

Case Report

Immunoglobulin D Multiple Myeloma Presenting as Spontaneous Fracture

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Keywords

Immunoglobulin D multiple myeloma · Spontaneous fracture · Multiple myeloma

Abstract

Immunoglobulin D multiple myeloma is a rare type of multiple myeloma that usually presents as bone pain, fatigue, or weight loss. We report a case of immunoglobulin D multiple myeloma in a 53-year-old Caucasian male patient with previous medical history of anaplastic oligodendroglioma status post-surgical resection who was evaluated for back pain while mowing the lawn. His physical examination showed tenderness over the lower thoracic vertebrae with no sensory or motor impairment. Initial lab investigations showed normocytic anemia and hypercalcemia with low parathyroid hormone. Magnetic resonance imaging of thoracic spine with and without contrast showed acute pathological fracture of the T12 vertebral body with enhancing soft tissue which extended into the left ventral epidural space and left T11–T12 neural foramen. Serum protein electrophoresis showed abnormal protein band in the gamma globulin. Free light chain assay showed serum free kappa which was elevated at 3,090.0 mg/L (reference range 3.3–19.4 mg/L). Immunoglobulin D was elevated at 566.0 mg/dL (reference range <15.3 mg/dL). The patient was successfully treated with standard chemotherapy and autologous peripheral blood stem cell transplant with complete remission 3 years after starting treatment. Advancement in the treatment of immunoglobulin D multiple myeloma urge clinicians to offer their patients new treatment options especially as of the earlier presentation of this subtype of multiple myeloma and the previous reports of worse prognosis.

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Introduction

Multiple myeloma is a common hematological malignancy that causes bony lesions. Rare subtypes of this relatively common disease are more frequently diagnosed in clinical practice. One of those subtypes is immunoglobulin D multiple myeloma which can have different clinical presentations. Advancement in the treatment of immunoglobulin D multiple myeloma is promising and should lead clinicians to earlier diagnosis for better outcomes.

Case Presentation

A 53-year-old Caucasian male patient with previous medical history of anaplastic oligodendroglioma status post-surgical removal and temozolomide therapy for 1 year in 2006 presented to the outpatient clinic on September 2013 complaining of back pain while mowing the lawn. Pain was described as severe and sudden and limiting the ability of regular daily activity. He denied any other site of pain. No fever, sweating, weight loss, or chills were reported. He was not taking any medications and he reported no allergy to any medication. He denied any tobacco use. He drinks alcohol occasionally and denies the use of any drugs including cocaine. His family history is significant for ovarian cancer in his mother and coronary artery disease in his father.

Physical examination showed tenderness over the lower thoracic vertebrae with no sensory or motor impairment. His neurological examination including gait assessment was normal. Initial lab investigations showed low hemoglobin of 12.5 g/dL (reference range 13.2–17.1 g/dL), normal vitamin D 25-OH of 47 ng/mL (reference range 30–100 ng/mL), high calcium level of 10.5 mg/dL (reference range 8.6–10.3 mg/dL), and low parathyroid hormone of 5 pg/mL (reference range 10–65 pg/mL). His creatinine was normal at 1.08 mg/dL (reference range 0.70–1.33 mg/dL), and he had mildly elevated alkaline phosphatase of 128 U/L (reference range 40–115 U/L). Aspartate aminotransferase and alanine aminotransferase were both within normal limits.

Serum protein electrophoresis showed total protein was normal at 6.9 g/dL (reference range 6.1–8.1 g/dL), normal albumin at 4.0 (reference range 3.5–4.7 g/dL), normal alpha 1 globulins, alpha 2 globulins, beta globulins, and gamma globulins, and showed an abnormal protein band in the gamma globulin region.

Urine electrophoresis showed normal 24-hour creatinine of 2.37 g (reference range 0.63–2.50 g/24 h) elevated total 24-hour protein of 3,050 mg (reference range <150 mg/24 h), elevated protein-to-creatinine ratio, and two abnormal protein bands detected in the gamma globulin region.

Free light chain assay showed serum free kappa elevated at 3,090.0 mg/L (reference range 3.3–19.4 mg/L) and low free lambda of 1.8 mg/L (reference range 5.7–26.3 mg/L), and free kappa/lambda ratio was elevated at >1,000.00 (reference range 0.26–1.65). Immunoglobulin D was elevated at 566.0 mg/dL (reference range <15.3 mg/dL). Magnetic resonance imaging (MRI) of thoracic spine with and without contrast showed acute pathological fracture of the T12 vertebral body with enhancing soft tissue which extends into the left ventral epidural space and left T11–T12 neural foramen.

Bone survey was done and showed diffuse patchy osteopenia with no focal lytic lesion. Bone marrow biopsy was obtained and showed 60% cellularity with about 54% cellularity consistent with atypical plasma cells with normal male karyotype. Fluorescent in situ hybridization was positive for gain of 11q; however, there was normal appearance of 11;14.

MRI of thoracic spine with and without contrast because of worsening back pain 2 weeks later showed a new T8 fracture (Fig. 1).

Diagnosis of spontaneous fracture secondary to immunoglobulin D myeloma was established. The patient was started on lenalidomide, bortezomib, and dexamethasone. He received a total of 6 cycles and his kappa light chain was down to 3.96 mg/L (reference range 3.3–19.4 mg/L) and his immunoglobulin D level was 0.7 mg/dL (reference range <15.3 mg/dL). He also received 9 treatments of high-dose radiation therapy to his thoracic spine for pain control. Indeed, he was receiving zoledronic acid on a monthly basis. After his fifth cycle, he was referred to a tertiary center for assessment of autologous peripheral blood stem cell transplant, which he underwent 6 months after the initial diagnosis.

Follow-up after 6 months of the transplantation showed complete remission, and the patient agreed to continue on lenalidomide maintenance therapy for a total of 3 years or as long as he is in remission.

Discussion

Multiple myeloma can present with spontaneous fracture [1]. Immunoglobulin D multiple myeloma is a rare disease with an incidence of 2% in all myelomas [2]. The main presenting features of immunoglobulin D myeloma were bone pain, fatigue, and weight loss [3]. A previous review of 77 cases reported bone pain as the most common presenting symptom, which happened in 77% of the cases with 89% lambda light chain predominance [4]. Median age of presentation was 57 years [4]. Though previously immunoglobulin D multiple myeloma was thought to have poor prognosis overall, a recent study reported median survival of 51.5 versus 50.7 months for patients with other subtypes [5]. Our case is peculiar as it represents a rare disease presenting in a young male patient with no identifiable risk factor. The presentation of spontaneous fracture is unusual for immunoglobulin D myeloma. Indeed, it shows excellent response to the standard chemotherapy used in multiple myeloma and to autologous peripheral blood stem cell transplant, which was done after complete remission. Previous reports of combining chemotherapy and autologous peripheral blood stem cell transplant showed improvement in prognosis in this rare disease [6].

Conclusion

Immunoglobulin D multiple myeloma can present as spontaneous fracture as initial presentation and it usually happens in young males. Advancement in the treatment of immunoglobulin D multiple myeloma urges clinicians to offer their patients new treatment options.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

The authors declare that there is no conflict of interest regarding the publication of this paper.

References

- 1 Kyle RA, Gertz MA, Witzig TE, et al: Review of 1027 patients with newly diagnosed multiple myeloma. *Mayo Clin Proc* 2003;78:21–33.
- 2 Bladé J, Kyle RA: Nonsecretory myeloma, immunoglobulin D myeloma, and plasma cell leukemia. *Hematol Oncol Clin North Am* 1999;13:1259–1272.
- 3 Bladé J, Lust JA, Kyle RA: Immunoglobulin D multiple myeloma presenting features response to therapy, and survival in a series of 53 cases. *J Clin Oncol* 1994;12:2398–2404.
- 4 Kim MK, Suh C, Lee DH, Min CK, Kim SJ, Kim K, Moon JH, Yoon SS, Lee G-W, Hang HJ, Kim S-H, Choi CW, Eom HS, Kwak J-Y, Kim HJ, Mun Y-C, Bang S-M, Lee K, Shin HJ, Lee JH: Immunoglobulin D multiple myeloma response to therapy, survival and prognostic factors in 75 patients. *Ann Oncol* 2011;22:411–416.
- 5 Zagouri F, Kastritis E, Symeonidis AS, et al: Immunoglobulin D myeloma: clinical features and outcome in the era of novel agents. *Eur J Haematol* 2014;92:308–312.
- 6 Pisani F, et al: IgD multiple myeloma a descriptive report of 17 cases: survival and response to therapy. *J Exp Clin Cancer Res* 2012;31:17.



Fig. 1. MRI of thoracic spine showing T8 fracture.