

Radiotherapy in Mediastinal/Intrapulmonary Malign Solitary Fibrous Tumor: A Case Report

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Received Date: November 30, 2020; **Accepted Date:** December 30, 2020; **Published Date:** January 08, 2021

ABSTRACT

Malignant solitary fibrous tumors are extremely rare and have an aggressive course. Priority treatment should be surgery, if possible. The role of radiotherapy and chemotherapy is controversial. Here, the treatment response after curative radiotherapy and the evaluation of the follow-up process of the patient who was evaluated with the pre-diagnosis of mediastinal mass and was diagnosed with malignant solitary fibrous tumor after subcarinal lymph node biopsy is mentioned.

KEYWORDS

Malign solitary fibrous tumor; Treatment; Radiotherapy; Acute treatment response

INTRODUCTION

Solitary fibrous tumors (SFTs) are extremely rare neoplasms originating from mesenchymal connective tissue. Although generally exhibiting benign features, 10%-15% of them are malignant or aggressive [1]. They mostly originate from visceral pleura. Those originating from the mediastinal pleura can be confused with primary tumors of the mediastinum. Most of the patients are asymptomatic, but nonspecific symptoms such as shortness of breath and cough may also be observed. They are usually encapsulated and vascularized tumors. The primary treatment is surgical resection. The main factors affecting the survival rate of the patients are tumor stage and complete surgical resection, but not tumor size [2]. However, total surgical resection is not always possible, and even a high recurrence rate has been reported despite surgery [3]. Although the role of radiotherapy (RT) is not

certain, it is stated that the combination of surgery and RT reduces the recurrence rate [4].

CASE REPORT

A mass located in the mediastinum and pleural effusion were detected in a 65-year-old male patient who presented with the complaint of dyspnea. Biopsy was taken from the subcarinal lymph node by bronchoscopy and its pathology was reported as "malignant SFT". Pleural fluid cytology was evaluated as reactive. In thoracic MRI taken for surgical evaluation, a mass lesion with the largest transaxial diameters of 11 cm × 15.5 cm located in the posterior mediastinum extending craniocaudally to the right hemithorax for approximately 18 cm, and invaded into heterogeneously contrasted carina, proximal sections of the right and left main bronchi, right main pulmonary artery, all pulmonary veins, anterior aspect of the vertebra, and on the right side to the corpus posterior part

Citation: Beyza Sirin Ozdemir, Radiotherapy in Mediastinal/Intrapulmonary Malign Solitary Fibrous Tumor: A Case Report. Cancer Med J 4(S3): 7-10.

of the right diaphragm was described so the patient was evaluated as an inoperable case.

Since distant organ metastasis was not detected in PET-CT, definitive RT and weekly carboplatin-paclitaxel chemotherapy were initiated. A total of 66 Gy RT was planned for the patient, with a fraction dose of 2 Gy/day using the "Image-guided Radiation Therapy" (IGRT)

device and 6 mv energy using the intensity modulated radiotherapy (IMRT) technique. However, due to the high doses of normal tissue, the total dose was kept at 50 Gy (Figure 1). If tumor shrinkage was detected during the treatment, weekly imaging (XVI) was performed on the treatment device with the hope of performing adaptive RT to increase the dose. However, no significant shrinkage was detected in the tumor during the treatment process.

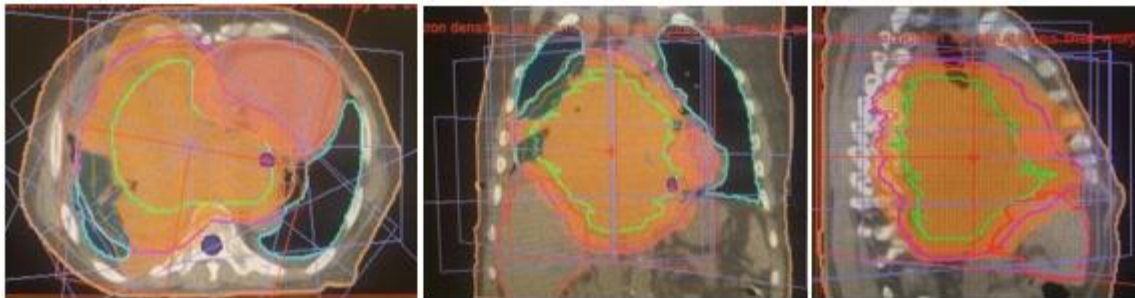


Figure 1: Planning images made with IMRT technique.

It was planned to be evaluated again with PET-CT three months after RT was completed (March 2020). After RT, the patient had no complaints. His general condition was very good. However, due to the COVID-19 pandemic in this process, the planned PET-CT could not be taken. The patient applied to the emergency service in April with complaints of shortness of breath and cough. No fever, but an elevation in CRP (177 mg/L) was detected and thorax computed tomography (CT) imaging and coronavirus 2019 (COVID-19) testing were performed. COVID-19 test was negative.

In thorax CT taken at the end of 4 months after the end of the treatment, a mass lesion located in the posterior mediastinum, extending to the subcarinal level, filling the right lung lower lobe and para-mediastinal area was reported. The mass with its the largest trans axial dimensions being measured as 10 cm × 7 cm was obliterating the bronchial branches going to the lower lobe of the right lung, and the plan between the mass and the esophagus could not be discerned (invasion). In addition, multiple bilateral metastatic lesions in the lung,

massive pleural effusion, and consolidated lung tissue (infective) located in the peribronchovascular area in the anterior of the left lower lobe of the left lung were detected. Upon development of pneumothorax after thoracentesis, a tube thoracostomy was opened and followed-up.

No improvement was observed in the general condition of the patient during follow-up. No bacterial infection was detected in tracheal aspirate smear, urine and pleural fluid cultures. Influenza test was performed and evaluated as negative. CRP (232 mg/L) remained at a high level. During the follow-up, the patient whose respiratory distress increased and his hypercarbia and hypoxia deepened was intubated. However, the general condition of the patient deteriorated further and died four months after RT.

DISCUSSION

Solitary fibrous tumors arise from CD 34-positive dendritic mesenchymal cells [5]. It can occur in any body parts such as mediastinum, pelvis, kidney, liver. It was first described as of pleural origin in 1931 [6]. In the

literature, it is generally observed to originate from the pleura.

Mediastinally located ones are in the form of slow-growing painless masses and are detected incidentally on chest X-ray. Especially malignant SFTs have a large volume and high recurrence rates. Although it is not a standard treatment, surgery is recommended primarily.

Although the place of chemoradiotherapy is not yet clear in the literature, it is stated that it can be used in patients who are inoperable or underwent partial resection [7,8]. Xue et al. applied adjuvant RT after partial resection in a case of SFT located in the paranasal region and stated that recurrence did not develop in their patients after 4.5 years of follow-up [9]. Saynak et al. delivered conventional external RT at a total dose of 60 Gy in 30 fractions to the patient who had recurrence in the 18th month after surgery with a diagnosis of pleurally located SFT. It has been reported that 60% response to treatment was achieved as demonstrated in thorax CT taken 6 weeks after completion of RT [10].

Vanfleteren et al. reported un-resectable mediastinal SFT treated with concurrent chemoradiotherapy with a complete metabolic and clinical response [11]. In the study, RT was started simultaneously with the first course of carboplatin and etoposide chemotherapy. RT was delivered as a total dose of 42 Gy in 28 twice-daily fractions of 1.5 Gy. In our study, we applied a total dose of 50 Gy RT to our patient with the conventional fractionation scheme. At the end of 4 months after treatment, we detected a partial response in the primary tumor. However, we observed a rapid deterioration in the general condition of the patient clinically due to the development of extensive lung metastases and pleural effusion within a short time.

CONCLUSION

In conclusion, since SFTs are rare tumors, it is necessary to evaluate the patients individually and offer the most appropriate treatment option. Although we do not forget that complete surgical resection is the most appropriate treatment option for these tumors, in the light of these and similar cases, definitive RT may be an evaluable option when surgery is impossible.

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