CASE REPORT

Solitary extraosseous plasmacytoma

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Key Clinical Message
Plasma cell neoplasms are characterized by a neoplastic plasma cell lineage which produces a monoclonal immunoglobulin. These neoplasms can present as a single lesion (solitary plasmacytoma) or as multiple lesions (multiple myeloma). Solitary plasmacytomas most frequently occur in bone (plasmacytomas of bone), but can also be found outside bone in soft tissues (extramedullary plasmacytomas).

Keywords
Extramedullary, multiple myeloma, plasmacytoma, solitary extramedullary plasmacytomas.

Introduction
We do not understand yet why some patients develop multiple myeloma and others a single plasmacytoma [1, 2], but that might be related to differences in the chemokine receptor expression profiles of the malignant plasma cells or cellular adhesion molecules [3]. Solitary extramedullary plasmacytomas (SEP) are extremely rare tumors that arise outside of the bone marrow in the absence of any sign of multiple myeloma [4, 5]. They are solitary lesions, and are often seen in the head and neck regions [6, 7], mainly in the upper aerodigestive and to a lesser extent in the gastrointestinal tract (GI) tract, bladder, central nervous system (CNS), thyroid, breast, testes, parotid gland, lymph nodes, as well as in skin.

Solitary extramedullary plasmacytomas tend to occur during the fifth and seventh decades of life, rarely in younger population [8].

Extramedullary plasmacytomas (EMPs) can arise in patients with multiple myeloma at any time during the course of the disease and in one-third of the cases, resulting in a worse clinical outcome that should not be confused with SEP [9, 10].

Case Presentation
This particular case involves a 40-year-old Hispanic male with past medical history of mental retardation, anxiety, and dyslipidemia who lives in a group home. Patient presented initially to his Primary care physician (PCP) for an annual physical. During routine examination, a neck mass was identified on the base of the lateral right side of the neck anterior to the sternocleidomastoid muscle. It measured 3 cm by 3 cm nontender with slight mobility. He denied any constitutional symptoms such as fever, chills, sweating, weight loss or change in diet or bowel habit, easy bruising, or hoarseness.

Patient was referred to ENT, and neck soft tissue CT scan with and without contrast was performed which showed enhancing 3.4 x 2.8 x 5.2 cm mass right retro-mandibular region just anterior to the sternocleidomastoid muscle with mass effect pushing the carotid vessels posteriorly with adjacent bony destruction; metastasis was not excluded by this test. Also, the scan was showing anterior adjacent mass approximately 0.8 x 1.1 cm that may present metastasis or mildly enlarged lymph node.

Upon seeing ENT doctor, Fine needle aspiration (FNA) was done on the site and referred to a medical oncologist.
for this suspicious mass indicating the malignant pathology. FNA result was not diagnostic as it showed cuboidal to columnar histologically benign appearing cells along with many small to medium size lymphocytes with differential diagnosis of: (i) salivary gland neoplasm, (ii) branchial cleft cyst, or (iii) possibility lymphoid proliferative disorder. Based on the FNA result, lymphoma could not be ruled out and surgical biopsy was recommended at that time. Initial blood work up was done showing monocyte was elevated of 11.3%.

Blood work up:

| Test     | Result |
|----------|--------|
| Cr       | 0.9    |
| WBC      | 5.9    |
| Hb       | 15.5   |
| Hct      | 45.3   |
| MCV      | 90.5   |
| Pt       | 283    |
| Granulocytes | 62.8% |
| Lymphocytes | 28.2% |
| Monocytes| 8.0%   |
| Glucose  | 87     |
| Na       | 142    |
| K        | 4.3    |
| AST      | 19     |
| ALT      | 27     |
| AlkPhos  | 80     |

Initial differential diagnoses by ENT physician involved Castleman disease versus plasma cell neoplasia. The report came back favoring plasma cell neoplasm “possibility of primary lymph node plasmacytoma by systemic involvement exclusion.”

Open biopsy was done with frozen section and it was found to have an extensive plasma cells with no carcinoma but some cells showing binucleated forms and atypical nuclei with final diagnosis of primary lymph node plasmacytoma (Figs 1 and 2).

There was kappa light chain restriction by Immunohistochemistry (IHC).

The following observations were made:

CD138 staining: positive flow cytometry: nondiagnostic.
Beta-2 microglobulin: normal range with no elevation immunofixation.
Quantitative immunoglobulins IgG/A/M panel: normal. Serum K/L light chains ratio: normal.
PET CT scan showing mildly enlarged right level 2B lymph nodes measuring 14 mm with SUV of 4.1 and few adjacent smaller level 2B lymph nodes which are a normal size with low-grade metabolic activity.
BM Bx was done and showed no myeloma, norm cellular marrow with maturing trilineage.
Hematopoiesis – no definitive morphologic or immunophenotypic evidence of clonal expansion of plasma cells or involvement by a mature B-cell non-Hodgkin lymphoma – mildly increased iron stores Kappa and lambda: stain a few polytypical plasma cells in normal number and distribution (Figs 3–6).

Initial recommendation by the oncologist was radiation therapy for definitive treatment for localized plasmacytoma of the neck which should likely provide adequate disease control on-site.
Patient received 4500 cGy in 25 fractions of radiation treatments on the right neck with 3060 cGy in 17 fractions. The primary tumor site received additional 1440 cGy in eight fractions of the total of 4500 cGy in 25 fractions.

After radiation, patient was observed and monitored periodically and clinically did not show any evidence of residual disease other than superficial skin changes from the radiation which resolved later.

The patient will be at risk for developing plasmacytoma at other locations, hence he will be under close follow up.

**Discussion**

In this report, we present a case of SEP with no involvement of the bony structure or bone marrow. The case is quite rare as it represents 2–10% of all the multiple myeloma cases [11, 12]. Solitary extra osseous plasmacytoma has a unique characteristic of being undetectable disease elsewhere on PET/CAT scan.

A plasmacytoma is a unique solitary mass of neoplastic monoclonal plasma cells in either bone or soft tissue (extramedullary).

Plasmacytoma has been classified into three subtypes [13]. The most common type is multiple myeloma, which is usually a disseminated disease and characterized by abnormal M protein. The other two types, solitary plasmacytoma of the bone and EMP of the soft tissue, are considerably less common. EMPs present in <5% of plasma cell neoplasms and often (>80%) originate in the head and neck region [16]. Anatomically, we can divide the SEP into two groups: (i) plasmacytoma of the skeletal system (SBP) and (ii) EMP [21, 24].

The diagnosis of EMP of the soft tissue has been based on the following criteria: (i) pathological tissue evidence of monoclonal plasma cells involving a single extramedullary site; (ii) no bone marrow involvement; (iii) no anemia, hypercalcemia or renal impairment caused by plasma cell dyscrasias; (iv) negative skeletal survey results; and (v) low serum or urinary levels of monoclonal immunoglobulin [2]. The etiology of this disease remains unknown, but factors, such as chronic irritation from inhaled irritants or viral pathogenesis, have been previously indicated [17, 18]. Radiotherapy remains the mainstay for the management of EMP. As it considered radiosensitive, with a local control rate of 90–100% with a rate of conversion into MM is 31%. Moderate dose RT of at least 40 Gy using limited radiation fields is recommended [14, 15, 17, 19, 20]. Because of the high rate of recurrence and progression to multiple myeloma, follow-up radiological and electrophoresis assessment is required.
following treatment [22]. The overall 10-year survival rate is ~70% [2, 16, 23]. A literature search revealed no publications supporting the use of surgery alone to treat EMP [24].

**Conflict of Interest**

None declared.

**References**

1. Jaffe, E. S., N. L. Harris, H. Stein, J. W. Vardiman, eds. 2001. World Health Organization Classification of tumours: pathology and genetics of tumours of haematopoietic and lymphoid tissues. IARC Press, Lyon, 352 pp, 185.
2. Soutar, R., H. Lucaft, G. Jackson, A. Reece, J. Bird, E. Low, and D. Samson; Guidelines Working Group of the UK Myeloma Forum; British Committee for Standards in Haematology; British Society for Haematology. 2004. Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. Br. J. Haematol. 124:717.
3. Hughes, M., A. Doig, and R. Soutar. 2007. Solitary plasmacytoma and multiple myeloma: adhesion molecule and chemokine receptor expression patterns. Br. J. Haematol. 137:486.
4. Mendenhall, W. M., C. M. Mendenhall, and N. P. Mendenhall. 2003. Solitary plasmacytoma of bone and soft tissues. American Journal of Otolaryngology 24:395–399.
5. Dimopoulos, M. A., L. A. Moulopoulos, A. Maniatis, and R. Alexanian. 2000. Solitary plasmacytoma of bone and asymptomatic multiple myeloma. Blood 96:2037.
6. Knowling, M. A., A. R. Harwood, and D. E. Bergsagel. 1983. Comparison of extramedullary plasmacytomas with solitary and multiple plasma cell tumors of bone. J. Clin. Oncol. 1:255–262.
7. Galeni, P., M. Cavo, A. Pulsoni, et al. 2000. Clinical outcome of extramedullary plasmacytoma. Haematologica 85:47–51.
8. Frassica, D. A., F. J. Frassica, M. F. Schray, et al. 1989. Solitary plasmacytoma of bone: Mayo Clinic experience. Int. J. Radiat. Oncol. Biol. Phys. 16:43.
9. Hallek, M., P. L. Bergsagel, and K. C. Anderson. 1998. Multiple myeloma: increasing evidence for a multistep transformation process. Blood 91:3–21.
10. Chen, H. F., T. Q. Wu, Z. Y. Li, H. S. Shen, J. Q. Tang, W. J. Fu, Z. G. Yuan, and J. Hou. 2012. Extramedullary plasmacytoma in the presence of multiple myeloma: clinical correlates and prognostic relevance. OncoTargets and Therapy 5:329–334.
11. Delauche-Cavallier, M. C., J. D. Laredo, and M. Wybier. 1988. Solitary plasmacytoma of the spine: long-term clinical course. Cancer 61:1707–1714.
12. Lasker, J. C., J. O. Bishop, and J. H. Wilbanks. 1991. Solitary myeloma of the talus bone. Cancer 68: 202–205.
13. Swerdlow, S., E. Campo, N. Harris, E. Jaffe, S. Liperi, H. Stein, et al. 2008. WHO classification of tumours of haematopoietic and lymphoid tissue. 4th ed. International Agency for research on Cancer, Lyon.
14. Hughes, M., R., Soutar, H. Lucaft, R. Owen, and J. Bird. 2009. Guidelines on the diagnosis and management of solitary plasmacytoma of bone, extramedullary plasmacytoma and multiple solitary plasmacytomas: 2009 update. British Committee for Standards in Haematology, London, UK.
15. Alexiou, C., R. J. Kau, H. Dieteltbiner, M. Kremer, J. C. Spiess, B. Schratzenstaller, et al. 1999. Extramedullary plasmacytoma: tumor occurrence and therapeutic concepts. Cancer 85:2305–2314.
16. Straetmans, J., and R. Stokroos. 2008. Extramedullary plasmacytomas in the head and neck region. Eur. Arch. Otorhinolaryngol. 265(11):1417–1423. doi: 10.1007/s00405-008-0613-0. Epub 2008 Feb 26.
17. Sasaki, R., K. Yasuda, E. Abe, et al. 2012. Multi-institutional analysis of solitary extramedullary plasmacytoma of the head and neck treated with curative radiotherapy. Int. J. Radiat. Oncol. Biol. Phys. 82: 626–634.
18. Sasaki, S., K. Hashimoto, S. Nakatsuka, et al. 2011. Plasmablastic extramedullary plasmacytoma associated with Epstein-Barr virus arising in an immunocompetent patient with multiple myeloma. Intern. Med. 50:2615–2620.
19. Chao, M. W., P. Gibbs, A. Wirth, G. Quong, M. J. Guinney, and K. H. Liew. 2005. Radiotherapy in the management of solitary extramedullary plasmacytoma. Intern. Med. J. 35:211–215.
20. Suh, Y. G., C. O. Suh, J. S. Kim, S. J. Kim, H. O. Pyun, and J. Cho. 2012. Radiotherapy for solitary plasmacytoma of bone and soft tissue: outcomes and prognostic factors. Annals of Hematology 91:1785–1793.
21. McIntyre, O. R. 1977. Current concepts in cancer multiple myeloma. N. Engl. J. Med. 301:193–196.
22. Moulopoulos, L. A., M. A. Dimopoulos, and D. Weber. 1993. Magnetic resonance imaging in staging of solitary plasmacytoma of bone. J. Clin. Oncol. 11:1311–1315.
23. Kar, M., R. Roy, J. Chakraborty, and S. Das. 2008. Extramedullary plasmacytoma – a rare presentation. Journal, Indian Academy of Clinical Medicine 9:298–301.
24. Chang, Y. L., P. Y. Chen, and S. H. Hung. 2014. Extramedullary plasmacytoma of the nasopharynx: a case report and review of the literature. Oncology Letters 7:458–460.