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Case Report

Cardiac angiosarcoma with lung metastasis presented with multiple halo signs: One case report and literature review ☆,☆☆

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ABSTRACT

Cardiac angiosarcoma is a rare aggressive malignancy with rapid progress and poor prognosis. Here we report 1 patient with cardiac angiosarcoma with lung metastasis, which presented as multiple halo signs and ground glass opacities. The patient underwent computed tomography (CT) guided lung biopsy and postoperative tissue histology confirmed the diagnosis of angiosarcoma. Transthoracic echocardiography (TTE) and cardiac Magnetic Resonance Imaging (MRI) identified the main tumor in the right atrium. Positron emission tomography/computed tomography (PET/CT) excluded intraabdominal lesions. The patient was given chemotherapy with nab-paclitaxel, cardiac radiation therapy and remained follow-up. Considering the rapid disease progression and poor prognosis, the present case report is intended to provide diagnostic insight into cardiac angiosarcoma with lung metastasis, especially with lung CT scans of multiple halo signs and ground glass opacities.

Introduction

Angiosarcoma is a soft tissue sarcoma that originates from vascular or lymphatic endothelial cells. Cardiac angiosarcoma is the most common primary cardiac neoplasm, which accounts for approximately 25%-30% of all primary cardiac malignancy [1]. It predominantly affects adults aged 40-50 years and most occur in the right atrium. Here, we present a case of a 41-year-old man diagnosed with primary cardiac angiosar-

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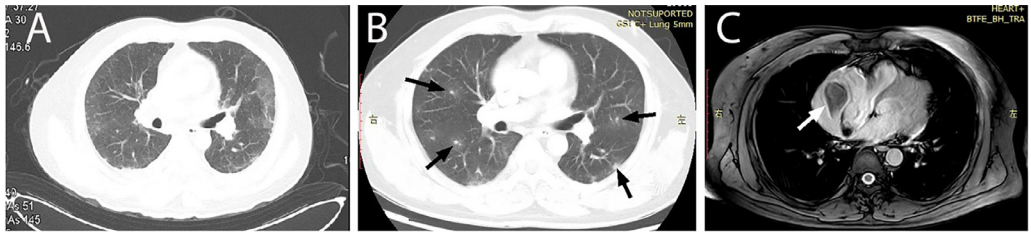

Fig. 1 – (A) Axial chest CT image showed multiple ground-glass opacities in bilateral lung fields (2024.3.13) (B) Axial chest CT image showed multiple halo signs and ground-glass opacities in bilateral lung fields (black arrows, 2024.5.16); (C) Cardiac magnetic resonance scan showed an immobile mass (white arrow) in the right atrium (white arrows).

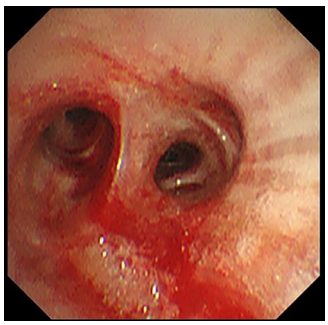

Fig. 2 – Bronchoscopy showed intrabronchial hemorrhage in right lower lobar bronchus.

coma with lung metastasis presented with multiple halo signs and ground glass opacities.

Case report

A 41-year-old smoking male driver presented with cough, exertional dyspnea, intermittent hemoptysis for 4 months. There was no fever, headache, weight loss, skin eruption, or chest pain. He visited several hospitals and underwent brain CT scan, PET/CT, gastroscopy and colonoscopy, which showed no obvious findings. TTE showed pericardial and pleural effusion. Cytological examination from pericardial and pleural drainage showed proliferative mesothelial cells. Physical examination revealed a body temperature of 36.8 °C, heart rate of 110 beats per minute (bpm), blood pressure of 123/84 mmHg, oxygen saturation of 95 %. Electrocardiography (ECG) showed a 2: 1 block atrial flutter with a heart rate of 140 bpm. Considering his presentations and frequent exposure to cigarette, as well as the multiple halo signs and ground glass opacities radiographic findings (Fig. 1A), infections like virus, mucorcosis, atypical or hypersensitivity pneumonitis was suspected initially. However, there was no improvement after empirical antimicrobial and corticosteroid treatments. Autoantibodies (including Extractable Nuclear Antigen (ENA) Antibodies Panel, antineutrophil cytoplasmic antibodies, Antiglomerular basement membrane (anti-GBM) antibody) were all negative. Diagnostic bronchoscopy were performed, which showed multiple intrabronchial hemorrhage (Fig. 2). Bronchoalveolar lavage fluid (BALF) cellularity and transbronchial lung biopsy (TBLB) tissue pathology were nonspecific. Initial cultures (blood, sputum, and lavage fluid) were negative. BALF targeted next generation sequencing (tNGS) test indicated aspergillus flavus, debaryomy ces hansenii, human beta herpesvirus 7 and human rhinovirus, which did not corelate with clinical menifestations. CT guided lung biopsy was then performed and postoperative tissue histological examination revealed poorly differentiated spindle-shaped cells. Immunohistochemical staining were positive for cluster of differentiation 31 (CD31), CD34, friend leukemia integration 1(Fli-1), ERG, Vimentin, P53, integrase interactor 1 (INI-1), but negative for cytokeratin (CK), thyroid transcription factor 1 (TTF1), Napsin-A, Cam5.2, epithelial membrane antigen (EMA), and carcinoembryonic antigen (CEA). Semiquantitative estimation of Antigen Kiel-67 (Ki-67) index was 40% (Figs. 3A-E). TTE and MRI (Fig. 1B) identified the main tumor in the right atrium. PET/CT excluded intra-abdominal lesions. High grade and stage IV right atrial angiosarcoma with pulmonary metastases was diagnosed. The patient was given chemotherapy with nab-paclitaxel, cardiac radiation therapy and remained follow-up.

Discussion

Angiosarcoma is a soft tissue sarcoma that originates from vascular or lymphatic endothelial cells. They are defined “high-grade” because of their aggressive behavior [2]. Angiosarcoma can occur in any in any part of the body and more commonly in older patients with cutaneous form. Primary cardiac angiosarcoma is the most common primary cardiac neoplasm, which accounts for approximately 25%-30% of all primary cardiac malignancy [1]. Because they are highly aggressive, most cases have already developed distant metastasis at the time of diagnosis. Metastasis are common, including lung, liver, lymph nodes, adrenal gland, bone, and brain, in which lungs being the most frequently affected site.

Symptoms of cardiac angiosarcoma are often nonspecific, which depend on local invasion and systemic metastasis, such as atrioventricular block arrhythmias, cough, hemoptysis, fever, dyspnea, and abdominal pain, making early diagnosis challenging. In the present case, the the patient was admitted to the hospital with dyspnea, occasional cough and hemoptysis, and was already with lung metastasis when con-

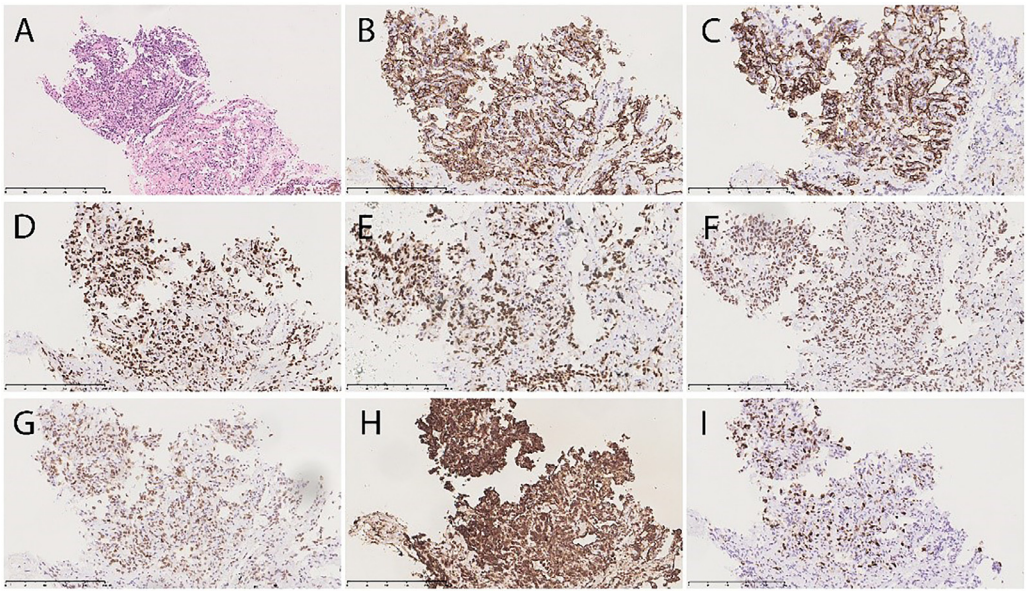

Fig. 3 – Pathological finding. Histological investigation showed poorly differentiated spindle shaped tumor cells (A, H&E stain) and immunohistochemically, they stained strongly positive for CD31 (B), CD34 (C), Fli-1 (D), ERG (E), INI-1 (F), P53 (G), and Vimentin (H); Semiquantitative estimation of Ki-67 index was 40% (I). Scale bars A, 100 μm; B-I, 50μm. CD = cluster of differentiation; Fli-1 = friend leukemia integration 1; INI-1 = integrase interactor 1, CK= cytokeratin, H&E = hematoxylin and eosin, TTF-1 = Thyroid transcription factor-1, Cam5.2 = anticytokeratin, CEA = carcinoembryonic antigen, Ki-67 = Antigen Kiel-67, (A 100 ×, B-I 200 ×).

firming the diagnosis. TEE has sensitivity of 75% identifying the cardiac mass. Cardiac MRI with contrast is helpful characterizing the tumor and its vascularization [3]. PET/CT is useful for potential metastasis evaluation. Nontheless, the golden standard for the diagnosis must be based on histopathological and immunohistochemical results.

Halo sign of lung CT scan is defined as parenchymal lung consolidation or nodules surrounded by ground-glass opacity attenuation, indicating alveolar hemorrhage surrounding by pulmonary infarction. Although halo sign was first described in immunosuppressed patients with Invasive Pulmonary Aspergillosis (IPA), other infectious conditions like mucormycosis, Cryptococcus, candida, respiratory syncytial virus, cytomegalovirus, and noninfectious conditions like lymphomatoid granulomatosis, sarcoidosis, primary lung neoplasm, lung metastases like angiosarcoma, melanoma, lymphoma should also be considered [4].

The histological features of angiosarcoma are characterized by pleomorphic, malignant endothelial cells. Immunohistochemical staining are positive for CD31, Fli-1, von Willebrand factor, vascular endothelial growth factor (VEGF), and Ulex europaeus agglutinin 1. Other markers like CD34, cytokeratin and vimentin can also be useful. Currently the treatment is very challenging because of rapid progress and poor prognosis, especially if there is metastatic stage. Surgical resection with involved margins is the primary treatment if there is local confined disease. For advanced or metastatic stage diseases, chemotherapy with anthracyclines, taxanes or ifosfamide is the first line of treatment. Adjuvant radiotherapy should also be considered. Angiosarcoma originates from vascular endothelium, however, anti-VEGF therapy with bevacizumab failed to gain benefit in a phase II study [5]. Genomic sequencing study showed facial and scalp angiosarcomas have high tumor mutation burden and ultraviolet light mutational signature that may have potential responsiveness to checkpoint inhibitors [6]. Immune checkpoint inhibitors (ICIs) are effective in some case series [7], however, in a retrospective study, patients with advanced cardiac angiosarcoma failed the ICIs treatment because of tumor progression [8]. Treatment with ICIs still need to be tested in a prospective trial for further study. For this case, we are undergoing chemotherapy and radiotherapy and follow up to see if there is durable responses.

Conclusion

From this case report, we could learn that early diagnosis is critical for the optimal treatment of cardiac malignancies. Lung metastasis with angiosarcoma should be noticed if there are halo signs, especially in patients with immunocompetent conditions.

Patient consent

Written informed consent was obtained from patient and his family for publication.

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