Recurrent intrathoracic solitary fibrous tumor: Remarkable response to radiotherapy

Min Liu, Bin Liu1, Lihua Dong, Bailong Liu

Abstract: Solitary Fibrous Tumor of the Pleura (SFTP) is an uncommon neoplasm which grows slowly. For some cases, surgery is warranted. However, for unresectable ones, the standard strategy has not been established yet. We presented a rare case of recurrent malignant intrathoracic solitary fibrous tumor. It was impossible to resect the tumor. Radiotherapy alone achieved a significant improvement effect.

Key words: Radiotherapy, recurrent, solitary fibrous tumor of the pleura

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Fibrous Tumor of the Pleura (SFTP) is an uncommon neoplasm which grows slowly. Surgery is the mainstay of treatment. Recurrent tumors should be strongly considered first for a repeated surgical resection. Little experience has been described in the literature with postoperative treatment and treatment for unresectable SFTP. Here, we present a case with recurrent SFTP which responded to radiotherapy remarkably.

Case Report

A 43-year-old man was presented in June, 2012 with a history of dyspnea for a month. He then received thoracentesis and about 200 ml bloody fluid was drained. Chest computed tomography (CT) with contrast demonstrated a huge mass about 10.3 × 15.8 cm in the left thorax [Figure 1]. Abdominal CT with contrast revealed a 2.4 cm lesion in right lobe of liver with an image of “fast in, fast out”. α-Fetoprotein (AFP) was normal. Hepatitis B surface antigen and hepatitis C antibody were negative. The biopsy of the lesion in the left thorax and liver was carried out respectively. The pathology of the former showed solitary fibrous tumor while the latter showed infiltration of a few inflammatory cells. In the surgery, a huge tumor was found occupying more than half of the left thorax. The tumor was removed completely. The definitive pathologic results were as follows: High-grade malignant solitary fibrous tumor with infarction and necrosis [Figure 2a]. The tumor was 17 cm × 16 cm × 12 cm and had a high mitotic rate (>20/10 high power field, HPF). The immunohistochemistry showed Ki-67 (+40%) [Figure 2b], Vimentin (+), CK (-), CD34 (focally+) [Figure 2c], P53 (-), Bcl-2 (+) [Figure 2d]. Then he recovered uneventfully. Given the high aggressiveness, adjuvant treatment was recommended. However, he refused.

In January, 2013, the patient found a lump on the back but neglected it. Two months later, he suffered from dyspnea again, getting worse progressively. His performance status was one according to Eastern Cooperative Oncology Group (ECOG) evaluation. Chest CT showed a giant (16.95 × 9.5 × 22 cm), irregular and heterogeneous mass occupying almost all the left thorax with apparently central necrosis. First, he went to see the thoracic surgeon. However, it was impossible to resect the tumor. The examination of liver demonstrated that the lesion of right lobe was slightly larger than a year ago, from 2.4 to 2.6 cm. In order to slow down the progression of the primary tumor, we carried out palliative radiotherapy to the thoracic tumor with a dose 50 Gy/25f. The tumor shrank remarkably to 10.7 × 7.5 × 17.5 cm, revealing a 44% response [Figures 3a and b] and dyspnea disappeared. The favorable response reflected high radiosensitivity originated from high aggressiveness of this tumor, but partially sacrificed by large amount of necrosis inside the tumor. Then we gave additional 10 Gy/5f to the shrunk tumor. The radiotherapy technology was as follows: 3D-CRT, 10MV, X-Ray, the mean dose and V20 of the lungs was 972.9 cGy and 15% respectively [Figures 4a and b]. In February, 2014, 8 months after radiotherapy, the thoracic CT showed the tumor was slightly smaller than that when he finished radiotherapy. The patient remains well and progression free survival has reached to 10 months.

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SFTP was reported first by Klemperer and Rabin in 1931. It is rare with an incidence of 2.8/10000. Commonly, it grows slowly without symptom. When it grows larger, patients may complain of cough, hemoptysis, dyspnea, and chest pain. There is hypoglycemia in less than 5% cases.

In 1981, Briselli started systemic research of SFTP, demonstrating that 88% of postoperative specimens were benign while 12% were malignant and eventually lead to death through local recurrence or metastasis. Basic fibroblast growth factor (FGF) and Ki-67 helped to diagnose malignant SFTP and predict the prognosis. Yuliang Sun et al., reported that Ki-67 expression in benign and malignant SFTP were 1.9% and 6.11%, whereas basic FGF expression in benign and malignant SFTP were 48.67% and 74.5% respectively with significant difference. CD34 and Bcl-2 were sensitive markers for SFTP. High mitosis rate (>4/10 HPF) signified malignancy. In our case, Ki-67 (>40%) and mitosis rate (>20/10 HPF) demonstrated highly malignant, consistent with early relapse after operation.

Surgery is the treatment of choice for SFTPs. For small tumors, Video-Assisted Thoracic Surgery (VATS) is recommended. While for large ones, standard thoracotomy should be performed. Given its paucity, the effect of adjuvant therapy is still unclear. Suter et al., reported one patient alive without evidence of disease over 20 years after subtotal resection of the tumor followed by radiotherapy. Relapse and metastasis still can take place after complete resection. Marc de Perrot recommended adjuvant therapy after resection of malignant sessile tumors, especially for recurrent ones. Our patient may benefit from adjuvant treatment after surgery given high malignance.

The treatment for unresectable malignant SFTP has not been established yet. In a case of huge pelvic malignant solitary fibrous tumor with lung metastasis, chemotherapy didn’t work while the tumor shrunk 12 months after 50 Gy irradiation of the pelvic region. Radiotherapy alone can attain 30-60% rate of control in sarcoma for patients refusing or unsuitable for surgery. In our case, irradiation alone achieved significant effect which was consistent with Saynak M’s report. Combined chemotherapy of Temozolomide and Bevacizumab might be effective in recurrent unresectable SFTP. Target therapy for SFTP is still in early stage. Malignant SFTP can
express PDGFRβ strongly and has missense mutation of 18 exons, providing promising treatment target for unresectable malignant SFTP.[17]

We can’t determine whether the lesion of liver is metastatic since an image of “fast in, fast out” was inconsistent with typical radiologic manifestation of liver metastasis and it increased only 0.2 cm after 1 year. So we should keep a close eye on it. Once it shows obvious enlargement, focal resection can be considered.

Our case verified the effect of palliative radiotherapy. After a period, hypoxia will grow again and make patient symptomatic. At that time, besides best support care, low dose of palliative reirradiation might be carefully tried since the irradiation dose of normal organs this time was low. Additionally, on the basis of comprehensive evaluation, chemotherapy might be tried. Given the high aggressiveness of this tumor and limited treatment options, the prognosis of this patient is poor.

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