

Pericardial effusion as an initial manifestation of multiple myeloma: A case report and literature review

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Abstract: Multiple myeloma, or Kahler's disease, is a neoplastic plasma cell disorder characterized by clonal proliferation of malignant plasma cells in the bone marrow. Although rare, it is the second most common hematologic malignancy. Multiple myeloma is associated with significant morbidity due to its end-organ destruction. Its clinical features are polymorphic and can be osteoarticular, neurological, hematological or visceral. However, cardiac involvement is uncommon and pericardial effusion secondary to multiple myeloma is very rare. Pericardial infiltration can be due to infection, amyloid deposits or plasmacytic infiltration and occurs at a late stage in the course of the disease. The majority of these patients are asymptomatic and pericardial tamponade is rare. The use of surgical drainage with intrapericardial injection of bleomycin has been reported in some cases, but the favorable response to systemic chemotherapy has been reported in only one case to date. Our case describes an unusual presentation of multiple myeloma, revealed by an isolated pericardial effusion.

Keywords: Multiple Myeloma, pericardial Tamponade, oligoclonal profile

INTRODUCTION

Multiple myeloma (MM) is a rare type of malignant hematological neoplasm, characterized by malignant clonal proliferation of plasma cells. It is the second most common blood cancer and represents 1% of cancers and 10% of hematologic malignancy. MM is associated with significant morbidity due to its end-organ destruction [1]. It affects men more than women. The average age at diagnosis is 60 years and can present with various clinical symptoms; it's most often discovered on the occasion of bone, hematological or renal manifestations [2-4]. Serosal infiltration is unusual and affects the pleural and peritoneal space [5, 6]. Cardiac and pericardial involvement is rare, usually observed at a late stage of disease; it's associated with a poor prognosis and an unsatisfactory response to chemotherapy [7]. Our case report is characterized by a first unusual presentation: isolated tamponade.

CASE REPORT

A 63-year-old patient, with no past medical history, presented at the emergency department with dyspnea stage IV of NYHA, progressing for 15 days, and chest pain. Initial vital signs were as follows: blood pressure 107/61 mmHg, heart rate 110 bpm, respiration rate 20/min, temperature 37.5°C, O₂ saturation 95%. On physical examination the patient presents signs of right heart failure: spontaneous turgor of the jugular veins, abdomino-jugular reflux and bilateral pitting edema of

the legs. Cardiac auscultation revealed muffled heart sounds.

Investigations:

1. Chest x-ray: reveals a generalized cardiac enlargement with a water-bottle shape (Fig 1)
2. Electrocardiogram shows sinus tachycardia with low QRS voltage.
3. Transthoracic echocardiography revealed a circumferential pericardial effusion of great abundance, with significant respiratory variations, notch of the right atrium, diastolic collapse of the right ventricle and the inferior vena cava (IVC) is distended with loss of normal respiratory variation (Fig 2a, 2b).

The patient was admitted to the Cardiovascular Intensive Care Unit. A pericardiocentesis was then performed, bringing 900cc of sero-hematic fluid. The chemical and cellular study revealed an exudative liquid without plasma cells, bacterial culture was negative. A biological check-up was also carried out: Complete Blood Count showed a normocytic normochromic anemia (hemoglobin at 08 g/dl), erythrocyte sedimentation rate (ESR) was accelerated at 116 mm at the first hour, LDH was elevated, renal function, total plasma calcium and thyroid function were normal, viral serologies were negative.

Transthoracique echocardiography showed a brilliant speckled appearance of the myocardium without signs of restrictive cardiomyopathy, regression of the pericardial effusion (7 mm in anterior and 10 mm in posterior pericardial sac), without significant respiratory variations nor collapse of the right cavities, IVC was compliant and not dilated. In view of this clinical picture (age, sex, anemia and accelerated ESR) and the absence of obvious etiology, we supplemented by a serum protein electrophoresis which revealed an oligoclonal profile of immunoglobulins at 31.1 g/L (Fig 3). Immuno phenotyping of plasma cells confirmed the presence of monoclonal immunoglobulins G Kappa (Fig 4). The myelogram showed medullary plasmacytosis up to 35% with dystrophic plasma cells (Fig 5). The radiological assessment was supplemented by standard radiographs of the skull, rachis and pelvis, which didn't show abnormalities.



Fig 2b: Transthoracic echocardiography, apical four-chamber view: circumferential pericardial effusion of great abundance



Fig1: Generalized cardiac enlargement with a water-bottle shape.

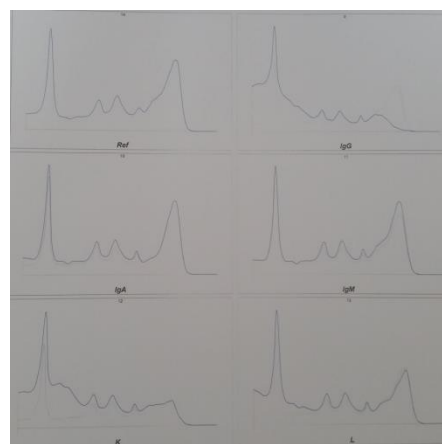


Fig 3: Oligoclonal profile of immunoglobulins at 31,1g/l g/L.

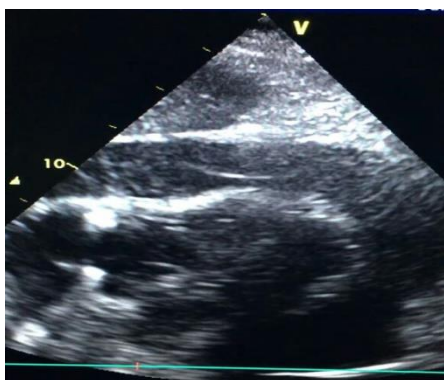


Fig 2a: Transthoracic echocardiography, subcostal view: pericardial effusion of great abundance

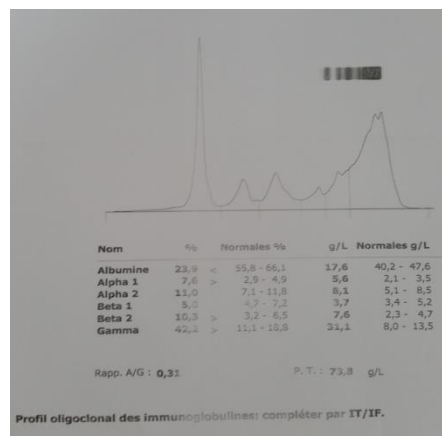


Fig 4: Monoclonal Gammopathy Ig G Kappa.

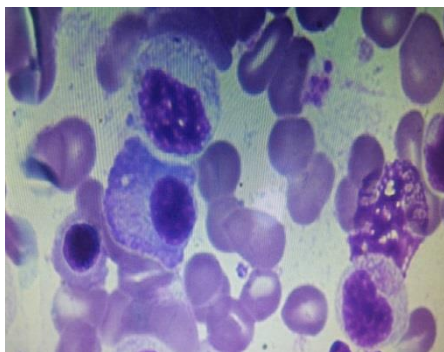


Fig 5: Plasmacytosis with dystrophic plasma cells.

DISCUSSION

Multiple myeloma or Kaler's disease is characterized by a proliferation of malignant plasma cells and usually presents with bone pain and organic damage. The most frequent presentation is anemia, hypercalcaemia, renal insufficiency and bone destruction [8, 9]. The immunoglobulins (Ig) involved are, in descending order of frequency: IgG, IgA and IgD. The type of immunoglobulin determines the presentation of the disease. IgM and IgG are more commonly associated with vascular symptoms and hyper viscosity syndromes [10, 11]. IgM is rarely associated with kidney damage; filtration in tubules is limited due to its large size. IgD is associated with extra-bone involvement, hepatosplenomegaly, lymphadenopathy and participation of serous membranes [10].

Our patient has a predominant Ig G Kappa band without extraosseous or renal involvement. Extraosseous localization usually involves the skin, soft tissues and liver. Extramedullary myeloma has been described; however, direct damage to the heart is extremely rare [12]. Cases of myeloma recurrences in the form of intracardiac or pericardial plasmacytomas were reported [13-14].

Although extramedullary involvement is rare, its incidence appears to be increasing [2, 15]. Its presence at the time of initial diagnosis or at the time of relapse is associated with a poor prognosis [2]. Recent evidence suggests that extramedullary myeloma occurs more frequently in high-risk patients, defined by cytogenetic and molecular markers, and has a poor response to treatment [15, 16]. Serous cavity invasion has ominous prognosis, with survival rate of less than 4 months as they have poor response to chemotherapy, shorter progression free interval and overall survival. Pericardial effusion in multiple myeloma may be secondary to cardiac heart failure due to amyloidosis or restrictive cardiomyopathy, infectious etiology and rarely plasma infiltration.

The largest study reviewed 869 patients with extramedullary presentation of multiple myeloma and found no patients with pericardial disease [3, 17]. Another study on 38 patients showed no pericardial

spread on autopsy [5, 18]. On further review of literature, 12 case reported pericardial effusion, out of which 5 cases had underlying heart failure secondary to amyloidosis, and the remaining 7 had isolated pericardial infiltration, while only 3 had bilateral pleural and pericardial spread [5]. Our case report is distinct as pericardial effusion with cardiac tamponade is rarely the initial presentation of multiple myeloma, especially in absence of typical presenting symptoms such as hypercalcemia and renal failure.

CONCLUSION

Diagnosis of multiple myeloma is possible even in the absence of bone damage, hematologic or renal complications. Pericardial involvement is getting more and more frequent. Extra-medullary disease is rare but unfortunately associated to sinister prognosis.

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