Endoscopic removal of an extensive sinonasal schwannoma

Norhafiza MAT LAZIM, Irfan MOHAMAD, Ramiza Ramza RAMLI, Shamim AHMED KHAN, Thin Thin WIN @ Safiya YUNUS

1Department of Otorhinolaryngology-Head & Neck Surgery and
2Department of Pathology, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia

ABSTRACT
Schwannoma is a rare, benign and slow-growing tumour which arises exclusively from Schwann cells. Essentially this tumour can be found in any part of the body. In the paranasal sinuses and nasal cavity, this tumour originates mostly from the ophthalmic and maxillary branches of the trigeminal nerve. The most common clinical presentation is unilateral nasal obstruction. Histological examination is crucial for diagnosis. The main modality of treatment is surgical excision. The type of surgery will depend on the location and extent of the tumour. Even though it is generally considered a benign tumour, there have been reports of potential malignant transformation. We report a case of a 52-year-old man with an extensive nasal mass which was removed endoscopically and histologically confirmed as schwannoma.

Keywords: Endoscopy, neurilemmoma, schwannoma, sinonasal, surgical endoscopies

INTRODUCTION
Schwannoma is a rare benign tumour of neurogenic origin. It can arise virtually from any myelinated nerves which include the peripheral and autonomic nervous systems as well as the myelinated cranial nerves. In the head and neck region, it can occur in sites such as the neck, intracranial, orbit, intraparotid, parapharyngeal and infratemporal fossa. The most common site is the vestibular nerve. The paranasal sinuses and nasal cavity are also frequently involved but to a lesser degree. For the sinonasal schwannoma, the clinical presentation varies and the symptoms include nasal obstruction, nasal discharge, facial pain and headache. The modality of treatment for this tumour is mainly by surgical excision and the type of surgery will depend on the site and extent of the tumour. Although it is considered a benign tumour, there have been reports on its potential malignant transformation. We report a case of a 52-year-old Malay gentleman with an extensive sinonasal schwannoma that was successfully removed by endoscopic resection.

CASE REPORT
A 52-year-old Malay gentleman presented with a history of right nasal blockage for more than a year. This symptom was associated
with unilateral nasal discharge, mild facial pain and frontal headache. The nasal discharge was sometimes thick and yellowish. There was no history of epistaxis. He had a history of a right polypectomy which was performed 15 years ago for right nasal polyposis. The medical and family histories were unremarkable.

Endoscopic nasal examination revealed polypoidal mass which filled almost the entire right nasal cavity. It extended posteriorly into the posterior choana. It was pressing part of the septum superiorly. The mass appeared greyish and fleshy and other parts appeared clear jelly-like (Figure 1a and 1b). Examination with a probe showed that the mass was most likely to have arisen from the lateral wall of the right nasal cavity.

A computed tomography (CT) scan of the paranasal sinus showed a homogenous soft tissue mass in the right nasal cavity. The mass extended posteriorly into the posterior choana. Laterally, the mass extended into the right maxillary sinus and was causing thinning and bowing of the superomedial wall of right maxillary sinus (Figure 2). It also caused obliteration of the right osteomeatal complex. Anterior and middle ethmoidal air cells were also involved.

Initial punch biopsy revealed an inflammatory nasal polyp. As the clinical suspicion was more in favour of an inverted papilloma, the patient was offered and agreed to undergo endoscopic right medial maxillectomy. Intraoperative findings showed that the tumour appeared to be arising from the axilla of the right middle turbinate. The mucosa of the maxillary sinus was polypoidal and filled with mucopurulent secretion. The medial wall of the maxillary sinus was sclerosed and irregular. Postoperative histopathological examination revealed a schwannoma with positive staining for S-100 (Figure 3a). The histopathology features of Verocay bodies were also present (Figure 3b).

The patient recovered well after surgery and is currently under regular follow up. There was no sign of recurrence at last visit in
sites such as the scalp, face, oral cavity, pharynx, larynx, trachea, parotid gland, middle ear and external auditory meatus could also be involved. 2 Essentially, this tumour could also arise from any cranial nerves with the exception of the optic and olfactory nerves. This is due the fact that these cranial nerves are ensheathed by glial cells instead of Schwann cells. In the sinonasal tract, the ophthalmic and maxillary branches of the trigeminal nerve including the sympathetic fibres to sinonasal mucosa are the sites of origin of this tumour. 1 In the head and neck region, the vestibular nerve is the most common site for the tumour to arise from. Nevertheless, other sites such as the scalp, face, oral cavity, pharynx, larynx, trachea, parotid gland, middle ear and external auditory meatus could also be involved. 2

**DICUSSION**

Schwannoma, which is also known as neurilemmoma is a benign, slow-growing tumour of neurogenic origin. It arises most exclusively from the Schwann cells which form the perineurium of the peripheral nerves. It can occur anywhere in the body. In the head and neck region, it has been reported to account for 25% to 45% of all schwannoma cases. However, it is rare in the sinonasal tract and accounts for only about four percent of cases. 1 In the head and neck region, the vestibular nerve is the most common site for the tumour to arise from. Nevertheless, other

---

**Figure 3:** a) Spindle cells are positive for S100 protein (S100 stain, x20), b) fragments of tumour tissue composed of spindle cells with the areas of nuclear palisading (Verocay Bodies) (H&E stain, x10).
This tumour has been reported to affect both genders equally with mean age of presentation between 20 and 50 years old. Nevertheless, there are reported cases in the young and elderly. The clinical presentations vary and depend on the site and the extent of the tumour. The most common symptoms associated with sinonasal schwannomas are unilateral nasal obstruction, epistaxis, hyposmia and pain. Exophthalmus, facial swelling and epiphora have also been reported but to a lesser degree.

Most patients with schwannoma present late to the hospital. This is mainly due to the similarities of their symptoms with that of other benign sinonasal conditions such as allergic rhinitis, chronic rhinosinusitis and nasal polyps. This results in delay in reaching the correct diagnosis in some of the cases. There are usually no distinctive clinical features to be noted during clinical examination; hence the diagnosis can only be made after histological examination. Radiological examination with CT scan normally does not show any bony involvement. However, in some cases, erosion of the neighboring bony structure may occasionally be observed. This is mainly due to pressure necrosis from a slow growing mass and does not necessarily imply malignant infiltration.

The treatment of choice for nasal cavity schwannoma is surgical excision. Depending on the site and extent of the lesion, various approaches can be utilised. These include mid-facial degloving, lateral rhinotomy or endoscopic approach. Mid-facial degloving and lateral rhinotomy are by far the most common operative procedures carried out for sinonasal diseases, particularly malignancy. Mid-facial degloving provides good exposure of the middle third of the face. It utilises four basic incisions, bilateral sublabial incisions from maxillary tuberosity to tuberosity, bilateral intercartilaginous incision, septocolumellar complete transfixion incisions and bilateral pyriform aperture incisions extending to the vestibule. In lateral rhinotomy, incision is made along the nasal bridge from the medial canthus down to the alar of the nose exposing the nasal cavity and the maxillary sinus. Though these procedures can offer wide surgical field exposure, they tend to leave surgical scars which may not be favourable to the patients. Endoscopic technique on the other hand can result in excellent outcomes if combined with other approaches. For example, endoscopic assisted maxillectomy can be offered for some nasal cavity tumours which extend to the maxillary sinus. This technique allows complete removal of a big tumour without leaving a surgical scar, as demonstrated in this case. Besides that, endoscopic approaches allows the tumour to be directly visualised on the monitor screen and the extent of tumour can be accurately assessed.

Histopathological examination of the tumour tissues is always warranted for the diagnosis of schwannomas. The characteristic histopathological features of this tumour are the presence of Antoni type A and Antoni type B areas as well as Verocay body. Antoni A areas consist of compact spindle shaped cells with twisted nuclei whereas Antoni B areas display loosely arranged spindle cells within a myxoid matrix that is prone to degeneration, haemorrhage and cyst formation. The positive staining for S-100 is specific for Schwann cells and is one of the histological criteria used for diagnosis of schwannoma.
though this tumour is considered benign, it has a potential to become malignant. Histologically, the malignant potential of this tumour can be predicted based on the presence of an unequivocal malignant foci manifested by increased cellularity, numerous mitoses, anaplastic cells, and invasiveness.  

This tumour is categorised as a radioresistant tumour. Radiotherapy is used only in cases where there is an incomplete removal of the tumour or in cases where patients are not suitable for surgery because of underlying medical illness or in a case of a very extensive tumour.

In conclusion, we showed that schwannoma even when it is extensive can still be removed successfully by an endoscopic approach. However, the true extent of the tumour needs to be assessed radiologically before considering endoscopic resection.

REFERENCES