MEGA JOURNAL OF CASE REPORTS

Case Presentation

Compiled Date: March 11, 2023

Low-Grade Myofibroblastic Sarcoma of the Lung: A Case Report

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Abstract

Low-grade myofibroblastic sarcoma (LGMS) is described as a distinct atypical myofibroblastic tumor often with fibromatosis-like features and predilection for the head and neck, especially the oral cavity and larynx. LGMS arising in the lung is extremely rare. Surgery is the treatment of choice. Postoperative chemoradiotherapy is feasible, but the therapeutic effect is limited and there are no standard guidelines. Adjuvant radiotherapy has been used for local control. We present a case of Low-grade myofibroblastic sarcoma of lung. Immunotherapy combined with anti-vascular targeting therapy has achieved good results.

Keywords: Low-grade myofibroblastic sarcoma; Immune checkpoint inhibitors; Angiogenesis inhibitors; Case report

Case Presentation

An 82-year-old male was admitted to our hospital because she has cough, expectoration with hoarseness for more than 1 year. The above symptoms were progressively aggravated. He began to show intermittent bloody sputum 3 days before admission. Physical examination revealed the breath sounds of both lungs were thick and there were no other special positive signs. No significant personal or family history was present. Computed Tomography (CT) showed subcarinal mass of trachea, involvement of left and right main bronchi, unclear boundary of local esophageal wall, considering the possibility of neoplastic lesions. Multiple hilar and mediastinal lymph nodes were enlarged and calcified **(Figure 1).** Under general anesthesia, the patients were examined by electronic bronchoscopy, mucosal biopsy and lung biopsy under ultrasonic bronchoscopy, and the wide and irregular new organisms in the lumen of the left main bronchus were found under bronchoscopy, the left main bronchus was obviously structured and the pathological tissue was taken from the new biological part of the left main bronchus, the ultrasonic bronchoscope was entered through the trachea catheter, the echo of the left Main Bronchus in the N7 group was lymphadenopathy low and uneven, the lymph nodes were biopsied in N7 group **(Figure 2).** HE stain (hematoxylin-eosin staining) findings suggested spindle cell tumor (N7 lymph node, left main bronchus neoplasia) **(Figure 3).** Immunohistochemistry revealed positive for smooth muscle

actin (SMA), focally positive for CD34 and CD68, while negative for S-100, Desmin, P53, nuclear β-catenin, ALK1, SOX10, MUC4, and STAT6. The imaging, histomorphological, and immunohistochemical features suggested a final diagnosis of LGMS of the lung **(Figure 4).** The patient was locally advanced; the tumor invaded the main bronchus and could not be operated on. Radiotherapy was used for this patient. Because the patient was old and could not tolerate chemotherapy, after general discussion, he was given arotinib 8 mg once daily and sindelizumab 200 mg once every 3 weeks. The patient underwent CT follow-up 3 months after therapy and CT showed the tumor shrank with necrosis **(Figure 5).** The patient had provided informed consent for publication of the case, and the study protocol was approved by Medical Ethics Committee of Peking University Third Hospital.

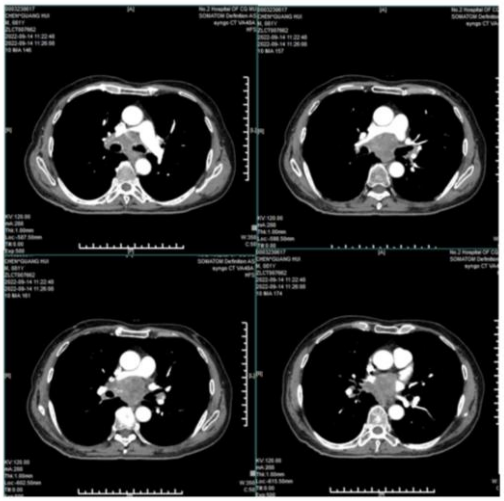

Figure1: Computed Tomography (CT) scan before treatment: There was an irregular soft tissue mass about 55 mm × 40 mm in size under the tracheal carina. The density of the mass was uneven, and the necrotic area and calcification were visible. The boundary of the mass was not clear, and the local wall of the bilateral main bronchi was not complete, locally, it protruded into the lumen of the left main bronchus, moved forward with the compression of the right pulmonary artery, and the boundary between the esophageal wall and the lesion was not clear.

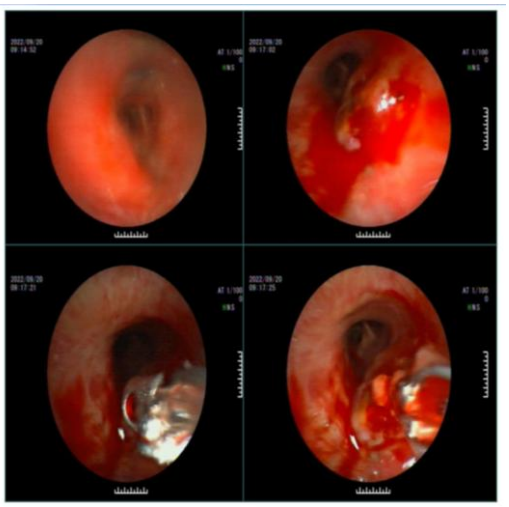

Figure 2: Bronchoscopic pictures：In the scope of endoscopy, we can see: Sharp Carina, mobility, wide and irregular new organisms in the lumen of the left main bronchus, resulting in obvious stenosis of the left main bronchus, and unobstructed openings of the right main bronchus and its lobes and segments, no definite new organism and stenosis were found in the lumen.

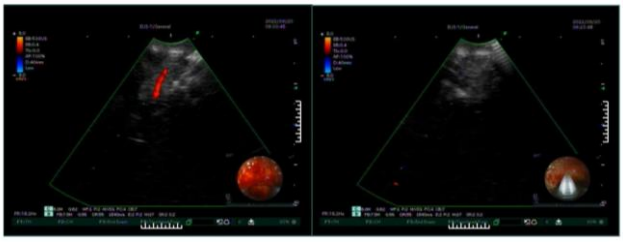

Figure 3: Ultrasonic bronchoscopy pictures：The results of eBUS showed that the lymphadenopathy in N7 group were hypoechoic and inhomogeneous, tissue fragments, necroses and strips of tissue were removed and sent to exfoliated cells and pathological biopsies.

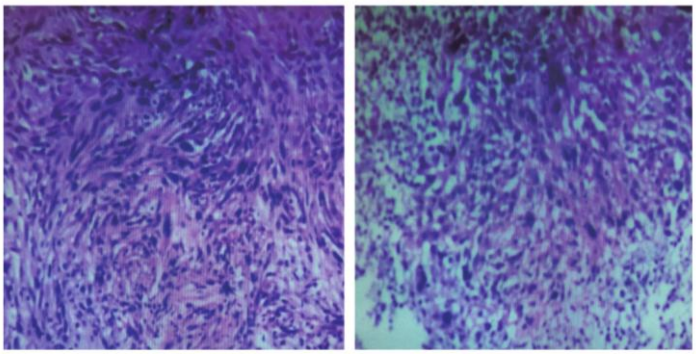

Figure 4: H& E staining of the tumour. Microscopically, the spindle cells were arranged in bundles, and the tumor cells were abundant, with moderate atypia at least in the local area of the nucleus, the cytoplasm of the tumor cells was light eosin, the cell boundary was unclear, the nucleus was wavy, and the tip of the tumor cells was pointed and thin, the nuclei were fat fusiform with vacuoles containing clear or unclear nucleoli.

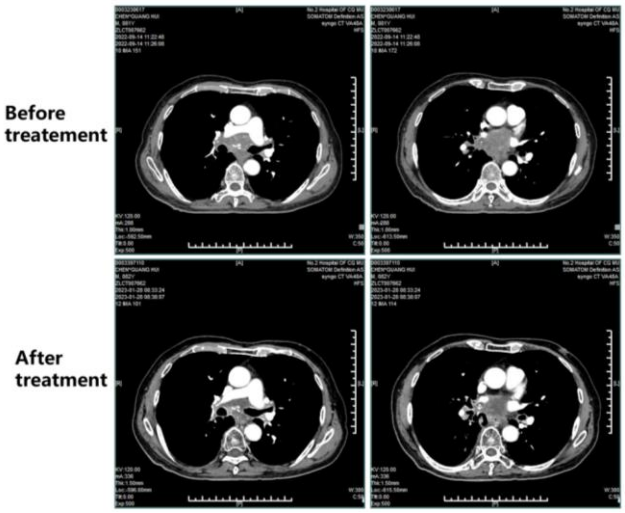

Figure 5: Comparison of CT images after treatment: The tumor area under the carina of trachea was smaller than before, the necrotic area was enlarged, and the main bronchus was invaded.

Discussion

Low-Grade Myofibroblastic Sarcoma (LGMS) was firstly described by Mentzel et al **[1],** which represents an atypical and extremely rare type of tumor composed of myofibroblasts. It was first classified as a new group of soft tissue and bone tumors by the WHO in 2002, and this classification was maintained in 2020 **[2].** LGMS has been reported to occur in deep soft tissues with predilection for the head and neck **[3].** However, according to previous study by Kim et al **[4],** the incidence of LGMS in the extremities or trunk may be higher than expected. Based on the rarity of this condition, this study aims to introduce a case report of LGMS in lung. LGMFS is a rare malignancy that is clearly classified as fibroblast/myofibroblast malignancy by the World Health Organization (WHO) in 2020 **[5].** The etiology of LGMFS is unknown. This solid infiltrating soft tissue tumor occurs in various parts of the body, particularly the tongue, followed by the mandible, neck, larynx, palate,

maxilla, and lip **[6].** The mean age of the patients was 40 years, but was reported in all age groups (7-85 years) **[7].** Because of the scarcity of published cases and the limited size of case series in the literature, it is unclear whether there is a gender bias **[8].** LGMFS is described as a slow-growing, painless mass and is often misdiagnosed as a benign lesion. At present, the treatment of choice for the disease is surgery, radiotherapy and chemotherapy is only adjuvant, and there is not much data. The patient was very old and the tumor invaded the main bronchus and esophageal wall directly, so it was difficult to operate, so the patient received radiotherapy first, but the patient was very old and his physical condition was poor, so he could not tolerate chemotherapy, how to choose effective and low-toxic drugs is a difficult problem. At present, Immunotherapy is the research hotspot of soft tissue sarcoma, cell-programmed death receptor-1/programmed death ligand-1(PD-1/PD-L1) MAB or combination therapy has shown some antitumor activity in some subtypes of soft tissue sarcoma, but the objective response rate of immune monotherapy was less than 10% **[9-11],** so the idea of combined antiangiogenic therapy came to mind. At present, amlotinib has been approved for soft tissue sarcoma indication, can be used as a very good combination of drugs. Amlotinib is a multi-target tyrosine-kinase inhibitor developed by our country. It can target to inhibit the kinase activities of VEGFR123, FGFR123, pdgfrαβ, c-Kit, Ret, etc., it has the effect of anti-angiogenesis and inhibiting the growth of tumor **[12].** In a phase II study of late-stage soft-tissue sarcoma with arotinib **[13],** PFS and OS were 5.5%. 6 months and 12. 3 months. Because of the low semi-inhibitory concentration on the above targets, its safety is better. The adverse reactions mainly include hypertension, hyperlipemia, hand-foot skin reaction and fatigue, most of which are grade 1-2, and can be tolerated or controlled. Amlotinib has been approved for the treatment of acinar soft tissue sarcoma, Clear-cell sarcoma sarcoma, and other advanced soft tissue sarcoma patients who have at least previously received anthracycline-containing chemotherapy for progression or recurrence; however, there is no report and study on the application of anlotinib in low-grade myofibroblastic sarcoma. After multi-disciplinary consultation and repeated communication with patients and their families, patients requested exploratory application. The patient was treated with sintilimab 200 MG Q3W in combination with oral amlotinib for 2 cycles, and the lesion size decreased, suggesting that anlotinib is effective for LGMFS. And patients with no significant adverse reactions, tolerance can be. It is still in continuous use and is expected to have a long-term effect. LGMFS is a rare malignancy with few clinical data and limited non-resectable treatment options. The combination of immuno-therapy with amlotinib may provide a new therapeutic option.

Conclusions

To the best of our knowledge, this is the first case report of LGMS arising in the lung. The Case showed combined immune checkpoint inhibitors and anti-angiogenic agents for the treatment of LGMS has certain clinical efficacy and acceptable safety, especially for patients who are inoperable.

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Citation of this Article

Zeng C and Zou M. Low-Grade Myofibroblastic Sarcoma of the Lung: A Case Report. Mega J Case Rep. 2023; 6: 2001-2007.

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