Sinonasal schwannoma of the middle turbinate

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ABSTRACT
Paranasal schwannomas are uncommon lesions, representing less than 4% of all head and neck schwannomas. They give rise to nonspecific symptoms such as nasal obstruction, epistaxis, and anosmia. Imaging features are generally nonspecific. Here, we present the radiologic features of a benign schwannoma of the middle turbinate with dural invasion in a 71-year-old woman.

Key words: • schwannoma • radiology • paranasal sinuses

Schwannomas are lesions that arise from the neural sheath of peripheral nerves, autonomic nerves, or cranial nerves. Nerve sheath tumors of the head and neck region mainly involve the eighth cranial nerve with only 4% occurring in the paranasal sinuses (1, 2). Occasionally, malignant schwannomas also occur in the paranasal sinuses. Sinonasal schwannomas do not have specific radiologic findings. The tumor rarely extends intracranially or intraorbitally, and imaging features can be similar to malignant neoplasms (3).

Case report
A 71-year-old woman presented with a 1.5-month history of medial deviation and diplopia of the left eye. She also had nasal obstruction but did not have epistaxis. An anterior rhinoscopy and an endoscopic examination showed a solid mass at the level of the middle turbinate and the roof of the nasal cavity. A thick, mucopurulent secretion was noted. Paranasal sinus computed tomography (CT) scan showed a 5 × 3.5 × 3 cm expansile soft tissue mass in the left ethmoidal sinus extending through the left frontal sinus superiorly and the right ethmoidal sinus medially. Erosion and destruction of the left ethmoidal sinus and dehiscence of the bony cribriform plate were seen. Bone fragments were seen within the mass. The left middle turbinate could not be separated from the mass. The nasal septum was deviated to the right. There were erosions in the lamina papyracea to the left of the mass, which caused an indentation of the left medial rectus muscle (Fig. 1).

Cranial magnetic resonance imaging (MRI) examination was performed to better assess intracranial extension. The lesion was hypointense on T1-weighted images and contained hypointense and hyperintense areas on T2-weighted images. There was heterogeneous enhancement after contrast injection. The left frontal bone and sinus were invaded by the tumor, but there was no dural enhancement or brain edema (Fig. 2). The differential diagnosis of the tumor included benign and malignant tumors of the sinonasal cavity such as squamous cell carcinoma, olfactory neuroblastoma, lymphoma, and schwannoma. Biopsy of the nasal mucosa revealed a schwannoma.

The endoscopy-assisted tumor resection was performed under general anesthesia. During the surgery, the mass could not be separated from the middle turbinate, from which it was thought to have originated (Fig. 3), and was excised with it. There was a 2.5 × 1 cm defect in the dura at the base of the skull. The dural defect was repaired with the auricular conchal cartilage and the septal bone grafts (Fig. 4). Pathologically, the tumor was a whitish-brown soft tissue mass, rubbery in consistency. Light microscopy showed spindle-shaped undulating cells in the loose myxoid stroma. The lesion contained cellular and hypocellular regions. There was ulceration and necrosis in the superficial part of the lesion.
Given these findings, a diagnosis of schwannoma was made.

Discussion

Sinonasal schwannomas have been postulated as arising from the ophthalmic and maxillary divisions of the trigeminal nerve, and from the autonomic nerves of sympathetic fibers of the carotid plexus and parasympathetic fibers of the sphenopalatine ganglion (4). Preferential locations have been reported as being the ethmoid sinuses, followed by the maxillary sinus, the nasal pits, and the sphenoid sinus. Nasal septal localization is rare. Schwannomas rarely show malignancy and more often develop in association with von Recklinghausen’s disease (5). The clinical symptomatology is varied and nonspecific but is similar to the signs of chronic nasal obstruction. Ethmoid sinus and nasal cavity tumors frequently present with epistaxis, whereas tumors of the maxillary sinus are usually associated with pain. As it may masquerade the more common sinonasal conditions such as chronic sinusitis or polyps, diagnosis of schwannoma is often delayed (6). Our patient presented with diplopia and medial deviation of the left eye.

Diagnosis is facilitated by endoscopy, CT, and MRI. Endoscopy typically reveals a unilateral polypoid nasal mass. CT reveals a unilateral nasal mass that may be expansile. During the CT evaluation of a sinonasal mass increased density within the sinus may well be interpreted as part of the tumor when in reality it simply represents fluid in the blocked sinuses. Schwannomas can cause bone remodeling by pressure and this behavior can lead to misdiagnosis as a malignant process. Preservation of bony margins can be helpful in differentiating schwannomas from malignant tumors, which tend to aggressively destroy bone. However, in the case of bony destruction and fragmentation, and intracranial or intraorbital extension, benign and malignant processes like esthesioneuroblastoma, fungal granuloma, nasoethmoidal carcinoma, and schwannoma cannot be differentiated by CT examination. MRI characteristics of schwannomas are typically isointense on T1-weighted images and hyperintense on T2-weighted sequences (6).

Some researchers have pointed out that T2-weighted MRI scans of minor...
Salivary gland tumors and neuromas of the paranasal sinuses give a bright signal compared with an intermediate T2-weighted signal for other sinonasal tumors (7). In addition, MRI is useful to determine the intracranial extension and better evaluate the cause of sinus obliteration (tumor versus inflammation) by contrast enhancement of the tumor (8). Our patient had atypical MR intensity features with cribriform plate destruction and bony fragments in the superior part of the lesion, and intraorbital extension. These features pointed toward malignancy. However, after contrast enhancement, no dural enhancement, brain edema, and extra- or intra-axial lesions were present, which supported a benign mass. Heterogeneous contrast enhancement of a tumor is usually associated with a cystic change or hemorrhage (9). Generally, linear dural enhancement is not specific for dural invasion but often represents reactive change. Dural enhancement with focal nodularity, dural thickening of more than 5 mm, and pial enhancement are highly suggestive of neoplastic dural invasion (10).

Schwannomas are usually described as being encapsulated; the capsule is assumed to derive from the perineurium of the nerve of origin. Some authors speculate that sinonasal mucosal autonomic nervous system fibers are devoid of perineural cells and, therefore, lack encapsulation. Encapsulation of schwannomas in this region is rare, which probably explains the rather aggressive growth pattern compared with schwannomas in other locations. The lack of encapsulation might make the tumor more difficult to define and extract completely (11). Our patient's tumor was not encapsulated either. Therefore, dural invasion and bone remodeling were associated with it.

The treatment of choice in schwannomas is surgical excision of the tumor. Surgical resection is usually curative. Radiation therapy is usually reserved for malignant nerve sheath tumors. If tumor is confined to the paranasal sinuses, the prognosis is excellent (6). Functional endoscopic sinus surgery has the advantage of lower morbidity, no external incision, and a shorter hospital stay when compared with traditional approaches. In our patient, septal cartilage and auricular conchal cartilage were used as a flap for the dural defect.

In conclusion, although sinonasal schwannomas are benign tumors, they have features of local erosion and destruction, which are suggestive of malignancy, making preoperative diagnosis difficult. In addition, they can show atypical MR intensity features. Excision is curative, and if a dural defect is present it can be repaired easily with native flaps.

References