Perspective

Multiple myeloma's pulmonary hypertension aetiology

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ABSTRACT

Pulmonary hypertension frequently exacerbates multiple myeloma through a variety of processes. These causes include concurrent cardiac amyloid-induced pulmonary hypertension, congestive heart failure, chronic thromboembolic pulmonary hypertension, high output heart failure caused by anemia or lytic bone lesions, and chronic thromboembolic pulmonary hypertension. This case study series demonstrates the numerous pathways via which people with multiple myeloma experience pulmonary hypertension. To determine the causes of pulmonary hypertension and how to treat it in patients with multiple myeloma receiving care at the University of California, San Diego. In order to identify individuals with multiple myeloma and pulmonary hypertension, a retrospective chart analysis was carried out. Patients also needed to undergo a right heart catheterization. Chart review provided information on pulmonary hypertension's demographics, comorbidities, clinical trajectory, and a etiology.

Key Words: *Pleural disease; Bronchoscopy; Interventional pulmonary medicine.*

INTRODUCTION

There are over new diagnoses of multiple myeloma each year, which accounts for over of all hematological malignancies. Multiple myeloma is defined by the neoplastic growth of plasma cells. Multiple myeloma can damage a variety of organ systems, including the circulatory system and pulmonary circulation, while being largely a hematological condition. There isn't much information available, though, about patients with multiple myeloma who also develop pulmonary hypertension. The World Health Organization classifies pulmonary arterial hypertension, pulmonary arterial hypertension due to left heart disease, pulmonary arterial hypertension due to lung disease/hypoxia, chronic thromboembolic pulmonary hypertension, and pulmonary hypertension due to multifactorial mechanisms into five groups based on a etiology. Although this risk is not clearly defined, patients with multiple myeloma are at risk for PH in all five groups. It has been hypothesized that MM patients experience PH at higher rates than people in general. According to earlier research, between and individuals with MM exhibited echocardiographic indications of PH at the time of diagnosis. This contrasts with the significantly lower prevalence of PH observed with other comorbidities. For instance, a recent analysis of over patients discovered that PH was only sometimes present in those with left heart disease, lung disease, and thromboembolic disease, showing that it is a relatively uncommon consequence of these widespread illnesses. There is no known cause for why patients with multiple myeloma have a higher incidence of PH than the general population, but potential causes include cardiomyopathy associated with MM, thrombophilia in MM leading to CTEPH, and unfavorable effects from chemotherapeutic drugs (dasatinib, carfilzomib) used to treat MM that can lead to PAH. Additionally, AL amyloid exacerbates up to of MM cases, and in at least of these patients, cardiac involvement can result in heart failure. Patients with MM are also at risk for high-output heart failure, which is believed to be a result of numerous arteriovenous fistulas in the context of severe bone involvement that may also result in group. With the exception of one tiny series of three patients, the majority of these earlier investigations lacked right cardiac catheterization data. Our case series seeks to identify the causes of pulmonary hypertension in multiple myeloma, as well as the course of treatment for the condition and its prognosis. Using a data extraction tool in the electronic health record, patients were found through a retrospective case review. Adult patients with pulmonary hypertension and multiple myeloma both primary and secondary pulmonary hypertension exist. We also required that a right heart catheterization be completed. From the electronic medical record, information on demographics, clinical laboratory data, drugs, imaging, procedure results, and clinical course was extracted and put

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into a safe database for analysis. A manual chart review was used to determine how accurately multiple myeloma and pulmonary hypertension were coded. Examining physician progress or clinic records, lab or biopsy data, and procedure notes allowed for the confirmation that patients satisfied all inclusion criteria. The initial search of the electronic medical record for patients with codes for multiple myeloma, pulmonary hypertension, and right cardiac catheterization yielded a total of individuals. From this group of patients, we eliminated those with normal pulmonary pressures, those with smoldering multiple myeloma as their diagnosis, those with monoclonal gammopathy of unknown significance, those who passed away before having a bone marrow biopsy to confirm their diagnosis, those who underwent right heart catheterization to implant a watchman device but did not have hemodynamic measurements taken, and patients. Diagnostic codes may have been used to label patients with multiple myeloma even if they did not have the condition. Upon reviewing their medical records, it was discovered that many of these patients had comparable diagnoses (such as amyloid without multiple myeloma) and that during the initial phases of diagnostic testing, doctors had recorded a number of tests under the ICD-code for multiple myeloma. The presence of multiple myeloma, pulmonary hypertension, and a right heart catheterization with measurement of intracardiac pressure were ultimately determined to be present in all patients who met the inclusion criteria. Two of the patients were diagnosed with PAH brought on by carfilzomib, two had PH from cardiac amyloid, one had high output heart failure, one had chronic thromboembolic pulmonary hypertension (CTEPH), two had congestive heart failure, and five more had both.

Orthotropic heart transplants were performed on both patients who had concurrent multiple myeloma and cardiac amyloid. The pulmonary artery pressures of one patient improved after receiving a heart transplant, but the pulmonary artery pressures of the second patient remained unchanged but improved subjectively. One patient was found to have high output heart failure brought on by lytic bone lesions and anemia. Alendronate was used to treat this patient's pulmonary hypertension, along with several other myeloma-specific treatments. As a result of severe cardiomyopathy and a poor ejection fraction, the patient with CTEPH was not found to be a candidate for pulmonary hromboendarterectomy (PTE) surgery. In its place, riociguat was started on the patient. After carfilzomib treatment for multiple myeloma began, two patients experienced pulmonary arterial hypertension. Pulmonary artery pressures in both cases decreased once carfilzomib was stopped. Nevertheless, a patient who had to permanently stop using carfilzomib needed to start PAH-targeted therapy. Additionally, she underwent a second autologous stem cell transplant when the multiple myeloma disease progressed after carfilzomib was stopped being used. Of the five patients who had congestive heart failure and developed pulmonary hypertension, four had heart failure with intact ejection fraction and one had heart failure with reduced ejection fraction. The cause of heart failure in the patient with heart failure is investigated to see if each patient underwent a right heart catheterization or echocardiography prior to the date of multiple myeloma diagnosis. Four of the eleven patients had not undergone a right cardiac catheterization or an echocardiography prior to being diagnosed with multiple myeloma. Prior to receiving a multiple myeloma diagnosis, one patient with CTEPH experienced pulmonary hypertension that was confirmed by a right heart catheterization. After receiving a multiple myeloma diagnosis, pulmonary hypertension worsened in both patients with likely medication-induced PAH brought on by carfilzomib. The diagnosis of multiple myeloma resulted in either new or worsened pulmonary hypertension in four patients with WHO group pulmonary hypertension caused by left heart dysfunction. This case series highlights the various ways that patients with multiple myeloma can become hypertensive, including pulmonary arterial hypertension brought on by chemotherapy, left heart failure frequently caused by concurrent amyloidosis, high output heart failure brought on by lytic lesions/anemia, and CTEPH from thrombophilia. A right cardiac catheterization was performed on each patient in this case series who had multiple myeloma and echocardiographic evidence of pulmonary hypertension in order to identify the origin of the condition. According to earlier research, pulmonary hypertension occurs in about one-third of individuals with multiple myeloma who receive a new diagnosis. This case series serves as an example of how it is crucial to identify the cause of pulmonary hypertension via right cardiac catheterization because it has significant therapeutic consequences. Invasive hemodynamics enables measurement of the cardiac output, pulmonary artery pressures, and pulmonary capillary wedge pressure to assist in differentiating the various etiologies of pulmonary hypertension. Patients should get a VQ scan as part of the evaluation for pulmonary hypertension in MM since the condition is linked to a very high risk for pulmonary embolism. This screening is to check for chronic thromboembolic pulmonary hypertension. All MM patients with pulmonary hypertension also require a complete medication review to find any drugs that may be linked to the emergence of PAH. Two individuals in our case series had pulmonary arterial hypertension as a side effect of MM treatment. When the offending medicine was stopped, their symptoms and PA pressures both improved (carfilzomib). Notably, some multiple myeloma drugs, including dasatinib and carfilzomib, as well as several monoclonal antibodies and immune regulators, have been linked to PH. One trial found improvements in NYHA functional class and pulmonary arterial pressures on repeat right heart catheterization, indicating that dasatinib-induced PAH can improve with medication withdrawal. However, around one-third of patients required long-term targeted PAH therapy since they continued to have PAH even after dasatinib was stopped. With the withdrawal of carfilzomib, hemodynamics have improved in two more small case series. However, there were patients who continued to experience PAH even after permanently stopping the offending medication, comparable to dasatinib-induced PAH. Leflunomide, on the other hand, is a popular immune modulator utilized in multiple myeloma treatment plans. Leflunomide has been linked to PAH in certain cases, however a recent study indicated that this association was rare and that individuals with additional PH risk factors were more likely to experience pulmonary hypertension when using leflunomide. Guidelines on cardio-oncology recently released by the European Society of Cardiology recommend baseline echocardiography prior to beginning treatment with dasatinib or proteasome inhibitors. All patients receiving treatments known to increase the risk of PAH should have long-term follow-up. Patients who exhibit indications or symptoms of pulmonary hypertension or heart failure should undergo repeat echocardiography. As more and more chemotherapeutics are created for the treatment of multiple

myeloma, drug-induced PAH in multiple myeloma may continue to rise in the coming years. Medication review is a crucial step to find potential offending drugs when PH is found in a patient with multiple myeloma. Concomitant cardiac amyloid is a significant factor in the development of PH in multiple myeloma patients. Multiple myeloma cases can become complicated by immunoglobulin light chain (AL) amyloid, with cardiac involvement happening in at least one of these patients and potentially leading to heart failure. Misfolded immunoglobulin light chains are accumulated as amyloid fibrils in the heart of these patients. Amyloid-suggestive echocardiographic characteristics include septal wall thickness, significantly reduced left ventricular tissue Doppler, and reduced global longitudinal strain with apical sparing pattern. The European Society of Cardiology (ESC) advises patients who have a clinical suspicion of cardiac amyloidosis to undergo serum and urine electrophoresis with immunofixation, serum free light chain ratio, and bone tracer cardiac scintigraphy. Cardiovascular scintigraphy in patients with cardiac ATTR amyloidosis demonstrates higher uptake of bone tracer, but it is often null in patients with AL cardiac amyloid. Patients with AL cardiac amyloid can be identified through either a positive end myocardial biopsy or an extra cardiac biopsy that meets both echocardiographic and cardiac MRI criteria as well as being positive for amyloid. Group PH can occur in people with heart failure who have cardiac involvement from AL amyloid (due to left heart failure).

As the median survival from the onset of heart failure is short, it is essential to recognize these patients early. It's crucial to remember that newer cardiac amyloid treatments, including tafamidis, should only be used to treat ATTR amyloid. The mainstay of treatment for AL amyloidosis is chemotherapy, which blocks the formation of light chains by plasma cells. Patients who meet certain criteria may be candidates for cardiac or multiple organ transplants. Two patients with concurrent multiple myeloma and cardiac amyloid were found in our series. Heart failure was the primary symptom of cardiac amyloidosis in both of these individuals, and an end myocardial biopsy confirmed the diagnosis. One patient had an end myocardial biopsy that was positive after an initial fat pad biopsy that had tested negative for amyloidosis. A confirmatory end myocardial biopsy was performed on the second patient, who had a cardiac MRI that was indicative with cardiac amyloidosis. An orthotropic heart transplant was performed successfully on both patients.