

Cicatricial Fibromatosis Diagnosis after Suspected Local Recurrence at the Bronchial Stump Following Lobectomy for Lung Cancer

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A mass excision surrounding the bronchial stump was performed to exclude malignancy in a 42-year-old man who had undergone a right lower lobectomy for lung cancer. The mass was identified as a cicatricial fibroma. Cicatricial fibromatosis, which is desmoid fibromatosis that arises in a surgical scar, is a well-known clinical condition. It consists of histologically benign neoplasms. Their occurrence after thoracic surgery is extremely rare. Biopsy or excision of suspicious lesions is very important for diagnosis. R0 resection remains the principal outcome for intrathoracic desmoid fibromatosis. We report that a cicatricial fibromatosis in the subcarinal space was removed after suspicion of local recurrence at the bronchial stump following lobectomy for lung cancer.

Key words: 1. Cicatricial fibromatosis
2. Lobectomy
3. Desmoid fibromatosis

CASE REPORT

A 33-mm mass was observed adjacent to the margin of the right lower lobe stump in the subcarinal space on chest computed tomography (CT) in a 42-year-old man who had undergone video-assisted thoracoscopic right lower lobectomy 18 months earlier for primary lung cancer (adenocarcinoma, acinar subtype). Owing to the size of the mass (32×25 mm), the pathologic stage of primary lung cancer was classified as pT2aN0M0. There had been no evidence of recurrence on chest CT taken nine months previously. The patient was relatively healthy with no other laboratory abnormalities present. The mass had increased uptake on F18 fluorodeoxyglucose

(FDG) positron emission tomography (PET)/CT imaging, with a maximal standardized uptake value (SUVmax) of 6.5 (Fig. 1). Therefore, we considered local recurrence of lung cancer at the bronchial stump.

Aspiration biopsy was conducted using ultrasound-guided bronchoscopy and ultrasound-guided esophagoscopy to confirm the diagnosis. Fiberoptic bronchoscopy findings were a mucosal bulging at the medial side of the right lower lobar stump; there was no evidence of a hypervascular and irregular mucosal layer. The esophagoscopy and endoscopic ultrasound revealed a 36-mm hypo-echoic round mass located 30 cm from the incisors with a relatively clear margin at the esophagus (Fig. 2). Both aspiration biopsies resulted in only

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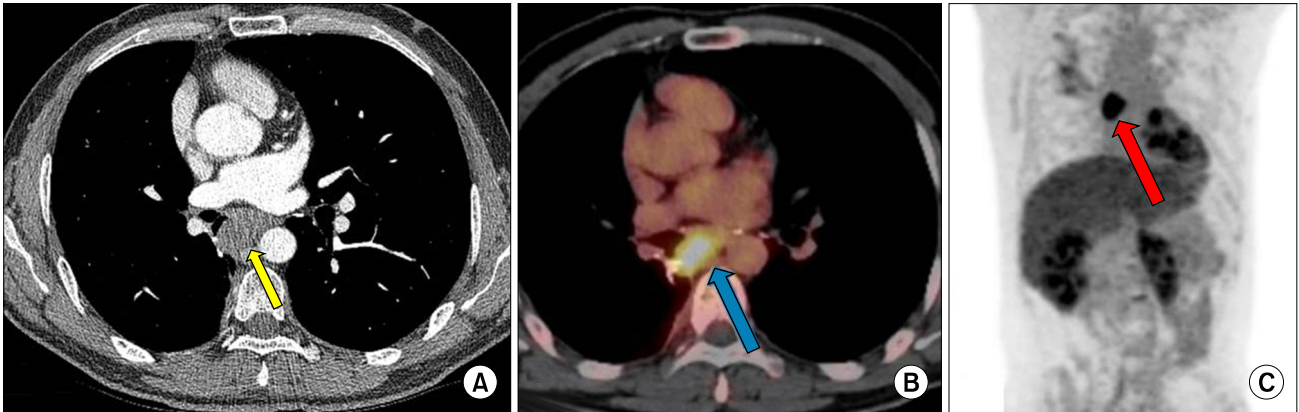


Fig. 1. (A) Mass (yellow arrow) compressing the bronchial stump and esophagus in the subcarinal space. (B) Increased uptake (blue arrow) in F18-fluorodeoxyglucose positron emission tomography (PET)/computed tomography. (C) Coronal PET maximum intensity projection image shows the increased uptake (red arrow).

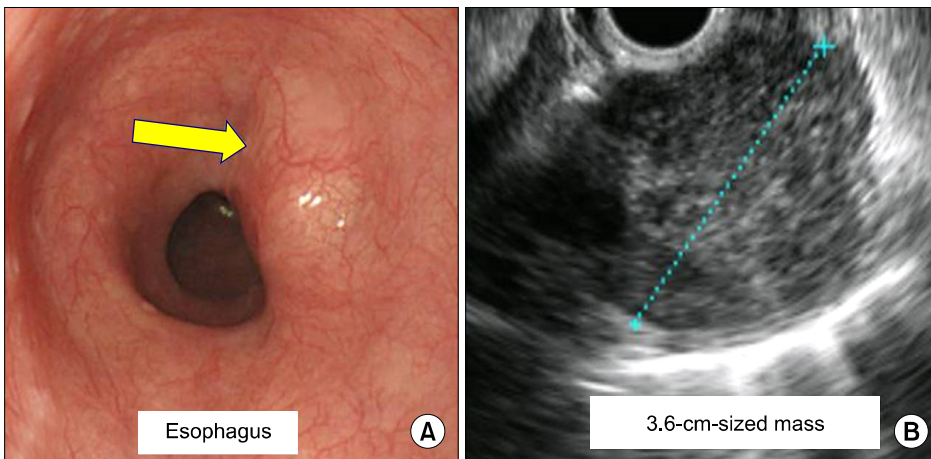


Fig. 2. The findings of esophagoscopy and endoscopic ultrasound. (A) The esophageal mucosa is bulging (yellow arrow) due to external compression and (B) 3.6-cm-sized hypoechoic mass was observed.

fibrotic tissue, without malignancy. Therefore, we decided to completely excise the subcarinal mass via right lateral thoracotomy. The mass was tightly adhered to the surrounding lung, bronchus, and esophagus tissues. The mass was removed with partial en-bloc excision of the esophageal longitudinal muscle and lung parenchyma. The postoperative follow-up was uneventful. The patient was discharged home on postoperative day 10.

The gross findings were a typical whorled appearance and an ill-defined margin. Microscopically, well-differentiated fibroblastic tissue with an infiltrative margin and long fascicles of spindle cells of variable cell density with few mitoses were observed (Fig. 3). Immunohistochemistry staining was weakly positive for Ki-67 and negative for C-kit, CD34, and

S-100 proteins. Therefore, cicatricial fibromatosis was diagnosed.

After discharge, the patient was monitored for recurrence or increase in size, with CT scheduled every six months for one year based on the recommended follow-up interval for his lung cancer. No recurrence has been found to date. If recurrence or increasing mass size is identified, we plan to inject triamcinolone guided by endobronchial ultrasonography for the first adjuvant therapy, and consider radiotherapy with intravenous tamoxifen injection for the second adjuvant therapy.

DISCUSSION

Cicatrical fibromatosis, which is desmoid fibromatosis that arises in a surgical scar, is a well-known clinical condition.

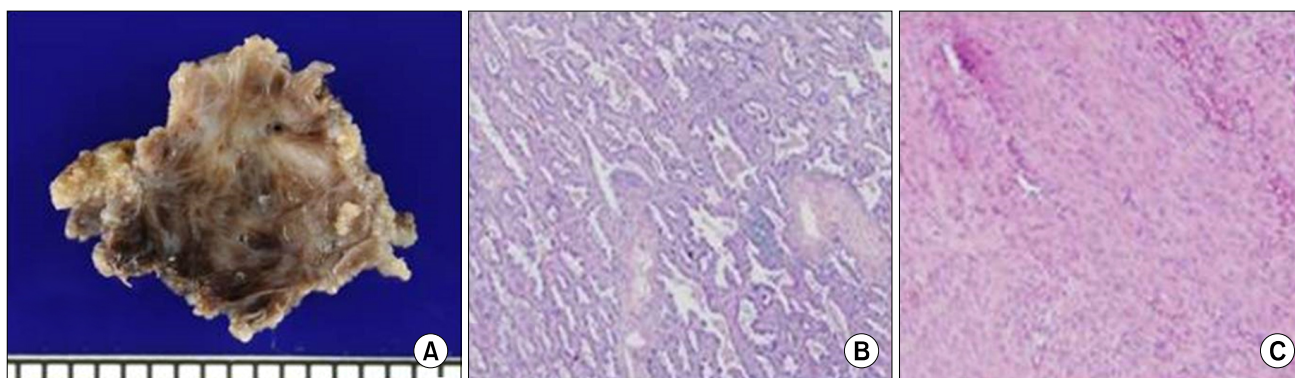


Fig. 3. (A) Firm and rubbery with typical whorled appearance and ill defined margin in gross findings. (B) Typical adenocarcinoma with acinar subtype lung cancer specimen which was obtained after initial lobectomy (H&E, $\times 100$). (C) Well-differentiated fibroblastic tissue at cicatricial fibromatosis (H&E, $\times 100$).

Desmoid fibromatosis is composed of histologically benign fibroblastic/myofibroblastic neoplasms that originate from musculoaponeurotic structures and are classified as deep fibromatoses [1,2]. Although desmoid fibromatosis can arise at any site, the extremities, abdomen, and retroperitoneum are the most common sites [1,2]. Fibromatosis of the chest wall occurs less frequently. The etiology of the tumor is still not understood. However, tumor development is strongly associated with genetic predisposition, physical trauma, and hormonal effects [1]; is often associated with female sex and familial adenomatous polyposis; and sporadically occurs at the site of previous trauma, scars, or irradiation [1,2]. Despite the benign histologic appearance and lack of metastatic potential, desmoid fibromatosis can cause aggressive local infiltration and compression of surrounding structures. A high recurrence rate exists, particularly in anatomic locations with restricted access for surgical resection [3].

The occurrence of cicatricial fibromatosis, especially adjacent to the bronchial stump that remained after thoracic surgery in the present case, is extremely rare. There are 16 reported cases of desmoid-type fibromatosis following thoracotomy [4], primarily at the chest wall or pleura. The uptake on F18 FDG PET/CT imaging (SUVmax, 6.5) in the present case was higher than the SUVmax of 1.9 previously reported for a tumor that originated at a thoracotomy site [4]. Because the tumor compressed and infiltrated the surrounding tissue of the lung parenchyma, esophagus, and bronchial stump, it was very difficult to distinguish from recurrent lung cancer.

Aggressive surgical management is the first treatment choice, with 95% of reported intrathoracic cases treated with surgical resection. However, radical resection is often a therapeutic challenge, owing to the anatomy of the mediastinum and high rate of recurrent tumors involving vital structures. Only 39% of radical resections were microscopically negative (R0). Surgical margins of 2–4 cm with en-bloc removal are optimal, when anatomic features make it possible. Optimal resection with chest wall involvement includes one unaffected rib above and below the lesion. Palliative surgical diminution might be lifesaving when vital organs are compressed. In this patient, the mass was located in the subcarinal area, adjacent to vital organs. Although wide radical excision was impossible, we attempted a clear margin; however, the tumor tissue adjacent to the right main bronchus and carina was microscopically positive.

Non-surgical care is the primary treatment for tumors that are radically unresectable. Monotherapy or combination therapy with anti-estrogens, non-steroidal anti-inflammatory drugs (NSAIDs), radiotherapy and chemotherapy can reduce tumor size. The optimal dose and duration of tamoxifen or other anti-estrogen therapy have not been determined. Indomethacin and sulindac are the NSAIDs of choice [5]. If positive margins or residual disease are evident postoperatively, adjuvant radiation therapy results in equivalent local control to that of surgery with negative margins. Radiation therapy alone results in durable local control rates of 70% to 80%. The role and efficacy of chemotherapy remains controversial, with poor

evidence.

The local recurrence of primary lung cancer after lobectomy was suspected in the present patient based on chest CT and F18 FDG PET/CT imaging; instead, a cicatricial fibromatosis was present in the subcarinal space. Because we failed to perform an R0 resection, the need for additional steroid therapy and radiotherapy should be considered.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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