Primary Leiomyosarcoma of the Anterior Mediastinum Encasing the Aortic Arch, Left Common Carotid and Left Subclavian Arteries

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A 66-year-old man presented with a one month history of hoarseness. Left recurrent nerve palsy and a left upper mediastinal mass were observed by an otorhinolaryngologist who referred the patient to our department. Chest computed tomography showed a superior mediastinal mass, which seemed to involve the left common carotid and left subclavian arteries from the greater curvature of the aortic arch. The innominate vein was compressed, and collateral circulation was well developed. The left upper lobe of the lung was also seemed involved. A mediastinal biopsy conducted via left thoracoscopy revealed a malignant spindle cell tumor. The mediastinum was irradiated (40 Gy), and surgical extirpation was subsequently undertaken 3 weeks later. The tumor was successfully removed without the use of extracorporeal circulation. Because only smooth muscle actin was focally but strongly expressed immunohistochemically, leiomyosarcoma was confirmed. The patient was discharged on day 14. A solitary left pleural metastasis was observed and resected 12 months after the surgery and the patient is well without further recurrence 16 months after the initial surgery.

Keywords: leiomyosarcoma, mesenchymal tumor, surgery, mediastinum

Introduction

Primary mediastinal leiomyosarcoma is an extremely rare entity. Because of its rarity, the details of this disease are still relatively unknown. The majority of the tumors are thought to arise from smooth muscle cells of the soft tissue in the mediastinum.1–3) The behavior of the tumor is often aggressive, and radiotherapy is not effective.4) Thus, the prognosis is very poor, and complete surgical resection is considered being the best and only curative treatment option.5,6) We hereby present a case of leiomyosarcoma involving the great vessels in the anterior mediastinum, which was successfully resected without the use of extracorporeal circulation.

Case Report

A 66-year-old man presented with hoarseness from which he had suffered for a month. He consulted an otorhinolaryngologist who observed left recurrent nerve palsy. A left upper mediastinal mass was also disclosed in a chest radiograph, and the patient was referred to our department. The patient smoked 15 cigarettes a day for 48 years and had been healthy without any history of chronic illness. He had lost 5 kg in weight in the last 3 months. He did not show any facial or peripheral edema.
Peripheral arterial pulses were not weakened. Lymph nodes could not be systematically palpated. The results of blood chemical studies, which included the analysis of serum tumor markers, such as carcinoembryonic antigen, squamous cell carcinoma antigen, cytokeratin 19 fragment, sialyl Lewis X antigen and progastrin-releasing peptide, were all within normal ranges. A chest X-ray showed a well-demarcated round-shaped mass of 7 cm in diameter in the left superior mediastinum. A chest computed tomography (CT) scan showed that the superior mediastinal mass involved the left common carotid artery and left subclavian artery from the greater curvature of the aortic arch (Fig. 1A). The innominate vein was compressed (Fig. 1B), and the development of collateral circulation was visualized. The left upper lobe of the lung seemed to be involved with the tumor, as well. A 3D-reconstructed view showed smooth but completely narrowed left common carotid and left subclavian arteries (Fig. 1C). A positron emission CT scan using fluorodeoxyglucose did not show any metastatic lesions but did show a high accumulation in the main tumor (the sum of the maximum standard uptake values was 12.33). A mediastinal biopsy was conducted by left thoracoscopy, which revealed a malignant tumor that consisted of spindle cells, suggesting the differential diagnoses of leiomyosarcoma, fibrosarcoma, a malignant peripheral nerve sheath tumor, monophasic fibrous synovial sarcoma, malignant hemangiopericytoma, malignant histiocytoma and the sarcomatous subtype of malignant mesothelioma.

From the findings of the enhanced CT, we thought that the occlusion of the left subclavian artery was impending. Thus, the tumor was irradiated with 40 Gy emergently. According to the result of histopathological examination of the biopsy specimen, vascular leiomyosarcoma, which is known as chemo-resistant malignancy, was highly suspected. The tumor size was gradually growing even after the radiotherapy. Thus, surgical extirpation was subsequently undertaken 3 weeks later without neoadjuvant chemotherapy.

A median sternotomy was done with a left neck collar incision. Videothoracoscopy was used for the manipulation in the left pleural cavity. A total arch replacement had been planned, and so extracorporeal circulation was standing by. The innominate vein was transected for a better view. The sternocleidomastoid, sternothyroid, and sternothyroid muscles were transected. In order to get a wider view, the left sternal edge was elevated with an internal thoracic artery-harvesting retractor. The left common carotid artery and left subclavian artery were secured just peripheral to the tumor. The left internal thoracic artery, left vagal nerve, and left phrenic nerve were transected. The tumor was easily dissected from the great vessels by thin fibrous layer. Finally, the adhered part of the lung was transected with a stapling device by thoracoscopy, and the tumor was then removed from the thorax without using the extracorporeal circulation (Fig. 2A). Plication of the left diaphragm was added for the subsequent diaphragm palsy. The total operative time was 297 minutes, and the bleeding was a total of 180 grams.

The gross size of the tumor was 6.5 cm in diameter. It had indentations that encased the left common carotid artery, left subclavian artery, and aortic arch (Fig. 2B).
The cut surface was grayish white and solid. Histopathologically, the tumor was not shown to have any connection to the pleura, lung parenchyma, thymus, great vessels and pericardium. Atypical spindle cells with swollen nuclei and an increased volume of chromatin were observed with a background of severe fibrosis and vascular proliferations (Fig. 3A). Mitoses were frequently noted. An immunohistochemical study was negative for epithelial markers such as cytokeratin AE1/AE3. Staining for the MIB-1 antibody against Ki-67 was focal, and its labeling index was 30%. Immunohistochemistry for mesothelial markers such as calretinin, WT-1, and cytokeratin 5/6 was also negative. D2-40 staining was faint and focal. Immunohistochemistry for the skeletal muscle marker MyoD1, the vascular endothelial marker CD34, and the neural marker S-100 protein was also negative. Only smooth muscle actin was strongly expressed and considered to be a positive finding, but it was also focal (Fig. 3B). Immunohistochemistry for other smooth muscle markers such as h-Caldesmon and desmin were negative. Because of these findings, leiomyosarcoma was confirmed. The origin was not identifiable histopathologically. However, the minor or medium vessels in the soft tissue of the anterior mediastinum were suspected as the origin. There was no metastatic disease in the subclavicular or mediastinal lymph nodes. There was no fibrosing or necrotic finding in the tumor tissue caused by preoperative irradiation.

The postoperative course was uneventful, and the patient was discharged on day 14 without any neurological deficits other than hoarseness. The patient gained 5 kg in weight 3 months after surgery. A solitary left pleural metastasis was observed 12 months after the surgery without locoregional recurrence, and the lesion was successfully removed by videothoracoscopic surgery. The patient is well without further recurrent disease 16 months after the initial surgery.

Discussion

Among mediastinal tumors, a mesenchymal tumor is a rare entity. Especially, leiomyosarcoma is rarely reported, as only 22 cases have been reported in the
English literature.\textsuperscript{1-9} Only Moran and coworkers have reported a series of 10 of these cases, and; otherwise, there have been only sporadic and single case reports. Abiko and coworkers have reported that 15 cases have been described in the Japanese literature, including his own single case.\textsuperscript{10} From these two reports involving 25 cases, the mean age of the patients was 61 (26–79) years old. Ten were men, and 15 were women. Six, 5, 2 and 12 tumors were located in the superior, anterior, middle and posterior mediastinum, respectively. Ten were asymptomatic. Chest pain, back pain, cough, dyspnea and hoarseness developed in 6, 2, 4, 3 and 1 patient(s), respectively (the numbers are overlapping). In 23 cases, the tumor did not seem to be connected to any organs of the mediastinum, and thus, their origins were unclear and were considered to originate in smooth muscles of the soft tissue. In two cases, the tumors originated in great vessels including the superior vena cava. Eighteen patients were treated by surgical resection alone. Surgery and chemotherapy, surgery and radiotherapy, surgery and chemoradiotherapy, chemotherapy alone, radiotherapy alone and chemoradiotherapy were performed in 1 patient each. One patient did not receive any treatment. The detected recurrent sites were the local region, the lungs, liver, brain and vertebrae. The reported longest survival of the patients was 7 years, which occurred in a patient who underwent complete resection alone for posterior mediastinal leiomyosarcoma.

Moran and colleagues described that the tumors of all 10 cases had a well-circumscribed surface. Because the tumor seemed to involve the great vessels in preoperative chest CT scans, we had prepared extracorporeal circulation in case total arch replacement was necessary. However, the tumor could be easily resected by dissection from the great vessels, including the aortic arch, left common carotid artery, and left subclavian artery. The tumor was not encapsulated, but a thin fibrous layer existed between the tumor and the great vessels. Thus, we believe that an aggressive surgical strategy of complete resection is the only curable treatment in patients with primary mediastinal leiomyosarcoma. Exploratory thoracotomy is recommended if complete surgical resection seems technically possible.

Acknowledgments

This study was funded by Kansai Rosai Hospital. We thank Professor Shigefumi Suehiro, department of cardiothoracic surgery, Osaka City University Graduate School of Medicine, for his kind help and advices in establishing the surgical strategy of this patient.

Conflict of Interest Statement

All authors have full control of all primary data and that we agree to allow the journal to review our data if requested. All authors have no conflict of interest.

References