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A Rare Case of Primary Malignant Pericardial Mesothelioma Diagnosed with Pericardiotomy

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ABSTRACT

Primary malignant pericardial mesothelioma (PMPM) is an extremely rare and lethal cardiac tumor. This article presents a 62-year-old man with recurrent pericardial fluid. The patient's clinical symptoms and imaging features were nonspecific. Initial diagnosis was constrictive pericarditis. After admission, the patient's symptoms worsened, and echocardiography indicated increased pericardial effusion. To diagnose and improve the patient's symptoms, pericardiotomy was performed; however, the procedure was unsuccessful because the pericardium was densely adherent to the myocardium. Histopathological examination, including immunohistochemical staining of the pericardial specimen revealed malignant mesothelioma. We recommended adjuvant therapy for the patient with cis-platinum and pemetrexed; however, the patient and his family refused treatment. The patient was discharged 11 days after surgery. The patient survived for more than 15 months with surgical treatment. In this report, the patient's symptoms improved, and the patient survived beyond the median survival after surgical treatment.

Conclusion: The definitive diagnosis of PMPM mostly has been obtained from specimens obtained by surgery. Surgery is an effective treatment method because it prevents cardiac tamponade and can improve symptoms or prognosis, but complete resection is impossible.

INTRODUCTION

Primary malignant pericardial mesothelioma (PMPM) is a rare and fatal condition that clinicians should be aware of owing to the variability of its clinical manifestations. The diagnosis may be delayed as a result of delayed treatment. Pathology remains the gold standard for the definitive diagnosis of PMPM. PMPM has no standard treatment. Because patients with PMPM frequently present late in the disease, surgical intervention usually is palliative. Here, we present a rare case of a 62-year-old man with PMPM diagnosed after pericardiotomy. Postoperatively, the patient's symptoms significantly improved and survival period was longer.

CASE REPORT

A 62-year-old man was admitted to our hospital with recurrent pericardial fluid on March 30, 2021. The patient had no incidental asbestos exposure. Blood chemistry revealed hepatocellular and renal injuries. Various examinations, including chest X-ray and echocardiography indicated pericardial effusions. Simultaneously, computed tomography revealed left pleural, pericardial, and pelvic effusions. (Figure 1) After admission, the patient's symptoms worsened, and echocardiography indicated increased pericardial effusion. The diagnosis of severe constrictive pericarditis causing decompensated heart failure was considered. After a preoperative discussion, our team decided upon a surgical intervention for the definitive diagnosis and to improve the patient's symptoms on April 12, 2021. Median sternotomy and pericardiotomy were performed. The bloody pericardial effusion was removed. Removing the left ventricular pericardium was difficult because it was closely adhered to the myocardium. The adhesions grew like tree roots. Histopathological of the pericardial specimen revealed malignant mesothelioma. (Figure 2) The patient's symptoms improved. We recommended adjuvant therapy with cis-platinum and pemetrexed for the patient after being diagnosed with PMPM, however, the patient and his family refused chemotherapy and opted for palliative care. The patient survived for more than 15 months at the follow up. The patient was informed that data concerning the case would be submitted for publication, and he consented.

DISCUSSION

PMPM is an extremely rare neoplasm and accounts for ~4% of primary heart and pericardial tumors [Apicella 2020]. Male patients dominate the PMPM population, with a median age of 46 years. PMPM often is found in the late clinical course or at autopsy [Massicotte 2019]. Unlike pleural mesothelioma, the relationship between PMPM and asbestos exposure is unclear [Mcgehee 2018]. Clinical features and imageological examinations are non-specific. PMPM is easily

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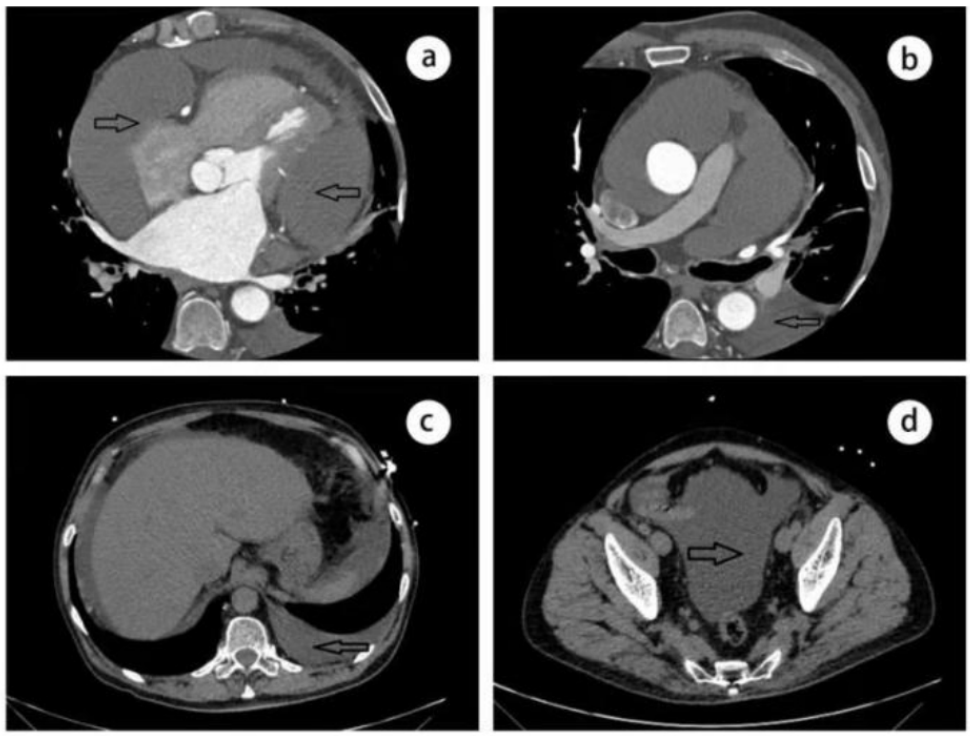

Figure 1. Computed tomography revealed massive pericardial effusion (A, arrows), left pleural effusion (B, arrows), seroperitoneum (C, arrows), and massive pelvic effusion (D, arrows).

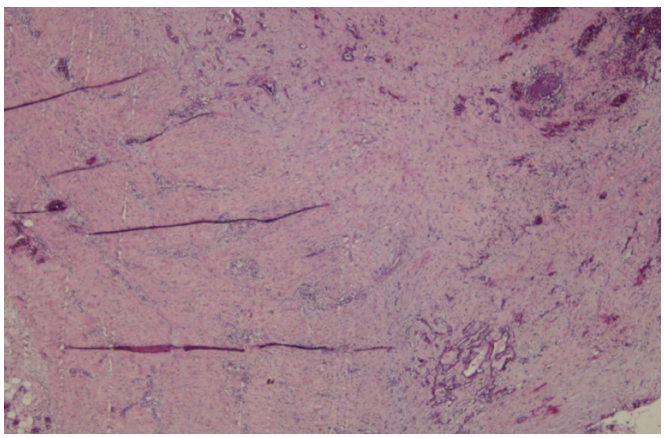

Figure 2. Histological analysis of the specimen: The tumor cells were large cuboidal or columnar cells with rounded nuclei, forming adenoid, cord-like, and papillary structures. The cells were atypical, and patches of bleeding were observed in the area.

confused with pulmonary diseases and general heart diseases. Therefore, the definitive diagnosis still depends on pericardial biopsy or postoperative pathological examination. In our case, initial diagnosis was constrictive pericarditis. Meanwhile, the patient's symptoms worsened, and the pericardial effusion rapidly grew after admission. Our team eventually decided to perform pericardiotomy for the definitive diagnosis and to improve the patient's symptoms. Histopathological examination of the pericardial specimen revealed malignant mesothelioma. Because of rarity, no standard treatment strategy has been established for PMPM. Surgery is the most important treatment. The operation includes pericardiectomy and pericardial window placement [Matsuyama 2020]. It is mostly useful for preventing cardiac tamponade or reducing tumor. Surgery and adjuvant therapies of chemotherapy or radiotherapy have become the fundamental treatment for selected patients, representing relatively prolonged survival. Even so, more than 30% of patients died during hospitalization, and the mortality rate at 6 months after discharge was ~75%. The prognosis of PMPM is extremely poor, and the fatality rate is high with a median survival of ~6 months [Kong 2017]. In this report, indication for surgery is difficult and it is not to cure the disease but to diagnose and save the patient's life.

CONCLUSION

PMPM is an extremely rare with a uniformly negative prognosis. The presence of recurrent pericardial fluid and relapsing cardiac tamponade or the resistance to effective anti-inflammatory or anti-tuberculous therapy should draw the attention of physicians to suspect PMPM. Surgery remains palliative in most cases and complete tumor excision scarcely is done. Nonetheless, surgical resection can be used to diagnose some difficult cases or may prolong patient survival.

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