Inflammatory Myofibroblastic Tumor of the Lung With Multifocal Metastases

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A 66-year-old man presented to the pulmonology clinic with cough and sputum that had been present for 3 weeks and for evaluation of a mass in the right lower lobe of the lung that was revealed on computed tomography (CT) of the chest. He was an ex-smoker with a smoking history of 40 pack-years. The physical examination was unremarkable, and laboratory evaluation only revealed an elevated erythrocyte sedimentation rate of 50 mm/hour and a C-reactive protein of 26.87 mg/L, suggesting the presence of an underlying inflammatory reaction.

CT scan revealed a locally invasive mass (4.0 cm × 5.3 cm) in the right lower lobe (Fig. 1A) and multiple lymph node metastases in both the supraclavicular regions, the mediastinal region, and the portocaval region. 18F-fluorodeoxyglucose-positron emission tomography–CT revealed substantial 18F-fluorodeoxyglucose-positron emission tomography avidity in all of these lesions (Fig. 1B, C, and D). T2-weighted magnetic resonance imaging of the thoracic spine revealed a peripherally enhanced mass-like lesion involving the T12 vertebra (Fig. 1E). On the basis of these findings, advanced lung cancer with distant metastases was suspected.

Pathologic examination of biopsy specimens obtained by percutaneous transthoracic needle biopsy reported the proliferation of fibroblasts with lymphoplasmacytic infiltration (Fig. 1F). Immunohistochemical staining was positive for smooth muscle markers such as actin and desmin and negative for the thyroid transcription factor-1, Immunoglobulin G4, and anaplastic lymphoma kinase-1. These findings were consistent with the diagnosis of inflammatory myofibroblastic tumor (IMT). Excisional biopsy of the right supraclavicular lymph node and the thoracic vertebra revealed no evidence of malignant cells. On the basis of these findings, a pathologist confirmed that these lesions were metastases from the pulmonary IMT.

On the basis of a careful review of previous reports on IMT, the patient was administered clarithromycin at a dosage of 500 mg twice a day.1 However, there was no obvious improvement in the follow-up chest CT scan after the initiation of the therapy. Subsequently, the patient was administered methylprednisolone at the dosage of 0.5 mg/kg once a day.2 Interestingly, a dramatic improvement in these IMT lesions was reported in the follow-up images 5 weeks after the initiation of the corticosteroi d therapy (Fig. 1G, H, and I).

IMTs are rare tumors that account for 0.04% to 0.1% of all pulmonary neoplasms.3 IMTs are composed of myofibroblastic spindle cells with an inflammatory infiltrate of plasma cells and lymphocytes. Although IMTs are benign histopathologically, they may invade surrounding structures, recur, or even metastasize clinically.4

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Complete surgical resection is the mainstay treatment of IMT of the lung. In the case of inoperable IMT, there is currently no established treatment. However, recent studies reveal the importance of molecular biological assessment of the disease and the therapeutic potential of crizotinib in the treatment of unresectable IMT harboring anaplastic lymphoma kinase gene rearrangement.

In this case, given the extensive involvement of the disease, surgical resection or targeted therapy was not appropriate for the patient. Instead, clarithromycin, followed by a systemic corticosteroid, was administered on the basis of sporadic case reports supporting its anti-inflammatory role. Although there was no discernible improvement with clarithromycin, a dramatic improvement was seen after systemic corticosteroid therapy in the follow-up images.

Based on our experience, a systemic corticosteroid may be one of the therapeutic options for unresectable IMT with extensive metastases, which has limited treatment options.

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