Extramedullary plasmacytoma arising from the nasal septum

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Abstract
We report a rare case of extramedullary plasmacytoma of the nasal septum in a 65-year-old woman. She presented with a 2-month history of left-sided nasal obstruction and intermittent blood-tinged nasal crusting. Nasal endoscopy revealed that a dark-red mass had arisen from the nasal septum; no evidence of invasion to adjacent tissues was seen. A biopsy specimen was diagnosed as a plasmacytoma (kappa light chain–type). Serum and urine electrophoresis failed to detect any myeloma component or Bence Jones protein. All other screening tests to rule out multiple myeloma were negative. These findings confirmed the diagnosis of extramedullary plasmacytoma. The mass was completely removed via an endoscopic approach. No recurrence was noted at the 2-year follow-up.

Introduction
Plasmacytomas are malignant tumors of monoclonal plasma cells. They originate in either bone (solitary osseous plasmacytoma) or in soft tissue (extramedullary plasmacytoma). Extramedullary plasmacytoma is a rare localized tumor. It has no characteristics of multiple myeloma, but the development of multiple myeloma has been observed in 8 to 36% of patients. Nearly 80% of all extramedullary plasmacytomas occur in the head and neck; most of these arise in the upper aerodigestive tract, probably because of the abundance of lymphatic tissue in this area. Extramedullary plasmacytoma represents less than 1% of all head and neck malignancies, and it accounts for 4% of all nonepithelial tumors of the nasal cavity, nasopharynx, and paranasal sinuses. We describe a case of extramedullary plasmacytoma that arose from the nasal septum.

Case report
A 65-year-old woman presented to our clinic with a 2-month history of left-sided nasal obstruction and intermittent blood-tinged nasal crusting. Nasal endoscopy detected a dark-red mass on the nasal septum (figure 1). No cervical lymph nodes were palpable. Computed tomography (CT) of the nasal cavities and paranasal sinuses showed that the mass protruded from the middle part of the septum but did not involve the underlying septal framework (figure 2). Histopathologic examination of the biopsy specimen revealed that the neoplasm exhibited a uniform population of plasma cells. On immunohistochemistry by the biotin-avidin immunoenzyme technique, these cells stained positive only for the kappa light chain (figure 3). Based on these findings, we presumptively diagnosed the tumor as a plasmacytoma.

Further clinical examinations were undertaken to look for multiple myeloma. These investigations included a complete blood cell count, measurement of serum calcium and creatinine levels, serum protein electrophoresis and immunoelectrophoresis, urinalysis for Bence Jones protein, evaluation of β2-microglobulin, a skeletal survey with plain films (skull, ribs, humeri, and femurs), abdominal ultrasonography, bone marrow aspiration, and biopsy. All findings were within normal limits. Accordingly, we ruled out multiple myeloma and confirmed a final diagnosis of primary extramedullary plasmacytoma of the nasal septum.

The septal mass was completely removed via an endonasal endoscopic approach. The mass was easily stripped from the underlying cartilage and bony portion of the nasal septum. At the 2-year follow-up, the patient exhibited no evidence of recurrence.

Discussion
The plasma cell dyscrasias are characterized by the clonal proliferation of plasma cells that produce a homogenous immunoglobulin protein. The major plasma cell dyscrasias are (1) multiple myeloma, (2) monoclonal gammopathy of unknown significance, (3) solitary plasmacytoma of bone, (4) extramedullary plasmacytoma, and (5) heavy-chain disorders.
EXTRAMEDULLARY PLASMACYTOMA ARISING FROM THE NASAL SEPTUM

Extramedullary plasmacytoma is a rare soft-tissue malignant neoplasm that is made up of monoclonal plasma cells. It can be either primary (without evidence of disease in other foci) or part of a systemic process during the course of multiple myeloma.

The first case of extramedullary plasmacytoma was reported in 1905 by Schridde. Between then and 1997, more than 400 articles addressing extramedullary plasmacytoma were published. In 1999, Alexiou et al reviewed all previous reports of extramedullary plasmacytoma and found 869 cases; 714 (82.2%) of them had occurred in the upper aerodigestive tract. The most frequently affected areas in the upper aerodigestive tract were the nasal cavity or paranasal sinuses (43.8%), followed by the nasopharynx (18.3%), the oropharynx (17.8%), and the larynx (11.1%).

Extramedullary plasmacytomas more commonly occur in males (ratio: 4 to 1), and 95% of cases occur in patients older than 40 years. Extramedullary plasmacytoma of the upper aerodigestive tract usually presents as a submucosal, reddish, sessile or polypoid tumor that is rarely ulcerated. Most occur as single lesions; 10 to 20% of cases feature multiple lesions.

The clinical presentation is primarily a function of the mass effect and varies according to the site of involvement. Because most of these lesions arise in the sinonasal or nasopharyngeal area, the most common symptoms are a nasal mass, nasal obstruction, and epistaxis. Other reported presenting symptoms include nasal discharge, pain, proptosis, cervical lymphadenopathy, and cranial nerve palsy.

The diagnosis of extramedullary plasmacytoma usually follows histologic examination. A diffuse or sheetlike proliferation of plasma cells of varying maturity and atypia is seen. Also, an eccentrically placed nucleus and a perinuclear hof (owing to a prominent Golgi apparatus) can be seen. Immunohistochemical staining will demonstrate the monoclonal nature of the plasma cells and confirm the neoplastic nature of the lesion. In addition, immunohistochemical study is used to differentiate extramedullary plasmacytoma from benign reactive plasmacytosis as well as other malignant disorders, such as undifferentiated carcinoma, melanoma, and esthesioneuroblastoma.

When a plasmacytoma is confirmed histologically, secondary diagnostic procedures must be carried out to exclude systemic involvement. This workup may include

Figure 1. Preoperative nasal endoscopy shows the red polypoid mass on the left side of the nasal septum. The mass extends to the inferior turbinate, but there is no sign of invasion into it.

Figure 2. Preoperative coronal (A) and axial (B) CTs show the well-margined soft-tissue mass in the middle portion of the nasal septum. No erosion of the underlying bone or cartilage is evident.
a complete blood count; measurement of calcium, serum urea nitrogen, and creatinine levels; serum and urine electrophoresis with immunofixation; a complete skeletal radiographic survey; and a bone marrow biopsy.

Although several reports regarding extramedullary plasmacytoma in the nasal cavity have been published, a finding of such a lesion in the nasal septum is quite rare. Malignant tumors of the nasal septum (including extramedullary plasmacytoma) are treated by surgery and/or radiotherapy.\(^\text{14}\) Although extramedullary plasmacytoma is known to be radiosensitive, most authors recommend a combination of surgery and radiotherapy for nasal cavity lesions.\(^\text{15-19}\)

The lesion in our patient was localized in the nasal septum, and there was no sign of invasion into the underlying bone or cartilage. Therefore, after we removed the tumor (while preserving the septal framework), we did not consider postoperative radiotherapy.

Local recurrence has been reported to occur in 8 to 30% of adequately treated cases.\(^\text{2,20}\) Conversion of extramedullary plasmacytoma to multiple myeloma has been reported in 8 to 36% of cases within 3 to 61 months.\(^\text{2,3}\) Therefore, long-term follow-up, including both local and systemic surveillance, is necessary. CT and measurements of serum immunoglobulin and urinary Bence Jones protein levels may be useful in detecting recurrence or conversion to multiple myeloma.

References