

CASE REPORT

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Relapse of multiple myeloma presenting as extramedullary plasmacytoma surrounding the aorta: A rare case report**Ömer Ekinci¹, Ali Dogan², Mehmet Aslan¹, Senar Ebinc², Cengiz Demir²**¹*Firat University, Faculty of Medicine, Department of Hematology, Elazig, Turkey*²*Yuzuncu Yil University, Faculty of Medicine, Department of Hematology, Van, Turkey*

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Abstract

Multiple myeloma (MM) is a hematological disease characterized by the malignant proliferation of plasma cells. MM can be concomitant with plasmacytoma, at diagnosis or during relapse. Extramedullary plasmacytoma (EMP) is rare and is encountered most frequently in the upper respiratory tract and nasopharynx. It is of much less frequent occurrence in the intraabdominal and thoracic regions. EMPs tend to have a poor prognosis with a characteristic of high relapse/refractory disease rates and a relatively short overall survival, despite the use of various novel medications. Here, we present a case of relapse of MM concomitant with a large EMP surrounding the aorta, which is an extremely rare pattern of involvement. Our case showed nearly complete remission with an aggressive chemotherapy regimen. MR imaging served as a guidepost in both the diagnosis and the post-treatment follow-up.

Keywords: Extramedullary plasmacytoma, multiple myeloma, plasmacytoma, aorta**Introduction**

Multiple myeloma (MM) is a hematological disease characterized by the malignant proliferation of plasma cells. MM usually presents with renal failure, anemia, hypercalcemia, lytic bone lesions, secondary immunodeficiency and fractures [1]. Plasmacytoma is defined as the massive infiltration of bone or soft tissues (extramedullary) by neoplastic plasma cells [2]. Extramedullary plasmacytoma (EMP) defines soft tissue tumors that are characterized by plasma cell infiltration and develop secondary to hematogenous spread, in an anatomical site distant from the bone marrow (usually liver, skin, central nervous system, pleura, kidneys, lymph nodes and pancreas) [3,4]. The prevalence of EMP in MM patients is approximately 6-8% at diagnosis, and approaches 10-30% during the course of the disease [5,6]. EMPs tend to have a poor prognosis with a characteristic of high relapse/refractory disease rates and a relatively short overall survival, despite the use of various novel medications [7]. Here, we present a case of relapsed MM concomitant with a large EMP surrounding the aorta, which is an extremely rare pattern of involvement.

Case Report

A 66-year-old male patient presented to our clinic with back pain and weakness in the legs. The patient had been diagnosed with IgG kappa multiple myeloma six years ago. In the initial diagnosis, he had been evaluated as an international staging system (ISS) stage-II, transplant eligible based on clinical and laboratory findings. He had received monthly zoledronic acid, two courses of vincristine, adriamycin, and dexamethasone (VAD) and two courses of bortezomib and dexamethasone (VD) regimens. Subsequent to complete response, he had undergone autologous hematopoietic stem cell transplantation (aHSCT) with high-dose melphalan for the purpose of consolidation. The patient had achieved complete remission under follow-up after aHSCT. The disease had relapsed approximately 4 years after the first aHSCT, and the patient had undergone another aHSCT with high-dose chemotherapy after a bortezomib, cyclophosphamide, and dexamethasone (VCD) chemotherapy regimen, and had been in complete remission under follow-up.

He presented with the complaints stated above 18 months after the second transplantation. On physical examination, bilateral lower extremities showed weakness and impaired sensation. Spinal vertebrae were examined with magnetic resonance imaging (MRI) in consideration of the history of MM. On MRI examination, there were diffuse lytic lesions involving all spinal segments and the

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sternum, and a soft tissue lesion that involved the aorta-vascular structures in the retrocrural space at the level of T7-L1 and extended to the spinal canal and involved the spinal cord at the level of T8-10 (Figure 1). An imaging-guided tru-cut biopsy was taken from the mass and the diagnosis was confirmed as plasma cell myeloma based on histopathological and immunohistochemical findings.

Laboratory tests at relapse were as follows: hemoglobin, 9.1 gr/dl; creatinine, 0.66 mg/dl; calcium, 8.9 mg/dl; M-spike on serum protein electrophoresis, (10.8%); gamma globulin, 8.24 g/dL. Total serum IgG was determined as 6720 mg/dL, β 2 microglobulin as 2.3 mg/L, albumin as 3.6 g/dL and lactate dehydrogenase as 314

IU/L. Although the patient underwent 2 courses of lenalidomide and dexamethasone (Len-Dex), and subsequently, 2 courses of lenalidomide, bortezomib, and dexamethasone (VRD), there was no reduction in the size of the plasmacytoma and the patient was considered non-responsive. As a more aggressive regimen, a combination of bortezomib, dexamethasone, thalidomide, cisplatin, adriamycin, cyclophosphamide, and etoposide (VDT-PACE) was administered. A very good partial response (VGPR) was obtained after two courses. The patient was not suitable for allogeneic HSCT because of poor performance status. The patient and his relatives were consulted, and it was decided to continue the treatment with chemotherapy agents.

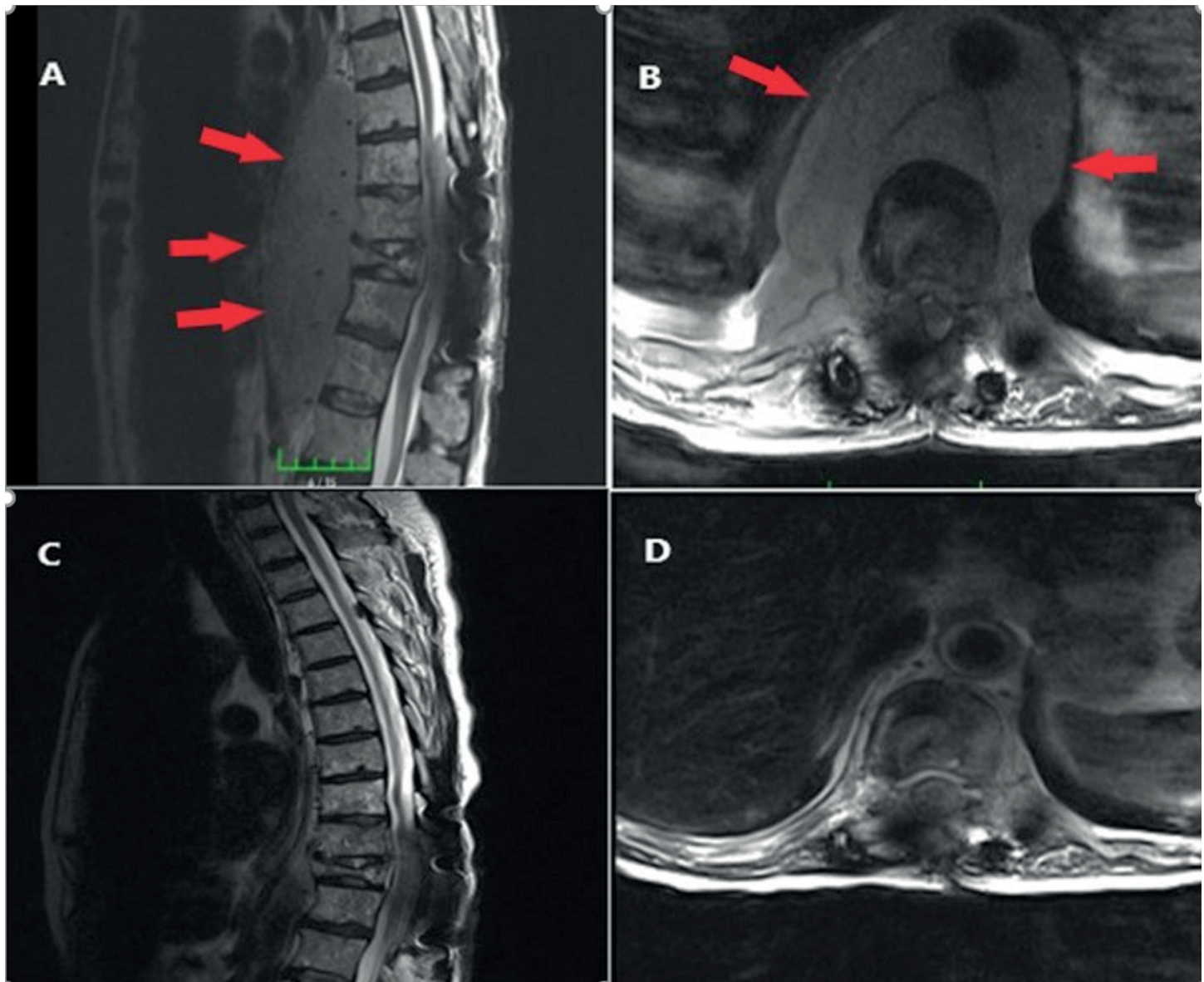


Figure 1. Magnetic resonance imaging showing lytic lesions involving all spinal vertebrae and a soft tissue lesion that involves the aorta-vascular structures in the retrocrural space at the level of T7-L1 and extends to the spinal canal and involves the spinal cord at the level of T8-10, on sagittal (A) and axial (B) planes. Nearly complete disappearance of the mass lesion in the same patient, on sagittal (C) and axial (D) planes.

Discussion

Plasma cell neoplasias include various disease groups such as multiple myeloma, plasma cell leukemia, solitary plasmacytoma of the bone, EMPs, Waldenstrom macroglobulinemia, primary

amyloidosis, light chain deposition disease and heavy chain disease [1]. EMP is a rare entity that is defined as the massive infiltration of plasma cells in organs or tissues other than the bone marrow, and comprises less than 10% of all cases [2,3]. EMPs are encountered most commonly in the upper respiratory tract

and nasopharynx [8]. Cases of EMP were also reported in the gastrointestinal tract, pleura, testis, skin, peritoneum, liver, brain, endocrine glands, kidney and lymph nodes [9-11].

EMP is being detected at higher rates, particularly due to the increase in the use of sensitive imaging methods such as PET-CT and MRI [12,13]. Our case also presented with a quite large EMP that involved the region surrounding the aorta, which is extremely rare. MR imaging provided useful data in both the diagnosis and post treatment of our case. Symptoms vary depending on the anatomical localizations of the masses or the dysfunctions that result from the direct mass effect or organ involvement. Our case also presented with neurological symptoms that appeared due to the mass that compressed the spinal cord and the surrounding nerves.

The presence of EMP, both at diagnosis and during follow-up, is linked to a poor prognosis and short survival [10]. The presence of plasmacytoma is associated with a poorer response to treatment and a higher relapse rate, although less pronounced in cases of bone-related plasmacytoma [14]. Our patient also did not respond to two different chemotherapy regimens administered after the second relapse and a more aggressive treatment had to be administered.

In conclusion, EMPs, although infrequently, are encountered during the course of multiple myeloma and its relapse. EMPs can be found in very rare localizations. In this regard, radiological, laboratory and histopathological evaluation of massive lesions during follow-up is important. Particularly, MRI can be effective as an imaging method in the diagnosis and close follow-up of patients with symptoms associated with extramedullary plasmacytomas.

Conflict of interests

The authors declare that they have no competing interests.

Patient informed consent

Written consent form was obtained from the patient.

Financial Disclosure

There are no financial supports.

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