Inflammatory Myofibroblastic Tumor: Role of Surgery from Uniportal VATS in Single Pulmonary Mass to Rigid Bronchoscopy in Bilateral Endobronchial Lesion

Padungkiat Tangpiroontham, MD1; Mana Rochanawutanon, MD2; Sawang Saenghirunvattana, MD3; Pimon Ruttanaumpawan, MD3

Abstract

A variety of clinical presentations are observed in pulmonary inflammatory myofibroblastic tumors. Diagnosis can be difficult and requires a high index of suspicion due to rarity of the disease. With the era of minimally invasive surgery, the authors wish to report two patients who were treated via thoracoscopic and bronchoscopic approach at Bangkok Hospital as options of investigation and treatment.

Keywords: inflammatory myofibroblastic tumors, single pulmonary nodule, pulmonary nodule, minimally invasive surgery

Inflammatory myofibroblastic tumor is a rare soft tissue tumor. It can be discovered in many organs but is most commonly found in the lung.1,2 Gold standard of diagnosis is tissue biopsy. Computed tomography (CT) and Positron emission tomography/computed tomography (PET/CT) has a role in diagnosis but sensitivity and specificity is not very high.3,4 Presentation may vary widely from single pulmonary nodule, endobronchial lesion, bilateral lung nodule or distant metastasis.2-11 Mainstay of treatment is complete resection which has a better survival rate and lower recurrence rate. Video-assisted thoracoscopic surgery (VATS) resection is one of the safest and most effective ways to treat the patient.9,11 In cases of endobronchial lesion, palliative endobronchial resection is utilized to restore patient’s airway.6,10

Case Report

Case #1

A 42-year-old Arabic male previously healthy presented with asymptomatic single pulmonary mass found from annual check-up chest x-ray. CT of Chest revealed 3.9 cm right upper lobe pulmonary mass with compression to posterior segmental bronchus of right upper lobe with possible right middle lobe invasion as shown in Figure 1A and 1B. Bronchoscopy with biopsy was discussed with the patient. He decided to proceed with surgical biopsy and resection given the concern of the progression of airway compression.

General anesthesia was performed by left sided double lumen tube. Uniportal VATS exploration revealed right upper lobe mass bulging out from pulmonary parenchyma with complete minor fissure as shown in Figure 1C. Given the proximity of the tumor to the main RUL bronchus, right upper lobe (RUL) lobectomy was undertaken through the same 4 cm uniportal incision. Frozen section showed spindle cell tumor with free bronchial and staple margin. The postoperative course was uneventful and the patient was discharged on postoperative day 1. One month postoperative chest x-ray showed full lung expansion without residual space as shown in Figure 1D.

Final pathology demonstrated inflammatory myofibroblastic tumor as proven by positive anaplastic lymphoma kinase (ALK) and smooth muscle actin and negative for CD34. All 2R, 4R, 7, 9 lymph node stations tested negative for tumor as shown in Figures 1E to 1H.
Figure 1A: shows chest x-ray, preoperative.

Figure 1B: CT chest, axial view shows central mass compressing apical segment of right upper lobe.

Figure 1C: Shows a bulging mass protruding under the intact visceral pleura.

Figure 1D: Shows good lung reexpansion on postoperative day 1.

Figure 1E: Cut surfaces of the mass reveals a solid, well demarcated grey-yellow mass without necrosis.

Figure 1F: Shows plump spindle cell tumor arranging in intersecting fascicles and storiform pattern with additional lymphoplasmacytoid cell infiltration.
**Case #2**

A 30-year-old Arabic previously healthy male presented with massive hemoptysis initially treated by a local hospital in his country and was found to have a bleeding RUL endobronchial mass. Chest x-ray revealed multiple bilateral pulmonary masses with endobronchial lesions, largest (6 cm) at right upper lobe as shown in Figures 2A-B. Complete resection is not possible due to insufficient pulmonary reserve. Biopsy demonstrated spindle cell proliferation with storiform and fascicular architectures with spindle cells stained diffusely for smooth muscle actin and focally desmin and caldesmon. Staining for cytokeratins, CD34, ALK, and ROS1. The conclusion was inflammatory myofibroblastic tumor given negative ALK and ROS1 are not positive in all adult cases. He was transferred to Bangkok Hospital Headquarters. Flexible bronchoscopy by pulmonologist revealed mild bleeding of endobronchial mass at right upper lobe bronchus with extension into right main bronchus causing near total occlusion. Rigid bronchoscope was introduced and tumor was cored out in right main bronchus as well as right upper lobe bronchus then bleeding was controlled by argon plasma coagulation as shown in Figure 2C. Pathology demonstrated inflammatory myofibroblastic tumor proven by diffusely positive smooth muscle actin, as shown in Figure 2D. Remaining intrapulmonary tumor was destroyed by microwave ablation in right upper, right lower and left lower lobe. Repeated flexible bronchoscopy was commenced 1 month later demonstrating the patency of right main and right upper lobe bronchi. On 6-month follow-up, he was found to have tumor metastasis at soft tissue area around left biceps muscle. Given no symptom of hemoptysis, we continued to follow-up without intervention. He continued to be symptom-free on 18 months follow-up with small progression of intrapulmonary tumor.
Discussion

Both case reports are examples of the variety this tumor may present. In early cases, complete resection is possible and can result in long term survival with low risk of recurrence. The 5-year and 10-year disease-free survival is 89% after complete resection. It can be found centrally located as high as 20%, therefore if left undiagnosed, it tends to invade bronchus causing curative resection to become very challenging. It can quickly progress to completely block main bronchus to the point that hemoptysis is one of the presentations. Early recannulation of airway is mandatory in this kind of case by either flexible or rigid bronchoscopy due to rapid progression. Furthermore, it can even metastasize to distant organs similar to the second case. The name of this disease itself might sound benign but it is indeed malignant in its nature. These patients have a tendency of being diagnosed at a young age, therefore surgical resection should be encouraged.

Conclusion

In early pulmonary inflammatory myofibroblastic tumor, thoracoscopic complete resection has an excellent short term outcome and may result in long-term survival. Even in metastatic cases, palliative airway control also has good short and mid-term outcomes.

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