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Case Report

Sternal Deformity Revealing a Multiple Myeloma in a Young Subject: A Case Report

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Abstract

Multiple myeloma (MM) or Kahler's disease is a malignant disorder of plasma cells which represents about 10% of hematological malignancies. The risk of developing a MM increases with age, peaking around the age of 65 years old, so it is rather rare among younger patients. We report the case of a 36-year-old man who presented with a 3 months history of dry cough, two episodes of low abundance haemoptysis, a stage IV dyspnea and bilateral chest pain. The patient also complained of lumbar sciatica and a 10kg weight loss in 3 months. The physical examination found rhonchus and crackling on pulmonary auscultation, a sternal depression and a painful palpation of the dorsal spine. The chest X-ray and the thoracic Computed Tomography (CT) showed an osteolytic mass of the sternum and diffuse LODWICK type IB and IC bone lesions of the dorsal spine and ribs with vertebral compression. Laboratory tests showed an inflammatory syndrome with hypergammaglobulinemia at 63,5 g/l (monoclonal) and the serum protein immunofixation found a type kappa monoclonal immunoglobulin IgG. The myelogram showed a rich marrow with 40% plasma cells and the presence of rare dystrophic plasma cells. The patient received chemotherapy with a good follow up.

Keywords: *Multiple Myeloma; Sternal Deformity; Young Subject*

Introduction

Multiple myeloma is a malignant lymphoid hematopathy, of unknown cause, defined by the combination of a proliferation of tumorous plasma cells and the secretion of monoclonal immunoglobulin. The disease can cause lytic bone complications, hypercalcemia, anemia, renal failure, repeated infections and its prognosis remains bad. It usually occurs in people aged between 60 and 70, with an average survival of 3 to 4 years. Less than 2% of myeloma patients are under the age of 40.

Patient and Observation

The patient was a 36-year-old man; non-smoker and worked as a bricklayer. He was exposed to cement dust, asbestos and passive smoking but had no medical history. The patient presented with a 3 months history of dry cough with 2 episodes of low abundance hemoptysis and exertion dyspnea. He also reported a bilateral chest pain associated to lumbar sciatica, anorexia, asthenia and a weight loss of 10 kg in 3 months. The clinical examination found rhonchus and crackling on pulmonary auscultation as well as a sternal depression and an intense pain on the palpation of the thorny apophyses of the dorsal spine. The chest X-ray showed enlarged upper and middle mediastina with a bayonet deformity of the trachea, diffuse reticular and micronodular opacities and a partial atelectasis of the right lung. The bone abnormalities consisted of diffuse lacunar lesions of the dorsal vertebrae while the skull X-ray showed geodes (Figure 1-3).

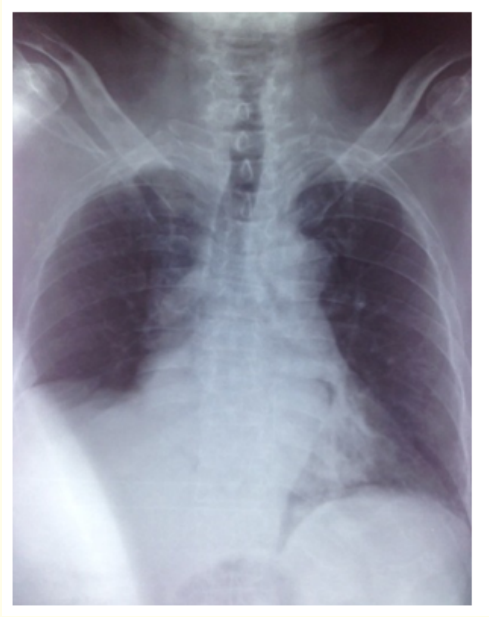

Figure 1: *Admission chest X-ray showing a Mediastinal enlargement.*

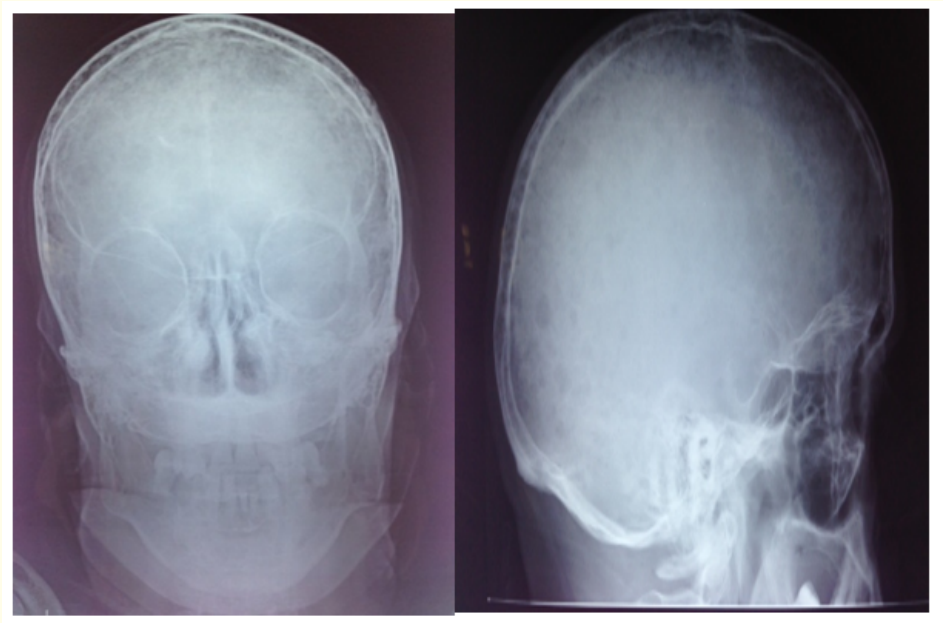

Figure 2: *Lacunar images on the skull radiography.*

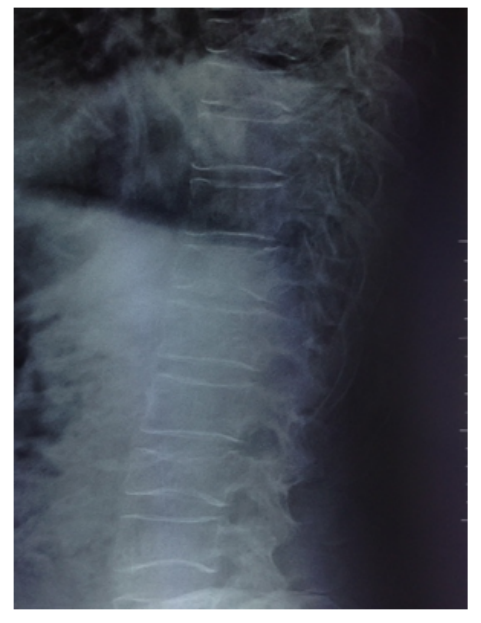

Figure 3: *Lytic images and vertebral compression.*

A Computed Tomography scan was done in order to better explore this mediastinal enlargement. It showed an enlarged sternum with lytic masses in the manubrium and the sternal body with a ruptured cortex. It also showed a heterogeneous appearance of the bone matrix with multiple diffuse lacunar LODWICK type IB and IC lesions of the dorsal vertebrae, the ribs and the shoulder blades with flattened and biconcave vertebral bodies T4, T5, T8, T9 and T1 (Figure 4). The CT aspect concluded to a chest localization of a multiple myeloma. A bronchoscopy was performed showing an extrinsic compression of the entire bronchial tree with thickening of all spurs. The results of the pathology study of the bronchial biopsies and the bronchoalveolar lavage were not conclusive.

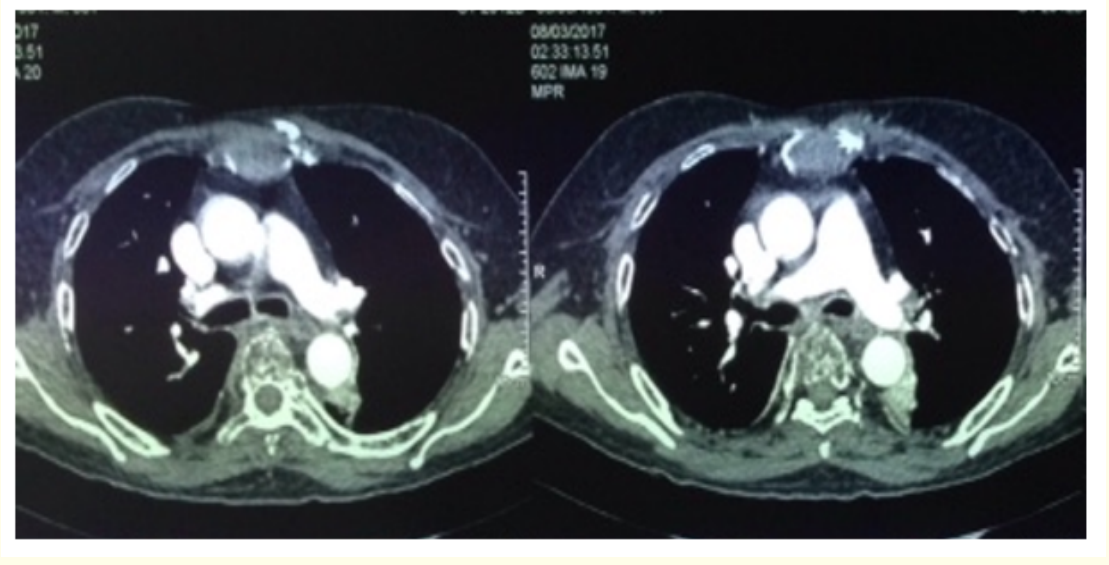

Figure 4: *Lytic sternal mass on the thoracic computed tomography.*

A CT scan guided biopsy of the sternal mass was done without incidents. Its anatomopathological examination showed a tumorous proliferation made of round plasmacytoid cells. The immunohistochemistry revealed a proliferation of lambda monoclonal plasma cells which confirmed a myeloma.

Blood tests revealed an inflammatory syndrome and an elevation of the lactate Dehydrogenase (LDH) at 396, a normal calcium level and a normal renal function. The protein electrophoresis found a hypergammaglobulinemia at 63.5g/l with a monoclonal peak and the protein immunofixation showed a monoclonal immunoglobulin of IgG Kappa type. The myelogram revealed a rich bone marrow with 40% plasma cells and the presence of rare dystrophic plasma cells. The patient received chemotherapy with good immediate follow-up.

Discussion

Multiple myeloma (MM) represents about 1% of all neoplasms in the Caucasian population, and 2% in the black one [1]. In Europe and the United States, the annual incidence of MM is about 4 per 100,000 people [1]. This incidence appears to be slightly higher in men than in women [1]. MM has a strong correlation with age and the average age of occurrence is over 60 years old. In the United States, the average age of onset of this disease is 68 for men and 70 for women [2], our patient is 36 years old. Multiple myeloma is rare among people under the age of 40, it represents about 2% of all myeloma cases and it is even rarer in patients under 30 years old [2]. A study by Bladé., *et al*. conducted on 3278 patients by the Mayo clinic in the USA, showed that the frequency of MM in patients under the ages of 40 and 30 is 2.2% and 0.3% respectively [1]. The study conducted by the National Cancer Institute (NCI) showed that out of 3815 myeloma patients, only 7 cases are less than 30 years old and therefore have a frequency of 0.18% [3]. Other studies have focused on MM in young subjects such as the one of Hewell and colleagues, which found a frequency of 1% in subjects under the age of 30 and it is even lower in those under 20 years old. Clinical and biological characteristics seem to be similar to those observed in older patients. In our reported case, MM was revealed by a sternal deformity with both dorsal and lumbar sciatalgia with a poor general condition. These are almost the same revealing symptoms of MM in the young subject reported in the literature. In the study by Usha and colleagues, of 14 cases of MM under the age of 40, 60% had dorsal and lumbar sciatalgia as well as asthenia. In the same study, the immunoelectrophoresis found IgG in 10 out of 14 cases, which is the case for our patient. Some studies have shown a higher proportion of MM with light Ig chains and IgD [4]. Regarding Bence Jones proteinuria (BJP), it has rarely been reported in the literature in young subjects, except in the study by Blade and al, which found BJP in 5 out of 10 patients [5], it was absent in our patient's case. The study conducted in the Mayo clinic revealed renal failure and hypercalcemia in 29% and 30% of the 72 patients respectively. Other more recent studies did not find any cases of renal failure, hypercalcemia or any other electrolyte disorder, which is consistent with the reported case [6]. On the imagery, our patient had vertebral fractures and multiple geodes of the skull, which is consistent with the literature reporting the frequency of lytic bone lesions in very young patients, particularly those under the age of 30. The study by Usha and colleagues showed lytic lesions of the bone in almost all cases with femoral and vertebral fractures in 28.5% of cases [7], extramedullary extension is also frequent. The study by Blade., *et al.* [1] found an extramedullary component in 14 out of 17 patients (82%) while in another study, it was found in three out of five patients (60%) [8]. Extramedullary extension was not observed in our case. The treatment of young patients with MM is no different from the one of elderly patients. Radiotherapy (RT) and chemotherapy are both indicated but it is clear that RT should be used with caution in very young subjects. Young patients with MM could benefit from intensive chemotherapy followed by stem cell transplantation. Survival is longer than that of older patients, especially those with good prognostic factors (normal renal function or low beta 2-microglobulin levels) and also those under 30 years of age. The average survival duration for patients with MM is between 2 and 3 years. In the study of Bladé and colleagues conducted on young subjects with MM, it was longer, around 87 months (more than 7 years) [2]. Our patient received chemotherapy cures with good 3 months follow-up after the start of treatment.

Conclusion

The average age of onset of MM is over 60 years old, rarely before the age of 40 and it never occurs among children. The clinical, biological, radiological characteristics and evolution of this disease do not differ significantly between the young subject and the elderly one, although some reviews of the literature have reported a more pronounced bone involvement.

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