Combined Treatment Approach for Rare Occipito-Cervical Spinal Lesions: Leiomyosarcoma

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Abstract
Leiomyosarcoma, a rare malignant tumor arising from smooth muscle, has rarely been reported to occur in association with spinal involvement. The treatment approach depends on tumor characteristics, staging, and presenting symptoms, and may include surgery with or without adjuvant radiation and chemotherapy. We present a case of primary leiomyosarcoma at the craniocervical junction in a 65 year old female treated by suboccipital craniectomy for tumor debulking followed by proton radiotherapy. The approach included a laminectomy of C1 and the superior aspect of C2 and accomplished the primary goal of sufficiently reducing the tumor volume. This permitted proton radiotherapy to the residual tumor while minimizing risk to the adjacent spinal cord. While rare, leiomyosarcoma of the vertebral column must be kept in the differential diagnosis of a craniocervical junction tumor and requires a multidisciplinary treatment approach to optimize outcomes.

Keywords
Leiomyosarcoma; Tumor; Craniocervical; Radiation; Proton

Introduction
Leiomyosarcoma is a malignant tumor of smooth muscle that typically presents in mid-to-late adulthood. Common primary sites include the retroperitoneum, abdominal cavity, and subcutaneous tissue of the extremities. Primary leiomyosarcoma of the bone is uncommon, with the majority of reported cases arising from the femur and tibia [1]. First reported by Lo et al. [2] primary lesions of the vertebrae are exceedingly rare. When occurring, primary or metastatic lesions of the vertebral column may be destructive and are a potential cause of pathologic fracture [3-6]. Neurologic manifestations may result from fracture deformity or from mass effect due to local invasion of tumors arising from the paraspinous musculature [7-9].

There is a paucity of published data on the optimum treatment of primary spinal leiomyosarcoma. Ziewacz et al. demonstrated that surgical intervention may improve pain and neurologic function in a series of eight patients undergoing decompression of metastatic leiomyosarcoma to the spine [10]. Adjuvant radiation and/or chemotherapy may help prevent local recurrence and enhance overall survival [8]. While gross total resection is preferred when possible, [11] this approach may result in excessive morbidity at the craniocervical junction due to adjacent neurovascular structures. This report describes a case of primary leiomyosarcoma at the craniocervical junction extending from the occiput to the superior portion of C2 with successful short-term results following subtotal surgical resection and postoperative proton radiotherapy.

Case Report
A 65-year-old female presented to an outside institution with the complaint of bothersome palpable soft-tissue mass on the posterior neck. The mass was initially painless, but had doubled in size and become painful over the course of approximately three months. Physical examination revealed a soft, fixed mass in the soft-tissues of the posterior neck; no neurologic deficit was present. Magnetic resonance imaging (MRI) obtained on presentation demonstrated a 5.3 x 3.0 x 4.9cm gadolinium-enhancing mass in the suboccipital region extending to the dural margin in the posterior fossa with invasion of the right inferior occipital bone. Inferiorly, the mass encased the posterior arch of C1 and abutted the superior aspect of the arch of C2 (Figure 1). At the outside institution an open biopsy of the lesion was performed, with initial pathologic findings suggestive of a malignant peripheral nerve sheath tumor. After consultation with medical and radiation oncology, the patient was subsequently referred to our institution for definitive treatment.

The treatment options were discussed in detail with the patient. She was counseled that wide en bloc resection theoretically would result in optimal survival and disease control; however, the morbidity of this approach was prohibitive given the location and infiltrative nature of the tumor. The patient subsequently elected to undergo suboccipital craniectomy for debulking of the tumor (Figure 2). The patient was positioned prone in a Mayfield head holder, and skin incision was made through the prior surgical site. The medial and superior aspects of the tumor were exposed and resected from the occiput to C2 in order to create the largest interval between residual tumor and neural elements. Resection goal was to minimize neural tissue exposure to the adjuvant radiation field. The lateral margin of the tumor, extending to the skull base, was not resected. Normal-waveform cerebral oximetry, noninvasive

Figure 1: Sagittal and axial T1-weighted images post-gadolinium administration demonstrating a 5.3 X 3.0 X 4.9cm mass at the craniocervical junction eroding the posterior arch of C1.
Discussion

Prior reports of treatment of osseous spinal leiomyosarcoma have appeared concerned metastatic lesions or primary lesions of the subaxial spine [5,6]. Sucu et al. reported on treatment of a primary leiomyosarcoma of C2 with partial spondylectomy, followed by anterior reconstruction with an expandable implant and posterior occipitocervical fusion with instrumentation [12]. The authors reported a good clinical outcome without evidence of local recurrence at one year postoperatively. The use of multimodal treatment with adjuvant radiation therapy has been reported on two prior occasions for cervical paraspinal leiomyosarcoma [8,13]. Lehman et al. reported local control and long-term survival following treatment of high-grade leiomyosarcoma of the cervical paraspinal musculature with metastases to the lungs and pancreas, following surgery, intensity-modulated radiation therapy, and chemotherapy [8].

In our case, complete resection of the tumor was deemed excessively morbid due to anatomic considerations. The goal of the resection was to debulk the tumor and decompress the neural elements while increasing the margins surrounding organs at risk for radiation-induced tissue damage, specifically neural tissue. The use of postoperative proton radiotherapy allowed for maximum dose delivery to the tumor site despite the proximity of the spinal cord. By minimizing the extent of bony resection, we avoided destabilization of the craniovertebral articulation and the need for occipitocervical fusion. This course of treatment was well-tolerated without evidence of local recurrence or metastasis over the follow-up period and may represent an option for patients with leiomyosarcoma at sites not amenable to complete en bloc resection.

References

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