

Extramedullary Multiple Myeloma in the Kidney: A Case Report

Sinan Demircioğlu^{1*} and Pembe Oltulu²

¹Department of Hematology, Van Yüzüncü Yıl University, Turkey

²Department of Pathology, Necmettin Erbakan University, Turkey

***Corresponding author:** Sinan Demircioğlu, Faculty of Medicine, Department of Hematology, Van Yüzüncü Yıl University, Turkey, Tel: 5554324474; Email: sinandemircioglu@gmail.com

Case Report

Volume 2 Issue 2

Received Date: July 26, 2018

Published Date: August 13, 2018

DOI: 10.23880/hij-16000128

Abstract

Extramedullary myeloma (EMM) is defined by the presence of plasma cells outside the bone marrow in a patient with multiple myeloma (MM). EMM is classified under four headings as bone-associated plasmacytoma, extramedullary disease (EMD), plasma cell leukemia and solitary plasmacytoma. EMD is more common in the nasopharynx, larynx, and upper respiratory tract. It is rarely seen in the kidney. We presented extramedullary disease of kidney, at relapsed myeloma patient.

Keywords: Multiple Myeloma; Extramedullary Myeloma; Extramedullary disease

Abbreviations: MM: Multiple Myeloma; EMM: Extramedullary Myeloma; ED: Extramedullary Disease; VAD: Vincristine-Adriamycin-Dexamethasone.

Introduction

Multiple myeloma (MM) is characterized by the proliferation of uncontrolled proliferating plasma cells, usually limited to the bone marrow. The extramedullary spread of MM may occur either during diagnosis or during the course of disease [1,2]. Extramedullary myeloma (EMM) has been recently classified by being divided into four groups. There are 1-Bone-associated plasmacytoma, 2- Extramedullary disease (EMD) with a hematogenous spread that occurs in the tissues remote from the bone marrow, 3- Plasma cell leukemia, 4- Solitary plasmacytoma [3-6]. It is necessary to demonstrate that the lesion is not associated with solitary plasmacytoma and bone to describe extramedullary disease [4].

Extramedullary disease and plasma cell leukemia have a worse course than the other 2 EMM subtypes. The presence of EMD in recurrence is associated with a survival of <6 months. This negative prognosis is less apparent in bone-associated EMM [7]. Prognosis is quite good after the treatment of solitary plasmacytoma with radiotherapy [8]. EMM is observed by around 6-8% during the first diagnosis of myeloma [9,10]. The incidence of EMM increases in the course of the disease, and this ratio increases to 10-30% [3,11].

Case Presentation

A 48-year-old male patient was diagnosed with multiple myeloma in 2005. He entered into remission after administering four cycles of VAD (vincristine-adriamycin-dexamethasone) chemotherapy, and radiotherapy to the thoracic region. He was followed up without medication since he did not accept the autologous

stem cell transplantation. During the ultrasonography performed due to abdominal pain while being followed up in remission for ten years, a mass was detected in the left kidney. Dynamic renal tomography was performed. The lesion which was approximately 52 mm in diameter in the lower middle part of the left kidney and observed to be slightly hyperdense from kidney parenchyma, contrast-enhancing in the arterial phase, and wash out in the late phase in the portal phase in unenhanced images was radiologically interpreted as renal cell carcinoma (Figure 1). Then, left radical nephrectomy was performed. The surgical pathology was evaluated as a kappa-positive plasmacytoma. It was determined that the leukocyte count was $5800/\text{mm}^3$, hemoglobin value was 11.6 g/dl , thrombocyte count was $263.000/\text{mm}^3$, immunoglobulins compressive, free kappa/lambda ratio was 5, and calcium and creatinine values were within normal limits. Monoclonal kappa light chain staining was observed in serum urine immunofixation. On PET CT, common lytic lesions were detected in the bone structure. Monoclonal plasma cell increase with kappa positive, lambda negative staining pattern was observed in bone marrow biopsy (Figure 2). Bortezomib, cyclophosphamide, dexamethasone chemotherapy, and zoledronic acid were initiated for the patient who was considered symptomatic MM.

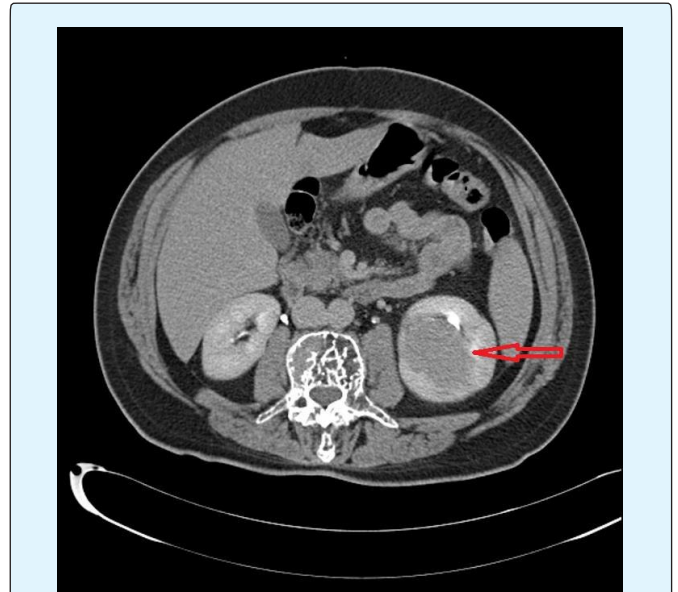


Figure 1: Lesion 52 mm in diameter which was observed to be slightly hyperdense from kidney parenchyma, contrast-enhancing in the arterial phase, and wash out in the late phase in the portal phase in unenhanced images.

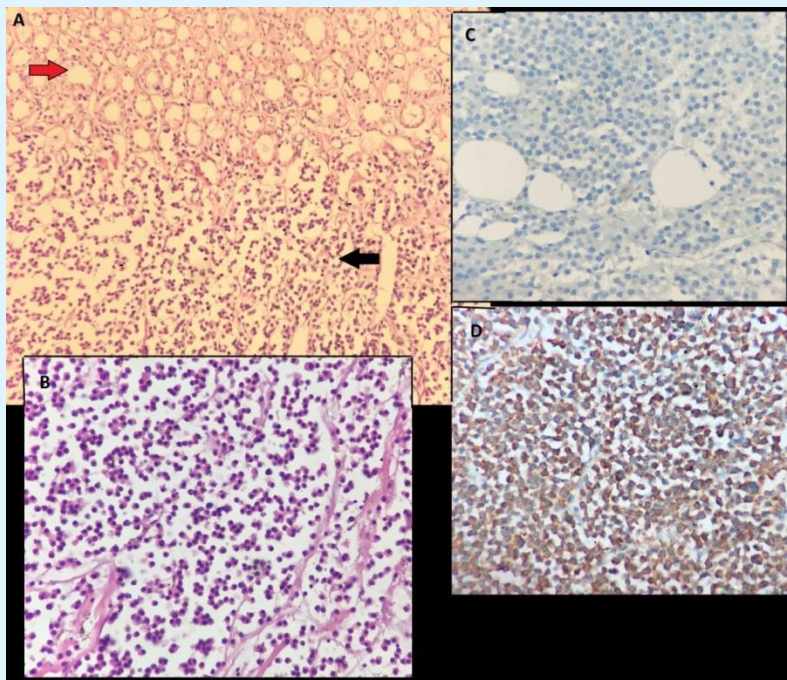


Figure 2: A) X100, H & E, atypical plasmocytic infiltration (red arrow) in kidney parenchyma (red arrow) B) X400, H & E, atypical plasmocytes C) X400, Lambda, Lambda negativity and D) X400, Kappa, kappa positivity.

Discussion and Conclusion

Although renal failure is observed in about half of the patients with multiple myeloma, extramedullary disease of the kidney is rarely observed [12]. EMD is more common in the nasopharynx, larynx, and upper respiratory tract [13]. Furthermore, it may also be observed in the gastrointestinal system, pleura, testis, skin, peritoneum, liver, brain, endocrine glands and lymph nodes [14-21]. In the literature, plasmocytoma in the kidney has been reported less than 30 cases without multiple myeloma [22].

In the study carried out by Weinstock, et al. on 663 MM patients, the researchers detected EMD in 55 (8.3%) patients. While it was detected in the head and neck region (31.6%), abdomen (26.3%), chest (21.1%) and central nervous system (12%) during diagnosis, it was observed to be common in the abdomen (40%) and chest (23,9%) in recurrent disease. In abdominal involvement, it was common in pancreas, peritoneum, kidney, and intestines during diagnosis. In recurrent patients, liver involvement was most common in the abdomen [23]. In another study, EMD was detected in 42 (9%) of 467 patients. While EMD was most commonly observed in the soft tissue and pleura/peritoneum, kidney involvement was not encountered in this study. In this study, it was shown that the cumulative incidence of EMD in MM was 9%, and that treatment responses were poor in the presence of EMD. Furthermore, it was also shown that the survival of patients with simultaneous EMD during diagnosis was even worse [24].

In conclusion, along with the available information, EMD may occur in many different tissues as in our case. Although MM-associated renal failure is common, it is rarely seen in the kidneys in plasmacytoma. Plasmocytomas can be detected with imaging methods in MM patients.

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