CASE REPORT

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Nasal schwannoma: a case report and clinicopathologic analysis*

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SUMMARY

According to the literature, half of the schwannoma cases occur in the head and neck areas and only less than 4% occur in the sinonasal tract. In this case, a 39-year-old male patient, with a-year-long progressive left side nasal obstruction and purulent rhinorrhea, is presented. The CT reveals a mass filling the left nasal cavity and nasopharyngeal space, with bony erosion of the inferior turbinate and medial maxillary bone. During surgical intervention, the mass is found to originate from the medial side of the left middle turbinate with maxillary sinusitis and inferior turbinate atrophy. The pathological examination reveals a noncapsulated tumor with palisading cellular arrangement and high cellular density. The pathological findings and nervous origin of the tumor are discussed after an extensive review of the literature.

Key words: nasal schwannoma, chronic paranasal sinusitis, nasal polyp

INTRODUCTION

Half of the cases with schwannoma occur in the head and neck region, but less than 4% occur in the sinonasal area. This tumor is derived from the schwann cell, which can be found in many kinds of nerves, including cranial nerves (except olfactory and optic nerves), peripheral nerves, sympathetic and parasympathetic nerves. Clinically, these patients are indicated to have unilateral nasal obstruction, frequent epistaxis, anosmia, and painful sensation. The characteristics of the tumor are polypoid, slow-growing, and encapsulated. With nasal schwannoma, however, some special pathological findings are specific, which are not found in tumors from other regions.

CASE REPORT

The patient was a 39-year-old male aborigine from Taiwan. He visited us because of a progressive left side nasal obstruction with intermittent purulent rhinorrhea for more than a year. No epistaxis, anosmia, or any other nasal symptom was mentioned. He denied any systemic disease and had never undergone any surgery. He was a worker in a dye-stuff factory, smoked one pack of cigarettes per day and chewed betel nuts for over 10 years.

During his visit, under anterior rhinoscopy, a large polypoid mass was noted in his left nostril, which occupied the left common meatus. Some yellowish mucopus was also found in the left nasal cavity. Posterior rhinoscopy revealed a large polypoid mass protruding from the left choana into the nasopharynx. A CT scan of the paranasal sinus showed a large soft tissue mass in the left nasal cavity with protrusion into the nasopharynx (Figures 1 and 2). Bony erosion of the nasal turbinates and

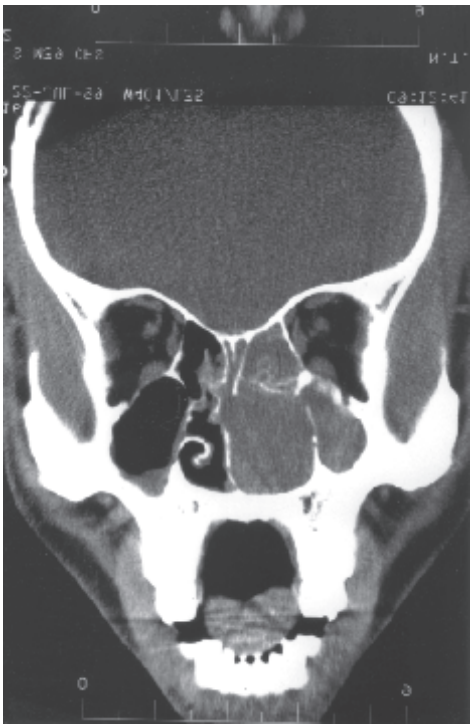

Figure 1. CT, large soft tissue mass in the left nasal cavity with bony erosion of inferior and middle turbinates and medial wall of maxillary sinus. Nasal septum deviated to right side due to mass effect of tumor. Total opacity in right maxillary and ethmoid sinus was noted.

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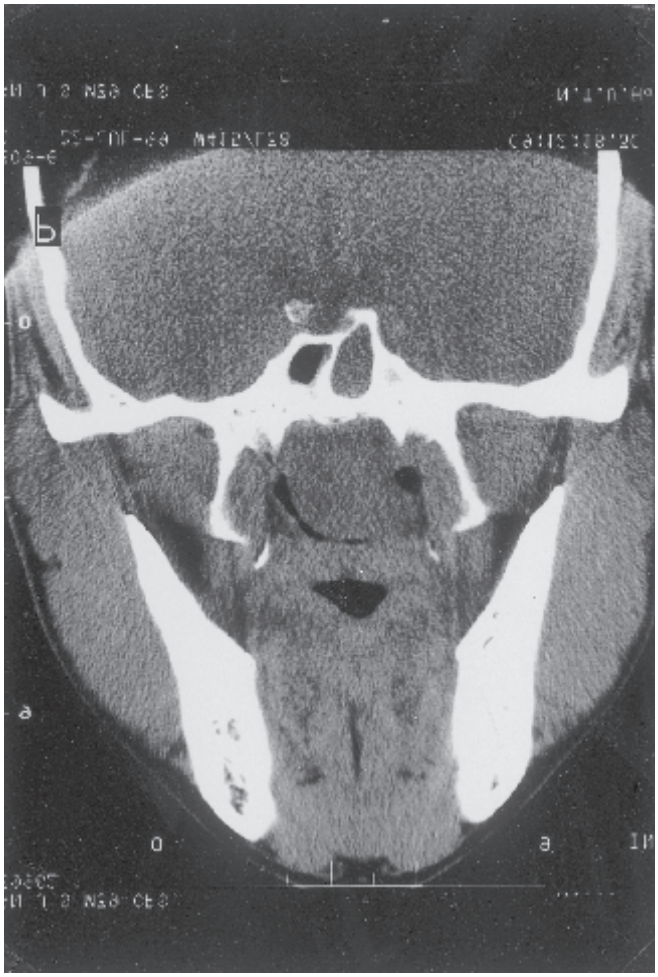

Figure 2. CT, a huge mass occupied the nasopharynx.

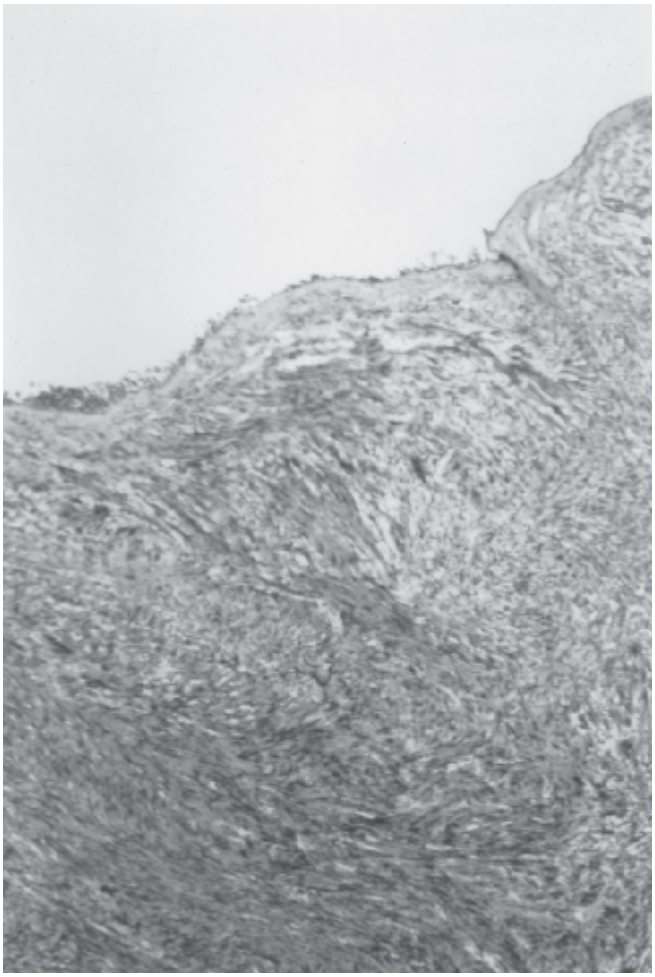

Figure 3. Non-encapusulated mass noted under microscope.

medial wall of the maxillary sinus was also noted. In addition, soft tissue density was also noted in the left frontal, maxillary, and ethmoid sinuses, which were compatible with sinusitis. Endoscopic sinus surgery was performed under general anesthesia with an impression of unilateral paranasal sinusitis with the left nasal polypoid mass. During the operation, the huge polypoid mass was found occupying nearly the entire left nasal cavity without much adhesion to nasal mucosa. Its insertion was at the medial side of the middle turbinate. A deviated nasal septum, a laterally displaced left middle turbinate, and an atrophic inferior turbinate were found. These seemed to be secondary changes from the mass effect. The bilateral middle meatus, especially on the left side, were narrow without polypoid tissue. En bloc extirpation of the mass was impossible because the tumor was so huge and was incased in the nasal cavity with fragile consistency. It was totally removed piece by piece, and the left middle turbinate where it originated was partially removed by a regular endoscopic sinus surgical procedure. The procedure was smooth and the patient's condition was uneventful.

The gross appearance of the specimen was yellowish white, soft, and polypoid. Microscopically, a section showed a non-encapsulated hypercellular tumor composed of round to ovoid cells with palisading or fasicular arrangement in fibrillary background (Figures 3 and 4). Verocay body-like structures were found in some sections (Figure 5). Nuclear atypia was absent and the mitotic index was low (average of less than 1/10 HPF).

Immunohistochemically, the tumor cells were strongly and diffusely positive for vimentin and S-100 staining. Neuron specific enolase and smooth muscle actin stainings were focally positive. Cytokeratin, epithelial membrane antigen, and desmin stainings were all negative. Therefore, benign nasal schwannoma was diagnosed.

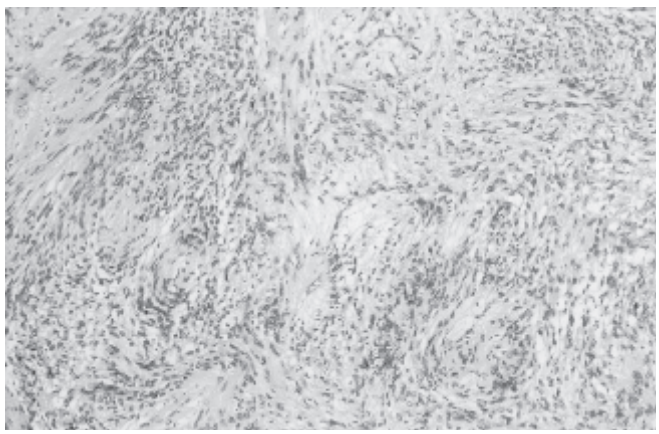

Figure 4. Weary spindle cell with high cellular density are arranged in palisading pattern.

No recurrence of the tumor was noted during follow up. The patient's condition had been uneventful in the 10 months following the operation.

DISCUSSION

Schwannoma is not a common tumor in the sinonasal tract. Only about 70 cases have been recorded in the literature (Hasegawa et al., 1997). A sinonasal schwannoma can be found in many sites, including the nasal septum, paranasal sinus, tip of the nose, turbinate, and nasopharynx (Khalifa et al., 1981; Pasic et al., 1990; Leakos et al., 1990; Lemmerling et al., 1998). The presenting symptoms of the tumor are always non-specific, depending on the site of the mass (Perzin et al., 1982). Generally, a unilateral nasal obstruction is the most common symptom, where patients usually feel a progressive unilateral nasal obstruction for a long period of time. Unilateral epistaxis is also a frequent complaint. Anosmia, painful sensation, and headache are noted because of the mass effect of the tumor (Verma et al., 1970; Kaufman et al., 1976; anonymous, 1995).

Grossly, the schwannoma is usually reported to be an encapsulated mass with a smooth surface. Microscopically, Antoni A and Antoni B arrangements are diagnostic for this tumor. Considering the neurofibroma as the major differential diagnosis in this area, the typical pathological finding of proliferating spindle cells within wide-spreading keloid collagen bundles with branching vessels is not found in this case. According to one report (Hasegawa et al., 1997), the pathological findings of schwannoma of the sinonasal tract are different from schwannomas in other regions. The differences include the loss of fibrous encapsulation and dominating hypercellularity. In this report, the pathological findings for our patient are compatible. No capsule was noted on the tumor surface. No typical Antoni B area was noted, and only interlacing weary cells with high density were found in all sections. These findings were compatible with the pathological findings of cellular schwannoma. However, no typical fibrous capsule of cellular schwannoma was found, but a Verocay body could be seen in some sections. These conflict the diagnostic criteria of cellular schwannoma (Casadei et al., 1995).

On account of the hypercellular pattern of nasal schwannoma, it is always important to consider the possibility of malignancy. However, a scanty mitotic change in the average high power view may support the diagnosis of benign schwannoma. Cellular schwannoma also has a benign clinical course (Casadei et al., 1995). The reason for the bony erosion of maxillary sinus in our patient is due to the effect from the pressure of the tumor instead of malignant cell invasion (Perzin et al., 1982; Casadei et al., 1995). In the bony lesion, there was no malignant cell infiltration, which further confirmed the diagnosis of its benign nature.

Immunohistochemical stains are important in making these differential diagnoses. Weary spindle cells are suggestive of nerve or muscle origin. Antibodies against vimentin, S-100, neuron specific enolase, smooth muscle actin, cytokeratin, epithelial membrane antigen and desmin were used. The tumor cells are strongly and diffusely positive for vimentin and S-100 stainings.

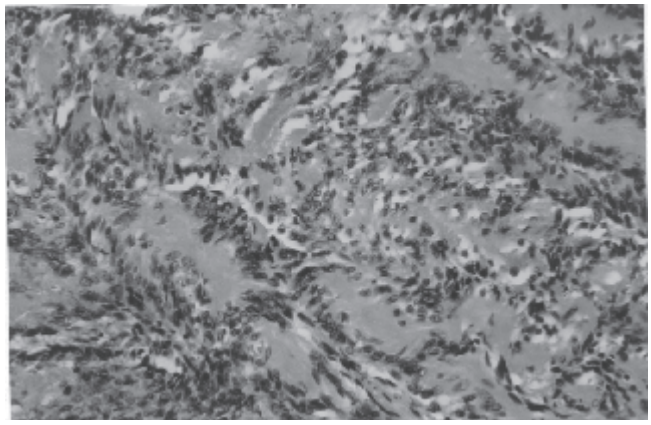

Figure 5. Verocay body-like structure in Antoni A area.

These are compatible with the diagnosis of either the typical or cellular schwannoma, but are not congruous with the differential diagnosis of juvenile angiofibroma, solitary fibrous tumor, hemangiopericytoma, fibroma, malignant peripheral nerve sheath tumor, or meningioma (Casadei et al., 1995; Hasegawa et al., 1997). Neuron specific enolase and smooth muscle actin stainings are focally positive, which revealed the possibility of a tumor of nerve or epithelial origin. However, antibodies against cytokeratin, epithelial membrane antigen, and desmin showed a negative result. Muscle origin is excluded after these stainings. These special staining patterns give a further confirmation of our diagnosis.

The tumor is noted as originating from the medial side of the middle turbinate. In this area, only the lateral superior posterior and lateral inferior posterior nasal branch from the sphenopalatine ganglion, and the inferior nasal branch of anterior ethmoidal nerve are within consideration. The first two branches are from the maxillary nerve, while the third nerve originates from the nasociliary nerve, branches of the trigeminal nerve. Both sympathetic and parasympathetic nerves are noted within the trigeminal nerve originating from the sphenopalatine ganglion. Therefore, there are several possibilities to the origin. According to one report (Hasegawa et al., 1997), schwannoma without a fibrous capsule has also been noted in gastric schwannoma. It is considered to be from the autonomic myenteric plexus because of the absence of a fibrous epineural sheath. Therefore, it is possible that the nasal schwannoma in our case is from autonomic nerve origin. Although the olfactory nerve is close to the location of the tumor, the lack of schwann cells in the olfactory nerve excludes this possibility.

In an overview of this case, we are reminded to include schwannoma in the clinical diagnosis when a patient presents with unilateral sinusitis and a large polyp. However, the extraordinary location, the lack of a fibrous capsule, and the presence of the Verocay body noted in this case are different from other reports (Casadei et al., 1995). Although a recurrence rate of 23% has been reported, nasal schwannoma usually has a benign clinical course (Casadei et al., 1995). Local wide excision of the tumor may be the first choice of management. In our case, endoscopic sinus surgery was enough for removal of the tumor because of its definite origin. On account of its gigantic

size and fragile consistency, en bloc resection is impossible. Complete removal is achieved by piecemeal resection. No recurrence or distant metastasis was noted during follow up. Nevertheless, considering the versatile entities of a unilateral polypoid mass, it is worthwhile to take a biopsy specimen before the operation for determining an appropriate surgical procedure. MRI evaluation before surgical exploration is recommended. Schwannoma presents as a solitary soft mass, with a high signal in the T2 weighted image in MRI. In some cases, the nerve is usually at the peripheral side of the mass (Lemmerling et al., 1998). These examinations promote better comprehension of the nature and the extent of the tumor. After clarification of the character and extent of the mass, endoscopic sinus surgery is enough for most benign lesions, otherwise, an external approach is the choice for unresolved cases. Finally, the findings in this case imply a possible relationship between the special pathological changes and the tumor origin, but the exact result needs further investigation.

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ANNOUNCEMENT