

## Schwannomas of ear, nose, throat and neck

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### Abstract

Schwannoma is a benign tumour of nerve sheath origin with latent malignant potential. All cranial nerves can give rise to schwannoma except for olfactory and optic nerves, which are devoid of Schwann cell. Schwannomas are usually asymptomatic and present late owing to compression of nerve of origin. We present our study of 19 cases of schwannoma arising from unusual sites in head and neck, having varied presentation and the challenges faced in management of these cases. These cases presented in detail to the department of Ear, Nose and Throat, KEM Hospital, and were thoroughly evaluated clinically and radiologically to formulate a management strategy. Schwannoma of the head and neck is a rare entity but should be considered as differential diagnosis in unilateral nasal mass cases, palatal masses, anterior and lateral neck masses. Nerve of origin may not always be clear preoperatively but the possibility of postoperative loss of nerve function should be kept in mind. Radiological investigations like computed tomography scan and magnetic resonance imaging play a pivotal role in management. In case of nonvascular neck tumours, fine needle aspiration cytology is crucial but has low accuracy in the diagnosis of neural tumors. Histopathology of excised tumour remains the gold standard in diagnosis.

### Introduction

Schwannoma is a benign tumour of nerve sheath origin. It is also named as neurilemmoma, neurinoma. 25 to 45% of schwannomas are located in the extra cranial head and neck region.<sup>1</sup> It may arise from cranial, peripheral or autonomic nerves.<sup>2</sup> All cranial nerves can give rise to schwannoma except olfactory and optic nerve which are devoid of Schwann cells.<sup>3</sup> Vagal nerve is most common site for origin of head and neck schwannoma followed by sympathetic plexus and cervical plexus.<sup>4</sup> Hypoglossal, facial nerves are the rare sites

of origin. Sinonasalcavity and intraoral schwannomas contribute only 4% and 1% of head and neck Schwannomas respectively.<sup>5,6</sup>

Schwannomas are usually asymptomatic. Symptoms of nerve involvement appear very late due to compression of nerve of origin. Such long latency period of nerve involvement makes it difficult to clinically diagnose neural origin of tumour.

### Case Reports

#### Case #1 – Facial nerve schwannoma

An 11-year-old male child presented with right side facial weakness of 6 months duration and decreased hearing since 4 months. Otoscopic examination revealed bulging tympanic membrane with a mass behind an intact tympanic membrane. High-resolution computed tomography temporal bone showed a soft tissue mass filling middle ear space and mastoid cavity with erosion of ossicles and mastoid air cells (Appendix Figure 1). Pure tone audiometry test showed moderately severe conductive hearing loss. Canal wall down mastoidectomy was done with excision of tumour arising from tympanic segment of the facial nerve. Facial nerve grafting was done using greater auricular nerve. Histopathology report was suggestive of facial nerve schwannoma. On 6-months follow-up, patient showed mild improvement in facial palsy with no sign or symptoms of recurrence.

#### Case #2 – Nasal middle turbinate schwannoma

A 32-year-old male patient presented with progressive left sided nasal blockage since 4 months and multiple episodes of epistaxis. Anterior rhinoscopy revealed left side smooth spherical nasal mass which was bleeding on touch. Computed tomography (CT) was suggestive of a 3×2 cm mass in the left nasal cavity extending into anterior ethmoid cells, frontal and maxillary sinus (Appendix Figure 3). Magnetic Resonance Imaging (MRI) of the lesion was suggestive of inverted papilloma or a neurogenic tumour (Appendix Figure 2). Endoscopic assessment revealed the mass was arising from middle turbinate and occupying middle meatus which was completely excised. Histopathology was suggestive of nasal schwannoma (Appendix Figure 4) Patient was followed up for 4 months with no recurrence.

#### Case #3 – Nasal septal schwannoma

A 38-year-old female presented with bilateral nasal blockage (Left >Right) since

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2 years which gradually progressed to cause complete blockage. It was associated with mucoid nasal discharge and hyposmia. There was a single episode of epistaxis which resolved with conservative management. On examination air blast was totally absent bilaterally. Anterior rhinoscopy showed left nasal polyp which was pale, smooth, non-tender and did not bleed on touch. The mass was pushing the septum to the right side and probe could be passed medially, superiorly and inferiorly.

MRI (Appendix Figure 7A and B, Appendix Figure 8 and Appendix Figure 10) of the paranasal sinuses showed a 54×42×54 mm heterogenous mass, hypointense on T1WI and hyperintense on T2WI with patchy central necrotic areas and hemorrhages, intensely enhancing on contrast. It caused lateral splaying of medial walls of both maxillary sinuses (Left>Right). Invasion of the ethmoidal, sphenoidal sinuses and the nasopharynx. The patient underwent endoscopic excision of the mass with posterior septectomy. The mass was dissected completely (Appendix Figure 11) and sent for histopathology examination. Endoscopic approach helped to prevent unwanted removal of healthy

nasal mucosa and the anterior and superior septum which helped to maintain the shape of the nasal dorsum. Patient was followed up after 3 months with diagnostic nasal endoscopy and CT scan which showed complete clearance with well epithelialized cavity and symptomatic relief for the patient.

Histopathological analysis of the tumour showed spindle cells arranged in Antony A and Antony B pattern. Verocay bodies were seen. There was a focal nuclear pleomorphism with tumour cells expressing S-100 protein but they were negative for HMB-45 (Appendix Figure 9A, B and C).

#### Case #4 – Palatalschwannoma

A 25-year-old male patient presented with swelling on palate since 2 months. Clinical examination of oral cavity showed 2×2 cm mass on right side of hard palate (Appendix Figure 13). CT scan showed 2×1.2 cm soft tissue mass of palate without bone erosion. Depending on clinical examination and CT report provisional diagnosis was made as minor salivary gland tumour. But on excision of palatal mass (Appendix Figure 12), histopathology report was suggestive of palatal schwannoma (Appendix Figure 18A and B).

#### Case #5 – Oropharyngeal schwannoma

A 52-year-old male patient presented with difficulty in swallowing for solids and change in voice since 1 year and regurgitation of food since 5 months. On examination, soft globular mass seen in oropharynx attached to left posterior pillar. CT scan showed a homogenous regular hypodense swelling (Appendix Figure 14) which slightly enhances on contrast.

Histopathology of the excised specimen revealed schwannoma most probably of glossopharyngeal nerve origin.

#### Case #6 – Vagal nerve schwannoma

A 13-year-old boy presented with a history of an asymptomatic swelling of the right side of neck since one year. Examination revealed a nontender firm swelling 5×6 cm situated anterior to the right sternocleidomastoid muscle and extending superiorly up to the right mastoid tip (Appendix Figure 15A). It was mobile in transverse plane.

Oropharyngeal examination revealed a smooth bulge on the right lateral wall of the oropharynx and the right tonsil and peritonsillar structures were pushed medially (Appendix Figure 15B).

The ultrasonographic examination revealed a 5×4×6 cm vascular mass in the right carotid space displacing the carotid

arteries anteriorly and the internal jugular vein laterally. The ultrasonography guided needle core biopsy report was schwannoma. The MRI of Neck showed well defined oval hypodense lesion in the right carotid space, extending from the angle of mandible to C4 vertebral level and enhancing on contrast (Appendix Figure 16A and B). Surgical excision of the lesion was performed (Appendix Figure 17) and histopathological examination confirmed the diagnosis (Appendix Figure 18A and B). Patient recovered completely without any morbidity.

### Discussion

Schwannoma is the benign tumour, arising from Schwann cell of nerve sheath. It is well encapsulated, solitary and a slow growing tumour. Verocay in 1908 was the first who established schwannoma as a pathological entity.<sup>7</sup> It can arise from cranial, sympathetic or peripheral nerves. An approximately 25-40% of total schwannoma cases arise in head and neck region. The most common site of the extracranial schwannoma in the head and neck region is the parapharyngeal space. Vagal nerve involvement is much common than sympathetic nerve involvement.<sup>8</sup> Other rare sites involve facial nerve, paranasal sinuses, intra oral, submandibular area.

In our experience with schwannoma, we came across few cases with rare sites of origin posing a diagnostic dilemma. Our series of schwannoma includes 12 cases (Appendix Figure 6) arising from lateral neck: vagal nerve schwannoma, spinal accessory nerve schwannoma (Appendix Figures 5 and 20-22), glossopharyngeal nerve schwannoma, cervical sympathetic chain schwannoma (Appendix Figure 19), 2 oropharyngeal schwannomas, one facial nerve schwannoma, one anterior neck thyroid gland schwannoma, one submandibular schwannoma, one branchial cleft schwannoma, one palatal schwannoma, one nasal septal and one middle turbinate schwannoma.

Facial nerve schwannoma compose only 0.8% of all intrapetrous mass lesions.<sup>9</sup> It can arise from any segment of facial nerve. Multiple segment involvement is more common than single segment involvement. Facial nerve is resistant to compression, so functional impairment is late. 27.3% of all patients with facial nerve schwannomas may present with normal facial nerve function.<sup>10</sup> Patient may present with conductive or sensory neural hearing loss depending on the segment involved. Radiology plays crucial role in diagnosis of facial schwannoma. It shows enhancement

on CT scan with contrast and high-signal intensity on T2-weighted MRI. Surgery is the only treatment for facial nerve schwannoma. Therefore patients are left with permanent facial dysfunction after surgery as tumour removal almost always requires total nerve resection.<sup>11</sup>

Sinonasal schwannoma is a rare entity which can present with nasal blockage, rhinorrhea, and epistaxis. It may clinically present like any other unilateral nasal mass. On radiological examination schwannomas may typically appear isointense in T1-weighted sequences and hyperintense in T2-weighted sequences. These radiological features may be variable in few cases so ultimately needs correlation with clinical and histopathological features.<sup>12,13</sup> In our case clinical and radiological examination was pointing towards inverted papilloma or neurogenic tumour but definitive diagnosis was only made on final histopathology report of excised mass. Histopathological examination of the schwannoma shows biphasic pattern of alternating Antoni A and B areas. Antoni A are the hypercellular areas composed of compact spindle cells arranged in short bundles or interlacing fascicles with nuclear palisading aligned around eosinophilic processes called Verocay bodies. Antoni B areas are hypocellular area shows spindle or oval cells arranged haphazardly in the loosely textured matrix. For definitive diagnosis on immunohistochemistry S100 is necessary.<sup>14</sup>

The most common location of intraoral schwannoma is the tongue and it rarely occurs in the hard palate. The palatal schwannoma was first reported in 1987 by Jones. In case of any slow tumour of palate most common differential diagnosis is salivary gland tumour.<sup>15</sup> The preoperative diagnosis as palatal schwannoma is quite difficult because it is an infrequent tumor and is not usually suspected in the oral cavity. MRI scan characteristics like low signal intensity on T1-weighted imaging and signal hyperintensity on T2-weighted imaging may be useful in diagnosis.

The hypoglossal nerve schwannoma is very rare tumour. Total reported number of cases of hypoglossal nerve schwannomas in literature are 121 and in only 12 cases extracranial part of the nerve was affected.<sup>16</sup> Surgical excision of tumour is the treatment of choice. Hypoglossal nerve schwannoma could present as submandibular region swelling or lateral neck swelling.<sup>17</sup> The most common differential diagnosis includes pleomorphic adenoma and carotid body tumour. Radiological investigations may help to rule out differential diagnosis but in few cases with variable MRI findings histopathology report remains gold standard

in diagnosis.

A total of 28 cases of spinal accessory nerve schwannomas have been reported, among which only two were located in the cervical region.<sup>18</sup> The schwannoma arising from the vagus is 2-5% of benign nerve tumour. It is the most common schwannoma of parapharyngeal space. Most cases of vagal schwannomas manifest between the third and sixth decades of the patient's life as it is a slow growing tumour. It is rarely seen in children as in our case. Radiological examination plays an important role to differentiate it from paraganglioma and also confirm its relation with surrounding structures. In case of nonvascular neck tumour, fine needle aspiration cytology (FNAC) is crucial but has low accuracy in the diagnosis of neural tumors.<sup>19</sup>

## Conclusions

Schwannoma is a rare entity but should be considered as differential diagnosis in unilateral nasal mass cases, palatal masses, anterior and lateral neck masses. The lower cranial nerves or sympathetic nerve schwannomas may present as lateral neck swelling without any particular nerve dysfunction. Nerve of origin may not always be clear preoperatively but the possibility of postoperative loss of nerve function should be kept in mind. Patient should be counseled about all possible complications of surgery. Preoperatively tumour and its relation with surrounding vital structures like carotid artery and other important nerves should be considered carefully. As it is a slow growing tumour which usually present without neuronal deficit, surgeon should evaluate risks and benefits of the surgery in every individual case. A meticulous dissection of tumour with intracapsular enucleation preserves the nerve and its function. Radiological investigations like CT scan

and MRI play a pivotal role in management. In case of nonvascular neck tumours, FNAC is crucial but has low accuracy in the diagnosis of neural tumors. Histopathology of excised tumour remains the gold standard in diagnosis.

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