Extramedullary plasmacytoma of the nasal cavity: Treatment perspective in a developing nation

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Abstract
Extramedullary plasmacytomas are uncommon tumors, with a worldwide annual incidence of 3 per 100,000 population. They account for 1% of all tumors of the head and neck and 4% of all nonepithelial tumors of the nasal tract. A variety of treatment options has been suggested. These treatments vary according to the site of presentation, the presence of locoregional spread, and the histologic picture. Radiotherapy has been widely used as a treatment modality, but little has been written about surgery as a single management modality. However, such an option assumes importance in a developing nation, where patient follow-up is erratic and treatment costs must be kept low. We discuss the feasibility of surgery in such a circumstance, and we describe our surgical treatment of a case of extramedullary plasmacytoma in an elderly woman who presented to our hospital in India.

Introduction
A plasma cell neoplasm represents a monoclonal proliferation of a B cell that has undergone (potentially malignant) transformation into a plasmacytoid cell. The three types of plasma cell neoplasms are multiple myeloma, medullary plasmacytoma, and extramedullary plasmacytoma. Extramedullary plasmacytomas occur in soft tissue outside the medullary bone—hence their name. Because of their rarity, they pose a diagnostic and treatment challenge to ENT surgeons. Treatment of multiple myelomas and medullary plasmacytomas is fairly standard, but such is not the case for extramedullary plasmacytomas. We describe a case of nasal extramedullary plasmacytoma that posed a dilemma in that it featured a number of treatment-limiting factors. These limitations compelled us to research the literature regarding the feasibility of surgery as the sole modality of treatment.

Case report
A 75-year-old woman presented with a 6-month history of right-sided nasal obstruction, a nasal mass, and blood-stained nasal discharge. Examination revealed that a gray-white mass with surface ulceration had filled the entire nasal vestibule. The probe test suggested that the mass was attached to the lateral nasal wall. Findings on posterior rhinoscopy were normal. Computed tomography (CT) showed that the 5 × 4-cm heterogenous mass had originated in the lateral nasal wall; the septum was tumor-free (figure 1). Punch biopsy identified the mass as a plasmacytoma. An undifferentiated carcinoma had been suspected initially, but it was ruled out by immunohistochemistry.

Evaluation of other mucosal surfaces of the upper aerodigestive tract detected no other significant masses. No cervical lymphadenopathy was present. A systemic workup for multiple myeloma included a bone marrow biopsy, bone scan, quantitative immunoglobulin assay, and measurements of hematocrit, white blood cells, blood urea, serum creatinine, serum electrolytes, serum calcium, and Bence Jones proteins in urine. All findings were within normal limits. Hence, a diagnosis of a solitary extramedullary plasmacytoma was established.

The patient's logistical and financial circumstances precluded radiotherapy, and we therefore performed a medial maxillectomy with adequate excision of the tumor margins. Operative findings showed that the tumor had arisen from the middle turbinate. Blood loss during the procedure was minimal, and the patient did not require any transfusion. Her postoperative course was uneventful, and she was discharged on postoperative day 6.

Grossly, the tumor was a gray-white nodular mass that measured 5 × 4 × 1.5 cm. A cut-surface analysis revealed that it was pale brown with foci of congestion. Microscopically, the mass was made up of monotonous sheets of neoplastic plasma cells with eccentric hyperchromatic...
nuclei and irregular chromatin distribution (figure 2, A). A second immunohistochemical stain confirmed that the lesion was not a carcinoma (figure 2, B). Binucleation and multinucleation were present. Microscopy confirmed that all resection margins, including the underlying bone and marrow, were free of tumor.

When last seen 3 years postoperatively, the patient exhibited no signs or symptoms of recurrence or progression to multiple myeloma.

Discussion
Approximately 80% of extramedullary plasmacytomas occur in the head and neck; of these, a similar percentage present in the upper aerodigestive tract and oral cavity.\textsuperscript{1,2}

In this area, most occur in the nose, paranasal sinuses, and nasopharynx (78% of cases).\textsuperscript{1,2} These tumors are three to four times more common in males than in females.\textsuperscript{1} Galieni et al.\textsuperscript{3} established five criteria for diagnosing extramedullary plasmacytoma:

- Biopsy of the tissue must reveal monoclonal plasma cell histology.
- Bone marrow plasma cell infiltration should not exceed 5% of all nucleated cells.
- Osteolytic bone lesions and other tissue involvement must be absent.
- Hypercalcemia and renal failure must be absent.
- A serum M protein concentration, if present, must be low.

Soft-tissue plasmacytomas arise from plasma cells located on the mucosal surfaces. They account for 3% of all plasma cell neoplasms.\textsuperscript{4}

In 1976, Wiltshaw\textsuperscript{5} described a staging system for extramedullary plasmacytomas:

- Stage I disease is defined by the presence of a tumor at only one extramedullary site.
- Stage II disease indicates involvement of regional lymph nodes.
- Stage III disease involves multiple metastases—in which case, the patient by definition no longer has solitary plasmacytoma.

The case described in this article posed a diagnostic dilemma. The physical appearance of the tumor—including...
the surface ulceration, which is rarely seen in plasmacytomas—was characteristic of a malignancy, which is usually the diagnosis for tumors at this site. It was only upon histopathologic examination and immunohistochemistry that we clinched the diagnosis.

Because most patients who present with extramedullary plasmacytoma are elderly and because it has been suggested that this rare tumor be treated as a malignancy, various treatment modalities in various combinations have been tried. The choice of treatment is also influenced by the extent of the tumor. Surgery is recommended for lesions that are localized and can be excised without much morbidity. Endoscopic excision, laser excision, and excision biopsy have been described for nasal and cervical lesions; these procedures are usually combined with adjunctive radiotherapy, which may either precede or follow surgery. One argument against aggressive management is the fact that local control is excellent regardless of tumor size; only 6 to 10% of patients experience a recurrence after adequate initial treatment. Moreover, because most of these tumors in the head and neck are in the vicinity of vital structures, extensive surgery for removal of large tumors with adequate margins would be disfiguring. Therefore, radiotherapy is considered by many the treatment of choice for this radiosensitive tumor.

In choosing the treatment strategy for our patient, we had to consider several factors. She lived in rural India, far from the hospital, and she was of low socioeconomic status. A 50-Gy course of radiotherapy at 2 Gy/day would have required 5 consecutive weeks of treatment and a prolonged hospital stay; the cost of this option was unacceptable to her. Also, the chance that she would be able to comply with a postradiation outpatient follow-up schedule was low.

Endoscopic clearance as a single-modality treatment was ruled out because it would have been very difficult to achieve adequate (1.5 cm) margins and because it is more effective when combined with radiotherapy. And again, regular endoscopic follow-up examinations would have been necessary to look for any recurrence.

Surgery was the only feasible option, and we decided to perform a medial maxillectomy with complete excision of the tumor. Surgical morbidity was minimal, and the patient was discharged within a week. Wound healing was good, and the patient experienced no procedure-related complications. The cost of this treatment was approximately one-half that of radiotherapy, with one-fifth the number of hospitalization days. Because spread to regional lymph nodes is not very common, occurring in 15 to 20% of cases, we did not administer any treatment to the neck.

There is some controversy in the literature over whether extramedullary plasmacytoma represents a neoplastic stage of multiple myeloma or if it is a distinct entity. The former view may hold for solitary plasmacytomas of bone, as 85% of such patients progress to multiple myeloma. However, only 15 to 20% of patients with extramedullary plasmacytomas progress to multiple myeloma or develop regional disease. A rise in baseline levels of serum myeloma monoclonal proteins or urine Bence Jones protein may signify a recurrence of the primary or the onset of multiple myeloma. Therefore, long-term follow-up for locoregional control and to look for progression to multiple myeloma is necessary.

In conclusion, it is our opinion that medial maxillectomy in appropriately selected patients with extramedullary plasmacytoma of the lateral nasal wall is an acceptable treatment. It has significant advantages over radiotherapy in terms of morbidity and cost.

References