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Case report

Giant primary liposarcoma of the thoracic esophagus

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Summary

Primary liposarcomas of the mediastinum are unusual tumors. We report herein a case of a 64 year-old man, who presented a liposarcoma involving the mediastinum and the neck. A complete resection was performed and we had a free-disease survival result of eighteen months.

Keywords: Esophagus; Mediastinal tumor; Sarcoma; Surgery

Un liposarcome géant de l'œsophage thoracique

Résumé

Les liposarcomes primitifs du médiastin sont rares. Nous rapportons le cas d'un patient de 64 ans de sexe masculin, présentant un liposarcome envahissant le médiastin et la région cervicale. Une résection complète a été réalisée et nous avons un recul d'évolution favorable de 18 mois.

Mots-clés: Chirurgie; Œsophage; Tumeur médiastinale; Sarcome

Introduction

Liposarcoma, the most common soft tissue sarcomas in adults [1], is characterized by multiple recurrences. No common approach in diagnosis and treatment of these tumors has been described. We report herein a removal of a giant primary liposarcoma of the esophagus extended to the pleural spaces and the neck. We had free-disease survival of eighteen months.

Case report

A 64 year-old man presented a vena cava syndrome in July 2004. Chest x-rays revealed a round opacity occupying the upper right of the chest (Figure 1). Flexible bronchoscopy was normal. The computed tomography (CT) scan confirmed a heterogeneous mass in the posterior mediastinum with fat density arising from the esophagus, merely invading the supra aortic trunks and the left cervical area (Figure 2). In intraoperative view the tumour involves the posterior wall of the esophagus. The patient underwent complete resection of this mass by a right posterolateral thoracotomy with a cervicotomy (Figure 3 and 4). We realised an esophagectomy with double exclusion and a transpyloric gastrostomy. A jejunostomy was performed to prepare a possible coloplasty. Sacrifice of the ipsilateral recurrent nerve was needed. Postoperative course was uneventful despite a left recurrent palsy caused by a radical systematic mediastinal lymphadenectomy of 4L station. Pathology results diagnosed an undifferentiated liposarcoma with incidental heterelogous of level II according to the grading of the Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) without lymph nodes invasion. Re-establishment of digestive continuity was carried out in October 2004 by a retrosternale gastroplasty with cervical esogastric anastomosis. A thyroplasty by silastic prosthesis was also realised in postoperative time because reintubation have compromised the plasty. In December 2005, the CT scan follow-up showed a pleural nodule in right basal hemithorax. The abdominal CT scan showed a high suspect hypodensity nodule in segment V of the liver which did not exist on previous scanners. The positron emission tomography scan in January 2006 confirmed a hyperfixation on the liver and a right pleural nodule. Chemotherapy was decided in February 2006 for the double hepatic and right pleural metas-

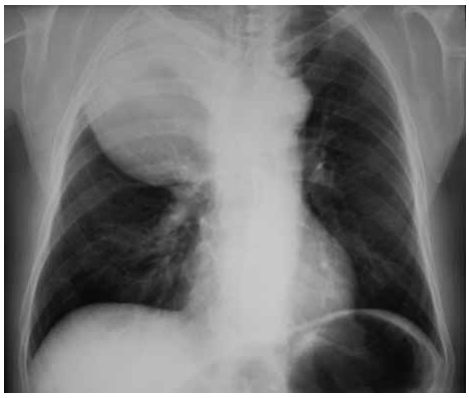

Fig. 1: Chest x-ray showing a mediastinal tumor

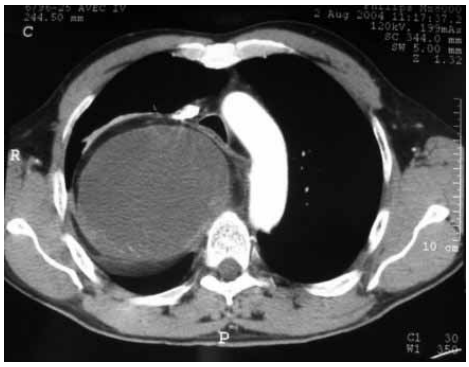

Fig. 2: Axial CT scan demonstrating posterior mediastinal tumor with pleural space invasion

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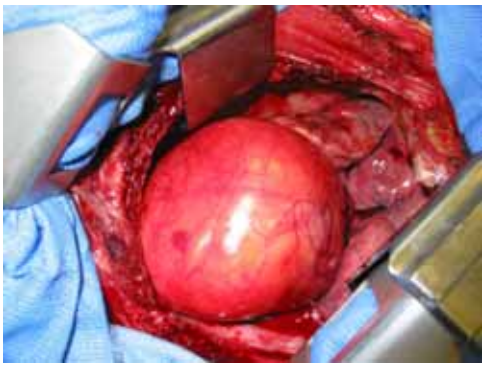

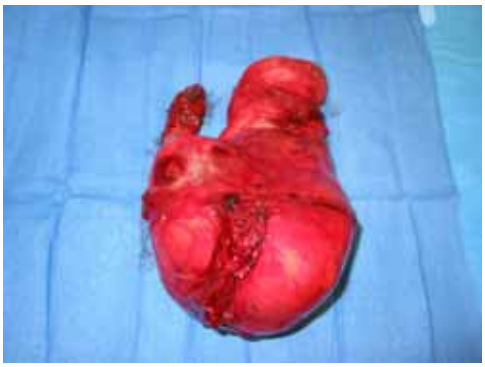

Fig. 3 and 4: Peroperative view of the esophagus tumor and the specimen after removal

tatic localization and there after a resection of the hepatic and pleural residual masses. Consequently the patient had a free-disease survival of eighteen months.

Comment

Mediastinal liposarcomas respectively constitute 2.7 and 0.12% of all liposarcomas and soft tissue sarcomas, [1,2]. Liposarcomas are almostly encountered in deeper structures as insidiously growing tumours. In our case, liposarcomas occurring in the posterior mediastinum extend into the pleural spaces and they may achieve a huge size before detection. [1]. Its signs and symptoms are related to size and direct invasion of contagious structures. In addition, asymptomatic cases discovered by radiological imaging also have been reported [3,4]. We described a liposarcoma arising from posterior wall of the oesophagus. Mediastinal liposarcomas, most commonly arise from the thymus-related fatty tissue in the anterior mediastinum also might occur in the posterior mediastinum [4,5]. A complete resection of such a mass is very difficult but feasible. However a differed re-establishment of digestive continuity was selected to prepare a possible coloplasty. Liposarcomas have very low response, both to radiotherapy and chemotherapy. Nowadays, therefore, complete surgical removal is the optimal treatment in this location [6,7]. The frequency of local recurrence is high (50% to 90%) [8]. We had a free-disease survival of eighteen months. Recurrence is common in deep-seated liposarcomas and it becomes apparent within the first 6 months in most cases, but it may be delayed within 5 to 10 years following the initial excision. Interestingly, surgical complete resection plus adjuvant therapy was suggested to be effective for treatment of a subset of diffusely infiltrating none capsulated mediastinal liposarcomas [9,10].

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