

# Schwannomas of the Paranasal Sinuses

## Case Report and Clinicopathologic Analysis

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• Schwann's cell tumors are common neoplasms arising from the neural sheath of autonomic, cranial, or peripheral nerves. We describe a case of destructive malignant schwannoma of the right ethmoid and sphenoid sinuses. The tumor was successfully treated by surgical excision, external radiotherapy, and radon seed implants. The patient remains symptom free 3 years after therapy ended, emphasizing the previously unrecognized benefits of radiotherapy for these types of tumors. The value of endoscopy in diagnosing and following schwannomas of the nasal and paranasal sinuses is highlighted in our review of the clinicopathologic features and radiologic findings of this tumor.

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Schwannomas of the paranasal sinuses are rare. Between 25% and 45% of schwannomas occur in the head and neck region, but only 4% involve the nasal cavity and paranasal sinuses. Perzin and colleagues<sup>1</sup> identified only

six cases of schwannomas among 430 000 patients.

The neoplasms develop from Schwann's cells of the neural sheath, and most are benign, well encapsulated, and amenable to surgical resection. However, the tumors are capable of undergoing malignant transformation. We present a rare case of malignant schwannoma of the ethmoid and sphenoid sinuses and we describe the clinical presentation, differential diagnosis, histologic findings, and treatment of schwannomas of the nasal or paranasal sinuses.

### REPORT OF A CASE

A 63-year-old woman presented to the regional hospital of Jackson, Miss, on March 26, 1987, with a 1-year history of postnasal discharge and right nasal obstruction. No pain or epistaxis were reported. During physical examination, she was found to have a polypoid mass, surrounded by purulent exudate, in the superior-posterior aspect of the right nostril. Indirect nasopharyngoscopy revealed that the mass was protruding into the nasopharynx.

Results of laboratory tests were normal. The computed tomographic (CT) scan of the paranasal sinuses showed diffuse opacification of the right anterior and posterior ethmoid sinuses and of the right sphenoid sinus.

After the diagnosis of right-sided chronic polypoid sinusitis was made, the patient was scheduled for right-sided intranasal polypectomy, ethmoidectomy, and sphenoidectomy on June 6, 1987. During surgery, there was

extensive bleeding after excising part of the polypoid mass. The procedure was terminated, and the nose was packed to stop the bleeding. The physicians awaited the results of the histologic examination before determining a subsequent course of treatment. A diagnosis of malignant schwannoma was made by the Vanderbilt pathology group, after which the patient was referred to the Otolaryngology Consultants of Memphis (Tenn).

Our CT scans of the paranasal sinuses disclosed a destructive mass in the right ethmoid and sphenoid sinuses, consistent with the findings for a malignant tumor. The mass was associated with bone destruction, particularly of the planum sphenoidale in the medial wall of the right orbit (Fig 1). Magnetic resonance imaging confirmed bony destruction by a malignant mass in the right posterior ethmoid and sphenoid sinuses (Fig 2).

Four-vessel carotid angiography showed a hypervascular tumor involving the right ethmoid sinus and nasal cavity. A chest roentgenogram was negative, except for a large superior mediastinal soft-tissue mass, which displaced the trachea posteriorly. This was a previously diagnosed thyroid goiter.

On June 2, 1987, a right-sided extended lateral rhinotomy was performed, removing the tumor from the nose and sinuses. Ligation of the right internal maxillary artery was also performed. The operative findings revealed a polypoid vascular tumor that involved the right ethmoid and sphenoid sinuses. The involved lamina papyracea was subtotally removed. A dehiscence of the posterior sphenoid wall was noticed, and although the frontal sinus was free of tumor,

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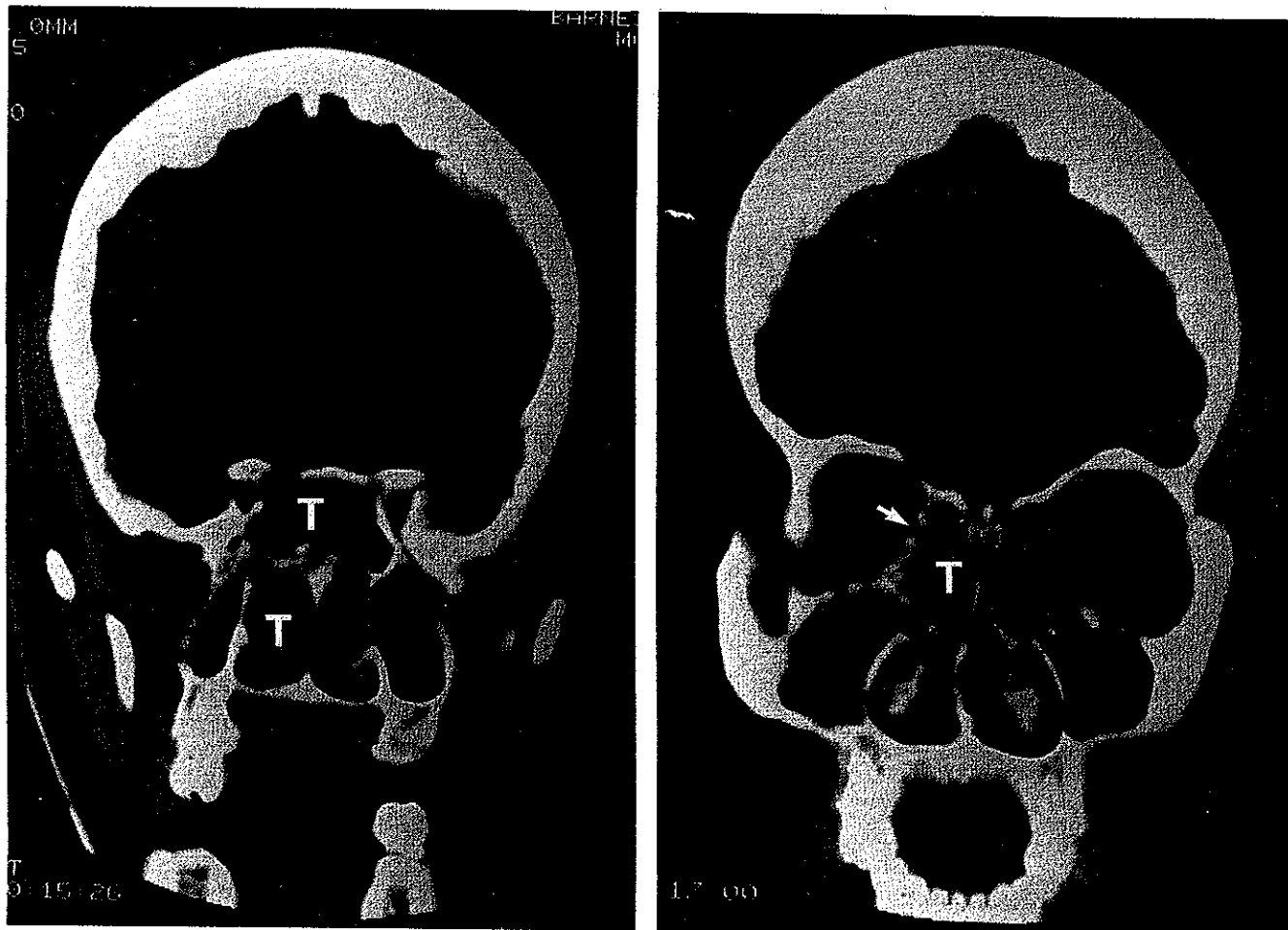


Fig 1.—Left, Coronal computed tomographic scan of the sinuses showing the tumor (T) filling the right ethmoid and sphenoid sinuses and the nasal cavity. Right, Coronal computed tomographic scan of the sinuses showing the tumor (T) and the involvement of the lamina papyracea (arrow).

the floor was partially excised to obtain an appropriate tumor margin. Analysis of the excised tissue confirmed the diagnosis of a malignant schwannoma.

Despite internal maxillary artery ligation, excessive bleeding occurred. Hemostasis was achieved by ligation and electrocautery. After an uncomplicated postoperative course, the patient was discharged 7 days after surgery.

A CT scan of the sinuses performed 3 days after surgery demonstrated a soft-tissue density in the right sphenoid, ethmoid, and maxillary sinuses, with involvement of the right nasal passage. It was difficult to determine whether this represented persistent tumor or a reactive tissue anomaly. After CT scans helped us to determine that that tumor remained in the sphenoid region, the patient was referred for adjuvant radiotherapy.

The patient received a total radiation dose of 64.8 Gy. Six weeks after radiotherapy ended, another sinus endoscopic examination was performed. No tumor could be detected.

However, a biopsy disclosed recurrent or residual tumor, and the patient was referred for implant radiation therapy.

Under direct vision, using a nasal endoscope, an iridium afterload catheter was placed in the involved right sphenoid-ethmoid region. The implant contained 10 seeds of iridium, each of which delivered 4.75 MCi. The implant was left in place for 42.5 hours, delivering a total dose of 25.5 Gy at a distance of 1 cm and 42.5 Gy at 6 mm from the iridium ribbon.

The patient has been followed up closely and remains free of symptoms almost 3 years after therapy.

#### COMMENT

First described in 1908 by Virchow, schwannomas are nerve sheath tumors that develop from the Schwann's cells that are derived from neuroectoderm.<sup>1,2</sup> Batsakis,<sup>2</sup> and Masson<sup>3</sup> who proposed the *schwannian* origin of

nerve sheath tumors, was later supported by the studies of Stout,<sup>4</sup> Del Rio-Hortega,<sup>5</sup> and Fisher and Vuzenski.<sup>6</sup>

Part of the difficulty in classifying and reporting nerve sheath tumors has been the vacillating nomenclature. Benign schwannomas have been called neurilemmas, neurolemomas, neurinomas, and peripheral fibroblastomas.<sup>7</sup> Malignant schwannomas have also been known as malignant neurilemmas, malignant neurinomas, malignant nerve sheath tumors, neurogenic tumors, neurogenic sarcomas, and neurofibrosarcomas.<sup>1</sup>

Primary Schwann's cell tumors arising in the nasal cavity and paranasal sinuses occur infrequently.<sup>1,2,4</sup> Only 4% of the nerve sheath tumors developing in the head and neck region occur in the paranasal sinuses.<sup>2</sup> Conley and

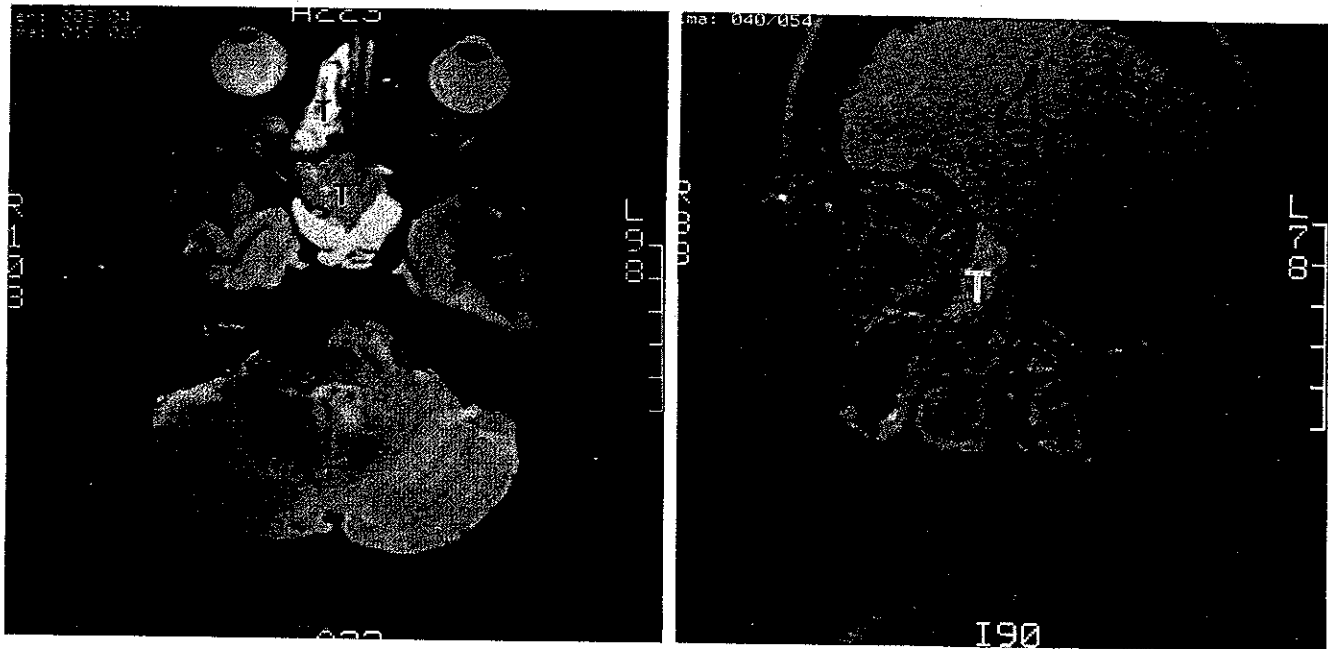


Fig 2.—Magnetic resonance imaging scan of the sinuses. Left, Axial view showing the tumor (T). Right, Coronal view.

Janecka,<sup>8</sup> in a study of 90 patients with schwannomas of the head and neck region, found only eight patients with tumors involving the upper respiratory passages.

The origin of schwannomas of the nasal and paranasal sinuses is presumed to be the sheath of the ophthalmic and maxillary branches of the trigeminal nerve and autonomic ganglia.<sup>7</sup> The olfactory nerve contains no Schwann's cells.<sup>2</sup> However, demonstrating the nerve origin is frequently impossible in these cases.

In descending order of frequency, the sites for schwannomas of the nasal and paranasal cavities are the nasal-ethmoid and maxillary area, intranasal area, sphenoid sinus, and frontal sinus.<sup>2,7,9</sup> Involvement of multiple sinuses is not uncommon. Age, sex, and race have no predictive value.<sup>7,10</sup>

The clinical presentation of a nasal or paranasal schwannoma is indistinguishable from any other tumor in this region. The signs and symptoms usually depend on the location and size of the tumor. Nasal obstruction, epistaxis, mucopurulent rhinorrhea, hyposmia, localized facial swelling, or pain, and proptosis can be caused by the tumor. Epistaxis is usually associated with ethmoid and nasal fossa involvement, and pain is usually experienced

with maxillary sinus tumors.<sup>2,11</sup> During the growth of sphenoid sinus tumors, third, fourth, or sixth cranial nerve palsies may develop.<sup>7</sup> Nonspecific clinical manifestations are produced by tumor necrosis or by impingement of the tumor mass.

The differential diagnosis of schwannomas of the nasal and paranasal sinuses includes mucoceles, gliomas, papillomas, esthesioneuroblastomas, sarcomas, carcinomas, and lymphomas.<sup>7</sup> The diagnosis can only be made by examination of biopsy specimens of tissue. The biopsy is frequently complicated by severe bleeding because of the extensive vascularization of most schwannomas, and the histologic diagnosis of schwannomas can be challenging. The pathologist must differentiate schwannoma from neurofibroma, fibromatosis, fibrosarcoma, fibrous histiocytoma, fibrous dysplasia, ossifying fibroma, and osteosarcoma.

Benign schwannomas are solitary, encapsulated masses that are attached to or surrounded by their presumed nerve of origin.<sup>1</sup> Histologically, they are characterized by organized cellular areas (Antoni A) and disorganized areas (Antoni B) in which myxoid or degenerative changes are often detected.<sup>1,7</sup>

Malignant schwannomas are highly

cellular, with significant epithelioid elements.<sup>1</sup> They are usually composed of numerous, closely packed, spindle-shaped cells that exhibit moderate or marked mitotic activity. Nuclear palisading may also be seen. Rhabdomyosarcomatous, chondrosarcomatous, melanocytic, or epithelioid differentiation may be pronounced. Malignant schwannomas are highly aggressive tumors that can infiltrate locally, and they have a high recurrence rate. Metastasis occurs commonly to the lungs and rarely to the regional lymph nodes.<sup>1,2</sup>

The prognosis is usually poor for malignant schwannoma. In a study of 15 patients with malignant schwannomas during an 18-year period, White<sup>12</sup> found that nine died within the first 20 months. Death is caused by direct intracranial invasion or pulmonary metastasis.

Malignant schwannomas are rare. In a review of 152 patients with benign and malignant neurilemmomas of the head and neck, Kragh and colleagues<sup>13</sup> found that only four of the tumors were malignant (2.6%). None of the malignant tumors arose in the paranasal sinuses. In reviewing patient records from a 70-year period, Perzin and associates<sup>1</sup> retrieved only four malignant schwannomas.

Advising a high index of suspicion for such a rare phenomenon cannot help the average physician. Instead, we recommend careful preoperative assessment for any tumor of the nose and paranasal sinuses.

High-resolution CT with an intravenously administered contrast agent is the best diagnostic modality for identifying the location and extent of intraorbital, intracranial, or soft-tissue involvement. Computed tomography delineates an image of the soft-tissue tumor and simultaneously outlines the skeletal margins well enough to rule out invasion.<sup>7</sup> Schwannomas usually demonstrate a mottled central lucency and peripheral intensification on contrast-enhanced CT scans. The heterogeneous appearance is ascribed to areas of neovascularity or increased vascularity with adjacent nonenhancing cystic or necrotic regions.<sup>7</sup>

Magnetic resonance imaging may be superior in defining soft-tissue tumors, but contrast-enhanced CT can better define intracranial invasion. However, because benign schwannomas can erode bone by pressure, bony erosion is not a criterion for malignancy, and, as our experience with magnetic resonance imaging increases, it may become the preferred modality for identifying these tumors. With either approach, the diagnosis must be confirmed by histologic examination of a tissue specimen.

Radical surgical resection is considered the treatment of choice for malignant schwannomas.<sup>1,2</sup> For nasal and paranasal sinus tumors, the approach is through a lateral rhinotomy incision with external ethmoidectomy. This allows excellent visualization of the sino-nasal region. However, en bloc resection is often impossible, and fractional excision is common. Large sphenoid

lesions may require a stage procedure, such as extradural craniotomy and lateral rhinotomy.<sup>7</sup>

In treating benign schwannomas, functional and cosmetic considerations should be taken into account, but they require careful attention because they usually impinge on critical structures. Treatment of benign tumors is often limited to debulking and to close follow-up, but surgical excision is encouraged if anatomic structures can be preserved.

Schwannomas were formally considered to be radioresistant.<sup>7</sup> This mistaken premise was probably due to the lower maximal tumor doses possible using only orthovoltage equipment and to the protracted regression seen after irradiation of certain neural tumors. In our case, radiotherapy and radon seed implants provided good adjuvant therapy. Although some malignant tissue remained after surgery, the disease has been well controlled for almost 3 years. We believe that radiotherapy, with or without radon seed implants, should be considered in the treatment of all such tumors, and we encourage further studies in this field.

Sinus endoscopy offers a dependable diagnostic and therapeutic tool for sinus tumors. By using sinus endoscopy, we precisely positioned the iridium implant, thus increasing its efficacy. Endoscopy also allowed us to obtain a tissue biopsy and to closely follow-up the patient's recovery.

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#### Editorial Footnote

The strength of this article is the presentation of a surgical option that was successful in managing this particular patient. External radiation and radon seed implantation were employed as a supplement to surgical excision through a lateral rhinotomy approach. The argument could be made that a skull-base approach from inferolaterally offers certain advantages in terms of visualization, access, and vascular control. In any event, we congratulate the authors for their success and for sharing their observations with us.

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