EDORIUM Journals

Int J Case Rep Images 2023;14(1):133–137.
www.ijcasereportsandimages.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Extramedullary plasmacytoma presenting as a rapidly growing lateral neck mass: A case report

Christopher G Salib, Cierra N Harper, Stephanie Carter, Therlinsie Fleurizard, Damirez Fossett

ABSTRACT

Introduction: We detail the rare clinical presentation of an extramedullary plasmacytoma as a rapidly growing lateral neck mass. The pathology and its management are discussed. A rapidly growing extramedullary plasmacytoma infiltrating the cervical spine.

Case Report: A 53-year-old male presented to our emergency room with a one-month history of worsening left-sided neck pain and an enlarging lateral neck mass. Evaluation with cervical imaging and laboratory analysis were performed. The patient underwent a soft tissue biopsy with subsequent spinal cord decompression and an occipital-cervical fusion. Pathology revealed the diagnosis of plasmacytoma. The patient was referred to oncology and radiation therapy for adjuvant treatment.

Conclusion: This case represents the unusual presentation of a rapidly growing extramedullary plasmacytoma to the soft tissues of the neck with subsequent erosion of the base of the skull and upper cervical vertebral spine. It was treated effectively with posterior cervical decompression, arthrodesis, and adjuvant radiation and chemotherapy.

Keywords: Chemotherapy, Extramedullary plasmacytoma, Occipito-cervical fusion, Radiation therapy

Christopher G Salib[1], MD, Cierra N Harper[2], BS, Stephanie Carter[2], BS, Therlinsie Fleurizard[1], MD, Damirez Fossett[1,2], MD

Affiliations: [1]Department of Neurosurgery, Howard University Hospital, Washington, DC, USA; [2]Howard University College of Medicine, Washington, DC, USA.

Corresponding Author: Damirez Fossett, MD, Department of Neurosurgery, Howard University Hospital, 2041 Georgia Avenue NW, Washington, DC 20059, USA; Email: damirez.fossett@howard.edu

Received: 10 March 2023
Accepted: 12 June 2023
Published: 30 June 2023

How to cite this article

Salib CG, Harper CN, Carter S, Fleurizard T, Fossett D. Extramedullary plasmacytoma presenting as a rapidly growing lateral neck mass: A case report. Int J Case Rep Images 2023;14(1):133–137.

Article ID: 101398Z01CS2023

****

doi: 10.5348/101398Z01CS2023CR

INTRODUCTION

Solitary plasmacytomas (SPs) are aggressive tumors characterized by the local proliferation of neoplastic monoclonal plasma cells without the systemic features of multiple myeloma [1, 2]. They are divided into two categories based on the affected tissue type: solitary bone plasmacytoma (SBP), found in bone, and extramedullary plasmacytoma (EMP), found in soft tissues. Solitary bone plasmacytoma most commonly presents in the axial skeleton, whereas EMP predominantly appears in the aero-digestive tract of the head and neck [3, 4]. Plasmacytomas involving the soft tissue of the cervical spine are relatively uncommon, and limited data exist on their management [5].

Solitary plasmacytoma is a rare condition with a cumulative incidence of 0.15 per 100,000 individuals. Only 30% of cases present as EMP [6–8]. Diagnosing EMP involves a tissue biopsy showing an isolated mass of monoclonal plasma cells in the soft tissue, and the absence of hypercalcemia, renal insufficiency, anemia, and bone lesions (CRAB). It is typically detected using positron emission tomography/computed tomography (PET/CT) or CT with contrast and is treated with moderate to high doses of radiotherapy with or without surgical intervention. Adjunctive chemotherapy remains a controversial treatment for SP and is not recommended

[8–11]. Historically, EMP has a significantly higher overall survival and progression-free survival when compared to SBP, and significantly lower rates of transition into multiple myeloma [2, 4].

Here we discuss the case of a 53-year-old male with an extramedullary plasmacytoma presenting as a rapidly growing lateral neck mass.

CASE REPORT

History

The patient is a 53-year-old male who presented to the Emergency Department of a tertiary academic medical center with a 4-week history of an enlarging left neck mass. The mass was first noticed after the patient sustained a mechanical fall at home and subsequently began to experience an increasingly painful region on the left side of his mouth without any limitation in range of motion. The pain was described as a dull ache, originating from inside of his mouth and around his left temple and jaw, and behind his left ear. The patient endorsed having had a dry cough with intermittent vomiting over the antecedent few months, along with an approximate 30-pound weight loss over a 5-month period. He also noticed increasing fatigue over the same time period but denied any other constitutional symptoms. His past medical history was significant for congestive heart failure, diabetes, hypertension, and hyperlipidemia, and his past surgical history was significant for a previous lumbar spine decompression and fusion. He denied smoking or illicit drug use but admitted to occasional alcohol use. He was admitted to the family medicine service.

Imaging was obtained including a contrast enhanced CT of the head and neck, and a non-contrast CT of the cervical spine. These images revealed a large left-sided soft tissue mass approximately 4 × 3 × 7 cm in dimension extending cephalad from the level of C4 to the base of the skull (Figures 1 and 2). There was considerable erosion of the clivus, the medial aspect of the mastoid process, the occiput, and the C1 and C2 vertebrae (Figures 3 and 4). The mass also extended into the left side of the spinal canal. Neurosurgical consultation was requested.

Examination

On physical exam a 2.5 cm × 3 cm firm, non-tender, non-mobile mass was palpated on the left lateral side of the neck at the angle of the mandible. The mass extended behind the ear. No focal neurologic deficits were noted. A thorough discussion with the patient and his family detailed the extent of the mass, and its' likely etiology as a tumor of undetermined origin and further oncologic work-up was recommended. Additionally, because of the gradually worsening neck pain, and imaging suggestive of aggressive erosion of the base of the skull and upper cervical vertebrae by tumor, a two-stage surgical procedure was also recommended. The initial procedure

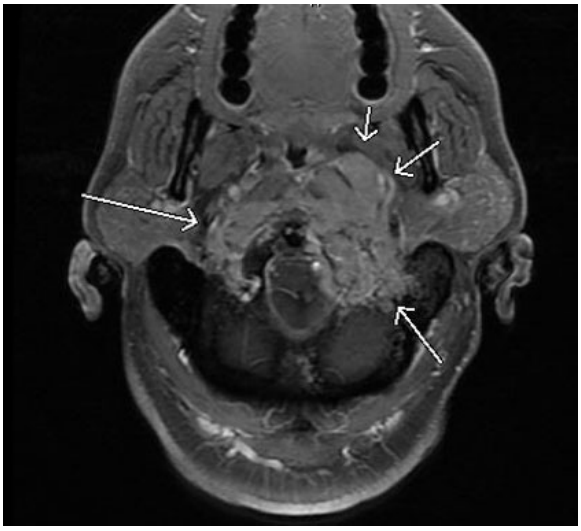

Figure 1: Preoperative axial computed tomography (CT) image of cervical spine demonstrating soft tissue mass.

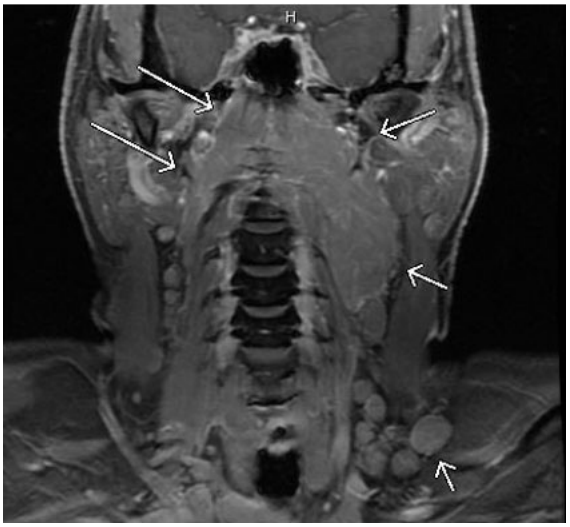

Figure 2: Preoperative coronal CT image of cervical spine demonstrating soft tissue mass.

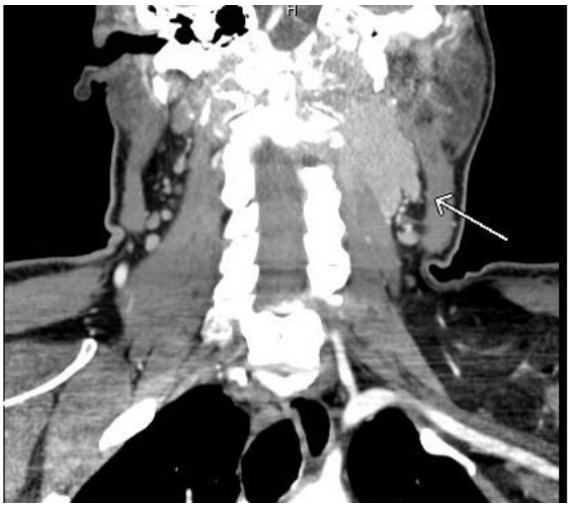

Figure 3: Preoperative coronal CT image of cervical spine demonstrating osteolytic invasion of soft tissue mass.

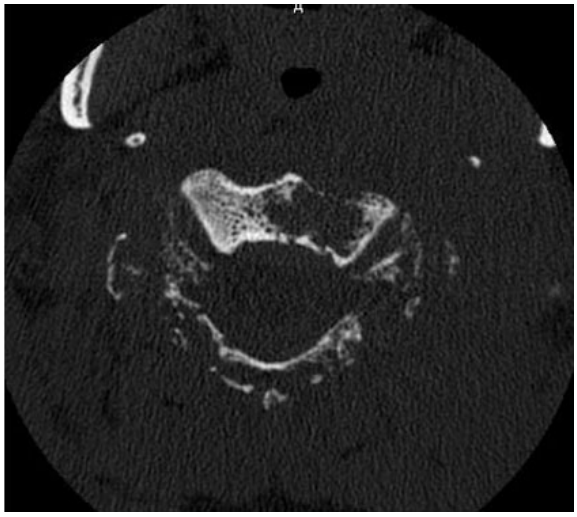

Figure 4: Preoperative axial CT image of cervical spine demonstrating osteolytic erosion of vertebral body by soft tissue mass.

described was a posterior cervical decompression, biopsy, and/or resection of the epidural component of the tumor, accompanied by spinal stabilization with instrumentation. The second stage, depending on pathology, would be resection of the soft tissue neck mass by the otolaryngology service. All preoperative labs were unremarkable.

Operation

Spinal stabilization with an occipital to C6 instrumented fusion was performed prior to spinal cord decompression. Under fluoroscopic visualization, 3.5 × 10 mm lateral mass screws were placed bilaterally at C3, C4, C5, and C6 utilizing the Magerl technique. An occipital plate was affixed utilizing two 8 mm screws lateral to the midline and a 12-mm screw in midline (Figures 5 and 6). Bilateral angulating rods were contoured and appropriately placed into the screw and plate heads respectively. Locking set nuts were placed and torqued to their appropriate final torquing tensions. Allograft and bone morphogenic protein (BMP) were utilized to augment the fusion. After occipital-cervical stabilization has been achieved, C1 and C2 posterior cervical laminectomies were performed to decompress the spinal cord. Tumor was noted to be densely adherent to the cervical dura on the left side of the spinal cord. No attempt was made to dissect it from the dura due to the potential risk of spinal cord injury from manipulation, and the risk of a dural tear leading to cerebrospinal fluid leakage. Likewise, no attempt was made to dissect tumor from the walls of the left vertebral artery since the right vertebral artery was noted to be atretic on preoperative imaging thereby rendering the left vertebral artery as the predominant blood supply to the posterior circulation. All tissue that was resected was sent to pathology including bone, muscle, and soft tissue. The patient recovered well in the post-anesthesia care unit and was later transferred to the floor.

Pathological findings

The final surgical pathology revealed CD38+ clonal plasma cells with cytoplasmic Kappa light chain restriction. The tumor was negative for CD19 or CD20 (Figure 7).

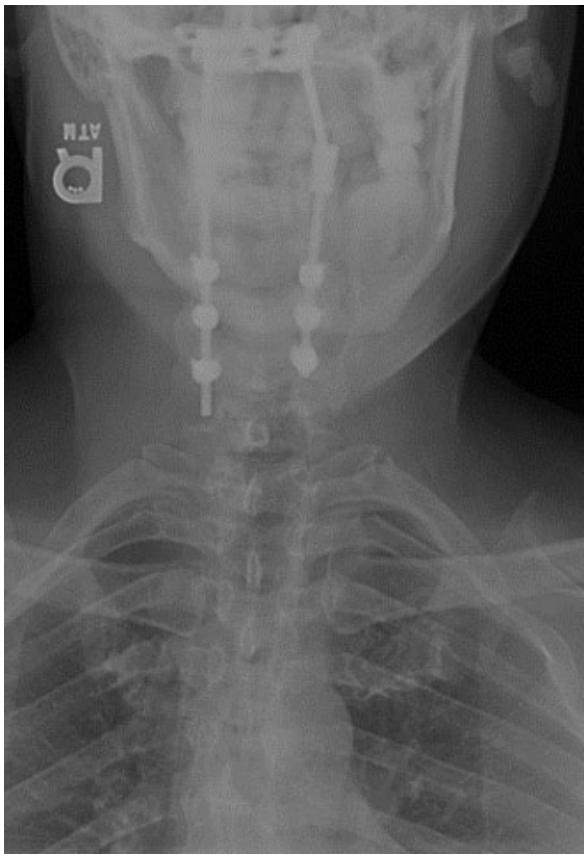

Figure 5: Postoperative anteroposterior (AP) radiograph of cervical spine demonstrating occipital-cervical fusion with instrumentation.

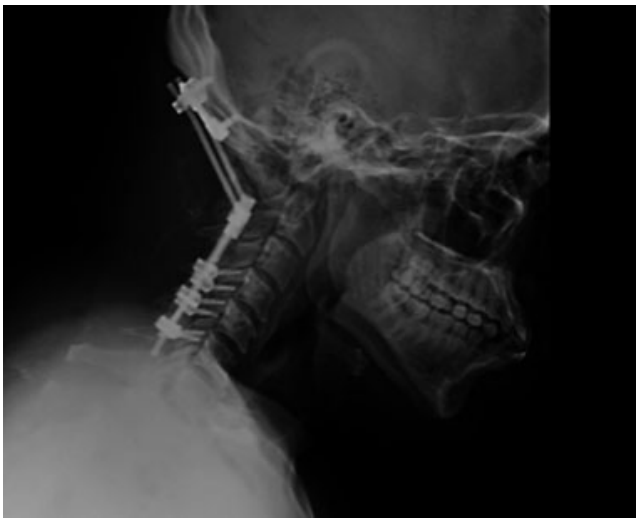

Figure 6: Postoperative lateral radiograph of cervical spine demonstrating occipital-cervical fusion with instrumentation.

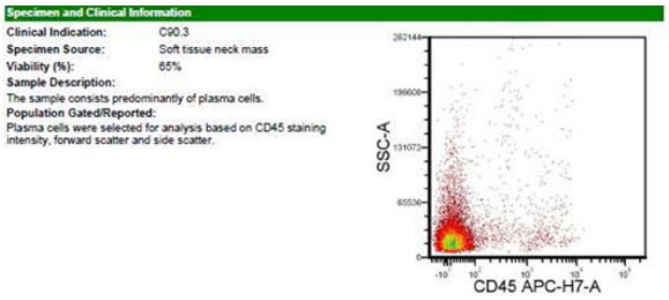

Figure 7: Flow cytometry from sampled surgically biopsied tissue demonstrating predominant plasma cells.

Postoperative course

The radiation therapy and medical oncology services were consulted, and adjuvant therapy was outlined. After the surgical wound was appropriately healed, a total of 3000 cGy of radiation in 200 cGy fractions was given to the region of the tumor, however, there was no resultant decrease in tumor size. Chemotherapy with 2.8 mg intravenous (IV) Bortezomib and Revlimid was subsequently initiated. Most recent follow-up at 29 months indicated that the patient's surgical scar had healed well, and there were no symptoms of head or neck pain or occipito-spinal instability. The left lateral neck mass had decreased in size, and the patient had significant clinical improvement. Unfortunately the patient has since passed from an unrelated cause.

DISCUSSION

Extramedullary plasmacytomas (EMPs) are a rare form of n-on-Hodgkin's lymphoma characterized by the local expansion of plasma cells into the soft tissue. These tumors comprise approximately 3% of all plasma cell neoplasms and arise in the aerodigestive tract in over 80% of documented cases in the literature [3, 12]. Cases of soft tissue cervical masses have not been reported to the authors' knowledge. The literature shows that EMPs affect males almost twice as much as females, with an average age of 55 years at the time of diagnosis [13]. The EMP is distinguished from other plasma cell tumors by the absence of systemic signs. This excludes the possibility of multiple myeloma, the most common plasma cell neoplasm. Extramedullary plasmacytomas are most definitively differentiated from other plasma cell tumors or lymphomas by a positive CD38 and monoclonal cytoplasmic light chain expression of malignant plasma cells obtained by surgical or needle biopsy, as in our patient [14].

Tumors of the neck which erode the bony cervical spine or invade the spinal canal can have devastating neurologic consequences. Bony collapse, and cord compression can result in significant neurologic compromise. Awareness of the symptoms of presentation: pain, progressive motor and sensory deficits, and myelopathy enhances the ability to make an early diagnosis and intervene surgically before catastrophic neurologic consequences occur [15]. Surgical techniques can vary based on the anatomic location and extent of local destruction caused by the neoplasm. This patient's soft tissue tumor was sub-totally resected to relieve spinal cord compression and provide tissue for diagnosis. Because of the immense potential for occipital-atlanto-axial instability, stabilization was accomplished with instrumented arthrodesis. Ultimately postoperative adjuvant therapy was also given. At our most recent follow-up, the patient had significant resolution of his pain as well as a decrease in his tumor burden. Extramedullary plasmacytomas are difficult to treat, however, multi-disciplinary management should be the paradigm used to provide optimal outcome.

CONCLUSION

This case represents the unusual presentation of an extramedullary plasmacytoma to the soft tissues of the neck with subsequent erosion of the base of the skull and upper cervical vertebral spine. A satisfactory therapeutic response was obtained with surgical stabilization followed by adjuvant radiation and chemotherapy.

REFERENCES

1. Kilciksiz S, Karakoyun-Celik O, Agaoglu FY, Haydaroglu A. A review for solitary plasmacytoma of bone and extramedullary plasmacytoma. Scientific World Journal 2012;2012:895765.
2. Finsinger P, Grammatico S, Chisini M, Piciocchi A, Foà R, Petrucci MT. Clinical features and prognostic factors in solitary plasmacytoma. Br J Haematol 2016;172(4):554–60.
3. Luh SP, Lai YS, Tsai CH, Tsao TCY. Extramedullary plasmacytoma (EMP): Report of a case manifested as a mediastinal mass and multiple pulmonary nodules and review of literature. World J Surg Oncol 2007;5:123.
4. Oertel M, Elsayad K, Kroeger KJ, et al. Impact of radiation dose on local control and survival in extramedullary head and neck plasmacytoma. Radiat Oncol 2019;14(1):63.
5. Baba H, Maezawa Y, Furusawa N, et al. Solitary plasmacytoma of the spine associated with neurological complications. Spinal Cord 1998;36(7):470–5.
6. Sato K, Fumimoto S, Fukada T, et al. Extramedullary plasmacytoma arising from the anterior mediastinum. Ann Thorac Surg 2017;103(5):e393–5.
7. Bladé J, Fernández de Larrea C, Rosiñol L, Cibeira MT, Jiménez R, Powles R. Soft-tissue plasmacytomas in multiple myeloma: Incidence, mechanisms of extramedullary spread, and treatment approach. J Clin Oncol 2011;29(28):3805–12.
8. Caers J, Paiva B, Zamagni E, et al. Diagnosis, treatment, and response assessment in solitary plasmacytoma: Updated recommendations from a European Expert Panel. J Hematol Oncol 2018;11(1):10.

9. D'Aguillo C, Soni RS, Gordhan C, Liu JK, Baredes S, Eloy JA. Sinonasal extramedullary plasmacytoma: A systematic review of 175 patients. Int Forum Allergy Rhinol 2014;4(2):156–63.
10. Wen G, Wang W, Zhang Y, Niu S, Li Q, Li Y. Management of extramedullary plasmacytoma: Role of radiotherapy and prognostic factor analysis in 55 patients. Chin J Cancer Res 2017;29(5):438–46.
11. Tsang RW, Campbell BA, Goda JS, et al. Radiation therapy for solitary plasmacytoma and multiple myeloma: Guidelines from the International Lymphoma Radiation Oncology Group. Int J Radiat Oncol Biol Phys 2018;101(4):794–808.
12. Bartl R, Frisch B, Fateh-Moghadam A, Kettner G, Jaeger K, Sommerfeld W. Histologic classification and staging of multiple myeloma. A retrospective and prospective study of 674 cases. Am J Clin Pathol 1987;87(3):342–55.
13. Galieni P, Cavo M, Pulsoni A, et al. Clinical outcome of extramedullary plasmacytoma. Haematologica 2000;85(1):47–51.
14. Dimopoulos MA, Kiamouris C, Moulopoulos LA. Solitary plasmacytoma of bone and extramedullary plasmacytoma. Hematol Oncol Clin North Am 1999;13(6):1249–57.
15. Pashayan R, Cavanaugh WM, Warshel CD, Payne DR. Plasmacytoma of the cervical spine: A case study. J Chiropr Med 2017;16(2):170–4.

****

Author Contributions

Christopher G Salib – Conception of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Cierra N Harper – Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Stephanie Carter – Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Therlinsie Fleurizard – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Damirez Fossett – Conception of the work, Design of the work, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.