Case Report

Primary Malignant Pericardial Mesothelioma Manifesting as Constrictive Pericarditis with Atypical Imaging Features: A Case Report

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ABSTRACT

Background: Primary malignant pericardial mesothelioma (PMPM) is an exceptionally rare and aggressive cancer originating from the mesothelial lining of the heart. This distinct and challenging oncologic entity is typically discovered at an advanced stage due to vague, nonspecific symptoms—most commonly chest pain and dyspnea. These clinical features often mimic more common conditions, making early detection difficult. Diagnostic confirmation usually requires detailed histopathologic analysis, often obtained through surgical procedures or postmortem examination. The disease's elusive nature and rapid progression contribute to its generally poor prognosis, underscoring the need for heightened clinical awareness and advanced diagnostic strategies.

Case Report: A 58-year-old patient was diagnosed with malignant pericardial mesothelioma, which progressed rapidly and resulted in death within 1 month of diagnosis.

Conclusions: PMPM remains a rare and highly aggressive cancer characterized by swift, fatal progression. Diagnosis is difficult because of nonspecific clinical symptoms. Accurate diagnosis is essential for subsequent treatment planning. Nonetheless, limited treatment alternatives, coupled with the minimal effectiveness of immunotherapy in advanced disease, highlight the pressing need for further research and therapeutic advances.

Keywords: Primary Malignant Pericardial Mesothelioma; Constrictive Pericarditis; Case Report

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Introduction

Malignant mesothelioma is a tumor that originates from the mesothelial lining, often found in the pleural space and peritoneum. Primary malignant pericardial mesothelioma (PMPM) is rare, aggressive, and typically has a poor prognosis. Chest pain and dyspnea are common early symptoms. Because of its unusual symptoms and imaging features, PMPM is often diagnosed at an advanced stage, with confirmation usually requiring histopathologic examination or postmortem findings.[1–3]

In this report, we describe the case of a 58-year-old patient diagnosed with malignant mesothelioma of the pericardium, which advanced rapidly and resulted in death within 1 month of diagnosis.

Case Report

The patient initially presented with severe dyspnea, nocturnal hyperhidrosis, and unintentional weight loss during the preceding 3 months, without a history of asbestos exposure, radiation therapy, or viral infections. There was no productive cough or chest pain. Two years earlier, the patient had idiopathic polyserositis, treated with NSAIDs, with no malignant cells found in previous imaging and histopathologic evaluations. On examination, the patient had sinus tachycardia (110 bpm) and hypotension. The severe dyspnea and unintentional weight loss raised concern about an underlying malignancy.

Laboratory investigations revealed hypochromic microcytic anemia (hemoglobin, 10 g/dL) and elevated D-dimer (1.76 mg/L). NT-proBNP was elevated (9751 pg/mL), along with alkaline phosphatase (253 U/L) and γ-glutamyl transferase (237 U/L). C-reactive protein was elevated (23 mg/dL), but procalcitonin was normal. The patient had reduced transferrin saturation (9%) and low serum transferrin (172 mg/dL), with elevated ferritin (3500 ng/mL), indicating anemia of chronic disease and iron deficiency. Additionally, β_2-microglobulin was slightly elevated (2.79 mg/L). Tests for hepatitis B, hepatitis C, and autoimmune diseases were negative, except for elevated anti-CCP antibody (200 U/mL). Electrocardiography showed sinus tachycardia (124 bpm), left axis deviation, persistent S wave, and slow R-wave progression.

The admitting diagnosis was heart failure (New York Heart Association functional class 3) and sinus tachycardia. Given the patient's medical history, atrial arrhythmia, and elevated C-reactive protein, constrictive pericarditis was considered. Coronary angiography performed 1 month earlier ruled out coronary artery disease. The chest radiograph showed bilateral pleural effusion without pulmonary infiltrates. Further investigation included echocardiography and thoracic computed tomography (CT).

Transthoracic echocardiography revealed a circumferential, organized pericardial effusion up to 12 mm, with thickening, adhesion, and biatrial enlargement, suggesting constrictive pericarditis (Figure 1).

These findings were confirmed by CT of the thorax, which showed pericardial thickening up to 3 cm and signs of constrictive pericarditis and pathologic mediastinal lymphadenopathy (Figure 2).

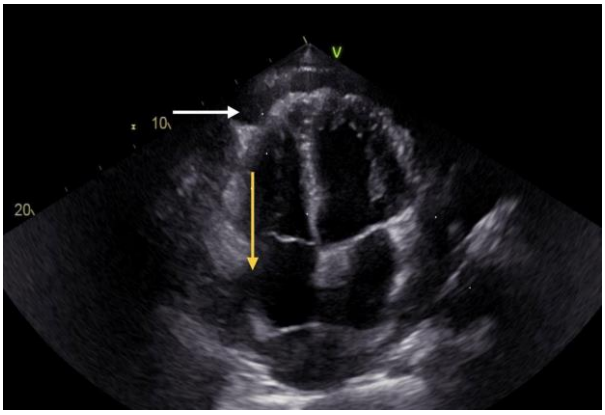

Figure 1. Transthoracic echocardiography shows a circular, organized pericardial effusion up to 12 mm in size without compromise of ventricular function (white arrow) and pericardial thickening, adhesion, and biatrial enlargement (yellow arrow).

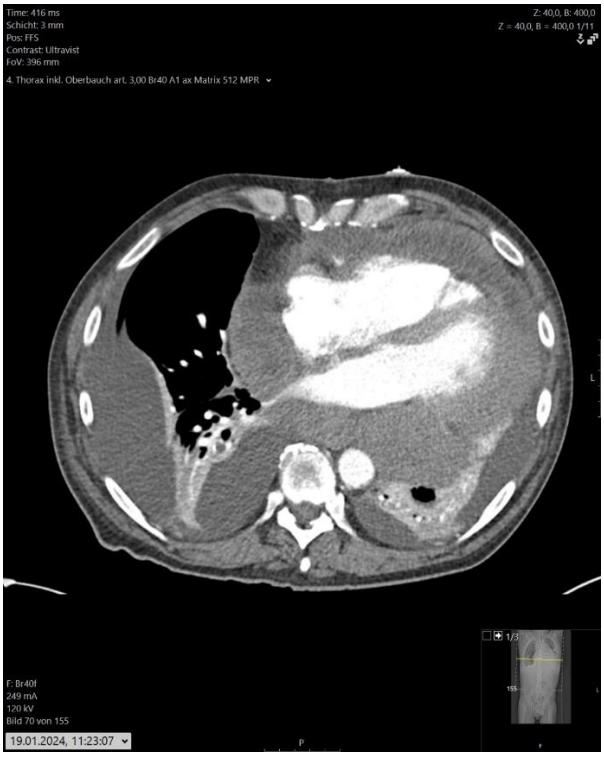

Figure 2. Computed tomography of the thorax shows a 3-cm pericardial effusion and signs of constrictive pericarditis.

Further investigations with colposcopy and gastroscopy showed no evidence of malignancy. A repeat CT scan 1 week later, prompted by a SARS-CoV-2 infection, revealed clear signs of pericardial tumor progression. There was substantial malignant pericardial infiltration and enlargement of right cervical, right axillary, and mediastinal lymph node metastases. Radiologic signs of superior vena cava syndrome were observed, although no clinical signs of engorged neck veins were present.

The patient was referred to a heart surgery center for further treatment. During surgery, a massive tumor encasing the entire heart and infiltrating the diaphragm was found. Complete resection was not possible because of the high risk of fatal hemorrhage. Histologic examination confirmed a diagnosis of malignant epithelioid mesothelioma of the pericardium.

Immunohistochemistry confirmed the diagnosis of a high-grade diffuse epithelial mesothelioma with 50% solid growth and no pleomorphic or rhabdoid morphology. The histologic samples were positive for pan-cytokeratin AE 1/3, with coexpression of WT-1, D2-40, and calretinin, while INI1 expression was preserved. The samples were negative for BerEP4.

The patient was referred to our hospital for systemic therapy. In agreement with the patient, immunotherapy with nivolumab and ipilimumab was started. Nevertheless, the patient died the following day.

Discussion

Malignant pericardial diseases are classified as primary or secondary tumors, with secondary tumors being more common.[4]

PMPM is a rare, aggressive tumor with a poor prognosis. Autopsy studies show an incidence of less than 0.0022%, accounting for approximately 1% of all malignant mesotheliomas.[1,2,5]

Because of its unusual clinical presentation and imaging findings, PMPM is often misdiagnosed. Definitive diagnosis relies on histologic examination of the pericardium, postoperative pathology, or autopsy findings.[1–3] The clinical misdiagnosis rate is high because PMPM symptoms are nonspecific and often mistaken for conditions such as tuberculous pericarditis, coronary artery disease, atrial myxoma, heart failure, and cardiomyopathy. Systemic symptoms of PMPM may include fever, night sweats, cough, and weight loss. Patients often present with dyspnea, chest discomfort, or chest pain due to pericardial constriction or cardiac tamponade.[5,6] In the present case, the patient initially presented with severe dyspnea, a history of nocturnal hyperhidrosis, and unintentional weight loss during the preceding 3 months, without a history of productive cough or chest pain.

Diagnosing pericardial conditions is often complicated and typically requires a combination of imaging techniques, such as echocardiography, CT, magnetic resonance imaging (MRI), and fluorodeoxyglucose positron emission tomography (FDG-PET).[7] The most common echocardiographic finding in PMPM is pericardial effusion. Hemorrhagic effusion is more likely to be malignant, but recurring effusion alone does not always indicate cancer. In this case, pericardial thickening, biatrial enlargement (constrictive pericarditis), and recurrent pericardial and pleural effusions suggested bleeding from tumor involvement of the pericardium. Therefore, recurrent effusions should not be solely relied on for diagnosis. Hemodynamic complications such as cardiac tamponade and constrictive pericarditis are common in PMPM. Some studies showed that cardiac tamponade increases the likelihood of malignancy, as seen in this case.[8] The patient was admitted because of recurrent cardiac decompensation and the onset of constrictive pericarditis.

Cardiac CT is crucial for assessing effusion density and calcification and for distinguishing malignancy from pericarditis by identifying solid nodules. It also helps evaluate the involvement of pericardial layers, epicardial fat, and myocardium. In this case, substantial progression was observed within 1 week, with increased malignant pericardial infiltration. Bilateral peripheral pulmonary embolism was also seen, which, along with other findings, suggests a malignant tumor and potential myocardial involvement, helping to differentiate PMPM from inflammatory pericardial lesions. Pericardial calcification is rare in malignant tumors, and while both PMPM and inflammatory diseases can cause pericardial and pleural

effusion, CT attenuation of the fluid is crucial because hemorrhagic density strongly indicates malignancy.

Cardiac MRI can evaluate abnormalities in cardiac hemodynamics and ventricular wall motion resulting from PMPM.[9] In this case, cardiac MRI was not performed because of worsening hemodynamic instability and increasing pericardial effusion, along with clear signs of malignancy on the CT scan.

Differentiating between PMPM and nonneoplastic lesions using imaging alone can be difficult, especially when diffuse thickening of the pericardium is present.[10] Accordingly, PMPM should be considered only after excluding other potential lesions.

Although clinical data and imaging techniques are crucial for diagnosing pericardial mesothelioma, a definitive diagnosis relies on pericardial biopsy or pathologic examination. Immunohistochemical analysis helps confirm mesothelioma and exclude other tumor types.[11]

Currently, no effective treatment for pericardial mesothelioma exists. However, prompt surgical intervention followed by postoperative adjuvant chemotherapy may help prolong patient survival.[1,2,12]

Tumor stage, pathologic findings, and genetic factors all influence prognosis. Approximately 60% of patients die within 6 months of diagnosis due to complications such as heart failure, pericardial constriction, and sudden tumor invasion, causing coronary thrombosis and myocardial infarction. The prognosis for this disease is poor, with median survival for patients with pericardial mesothelioma being approximately 6 months.[13]

Conclusions

PMPMs are rare, sporadic tumors. They should be considered and treated appropriately in cases of recurrent pericardial effusion or constriction, especially in patients unresponsive to pericardiocentesis or with unexplained superior vena cava thrombosis. Combining clinical data with imaging findings can provide diagnostic insight. This case report emphasizes the limited treatment options and poor prognosis associated with PMPM.

Declarations:

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Conflict of Interest

The authors report no conflict of interest.

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