Rare occurence of Extramedullary Plasmacytoma with history of Solitary Bone Plasmacytoma

Yen-Chung Wang, DO; Kunal Elete, DO; Srujana Dasari, MD; Harris Naina, MD

Medical City Fort Worth

Introduction

Plasmacytomas are rare hematologic malignancies that usually arise as solitary tumors formed by a proliferation of a single clone of a plasma cell. Tumors are histologically similar to multiple myeloma (MM), but a defining feature is the lack of marrow involvement. A plasmacytoma may present as either a single bone lesion, solitary bone plasmacytoma (SBP), or less commonly as a soft tissue mass, an extramedullary plasmacytoma (SEP). Here, we present a case of an SEP in a patient with a prior history of an SBP.

Case Description

Patient is a 74-year-old male with a past medical history x of hypertension, hyperlipidemia, and left sinonasal plasmacytoma s/p radiation treatment in Fall 2020, who was initially lost to follow up but presented to the clinic with a three week history of lower back pain and left leg weakness. MRI imaging (Fig. 1) revealed a large mass within the left paraspinal musculature with osseous destruction of the left L2-L4 transverse processes. Biopsy revealed a proliferation of atypical plasma cells (Fig. 2). Patient then presented to the ED a few days after the biopsy due to worsening intractable back pain. Due to the location of the tumor and focal muscle weakness, surgical oncology and neurosurgery were both consulted for evaluation. Patient then underwent surgical debulking of the mass, which was complicated by hemorrhagic shock and hematoma formation. Once stabilized, the patient was discharged with a plan for outpatient radiation treatment.

Images

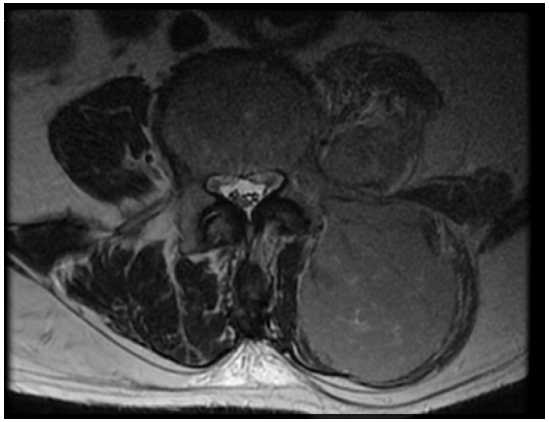

Fig 1: A bulky, predominantly soft tissue mass, with bone destruction, most apparent involving the left L3 and early involvement of left L2. It measures 15 cm craniocaudal and 5.5 x 6 cm in the axial plane.

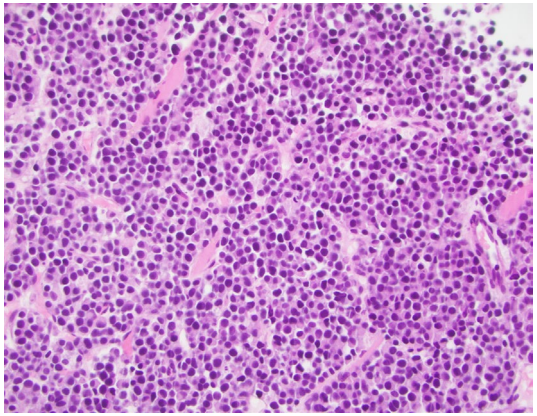

Fig 2: This histology slide of the lumbar mass depicts the extensive plasma cell infiltration of the tissue.

Discussion & Conclusion

The incidence of SEP is only 1.7-4.5%, and it's unclear why some individuals are predisposed to developing a plasmacytoma vs. a MM. Interestingly, a study by Hughes et al, suggests that differences in cellular adhesion molecule or chemokine receptor expression profiles may play a role. SBPs and SEPs are distinguished from MM by lack of the following criteria: anemia, hypercalcemia, renal insufficiency, or lytic bone lesions attributable to MM. Plasmacytoma may also still be diagnosed if marrow involvement shows <10% of plasma clonal cells. Our patient had none of the aforementioned laboratory or imaging abnormalities, but he did have 3-5% of clonal cells in his marrow. In regards to management, localized XRT is the treatment of choice for plasmacytomas, but surgical debulking may precede in those with a large tumor burden. The prognosis of those with a SBP or SEP is quite similar, with median survival of 10-11 years. Despite appropriate XRT, there exists a significant risk of progression to MM. Moreover, only 2% of patients with SBP have been found to develop sequential plasmacytomas, as seen in our patient. We hope to bring more attention to these uncommon plasma cell dyscrasias and emphasize the importance of regular follow-up.

References

1. Soutar, R., Lucraft, H., Jackson, G., Reece, A., Bird, J., Low, E. and Samson, D. (2004), Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. British Journal of Haematology, 124: 717-726. 2. Hughes, M., Doig, A. and Soutar, R. (2007), Solitary plasmacytoma and multiple myeloma: adhesion molecule and chemokine receptor expression patterns. British Journal of Haematology, 137: 486-487. 3. Goyal G, Bartley AC, Funni S, Inselman J, Shah ND, Marshall AL, Ashrani AA, Kapoor P, Durani U, Hashmi SK, Siddiqui MA, Buadi FK, Go RS, Kyle RA, Kumar S, Gonsalves WI. Treatment approaches and outcomes in plasmacytomas: analysis using a national dataset. Leukemia. 2018 Jun;32(6):1414-1420. 4. Alexiou C, Kau RJ, Dietzfelbinger H, Kremer M, Spiess JC, Schratzenstaller B, Arnold W. Extramedullary plasmacytoma: tumor occurrence and therapeutic concepts. Cancer. 1999 Jun 1;85(11):2305-14. PMID: 10357398.

This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.

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