Solitary and Extramedullary Plasmacytoma

Note: Consider Clinical Trials as treatment options for eligible patients.

INITIAL EVALUATION

- History and physical
- CBC with differential, BUN, creatinine, electrolytes, albumin, LDH, calcium, beta-2-microglobulin, serum quantitative immunoglobulins, serum protein electrophoresis (SPEP), serum immunofixation (SIFE), and serum free light chains (sFLC) including involved:uninvolved sFLC ratio
- 24-hour urine protein electrophoresis (UPEP) and urine immunofixation (UIFE)
- Bone marrow biopsy and aspirate with flow cytometry
- PET/CT of whole body or MRI of whole body
- If PET/CT of whole body or MRI of whole body is unavailable, then perform skeletal survey and MRI of the cervical, thoracic, lumbar and sacral spine. Consider CT or MRI of the affected area.
- In select settings, other imaging studies may be considered, such as ultrasound for superficial masses
- Lifestyle risk assessment[1]

Patient's initial evaluation findings meet criteria for multiple myeloma treatment? (see below)

TREATMENT

Yes → Treat as multiple myeloma (see Multiple Myeloma algorithm)

No →
- Treat as plasmacytoma, see Page 2
- Progression to multiple myeloma risk assessment, see Page 2

Criteria for multiple myeloma treatment:
- Anemia, hypercalcemia, renal failure due to multiple myeloma **and/or**
- Bony lytic lesions due to multiple myeloma in a skeletal survey **and/or** MRI of whole body and/or PET/CT of whole body **and/or**
- sFLC involved:uninvolved ratio ≥ 100 **and/or**
- Greater than one focal lesions on MRI (each focal lesion must be 5 mm or more in size) **and/or**
- Percentage of clonal plasma cells is ≥ 60% in the core biopsy by CD138 immunohistochemistry

Note: Treatment may be considered if percentage of clonal plasma cells is ≥ 10% in the core biopsy by CD138 immunohistochemistry

[1] See Physical Activity, Nutrition, and Tobacco Cessation algorithms; ongoing reassessment of lifestyle risks should be a part of routine clinical practice

Department of Clinical Effectiveness V6
Approved by the Executive Committee of the Medical Staff on 04/19/2022

Solitary and Extramedullary Plasmacytoma

Note: Consider Clinical Trials as treatment options for eligible patients.

CLINICAL PRESENTATION

Solitary plasmacytoma of bone (SPB)

Extramedullary plasmacytoma (EMP)

PRIMARY TREATMENT

- Treat with involved site radiation therapy (ISRT) to a total dose of at least 40 Gy (regardless of site)[1]
- Multiple myeloma progression risk assessment[2]

FOLLOW-UP SURVEILLANCE

Evaluate response every 3 months for the first 2 years, then every 6-12 months thereafter:

- CBC with differential, BUN, creatinine, electrolytes, albumin, LDH, calcium, beta-2-microglobulin, serum quantitative immunoglobulins, SPEP, SIFE, and sFLC
- 24 hour urine for total protein, UPEP, and UIFE
- Consider bone marrow biopsy and aspirate, if clinically indicated
- MRI and/or PET/CT at 3 months after completion of ISRT to assess treatment response. Consider skeletal survey and/or MRI and/or CT and/or PET/CT every 6-12 months, if clinically indicated.

Complete response

- Complete disappearance of paraprotein in serum or urine by immunofixation and normalization of sFLC
- No new bone lesions or other features of multiple myeloma (see Multiple Myeloma algorithm)

→ Re-evaluate as indicated

Persistent presence of serum paraprotein 1 year after treatment → Consider more frequent monitoring

Progressive disease →

- Restage with myeloma workup
- See Multiple Myeloma algorithm

[1] Historically, the recommended dose has been at least 40 Gy. More recent data suggests that lower doses may be sufficient. Refer to suggested readings for data regarding ISRT dose.

[2] Risk factors:

- Plasmacytoma size ≥ 10 cm at diagnosis
- Persistent presence of serum paraprotein 1 year after treatment

Rates of progression to multiple myeloma for patients with 1 or 2 risk factors:

- 3 years from diagnosis – 65%
- 5 years from diagnosis – 70%
- 10 years from diagnosis – 82%

Department of Clinical Effectiveness V6
Approved by the Executive Committee of the Medical Staff on 04/19/2022

SUGGESTED READINGS

Cavo, M., Terpos, E., Nanni, C., Moreau, P., Lentzsch, S., Zweegman, S., . . . Zamagni, E. (2017). Role of 18 F-FDG PET/CT in the diagnosis and management of multiple myeloma and other plasma cell disorders: A consensus statement by the International Myeloma Working Group. *The Lancet Oncology*, 18(4), e206-e217. https://doi.org/10.1016/S1470-2045(17)30189-4

Dimopoulos, M., Moulopoulos, L., Maniatis, A., & Alexanian, R. (2000). Solitary plasmacytoma of bone and asymptomatic multiple myeloma. *Blood*, *96*(6), 2037-2044. https://doi.org/10.1182/blood.V96.6.2037

Liebross, R. H., Ha, C. S., Cox, J. D., Weber, D., Delasalle, K., & Alexanian, R. (1999). Clinical course of solitary extramedullary plasmacytoma. *Radiotherapy and Oncology*, *52*(3), 245-249. https://doi.org/10.1016/S0167-8140(99)00114-0

Manasanch, E. E., Kunacheewa, C., Claussen, C. M., Lee, H. C., Thomas, S. K., Gunther, J., . . . Weber, D. M. (2021). Serum paraprotein persistence and size determine outcome in a cohort of patients with a modern definition of plasmacytoma with up to 19 years of follow up. *Blood Cancer Journal*, *11*(17), 1-5. https://doi.org/10.1038/s41408-021-00419-1

Mendenhall, C. M., Thar, T. L., & Million, R. R. (1980). Solitary plasmacytoma of bone and soft tissue. *International Journal of Radiation Oncology, Biology, Physics*, *6*(11), 1497-1501. https://doi.org/10.1016/0360-3016(80)90006-1

Ozsahin, M., Tsang, R. W., Poortmans, P., Belkacémi, Y., Bolla, M., Dinçbas, F. O., . . . Zouhair, A. (2006). Outcomes and patterns of failure in solitary plasmacytoma: A multicenter Rare Cancer Network study of 258 patients. *International Journal of Radiation Oncology, Biology, Physics*, *64*(1), 210-217. https://doi.org/10.1016/j.ijrobp.2005.06.039

Reed, V., Shah, J., Medeiros, L. J., Ha, C. S., Mazloom, A., Weber, D. M., . . . Dabaja, B. S. (2011). Solitary plasmacytomas. *Cancer*, *117*(19), 4468-4474. https://doi.org/10.1002/cncr.26031

Soutar, R., Lucraft, H., Jackson, G., Reece, A., Bird, J., Low, E., & Samson, D. (2004). Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. *British Journal of Haematology*, *124*(6), 717-726. https://doi.org/10.1111/j.1365-2141.2004.04834.x

Tsang, R. W., Gospodarowicz, M. K., Pintilie, M., Bezjak, A., Wells, W., Hodgson, D. C., & Stewart, A. K. (2001). Solitary plasmacytoma treated with radiotherapy: Impact of tumor size on outcome. *International Journal of Radiation Oncology, Biology, Physics*, *50*(1), 113-120. https://doi.org/10.1016/S0360-3016(00)01572-8

Weber, D. M. (2005). Solitary bone and extramedullary plasmacytoma. *American Society of Hematology Education Program Book*, *2005*(1), 373-376. https://doi.org/10.1182/asheducation-2005.1.373

Wilder, R. B., Ha, C. S., Cox, J. D., Weber, D., Delasalle, K., & Alexanian, R. (2002). Persistence of myeloma protein for more than one year after radiotherapy is an adverse prognostic factor in solitary plasmacytoma of bone. *Cancer*, *94*(5), 1532-1537. https://doi.org/10.1002/cncr.10366

DEVELOPMENT CREDITS

This practice algorithm is based on majority expert opinion of the Myeloma Center providers at the University of Texas MD Anderson Cancer Center. It was developed using a multidisciplinary approach that included input from the following:

Core Development Team Leads

Elisabet E. Manasanch, MD (Lymphoma/Myeloma)

Workgroup Members

Bouthaina S. Dabaja, MD (Radiation Oncology)
Penny Q. Fang, MD (Radiation Oncology)
Wendy Garcia, BS♦
Kevin W. McEnery, MD (Musculoskeletal Imaging)
Chelsea Pinnix, MD, PhD (Radiation Oncology)
Donna M. Weber, MD (Lymphoma/Myeloma)
Milena Zhang, PharmD♦

♦Clinical Effectiveness Development Team