Liposarcomas are extremely rare in the mediastinum. They may achieve considerable size before causing any symptoms. Mediastinal liposarcomas may invade surrounding structures like the pericardium or the superior vena cava. Complete surgical excision is the optimal treatment in resectable cases. Excision of adjacent structures like the pericardium may be needed if the tumor infiltrates them. We report on a case of a giant liposarcoma of the mediastinum involving both hemithoraces and extending into the neck, which was successfully managed by complete surgical excision. (Ann Thorac Cardiovasc Surg 2006; 12: 425–7)

Key words: mediastinal, liposarcoma

Introduction

Liposarcomas are the most common soft tissue sarcomas in adults. However, mediastinal liposarcomas are extremely rare. They constitute 2.7% of all liposarcomas and less than 1% of all mediastinal tumors. We report on a rare case of a giant liposarcoma of the mediastinum involving both hemithoraces and extending into the neck.

Case Report

A 62-year-old man presented to us with dyspnoea at rest, dysphagia to solids and change of voice. Physical examination revealed vague fullness in the neck, predominantly on the left side. The chest was dull to percussion and air entry was reduced in the right upper zone. The trachea was shifted to the right. Barium swallow revealed a large retropharyngeal prevertebral mass displacing the posterior pharyngeal wall, trachea and postcricoid region anteriorly, with intrathoracic extension of the mass pushing the esophagus laterally; the esophageal lumen appeared regular. Esophagoscopy and endoscopic ultrasound (EUS), showed a large well encapsulated hyperechoic mass extending between the vertebral space and the esophagus from 15 to 30 cm. This mass displaced the aorta and compressed the esophageal lumen. However, it was free from the esophageal wall and other surrounding structures. Fibreoptic bronchoscopy (FOB) showed decreased movement of the left vocal cord with a bulge in the posterior wall of the pharynx, posterior wall of the trachea and both main bronchi. The tracheal lumen was compromised by about 50% and the posterior wall of the trachea and both bronchi were mobile.

A computed tomography (CT) scan revealed a large, well-defined mass. This mass extended from the deep cervical space in the neck at C2 vertebral level down through the posterior mediastinum to the level of the carina on both sides of the vertebral column (Figs. 1A–1C). The mass displaced the left half of the thyroid gland, the sternocleidomastoid and the common carotid artery in the neck and trachea and esophagus in the thorax. A fine needle aspiration cytology (FNAC) attempted from the neck was nondiagnostic. The patient had adequate pulmonary function and surgical exploration was carried out. The neck was first explored and the mass was carefully dissected and separated from the esophagus and the carotid sheath. A right thoracotomy (Fig. 2) was then performed and the mass was separated from the esophagus, trachea and superior vena cava and delivered en masse. The patient tolerated the surgery well and had an uneventful postopera-
tive recovery. The final histopathology showed a spindle cell tumor with lipogenic character suggesting a well-differentiated liposarcoma (Fig. 3). On immunohistochemistry, it expressed S-100 focally.

**Discussion**

Liposarcomas arise from primitive mesenchymal cells. They are almost always encountered in deeper structures as insidiously growing tumors. Mediastinal liposarcomas may extend into the pleural spaces and achieve a large size before detection. The presenting signs and symptoms are related to size and direct invasion of contiguous structures like the pericardium or superior vena cava. Dyspnoea, chest pain and tachypnea are the most common symptoms. Asymptomatic cases discovered by radiologi-
Mediastinal liposarcomas arise from the thymus-related fatty tissue in the anterior mediastinum but also might occur in the posterior mediastinum. On CT, the appearance of mediastinal liposarcomas varies from a predominantly fat-containing mass to a solid mass. Soft tissue or solid densities are related to the necrosis, heterogeneity and soft tissue component in liposarcomas. On magnetic resonance imaging (MRI), T1-weighted images show the fatty tissue with high signal intensity, whereas the signal intensity diminishes in T2-weighted images. MRI is better than CT scanning in ruling out invasion of vessels in the mediastinum and thoracic inlet.

Surgical removal is the optimal treatment for a mediastinal liposarcoma, as in other sites. Partial excision or debulking of the tumor may relieve the compression effect of more advanced and infiltrating tumors. Excision of adjacent structures like pericardium may be done if the tumor infiltrates them. Liposarcomas have very low sensitivity to radiotherapy and chemotherapy. High doses of radiation may result in mediastinal fibrosis, thus precluding radiotherapy in this location. In a single case report postoperative radiotherapy resulted in long-term survival of 5 years. The role of chemotherapy is very limited.

Recurrence is common in deep-seated liposarcomas and becomes apparent within the first 6 months; in most cases it is probably related to the incomplete excision at the time of primary surgery. The paucity of reported cases in literature precludes strong recommendations on adjuvant treatment and prognostication.

References