Case report

Long-term survival after multiple resections of a fibrosarcoma involving the lung and chest wall

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Abstract

We report on the case of a 61-year-old male patient who developed a giant fibrosarcoma involving both the lung and chest wall. This patient underwent three extended resections including the chest wall in each case. Radiotherapy was administered after the last resection, when the tumor was obviously not completely removed. The patient lives a normal life with no signs of recurrence 5 years after his last resection. Multiple extended resections of large and aggressive sarcomas can result in long-term survival, with good quality of life, in adequately selected patients.

Keywords: Fibrosarcoma; Lung; Chest wall

1. Introduction

Fibrosarcomas of the lung are rare tumors accounting for less than 0.05% of all primary lung malignancies [1]. Their clinical behavior is often unpredictable, but most of the patients die within 5 years after diagnosis if the tumor is invasive [2]. We report on a patient who is alive and disease-free 5 years after the third resection of a giant invasive fibrosarcoma. This case stresses the importance of considering surgery in selected patients with large and aggressive sarcomas.

2. Case report

A 61-year-old male patient, smoker, presented in 1993 with an enlarging right lung mass causing persistent chest pain. A pulmonary nodule had been detected in his right upper lobe in 1985, but the patient declined surgical treatment at that time. Despite the long evolution time, the patient did not have metastasis and eventually underwent a right upper lobectomy with localized chest wall resection. The tumor measured 9.6 cm in greatest diameter and was well demarcated. Histologically, the tumor was densely cellular with rare fibrotic areas. The cells were spindle shaped with pleomorphic hyperchromatic elongated nuclei and were arranged in a classical herringbone pattern. A high mitotic count of about 20 per 10 high power fields (HPF) and necrotic areas were observed. All resection margins were histologically free of tumor. Immunohistochemistry was positive for vimentin, but negative for keratin, S-100 protein, desmin, and actin, suggesting the presence of a sarcoma rather than a mesothelioma. One year later, the patient presented with a large anterior right-sided mediastinal mass and was referred to our institution for further treatment. On computed tomography (CT) and magnetic resonance imaging (MRI), the tumor extended from the aortic arch down to the diaphragm, invading the anterior chest wall and sternum, compressing the superior vena cava, ascending aorta, and right atrium (Fig. 1). Surgery was performed through a hemi clamshell incision in the fourth intercostal space. The tumor was resected en bloc with six ribs, the upper two thirds of the sternum, parts of the right major pectoralis muscle, and the right middle lobe. The mediastinal structures were not invaded, but severely compressed. The central venous pressure dropped from 24 to 10 mmHg immediately after the tumor was lifted off the superior vena cava. The chest wall was reconstructed with Marlex mesh and methyl-methacrylate (sandwich technique). Histologically, the tumor was identical to the previous one. Immunohistochemistry was once more positive for vimentin and negative for keratin. Additional immunostaining including Cam 5.2, epithelial membrane antigen (EMA), ...
CD34, and α-smooth-muscle actin (α-SMA) was performed and remained negative.

Six months later, the patient developed another recurrence, this time located in the right posterior costovertebral sulcus. No metastatic disease was observed on CT of the head, chest, and abdomen and on the bone scan. A third resection was undertaken through a right posterolateral thoracotomy. Part of the posterior chest wall was resected, but the tumor could not be completely removed because of invasion of the intervertebral foramen. Therefore, external radiotherapy was administrated postoperatively with 6000 cGy to the residual site. The patient is now doing well 5 years after this last resection with a satisfying quality of life and no restriction to his daily activities (Fig. 2). Recent CT and MRI scans of the chest showed no signs of recurrence.

3. Comment

Fibrosarcomas are potentially highly malignant tumors originating from mesenchymal cells and occurring in various anatomic sites including the lung and chest wall [2–4]. In the current case, both the lung and the chest wall were involved at the time of the first resection, therefore the primary origin of the tumor remains speculative. In the chest, fibrosarcomas must be differentiated from fibrous mesotheliomas, malignant fibrous tumors of the pleura, and other sarcomas such as myxofibrosarcoma, synovial sarcoma, or nerve sheath sarcoma [2,3]. Over the last few years, immunohistochemistry has become extremely helpful to differentiate between these tumors. Mesotheliomas always stain positive for keratin, whereas fibrosarcomas, fibrous tumors of the pleura, and most other sarcomas remain negative for keratin and stain positive for vimentin.

Fig. 1. MRI of the first recurrence. The tumor extended from the aortic arch down to the diaphragm invading the anterior chest wall and the sternum, and compressing the superior vena cava, the ascending aorta and the right atrium (T, tumor; H, heart; LL, left lung).

Fig. 2. Patient’s photograph (A) and chest-X-ray (B) showing successful reconstruction of the chest wall with satisfying anatomical results after the third resection.
Additional markers such as S-100 protein, desmin, or α-SMA allow further differentiation between the different types of sarcoma [6]. The distinction between fibrosarcomas and solitary fibrous tumors of the pleura can be performed by staining for CD34, which is a transmembrane cell surface glycoprotein, originally described as a marker of human hematopoietic stem cells, and now ubiquitously observed on a novel family of interstitial spindle cells characterized by slender dendritic prolongation of their cytoplasm and involved in antigen presentation [7]. Although the absence of positive immunostaining for CD34 suggests the diagnosis of fibrosarcoma, a solitary fibrous tumor of the pleura with negative staining for CD34 cannot be completely excluded in the current case [7].

The 5-year survival of patients operated for primary sarcoma of the lung usually ranges between 40 and 50%, but the grade of malignancy, the size of the tumor, and the stage of disease have been shown to impact on the prognosis [2,3,8]. The current tumor was of high-grade malignancy and was locally extremely aggressive as demonstrated by the size of the tumor and its infiltration of the lung and chest wall. The tumor had been growing for several years before its surgical resection and, thus, may have degenerated and become progressively more aggressive over time.

The role of adjuvant therapy in primary pulmonary sarcoma is not well established and is usually recommended for high-grade malignancy and/or when the resection margins are not microscopically free of tumor [3,8]. In the current case, adjuvant radiotherapy was administered after the last resection because of tumoral extension into the intervertebral foramen. Since the patient is alive and disease-free 5 years later, the tumor must have been radiosensitive and one may speculate that adjuvant radiotherapy may already have been beneficial to prevent or delay local recurrence after the first surgical procedure.

In conclusion, the current case stresses that multiple resections of large and aggressive sarcomas can result in long-term survival with good quality of life in adequately selected patients.

References