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Case Report
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Primary Malignant Pulmonary Glomus Tumor: A Case Report and Literature Review

*Luo S[1], Fu T[1], Liu F[2] and Wang Z[3]**

[1]Department of Radiology, Hainan General Hospital (Hainan Affiliated Hospital of Hainan Medical University), P. R. China

[2]Department of Pathology, Hainan General Hospital (Hainan Affiliated Hospital of Hainan Medical University), P. R. China

[3]Department of Radiology, Hainan Hospital of Traditional Chinese Medicine, China

Abstract

Malignant glomus tumors originating from the lungs are relatively rare. Here, we report a case of primary malignant pulmonary glomus tumor. The patient was a 41-year-old female. Chest CT revealed an 8 cm × 6 cm mass in the upper lobe of the right lung with multiple spotted calcifications and uneven enhancement on enhanced scans. Multiple metastatic nodules were seen in both lungs, and enlarged lymph nodes were observed in the mediastinum and right hilar lobe. The pathological diagnosis after right lung puncture was malignant pulmonary glomus tumor. Chemotherapy and targeted therapy with arotinib hydrochloride were used, and the patient was still alive after 36 months of follow-up. The clinical manifestations of primary malignant pulmonary glomus tumor lack specificity. The imaging manifestations are well-defined nodules or masses, with peripheral enhancement on enhanced scans. The imaging manifestations of benign and malignant pulmonary glomus tumors overlap with those of other lung tumors, making differential diagnosis difficult. The diagnosis is mainly based on pathology and immunohistochemical staining.

Keywords: Glomus tumor; Pulmonary; Malignant; Computed tomography

Introduction

Glomus tumors are rare soft-tissue mesenchymal tumors originating from perivascular glomus cells of arteriovenous anastomoses and account for about 1.6% of soft-tissue tumors [1]. They were first reported by Masson in 1924 [2]. Glomus tumors tend to occur in the dermis and subcutaneous tissues of the extremities with abundant glomus cells; most glomus tumors are benign and the proportion of malignant glomus tumors is less than 2% [3]. Glomus tumors can also occur in other areas, including the gastrointestinal tract, female reproductive tract, and the cardiovascular and respiratory systems [4-6]. Glomus tumors of the respiratory tract typically occur in the trachea and bronchial airway, and malignant glomus tumors originating in the lung are rare [7,8]. We report a case of primary malignant pulmonary glomus tumor confirmed by pathological biopsy and immunohistochemistry. We also review the relevant literature and highlight the imaging features of the tumor.

Case Presentation

The present study was approved by the Ethics Committee of Hainan General Hospital (Hainan, China) does not require approval for case report, and informed consent was received from the patient prior to the study.

The patient was a 41-year-old female. She had experienced cough, sputum, and chest pain for two years, accompanied by paroxysmal needle-like chest pain, as well as shortness of breath after activity. Local treatment for repeated attacks of pneumonia resulted in no improvement. Six months before presentation, the symptoms worsened, with increased coughing and small amounts of bright red blood in the sputum over three days. Chest CT showed the presence of a soft-tissue density mass shadow in the upper lobe of the right lung, with a maximum cross-section of about 8.8 cm × 6.8 cm. Multiple speckled calcification foci were seen within the mass, and the boundary with the right mediastinum was unclear. The trachea was slightly compressed and the bronchus in the upper lobe of the right lung was narrowed and blocked. The enhanced scan was uneven, showing compression of the superior vena cava (Figures 1-3) and the presence of a low-density shadow in the upper

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Correspondence:
Zhenping Wang, Department of Radiology, Hainan Hospital of Traditional Chinese Medicine, NO. 47, Heping North Road, Meilan DIC, Haikou, Hainan, 570203, P. R. China,

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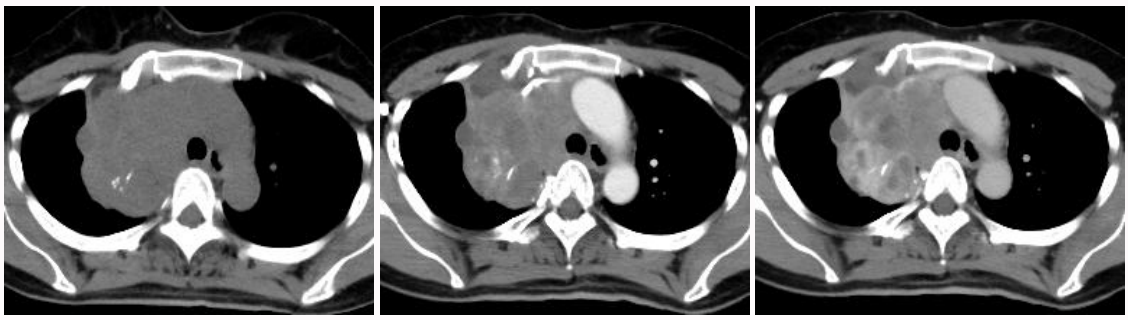

Figures 1-3: Initial chest CT. A mass in the upper lobe of the right lung, with multiple speckled calcification. The enhanced scan showed uneven enhancement in the arterial phase and progressive enhancement in the venous phase.

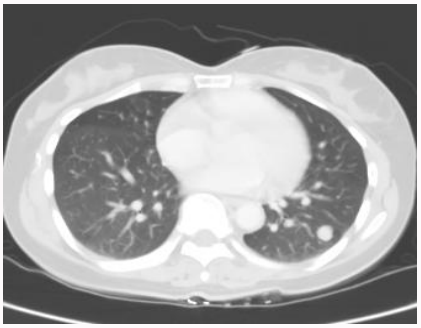

Figure 4: Metastasis was observed in the lower lobe of the left lung.

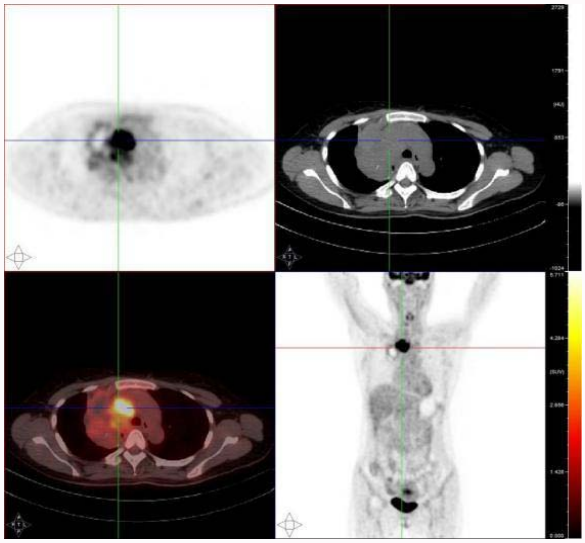

Figure 5: PET-CT shows a mediastinal mass in the right superior lobe of the lung, with enlarged lymph nodes in the mediastinum and right hilum of the lung, and increased FDG metabolism in the lesion.

lobe of the right lung. Multiple nodules were observed in both lungs, with a larger nodule located in the lower lobe of the left lung (Figure 4). The diameter of this nodule was approximately 1.3 cm. Enlarged lymph nodes were seen in the mediastinum and right hilum, and the right upper pleura was thickened.

^{18}F-FDG PET-CT showed a mass beside the mediastinum of the superior lobe of the right lung. The mass had a size of 8.4 cm × 6.5 cm. The density of the mass was not uniform, with multiple low-density shadows and scattered calcification foci, and the boundary with the mediastinum was not distinct. Further investigation showed multiple high-metabolic foci of FDG concentration in the lesion, with the largest located near the aortic arch. The size was approximately 4.1 cm × 2.5 cm, with an SUVmax of about 11.2 g/mL (Figure 5). Multiple nodules could be seen in both lungs, with the largest in the lower lobe of the left lung with a size of about 1.5 cm × 1.1 cm, showing an SUVmax value of 6.5 g/mL. There were enlarged lymph nodes in the mediastinum and right lung hilum, together with increased FDG metabolism and an SUVmax value of 4.3 g/mL.

Biopsy of the right lung showed that the tumor was rich in thin-walled blood vessels, with irregular lumens, visible branches, and swelling. Tumor cells were seen growing diffusely around the blood vessels. The tumor cell boundaries were clear, with round, polygonal, or partially fusiform shapes. The cells contained abundant cytoplasm and showed acidophilic granulosis or light staining. The nuclei were obviously atypical, with some having nucleoli, as well as evidence of inclusion bodies, nuclear division (4/50HPF) and pathological nuclear division (Figure 6, 7). Immunohistochemical staining indicated swelling. Tumor cells showed SAM, Caldesmon and Vimentin (diffuse +), Syn (partial +), CD34 (blood tube +), Ki-67 (20%), EMA, CK, CgA, S-100, Desmin, LCA, MelanA, HMB45 both (-). The pathological diagnosis was malignant glomus tumor of the right lung.

Due to metastasis in both lungs and the mediastinal lymph nodes, the patient was reluctant to undergo surgery and received chemotherapy. At the follow-up six months later, chest CT showed that the mass and intrapulmonary metastases in the upper lobe of the right lung were larger than before. Anlotinib hydrochloride was then self-administered After targeted therapy, chest CT at 17, 28, and 36 months follow-up showed that the right lung mass and intrapulmonary metastasis were smaller than before (Figure 8, 9). There were no new metastases.

Discussion

Glomus tumors originate from glomus cells, a type of mutated smooth muscle cell located at arteriovenous anastomoses [9] as well as under the nail bed of the fingers and toes. The tumors are thus most likely to occur in the palm, wrist, forearm, and foot, and are mostly benign. Tumors have also been reported in the nasal cavity, trachea, lung, thyroid gland, esophagus, stomach, colon, rectum, and other parts [1,3-8]. Due to the lack of angio-spheroids, glomus tumors originating in the lungs are very rare. To date, only 40 cases of pulmonary glomus tumors have been reported in the Chinese and foreign literature [10-12]. The age of onset ranges from 9 to 83 years, occurring mostly in middle and older ages, with a similar occurrence

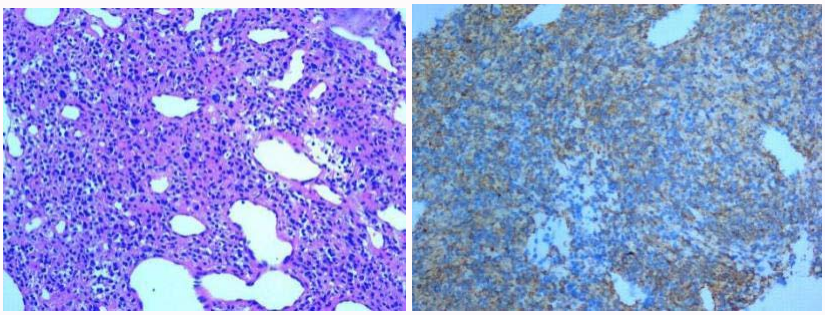

Figure 6, 7: Histological and immunohistochemical findings. Figure 8: The tumor were polymorphic with obvious nuclear atypia and pathological nuclear division. H&E stain, original magnification x200. Figure 9: The tumor cells were positive for α-SMA. EnVision method, original magnification x200.

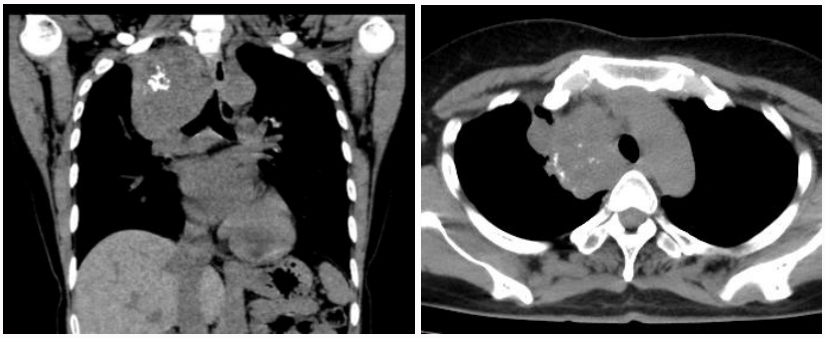

Figure 8, 9: Chest CT after 6 months showed that the mass in the upper lobe of the right lung was smaller than before.

between men and women [11,12]. There are no specific clinical features, and the common symptoms are cough, sputum, hemoptysis, chest pain, and dyspnea, among others.

According to the classification of soft tissue tumors by the World Health Organization in 2013 [13], glomus tumors are classified as benign glomus tumors, glomus tumors with uncertain malignant potential, malignant glomus tumors, and glomus tumors. The diagnostic criteria for malignant glomus tumors are (1) significant nuclear atypia and any level of nuclear division, and (2) atypical nuclear division. Tumors that do not meet the diagnostic criteria for malignant lesions and have at least one feature of nuclear atypia should be diagnosed as hemangioma with uncertain malignant potential. Immunohistochemical markers indicate diffuse hemangioma expression of SMA, muscle-specific actin, vimentin, Caldesmon, Calponin, and type IV collagen, with no expression of S-100 and cytokeratin [14]. Slightly different from soft tissue glomus tumors, glomus tumors of the lung and trachea are more malignant. It has been reported in the literature [11,12] that the majority of primary glomus tumors of the lung and trachea are benign, accounting for about 74%, followed by malignant glomus tumors (about 21%) and glomus tumors with uncertain malignant potential (about 5%). The reason may be related to the deep location of lung and trachea tumors, preventing early detection if the patient has no symptoms or does not undergo regular physical examinations.

The CT findings of pulmonary hemangioma show nodules or masses, occasionally coin-like nodules, ranging in size from 1.0 cm to 9.7 cm, with uneven densities and no calcification or cavities. Enhanced CT shows uneven peripheral enhancement, with no obvious central enhancement. Enhanced CT is able to reflect the distribution of the peripheral blood vessels of the tumor [10-12,15-21]. Cunningham et al. [10] considered that both benign and malignant pulmonary glomus tumors show no specific imaging features, and the sizes of benign and malignant tumors overlap considerably. Tumors with the largest diameters of 9.7 cm were found to be pathologically benign [11,12]. MRI findings of glomus tumor are rarely reported but show equal signals on T1WI and high signals on T2WI, as well as high signals in the central area on both T1 and T2 images. Enhanced MRI shows significant peripheral enhancement in the early stage, gradually extending to the central part of the tumor, with no enhancement in the center of the tumor [19]. It has been reported that FDG PET of pulmonary glomus tumors shows round or irregular soft-tissue masses with low- to moderate-intensity FDG accumulation [10,22], with SUVmax values ranging from 4.5 to 5.2, while malignant glomus tumors may also present with low to moderate uptake.

The imaging characteristics of pulmonary malignant glomus tumors have not been described. The differential diagnosis includes carcinoid and hemangiopericytoma/solitary fibroma, smooth muscle tumor (especially epithelioid leiomyoma), Primitive Neuroectodermal Tumor (PNET), paraganglioma, and metastatic tumors, among others [8]. Histological and immunohistochemical staining can be used to distinguish these tumors effectively.

Surgical treatment is the first choice for pulmonary hemangioma. Benign hemangioma has a good prognosis. No postoperative recurrence and malignant transformation have been reported. The prognosis of malignant pulmonary vascular balls is still unclear, and distant metastasis is the main cause of poor prognosis and death. Metastasis usually occurs 3 to 4 years after surgery, with metastasis rates of 31.2% to 38.0% [23]. DeCocker et al. indicated that sub-lobectomy by wedge, anatomical segmentectomy, or sleeve resection are the preferred treatments for malignant pulmonary glomus

tumors [7,24,25], which can be combined with mediastinal lymph node dissection and postoperative chemotherapy. The present case of pulmonary malignant glomus tumor was treated with chemotherapy and anti-nilotinib hydrochloride, and has survived after 36 months of follow-up. However, the mechanism of action of anrotinib in patients with malignant pulmonary glomus tumors still needs to be supported by clinical studies with larger sample sizes.

To sum up, primary malignant pulmonary glomus tumor is a rare tumor that lacks specific clinical manifestations. Imaging manifestations are nodules or masses with clear boundaries and peripheral enhancement in enhanced scans. The imaging findings of benign and malignant pulmonary glomus tumors overlap with those of other lung tumors, making differential diagnosis difficult and thus mainly reliant on pathological and immunohistochemical staining. The main treatment for malignant pulmonary glomus tumors is lobectomy, supplemented by chemotherapy after surgery. The long-term prognosis is not clear.

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