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Short Communication

Solitary Extramedullary Plasmacytomas: The Demographic Profile and Clinical Outcome in a Tertiary Care Centre

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ABSTRACT

Background: Plasmacytomas are the clonal proliferation of plasma cells that are cytologically and immunophenotypically identical to those of plasma cell myeloma but manifest a localized growth pattern. These patients are treated with radiotherapy, however large studies from India about the outcome of this disease is lacking. **Objectives:** To analyse the demographic & clinical profile of patients with solitary Extramedullary plasmacytoma. **Methods:** sixteen patients who were diagnosed and treated for solitary EMP during 2006 to 2010 at our institute were considered for this retrospective study. Histopathology was used to confirm the diagnosis. All these patients received curative dose of radiotherapy in form of EBRT (dose range 30-60 Gy, over 4-6 weeks). These patients were then followed up every three monthly and their outcome was correlated with tumor size (> 5cms or less), and dose of radiotherapy received (>40Gy or less). **Results:** EMP commonly affected males with median age of 56 years. The most common site of involvement was head and neck region. Twenty five percent had elevated 'M' protein. At median follow up of 5 years (range 3-7 years), three patients developed multiple myeloma and two patients developed local recurrence. Local control rate was significantly better ≥ 40Gy when compared to <40Gy (p=0.0256). Size >5cms did not have poor outcome (p= 0.1058). 5 year EFS was 71±4.8%. **Conclusion:** EMP is a highly radiosensitive tumor. With proper diagnosis and management excellent outcome can also be achieved in our scenario.

***Keywords*:** Solitary Extramedullary Plasmacytoma

INTRODUCTION

Plasmacytomas usually represent localized growth of plasma cells. These can arise in association with bony structures (medullary) or without (extra-medullary). Solitary Extramedullary plasmacytoma (EMP) are rare tumors which accounts for less than 5% of all plasma cell neoplasm. [1] These tumors are differentiated from multiple myeloma by lack of CRAB (increased calcium, renal failure, anaemia or multiple bone lesions). [2]

In this study we have analysed the demographic profile, therapy received and outcome of EMP. To date there are few studies of EMP from India, and to the best of our knowledge this is the largest study of outcome of EMP in India.

MATERIALS AND METHODS

Sixteen patients who were diagnosed and treated for solitary EMP during 2006 to 2010 at our institute were considered for this retrospective study. The case records of these patients were analysed in detail for demographic profile, clinical features, treatment and outcome. The pathologic material was reviewed by a single pathologist. The criteria used to diagnose EMP were 1) Tissue biopsy showing monoclonal plasma cell histology [figure 2]. 2) Absence of lytic bone lesion, anaemia, renal insufficiency, hypocalcaemia and absence or low monoclonal protein in serum/urine. [3] The histological diagnosis was confirmed using CD38+ve by IHC, and monoclonal light chain expression of plasma cells. These patients also underwent complete blood examination, serum biochemistry to rule out hypercalcemia and renal failure, bone marrow aspiration and biopsy, skeletal X-ray survey, serum protein electrophoresis, immunoglobulin quantification, and immune- electrophoresis.

The institution policy for treatment of these patients included radiotherapy alone or combined with surgery. Radiotherapy (EBRT) was given by conventional techniques with dose range of 30-60Gy over 4-6weeks. Radiotherapy was given to gross tumor plus 2cms margin in head and neck area. Adjacent lymph nodal irradiation was not done.

The response to treatment was defined as CR (complete remission) if disappearance of tumor mass and clinical symptoms as well as M component if present. PR (partial remission) as reduction in signs and symptoms related to disease and no response if clinical features were unmodified with therapy. These patients were then followed up every 3 monthly for local failure, multifocal extramedullary relapse or myeloma. The outcome was correlated with tumor size (> 5cms or less), and dose of radiotherapy received (>40Gy or less). The event free survival was evaluated for all patients using Kaplan Meier curve (SPSS 19-SPSS Inc, USA).

RESULTS

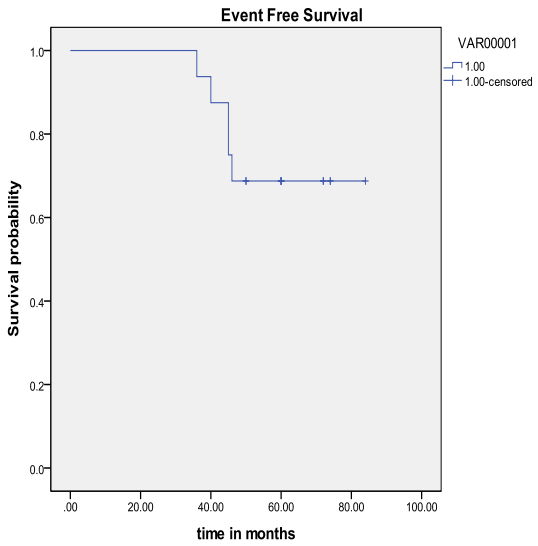
Table 1. Patients' profile.

Study	Shih et al [6]	Galieni et al [7]	Present study
Number of patients	10	46	16
Male: female	9:1	1.7:1	1.6:1
Median age	63	55	56
Site- head and neck lungs pelvis testis bladder skin thyroid brain GIT lymph node	 8 1 1	 36 1 1 1 1 4	 10 2 2 1 1
Monoclonal component	60%	21%	25%
Recurrence/myeloma	40%	22%	31.25%

The median age of presentation was 56 years (range 40-72). The clinical features are given in table 1. Head and neck region was the most common site of disease with presenting symptom ranging from mass effect due to lesion, epistaxis, nasal obstruction, cheek swelling. In the head and neck region, the sites of involvement were orbit (4 cases), Maxilla (2 cases), Nasopharynx (2 cases) and nasal cavity in 2 patients. M-spike was present in four patients at diagnosis. Fourteen patients were treated with radiotherapy alone and 2 patients received both surgery and radiotherapy. 14 patients achieved complete remission while partial remission was seen in two patients. The median follow up was 5 years (range 3 to 7 years). Two patients (12.5%) developed local recurrence, and three patients(18.7%) developed multiple myeloma. Local control rate was significantly better with ≥ 40Gy when

compared to <40Gy (p=0.0256). Size >5cms did not have poor outcome (p= 0.1058). The 5 year EFS was 71±4.8%.(figure 1). Two patients who experienced local relapse were again treated with local radiotherapy (46 and 44Gy) since they had received low dose RT (30 Gy) upfront. Three patients with multiple myeloma were treated with thalidomide, dexamethasone with bisphosphonates.

Figure 1: Event free survival



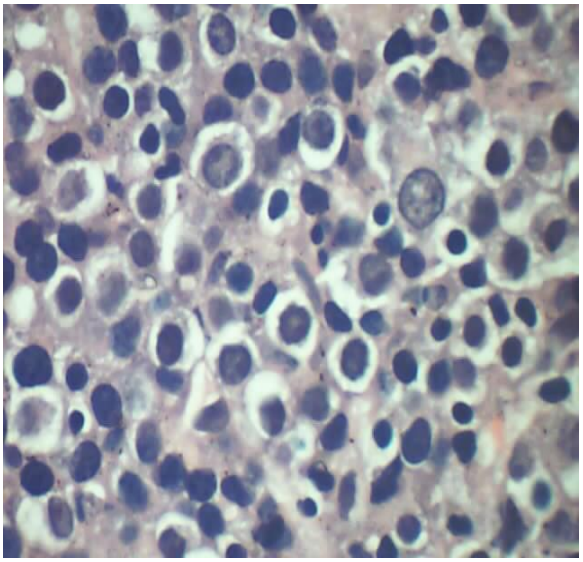


Figure 2. Showing plasma cells in a tissue biopsy.

DISCUSSION

EMP occurs when soft tissue is infiltrated by clonal plasma cell. As these tumors are highly curable with radiotherapy, differentiating them from other lesion in elderly such as poorly differentiated neoplasm, immunoblastic lymphoma is important. This study highlights the need for proper diagnosis and management, further avoiding unnecessary chemotherapy in EMP.

As seen in our study, EMP is a disease of elderly with a slight male predominance. This has been reported in other larger studies as well. [4] A review of literature worldwide suggest that the head and neck region is the most common site of origin, with gastrointestinal tract being the next most frequent site. [5] Even in our study head and neck region was the commonest site but lung and pelvic involvement were the next most frequent sites. The comparison of clinical features of our study with other studies is given in table 1. [6, 7]

As seen in our study and World Series EMP has excellent local disease control and long term disease free survival with radiotherapy. However the optimal radiotherapy dose and need for elective nodal irradiation is still a matter of controversy. The 5 year local control rate exceeds 90% with ≥40 Gy, while its 40% with <40Gy. [8] Better local control with ≥40Gy has been seen our study as well as in studies also. [9] Size >5cms tumor did not have poor outcome; this is in contrast to study from Princess Margaret Hospital where larger tumor had poor outcome. [10] This difference may be due to combining both solitary plasmacytoma of bone and EMP in their study. As size is a definite poor risk in solitary plasmacytoma of bone, but not in EMP. [11]

The local failure rate and conversion to myeloma rate was 12.5% and 18.7% respectively in our study, this is similar to

other studies. [6, 7] Long term disease free survival approaches 70-80% indicating an excellent response to radiotherapy. [4]

CONCLUSION

In conclusion EMP is a highly radiosensitive tumor with excellent outcome. Larger trials are needed to define the role newer modalities for diagnosis and follow-up like PET scan and role of nodal irradiation in treatment.

REFERENCES

1. Dimopoulos MA, Kiamouris C, Moulopoulos LA. Solitary plasmacytoma of bone and Extramedullary plasmacytoma. Hematol OncolClin North Am 1999;13:1249-57.
2. Kilciksiz S, Karakoyun-Celik O, Agaoglu FY, Haydaroglu A. A review for solitary plasmacytoma of bone and extramedullaryplasmacytoma. Scientific WorldJournal. 2012;2012:895765.
3. Galieni P, Cavo M, Pulsoni A, Avvisati G, Bigazzi C, Neri S, et al. Clinical outcome of Extramedullary plasmacytoma. Haematologica 2000;85: 47-51.
4. Ozsahin M, Tsang RW, Poortmans P, Belkacémi Y, Bolla M, Dinçbas FO, et al. Outcomes and patterns of failure in solitary plasmacytoma: a multicenter Rare Cancer Network study of 258 patients. Int J RadiatOncolBiol Phys. 2006;64:210-7..
5. Dimopoulos MA, Kiamouris C, Moulopoulos LA. Solitary plasmacytoma of bone and extramedullaryplasmacytoma. Hematol OncolClin North Am 1999;13:1249-57.
6. Shih LY, Dunn P, Leung WM, Chen WJ, Wang PN. Localised plasmacytomas in Taiwan: comparison between extramedullaryplasmacytoma and solitary plasmacytoma of bone. Br J Cancer 1995;71:128-33.
7. Galieni P, Cavo M, Pulsoni A, Avvisati G, Bigazzi C, Neri S, et al. Clinicaloutcome of Extramedullary plasmacytoma. Haematologica 2000;85: 47-51.
8. Tournier-Rangeard L, Lapeyre M, Graff-Caillaud P, Mege A, Dolivet G, Toussaint B, et al. Radiotherapy for solitary extramedullaryplasmacytoma in the head-and-neck region: A dose greater than 45 Gy to the target volume improves the local control. Int J RadiatOncolBiolPhys 2006;64:1013-7.
9. Creach KM, Foote RL, Neben-Wittich MA, Kyle RA. Radiotherapy for extramedullaryplasmacytoma of the head and neck Int J RadiatOncolBiol Phys. 2009 Mar 1;73(3):789-94.
10. Tsang RW, Gospodarowicz MK, Pintilie M, Bezjak A, Wells W, Hodgson DC et al.. Solitary plasmacytoma treated with radiotherapy: impact of tumor size on outcome. Int J RadiatOncolBiolPhys 2001;50:113-20.
11. Knobel D, Zouhair A, Tsang RW, Poortmans P, Belkacémi Y, Bolla M, et al. Prognostic factors in solitary plasmacytoma of the bone: a multicenter Rare Cancer Network study. BMC Cancer. 2006 May 5;6:118.

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