

Cardiac Tamponade as First Manifestation of Multiple Myeloma: a Case Report With Literature Review

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Abstract

Background: Multiple myeloma is a hematologic malignancy characterized by clonal proliferation of plasma cells, mainly in bone marrow. Extramedullary disease is reported in many cases and may occur at diagnosis, at progression, or during relapse phase. Pericardial involvement is a rare condition that usually occurs with advanced-stage disease. We report a rare case of plasma cell-based pericardial effusion with cardiac tamponade as a form of presentation of multiple myeloma.

Case presentation: A 76-year-old woman was admitted to the emergency department for worsening shortness of breath, on segmental examination, she had dyspnea, engorged jugulars, muffled heart sounds and lower limb edema. A Chest X-ray demonstrated cardiomegaly with a mild left pleural effusion, laboratory data showed immunoglobulin G lambda, diagnosis of plasma cell-based pericardial effusion was established by pericardial fluid cytology. The patient received systemic chemotherapy, according to Melphalan-Prednisone and thalidomid protocol.

Conclusion: Multiple myeloma with cardiac effusion should be considered for differential diagnosis of patients with recurrent and unexplained pericardial effusions.

Background

Multiple myeloma (MM) is a malignant plasma cell dyscrasia, characterized by clonal proliferation of plasma cells in the bone marrow and other organs. It is the second most frequent hematologic disease that accounts for 10% of all hematologic malignancies and primarily affects patients over 65 years of age [1].

Extramedullary disease in MM occurs in 6 to 20% and it is characterised by a poor prognosis and drug resistance [2]. It has various clinical manifestations and different sites of organ involvement. Pericardial involvement is a rare complication caused by amyloidosis, infections, bleeding dyscrasia or plasma cells infiltration. It usually occurs later during the disease course. It has been described in only a few single case reports as an initial presentation of MM [2–4].

We report a rare case of MM with pericardial effusion associated with cardiac tamponade as a form of presentation of the disease treated by systemic chemotherapy according to MPT regimen.

Case Presentation

A 76-year-old woman, with no significant past medical history and no history of familial neoplasia, was admitted to the emergency department on December 2019 for worsening shortness of breath, abdominal distension and lower limb swelling which has been evolving for 18 days.

The patient denied having chest pain, fever, syncope or bone pain. On the initial physical examination, she was afebrile and tachycardic at 101 beats per minute. Her blood pressure was at 92/60mmhg. On

segmental examination, she had exertional dyspnea, engorged jugulars, muffled heart sounds and lower limb edema without hepatosplenomegaly or lymphadenopathy.

A Chest X-ray and thoracic tomodensitometry demonstrated cardiomegaly with a mild left pleural effusion (Fig. 1), but no abnormalities in the lung fields. An electrocardiogram (ECG) showed a low QRS voltage in the precordial leads. Bedside ultrasound revealed a large circumferential pericardial effusion of 27 mm in diastole and diastolic collapse of the right ventricle and inferior vena cava. Pericardiocentesis was performed without delay and 800 ml of serosanguinous fluid was drained. Pericardial fluid chemistry revealed a sterile exudate effusion and bacterial and acid fast cultures were negative.

Laboratory data showed white blood cells of 4.4 G/L without plasma cells, platelets 85 G/L and haemoglobin 13.5 g/dl. Serum protein electrophoresis showed a monoclonal spike 54 g/L.

The patient was readmitted on January 2020 with clinical signs of a recurrent cardiac tamponade, and consequently was treated with surgical pericardial window. A pericardial biopsy was performed while pericardial window was created.

Cytological examination of the pericardial fluid revealed plasma cells (Fig. 2,3), without pericardial infiltration by mature plasma cells on histological examination of pericardium.

Bone marrow aspirate and biopsy (Fig. 4) examination revealed a hypercellular marrow with 19% of mature plasma cells. Serum total protein was 67 g/L and erythrocyte sedimentation rate was 85. Serum creatinine level was 7 mg/dl, serum calcemia 95mg/L, LDH 268 mg/L, B2M 6.2 mg/L. Serum immunoelectrophoresis showed immunoglobulin G lambda and urine was negative. Ratio Free light chain (FLC) was 181. Initial skeletal radiological survey revealed two osteocondensing lesions in the skull 2 and 7 mm. karyotype was not done. A diagnosis of multiple myeloma was established with ISS (international scoring system) stage III. The patient was treated according to MPT regimen (Melphalan, prednisone and thalidomide). After the fourth cycle of MPT, she had a FLC ratio at 24. After the sixth cycle, she presented to the emergency department with oliguria and acute renal failure requiring urgent dialysis. Her serum calcemia at was 116mg/L. She died from hyperkalaemia-induced cardiac arrhythmia few hours later.

Discussion

Extramedullary disease in MM is a recognised manifestation of MM characterised by involvement of different anatomical sites distant from the bone marrow including lymph nodes, skin, breast, liver, central nervous system [5–13]. Pericardial involvement is particularly rare with myelomatous pericardial effusions present in less than 1% of all cases of MM [3]. The first case reported in literature was in 1949 by Chorg [4]. In our case, no symptoms of MM were found. The monoclonal spike discovered in the card of the etiological assessment of tamponade, which directed us to the search for multiple myeloma.

Ig A type MM appears to have more high-risk of extramedullary disease in general and pericardial involvement in particular [14]. Cardiac tamponade secondary to myelomatous effusion of the

pericardium is exceptional with only 29 cases including ours, reported in the literature and most cases are diagnosed retrospectively at postmortem examination, it is usually the result of metastatic or relapsed myeloma [3, 15–20]. As myelomatous effusion is associated with poor prognosis with an average survival of 13.5 weeks [3], an efficacy, less invasive procedure that can establish an accurate diagnosis with a minimum risk and discomfort is highly desirable. Positron emission tomography with 18F-fluorodeoxyglucose (18F-FDG) and computed tomography (PET-CT) are used in valid scores established in differentiating malignant pleural effusion from benign effusions [21, 22]. However, the literature on the use of PET/CT in pericardial disease remains very limited, and this area warrants further investigation [22, 23].

No current consensus exists on the management of pericardial involvement in MM. Drainage is required for initial management of symptomatic effusions and can be completed by pericardiocentesis or pleuropericardial window. Beyond pericardiocentesis, intrapericardial therapeutic administration of cytotoxic agents (bleomycin or cyclophosphamide), systemic chemotherapy (corticosteroid, vincristin, adriamycin, cyclophosphamide, imid, bortezomib) and radiation therapy have all been used but with little therapeutic benefits, no case of hematopoietic stem cell transplant was published [17–20].

Our patient was given MPT regimen and her results after induction chemotherapy were promising (ratio FLC: 86%, no recurrence of pericardial effusion). However, her myelomatous pericardial effusion, her renal failure and hypercalcemia causing her death despite her ongoing treatment reflect the highly aggressive disease presentation at both the time of diagnosis and progression.

The present case is unique as the plasma cell-based pericardial effusion leading to cardiac tamponade is the initial presentation of MM in a patient with no signs or symptoms of myeloma.

It highlights the need of hematologists to be aware of such a possibility and investigations on pericardial fluids are necessary despite the absence of symptoms of myeloma.

Conclusion

Pericardial effusion in multiple myeloma is rare; it can be present at any time of diagnosis. It should be considered for differential diagnosis of patients with recurrent and unexplained pericardial effusions.

Abbreviations

MM: Multiple myeloma

MPT: Melphalan, prednisone and thalidomid

LDH: lactate dehydrogenase

B2M: beta 2 microglobulin

FLC: free light chain

Ig A: immunoglobulin A

Declarations

- Ethical approval and consent to participate: Not applicable
- Consent for Publication: Written informed consent was obtained from the patient's next of kin for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal
- Availability of data and material: yes
- Competing interests: No
- Funding: No
- Authors Contribution:

ML: editing

KK: editing

FL: editing and correction

IT: Chef, correction

- All authors have read and approved the manuscript
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Figures

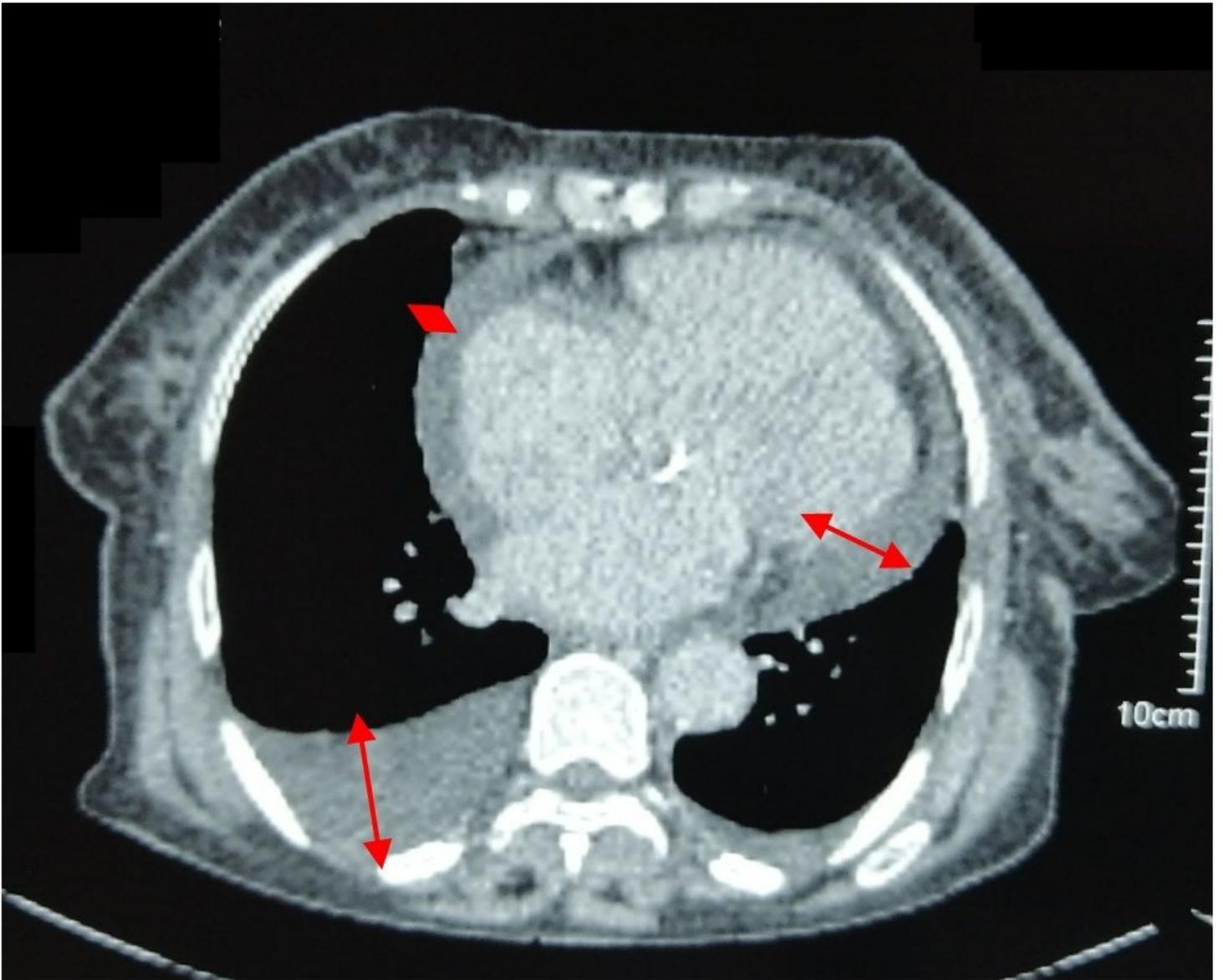


Figure 1

Thoracic tomodensitometry shows a cardiomegaly, left pleural effusion and cardiac effusion

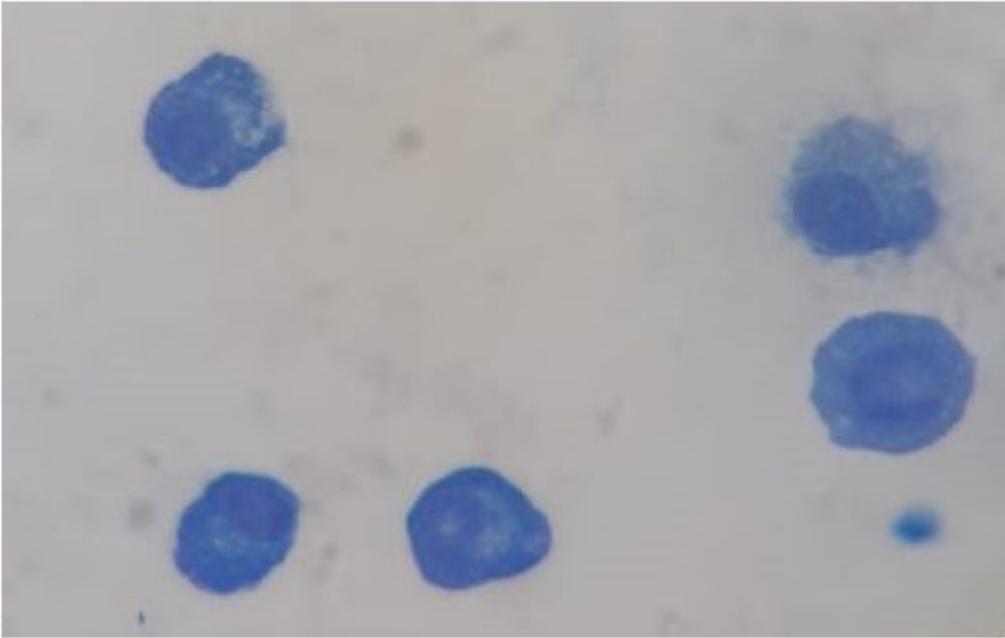


Figure 2

plasma cells

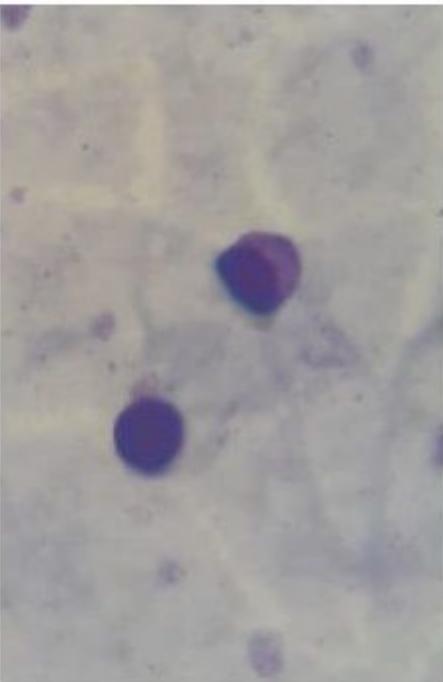


Figure 3

plasma cells

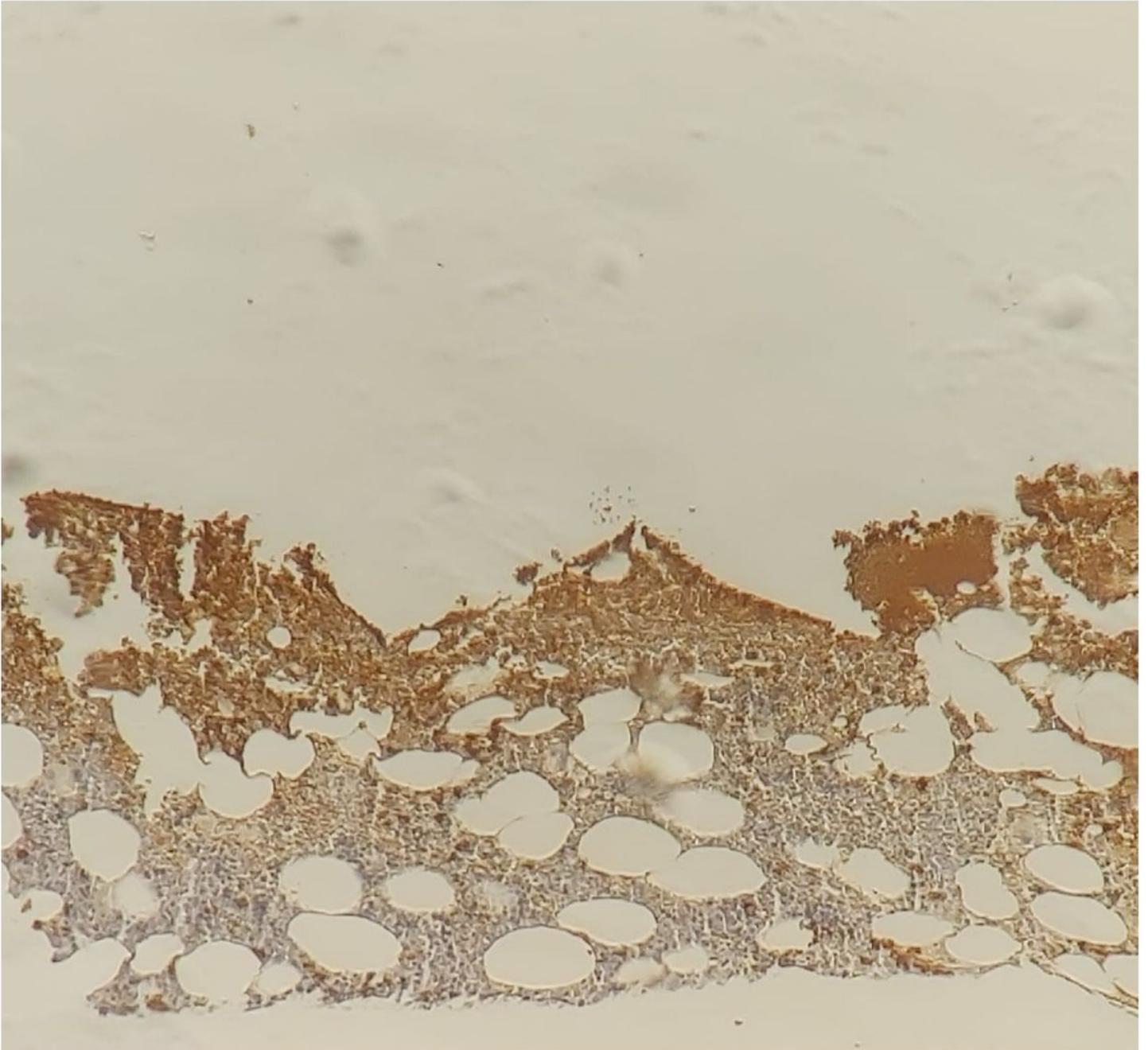


Figure 4

cytoplasmic and membrane expression of the anti lambda antibody

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