Malignant peripheral nerve sheath tumor of the paranasal sinuses

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Abstract
Malignant peripheral nerve sheath tumors of the nose and paranasal sinuses are extremely uncommon. We report the case of a 65-year-old woman who presented with a rapid emergence of "nasal polyps" that completely obstructed her nasal passages. Six years earlier, she had been assessed elsewhere for nasal polyps. At presentation, the patient exhibited gross polyposis, with lesions protruding from both nostrils. Histology confirmed a diagnosis of a malignant peripheral nerve sheath tumor. This case demonstrates that symptoms of nasal obstruction are not always secondary to simple causes. We discuss the clinical picture of nasal and paranasal malignant peripheral nerve sheath tumor, its pathology, and its treatment.

Introduction
Malignant peripheral nerve sheath tumor—also known as neurogenic sarcoma, malignant neurilemoma, malignant schwannoma, and neurofibrosarcoma—is an uncommon neoplasm that originates in Schwann's cells of the nerve sheath. Primary neurogenic tumors of the nose and paranasal sinuses are rare, accounting for only 4% of all neural tumors of the head and neck.

In this article, we describe a case of malignant peripheral nerve sheath tumor of the paranasal sinuses, and we review the clinical presentation, histologic findings, differential diagnosis, and treatment of this tumor.

Case report
A 65-year-old woman presented to our department in May 2002 with rapidly developing "nasal polyps" that completely obstructed her nasal passages. The previous year, she had been diagnosed with metastatic colon adenocarcinoma. In 1996, she had been assessed elsewhere for nasal polyps when she complained of right nasal obstruction. Examination under anesthesia at that time had revealed that her right nasal cavity was occluded by a firm, fleshy growth. Histology of biopsy specimens suggested that she had a low-grade malignant peripheral nerve sheath tumor. Unfortunately, the patient did not return for further investigation and follow-up until she presented to our department 6 years later.

Our examination revealed that the patient had large polyps protruding from both nostrils (figure 1). She reported that the lesions occasionally bled. She also had right-sided otitis media with effusion. Computed tomography (CT) of the sinuses revealed that an expansile soft-tissue mass had filled the right nasal cavity and extended back to the choana and forward to the nares (figure 2). The nasal septum was perforated anteroinferiorly. The osteomeatal unit on the right was markedly enlarged, and all of the right-sided sinus groups were completely opaque. The left sphenoid sinus was also involved. No orbital or intracranial extension was observed. Biopsy confirmed that the mass represented a recurrence of the low-grade malignant peripheral nerve sheath tumor.

The patient underwent a right medial maxillectomy via a Denker extended maxillary anterior antrostomy approach. The tumor was visualized in the antrum extending up to and throughout the nasal cavity (figure 3). The tumor was removed along with the lateral nasal wall, which was incised from below the inferior turbinate to above the middle turbinate and anteriorly at the nasal valve area. The nasal cavity was packed with bismuth iodoform paraffin paste; the packing was removed 2 weeks later with the patient under general anesthesia.

On histologic examination, the excised specimen was identified as a cellular spindle-cell lesion with small plump nuclei and a few larger pleomorphic cells (figure 4). In addition, prominent vascularity and many mast cells were seen. The spindle cells exhibited no particular architectural pattern. Immunohistochemistry studies demonstrated focal
positivity in the spindle cells with desmin, smooth-muscle actin, and vimentin. A few areas of focal positivity for S-100 and MIB-1, a marker for proliferation, were also seen.

The patient returned for follow-up 6 months postoperatively and was progressing well with no complications. Her airways were patent and her sense of smell had returned to normal. However, she died 1 month later from the metastatic colonic adenocarcinoma.

Discussion

Most nerve sheath tumors of the nose and paranasal sinuses arise from the ophthalmic and maxillary branches of the trigeminal nerve and its terminal branches, although it is often difficult to identify exactly which nerve is involved.\(^2\) The most common sites of these tumors are the ethmoid and maxillary sinuses.\(^3\) Other sites are the intranasal cavity and the sphenoid sinus. The frontal sinus is the least common site.

Malignant peripheral nerve sheath tumors can arise de novo or as a result of the malignant transformation of a neurofibroma (von Recklinghausen’s disease).\(^4,5\) Cases of this tumor have been reported in all age groups from children to the elderly, but the peak incidence occurs between the second and fourth decades of life; the tumor has no predilection for sex and race.\(^6\)

**Clinical presentation.** The clinical presentation differs according to the site involved, but the most common symptoms are unilateral nasal obstruction, epistaxis, pain in the facial region, and swelling of the facial and orbital region.\(^7\) Other presenting complaints include mucopurulent rhinorrhea, hyposmia, and headache.

Malignant peripheral nerve sheath tumors can be aggressive, and they have a tendency to erode into adjacent bone and soft tissue.\(^8,9\) Bony erosion did not occur in our patient. The primary mode of tumor spread is hematogenous. Pulmonary metastasis is common.

**Histologic findings.** Histologically, these tumors have no defined,classic appearance. Commonly described findings are the presence of spindle cells with a high mitotic rate and indistinct cytoplasmic borders arranged in bundles or fascicles.\(^7\) Immunohistochemistry plays an important part in diagnosis and in excluding fibrosarcoma, synovial sarcoma, and fibrous histiocytoma. Malignant peripheral nerve sheath tumors are immunoreactive for S-100 protein and vimentin. They are negative for neurofilament antibody and epithelial membrane antigen.

**Immunohistochemistry**

- **S-100 protein** is a calcium-binding protein found in nerve sheath cells and melanocytes. It is a sensitive marker for the diagnosis of peripheral nerve sheath tumors.
- **Vimentin** is a cytoskeletal protein found in many types of connective tissue and is a useful marker for the diagnosis of sarcomas. It is often expressed in sarcomas arising from the sympathetic nervous system.
- **Desmin** is a muscle-specific actin-binding protein found in smooth muscle, cardiac muscle, and skeletal muscle. It is a useful marker for the diagnosis of smooth muscle tumors.
- **Smooth-muscle actin** is a muscle-specific actin-binding protein found in smooth muscle cells. It is a useful marker for the diagnosis of smooth muscle tumors.

**Figure 1.** At presentation, the polyps can be seen protruding from the nostrils.

**Figure 2.** Preoperative coronal CT shows the extent of the mass.

**Figure 3.** Intraoperatively, the tumor is seen in the antrum.
nerve sheath tumors specifically demonstrate S-100 positivity. They are immunoreactive for vimentin and not immunoreactive for HMB-45.

**Differential diagnosis.** There are several differential diagnoses for malignant peripheral nerve sheath tumors of the nose and paranasal sinuses—nasal polyps, mucoceles, gliomas, papillomas, esthesioneuroblastomas, sarcomas, carcinomas, and lymphomas. To reach a definitive diagnosis, biopsy of the specimen must be performed. Obtaining a biopsy can be complicated by severe bleeding secondary to the extensive vascularization of these tumors. CT and magnetic resonance imaging may not provide a definitive diagnosis, but they can play an important part in assessing, staging, and monitoring the progression of the disease.

**Treatment.** The preferred treatment for malignant peripheral nerve sheath tumors of the nose and paranasal sinuses is wide local excision. For our patient, we opted to perform a medial maxillectomy and sphenoethmoidectomy via a Denker anterior maxillary antrostomy approach. This technique provides excellent exposure and avoids a facial scar. Regional lymph node clearance is unnecessary.

The role of chemotherapy and radiotherapy in the treatment of this tumor is debatable. The tumor has been reported to be radioresistant. Some authors have reported using radiotherapy postoperatively, but there is no clear evidence of definite benefit. Chemotherapy has been used in the treatment of unresectable recurrent disease and metastases.

**Prognosis.** Patients with a malignant peripheral nerve sheath tumor have a poor prognosis, as reported 5-year survival rates range from 50 to 65%. The prognosis is worse in patients who have associated von Recklinghausen’s disease—30% survival at 5 years.

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**References**