had been troubling her for some weeks, and with recent onset of dysphagia for solids and liquids and chest pain. A radiography of the chest showed the presence of an enlargement of the cardio-mediastinal shadow with opacity that completely occupied the posterior mediastinum and gave rise to atelectasis of the lower pulmonary lobes. The patient reported the presence of a radiologically evident mediastinal alteration detected 4 years earlier for which no documentation was available. The patient had had neither fever nor weight loss in the preceding months. Her case history included a bilateral hip prosthesis operation for coxarthrosis and post-transfusional hepatitis C. A reduction of vesicular murmur was clinically evident at the pulmonary base bilaterally. The patient underwent a CT scan of the chest which showed the presence of a voluminous hourglass-shaped mass of the posterior mediastinum measuring 21x12 cm, which was unhomogeneous and primarily composed of adipose tissue, affecting both hemithoraces on passing from the aortic arch to the posterior diaphragm recess. The mass was compressing and dislocating the main bronchi, the segments and sub-segments of the basal pyramid, and the heart towards the back causing atelectasis of the compressed pulmonary parenchyma. There were no macroscopic signs of infiltration (Fig. 1). A CT scan of the upper abdomen confirmed the presence of a voluminous mass of fatty density with retrodiaphragmatic extension and anterior dislocation of the liver and spleen. Oncological markers and CT scan of the brain yielded negative findings. A bronchoscopy revealed a widespread picture of chronic bronchitis and a right trans-bronchial needle aspiration showed the presence of fatty tissue. Respiratory function tests revealed a picture of mixed ventilatory insufficiency of significant magnitude. In order to obtain a pre-operative diagnosis we carried out an ultrasonically guided true-cut transthoracic biopsy. The biopsy sample was composed primarily of adipose tissue, with mildly atypical cytological features. As the possibility of a lipomatous tumour was suggested, surgical removal was recommended. By means of a right thoracotomy of the sixth intercostal space we carried out the complete removal of the neoplasm through the posterior mediastinum. The neoplasm appeared to be cleavable from the adjacent structures, with no macroscopic residues at the end of the operation. The neoplasm also appeared to be macroscopically encapsulated and was made up primarily of adipose tissue and weighed approximately 1800 g. On cut section it appeared yellowish with focal necrotized and haemorrhagic areas (Fig. 2). Microscopically, the tumour was composed of adipocytes exhibiting marked variation in cell size. Numerous hyperchromatic stromal cells and scattered lipoblasts were encountered, together with areas of ischaemic necrosis.
posterior position. There is no statistically significant difference between the incidence in men and women and most patients are over 40, with an average age of 45\textsuperscript{2}. There is, however, a very low incidence of childhood cases, only a few of which have been described in the literature\textsuperscript{2}; even rarer at this age is the mediastinal location\textsuperscript{6}.

Enzinger and Weiss\textsuperscript{1} classify these tumours as belonging to 5 types: 1) well-differentiated, 2) myxoid, 3) round cell, 4) dedifferentiated, and 5) pleomorphic. Well-differentiated tumours are further classified (8) as: 1) adipocytic (lipoma-like), 2) sclerosing, 3) inflammatory, and 4) spindle-cell. The most common of these is the myxoid liposarcoma, which accounts for 40\%-50\% of cases. Unlike the well-differentiated types, whose low grade of malignancy has recently led to the proposal that they should be called lipomatous tumours\textsuperscript{6}, the other forms are more aggressive.

From a diagnostic viewpoint, CT scans and MRIs are unable to distinguish between the benign and malignant forms except when it is a question of poorly differentiated forms\textsuperscript{1}. It is recommended, when possible, to perform an ultrasonically or CT-guided tru-cut transthoracic biopsy, which may be of assistance when choosing the therapeutic course. In the case described here, in fact, the tru-cut biopsy provided a diagnosis of lipomatous tumour and prompted us to perform surgical excision of the mass with the aim of oncological radicality. Whenever possible, the choice of treatment should be to completely excise the tumour surgically, as subtotal resections are often subject to early recurrence. Adjuvant chemotherapy and postoperative radiation therapy are of limited benefit and their usefulness has been confined to cases of incomplete surgical resection\textsuperscript{9}.

We opted for a right thoracotomy in order to excise the bulkier part of the tumour, intending to excise the left residue later. However, while operating, the mass presented as completely encapsulated and well cleavable from the adjacent structures. This being the case, it was possible to perform a complete bilateral excision of the neoplasm through the posterior mediastinum with no evidence of macroscopic residues.

The prognosis of liposarcoma is essentially linked to its histology, to the presence of pseudocapsules, and to tumour differentiation. Patients with nonencapsulated forms or with tumours that are less well differentiated have a poor prognosis, with an average survival of approximately two years\textsuperscript{6}. Myxoid liposarcomas, the most common form of lesion, develop local recurrences in 50\% of cases.
Conclusion

The case we describe here is a voluminous encapsulated liposarcoma of the posterior mediastinum made up primarily of circumscribed tumour tissue, for the most part well defined, with a lipoma-like, sclerosing appearance and myxoid focal areas. The excision was complete without any apparent macroscopic residues, although its myxoid nature places this lesion among those presenting the highest rate of local recurrence. The patient is currently in good health and a follow-up chest CT scan revealed no signs of local recurrence of the disease 6 months after the operation.

References