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Management Of Extracranial Schwannomas In Head And Neck Region - An Observational Study

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Abstract

ABSTRACT

Background: Schwannomas, benign tumors arising from Schwann cells, often manifest as slow-growing lesions in the peripheral nerve sheath. While typically asymptomatic, they can affect cranial and peripheral nerves. Surgical excision is the primary treatment, but preserving nerve function poses challenges.

Methods: This retrospective study analyzed 12 cases of benign head and neck schwannomas diagnosed at Department of ENT, SCB Medical College, Orissa, India between 2021 and 2023. Data encompassed patient demographics, tumor characteristics, diagnostic methods, surgical approaches, histopathology, and follow-up outcomes. Pre-operative investigations included Fine Needle Aspiration Cytology, Ultrasonography, and imaging.

Results: Predominantly middle-aged and male patients presented with painless swelling, commonly in the cervical region, tongue, nose, and hard palate. Mean symptom duration was 8.5 months. Imaging depicted characteristic features, guiding surgical planning. Various approaches ensured complete excision, preserving nerve function. Histopathology confirmed the diagnosis, with positive S-100 staining. No cases showed malignancy or recurrence during follow-up.

Conclusions: Head and neck schwannomas, though rare, present diagnostic and management challenges. Pre-operative diagnosis relies on imaging and biopsy, with surgical excision essential for treatment. Nerve preservation minimizes post-operative complications. Despite diagnostic difficulties, maintaining a high index of suspicion for schwannomas in patients with painless, slow-growing swellings is crucial.

Keywords

Schwannoma, Extracranial, Antoni A, Antoni B

INTRODUCTION

Schwannoma is also called as neurilemmoma or neurinoma which originates from Schwann cells of the peripheral nerve sheath.[1] Notably, approximately 25%–45% of schwannomas originate in the head-and-neck region, with around 10% originating from either the vagal or sympathetic nervous system.[2] They are benign, slow-growing, well-differentiated, and solitary lesions. The hallmark of schwannoma is a biphasic cellular appearance with hypercellular Antoni A areas with verocay bodies and Anton B areas.[3] They are usually asymptomatic and diagnosed only after histopathological evaluation. The cranial nerves such as V, VII, X, XI, XII, sympathetic or peripheral nerves can be involved.[4] They are usually encapsulated except for a few locations like the central nervous system, nasal cavity & nasopharynx. It is always a challenge to excise these encapsulated tumor masses without getting any neurological deficit. Therefore, it is important to determine the nerve of origin, site, and extension of the tumor to prevent complications. In this study, we describe very rare types of schwannomas occurring in the head and neck region along with their surgical management.

OBJECTIVES

This retrospective study aims to comprehensively analyze the clinical presentation, diagnostic methods, surgical approaches, histopathological characteristics, and follow-up outcomes of patients diagnosed with extracranial head and neck schwannomas. The objectives encompass evaluating the demographic profile of affected individuals, assessing the efficacy of pre-operative diagnostic techniques, analyzing surgical approaches with a focus on nerve preservation, examining histopathological features including immunohistochemical staining, and investigating post-operative outcomes such as recurrence rates and nerve function preservation. Through this study, we seek to contribute valuable insights into the diagnostic and management

challenges posed by head and neck schwannomas, aiming to enhance clinical understanding and optimize patient care in this rare yet clinically significant condition.

MATERIALS & METHODS

This retrospective analysis examines 12 cases of benign schwannoma in the head and neck region, diagnosed at the Department of ENT & Head and Neck Surgery, S.C.B Medical College, India, from 2021 to 2023. The study adhered to Institutional ethical committee approval and the Helsinki Declaration of 1975, revised in 2013. Patients diagnosed postoperatively with schwannoma after histopathological examination were included. Data on patient demographics (age, sex), tumor characteristics (location, size), lesion duration, and presenting symptoms were collected. Pre-operative investigations comprised Fine Needle Aspiration Cytology, Ultrasonography, Contrast-enhanced CT scan, or MRI scan. Each patient underwent surgical excision with preservation of the nerve of origin. Statistical analysis was conducted to assess patient demographics, symptom duration, and tumor size using descriptive statistics (mean, median, range) and inferential statistics for variable associations such as tumor location and symptoms. Post-operative complications, histopathological findings, immunohistochemistry results, and follow-up details were documented.

RESULTS

Most of the patients affected were in the age group of 31-45 years old. One patient (8.3%) was aged 15 years or below, seven (58.3%) were between 16 and 45, three (25%) were between 46 and 60, and one (8.3%) was over 60. The mean and median age of presentation of schwannoma and

range were 38.5, 39, and 15 to 64 years respectively. Eight cases were male and four cases were female. (Table-1)

Table 1: DEMOGRAPHIC PROFILE

Age (Years)	**No. of Patients (n=12)**
<15	1
16-30	3
31-45	4
46-60	3
>60	1
Sex	
1. Male	8
2. Female	4

In none of our cases, family history of von Recklinghausen's disease, neurofibromatosis type 2, schwannomatosis, or Carney's complex was present. The tumors occurred most commonly in the cervical region (33.3%), tongue (25%), nose (25%), hard palate (8.3%) and posterior pharyngeal wall (8.3%). (Fig.1,2,3,4) The duration of symptoms ranged from 4 days to 12 months (mean=8.5months, median=8.5months) and gross tumor size ranged from 1.5x1x 1 to 6.4 x5 x 4cm (mean=3.64 x 2.98 x 2.20 cm). Most of the patients presented with a painless swelling. 3 patients (25 %) underwent only contrast-enhanced CT scan, 4 patients (33.3%) underwent MRI

and in 1 patient (8.3%) both contrast-enhanced CT scan & MRI was done. (Fig.3,4) Most tumors appeared isointense on T1-weighted images & heterogeneously hyperintense on T2-weighted images. (Table-2)

Table-2: CLINICAL PROFILE

Sl no.	**Location**	**Presenting Complaints**	**Duration of Lesion (months)**	**Gross Tumor Size (cm)**
1.	Dorsum of tongue	Painless swelling	9	2 x 1.5 x 1
2.	The right lateral border of the tongue	Painless swelling	7	2 x 2 x 1
3.	Base of Tongue	Pain on swallowing, Globus sensation Change in voice	11	4 x 3.5 x 3
4.	Hard Palate	Pain on chewing	4	1.5 x 1 x 1
5.	Nasal cavity and paranasal sinuses	Nasal obstruction, intermittent nasal bleeding, hyposmia	10	6.4 x5 x4
6.	Nasopharynx	Nasal obstruction Voice change	7	3.5 x 3 x 2.5

7.	Vagus Nerve	Painless slow growing neck swelling	8	4 x 4 x 3
8.	Ansa cervicalis	Painless slow growing neck swelling	9	4.8 x 3.6 x 3
9.	Brachial Plexus	Painless slow growing neck swelling	8	5 x 4.2 x 3
10.	Posterior pharyngeal wall	Foreign body sensation	12	2 x 1.5 x 1
11.	Parapharyngeal space	Painless neck swelling	12	3.4 x 2.5 x 2
12.	Retropharyngeal space	Difficulty in swallowing	6	4 x 3.2 x 2

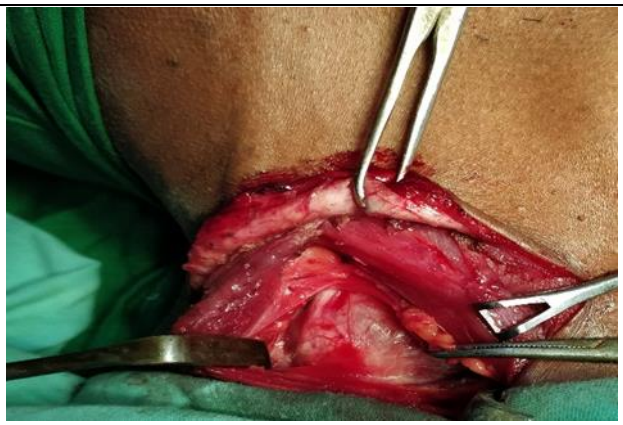

Figure 1 Brachial Plexus Schwannoma

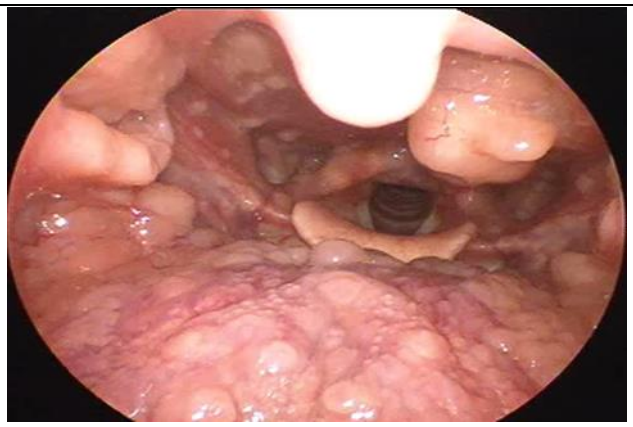

Figure 2 Posterior pharyngeal wall Schwannoma

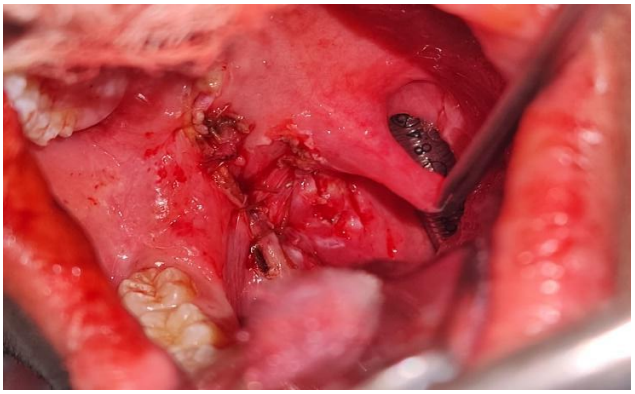

Figure 3 Parapharyngeal Schwannoma

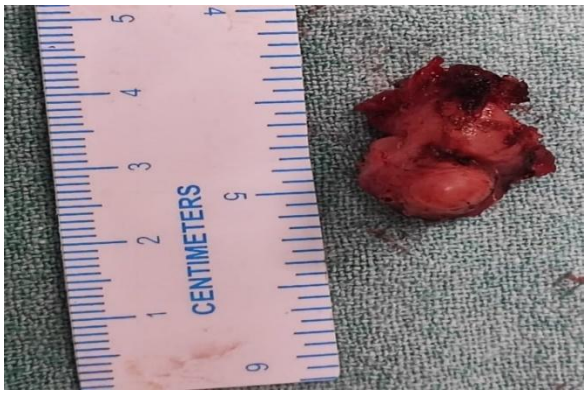

Figure 4 Excised mass

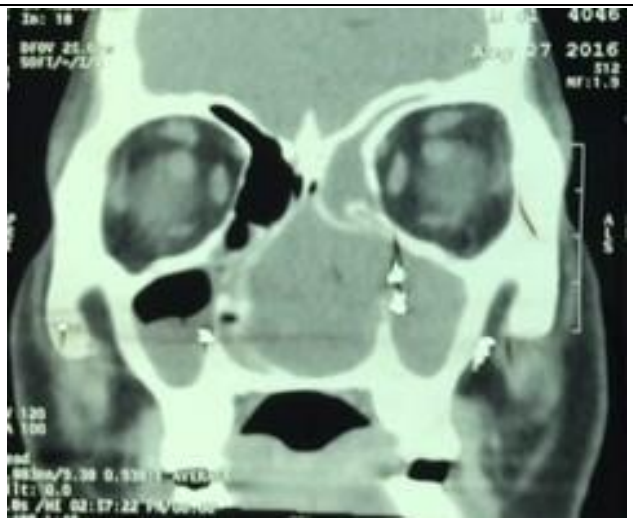

Figure 5 Coronal view of CT Scan PNS showing a well defined homogenous mass involving nasal cavity, maxillary, and ethmoid sinus.

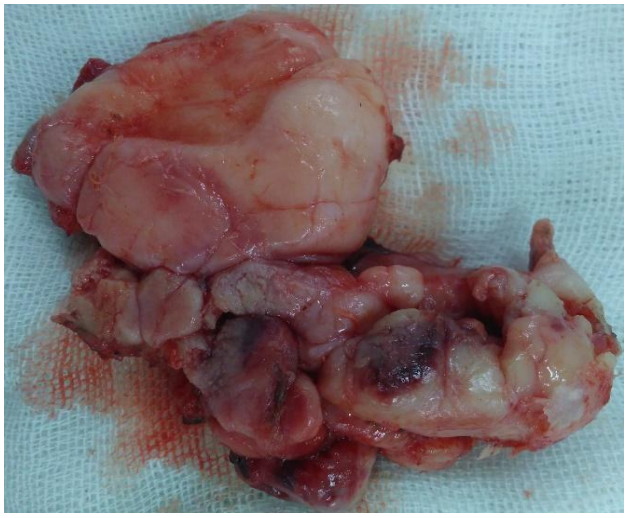

Figure 6 Excised Sinonasal Schwannoma.

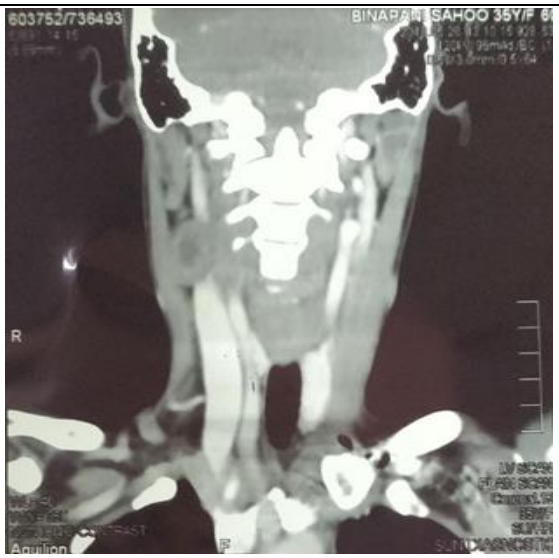

Figure 7 Well defined oval shaped mass located between sternocleidomastoid muscle & great vessels

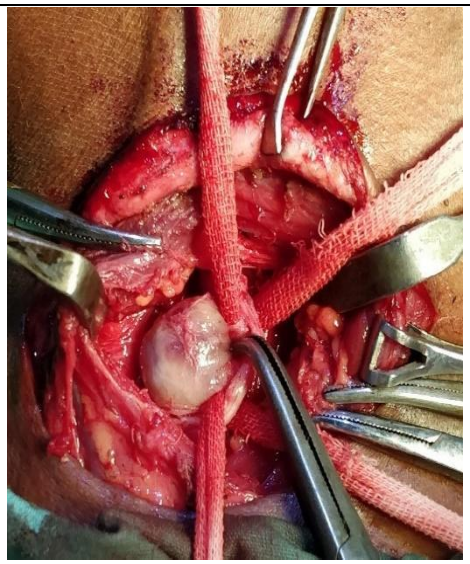

Figure 8 Vagus Nerve Schwannoma

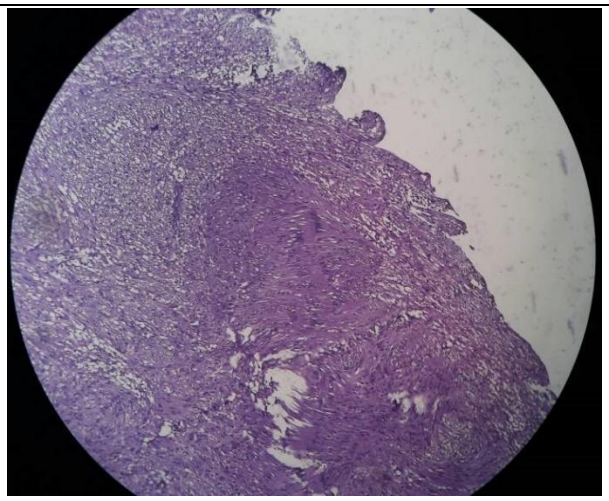

Figure 9 Antoni A areas with spindle cells in Fascicles.

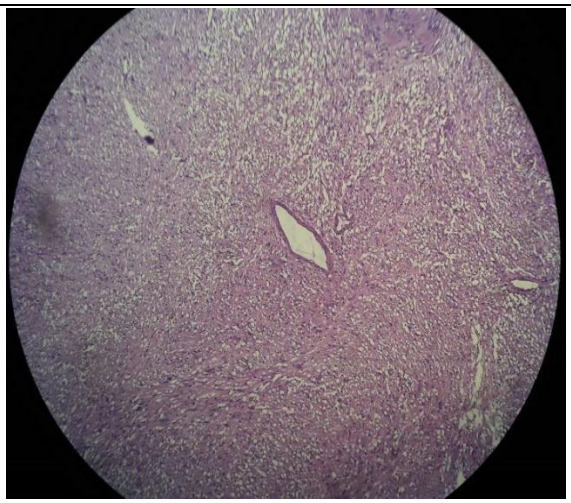

Figure 10 Antoni B areas with microcystic space & vessels.

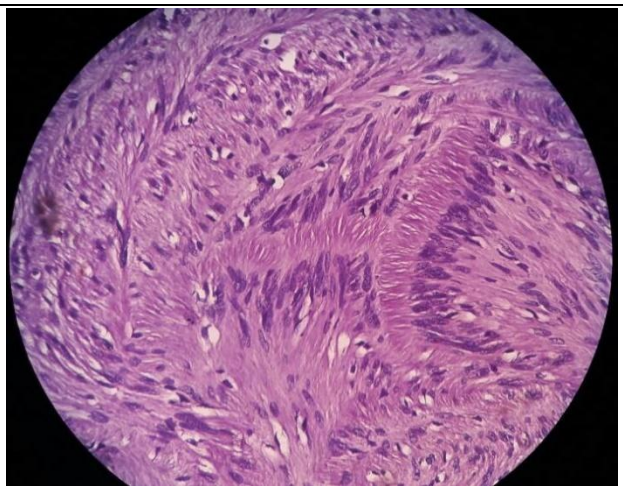

Figure 11 40 X showing spindle cells & a typical Verocay body.

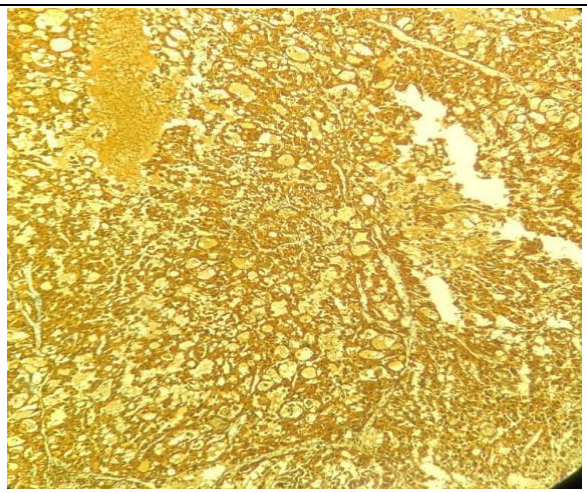

Figure 12 S-100 positive on Immunohistochemistry.

Complete intracapsular excision of the tumor mass with preservation of the nerve was done by intraoral (41.6%), endoscopic (16.6%), endoscopic with intraoral (8.3%), transcervical (25%), rigid laryngoscopy (8.3%) approach. The final diagnosis of schwannoma was determined by fine needle aspiration cytology followed by excisional biopsy in 5 patients (41.6%), incisional & excisional biopsy in 1 patient (8.3%), punch & excisional in 1 patient (8.3%) and only excisional

biopsy in 4 patients (33.3%). The biphasic appearance of Antoni A and Antoni B areas was seen in all patients on histopathology. But verocay bodies appeared in only 5 cases (41.6%). It is difficult to differentiate these nerve sheath tumors into benign or malignant on FNAC. Immunohistochemical staining for S-100 protein was positive in all cases. In none of our cases, there were features of malignant transformation and any evidence of recurrence.

Table-3: IMAGING & BIOPSY

Sl No.	**Location**	**Radiological investigations**	**Type of biopsy confirming Schwannoma**
1	Dorsum of Tongue	Not done	Excisional Biopsy
2	Lateral border of tongue	Not done	FNAC & Excisional Biopsy
3	Base of Tongue	CEMRI	Incisional & Excisional
4	Hard Palate	CECT	FNAC & Excisional
5	Nasal cavity and paranasal sinuses	CECT	Punch biopsy & Excisional biopsy
6	Nasopharynx	CECT	Punch biopsy & Excisional biopsy
7	Vagus Nerve	CECT, CEMRI	FNAC & Excisional biopsy
8	Ansa Cervicalis	CEMRI	FNAC & Excisional biopsy
9	Brachial Plexus	CEMRI	Excisional

10	Posterior pharyngeal wall	Not Done	Excisional
11	Parapharyngeal Space	CEMRI	FNAC & Excisional biopsy
12	Retropharyngeal space	CECT, CEMRI	Excisional biopsy

Table-4: TREATMENT & FOLLOW UP

Sl No.	**Location**	**Surgical Approach**	**Post-operative complications**	**Follow up**
1	Dorsum of Tongue	Intraoral	No	2 months - No recurrence
2	Lateral border of tongue	Intraoral	No	2-4 months - No recurrence
3	Base of Tongue	Intraoral with Coblation	No	3-12 months - No recurrence
4	Hard Palate	Intraoral	No	4 months - No recurrence
5	Nasal cavity and paranasal sinuses	Endoscopic sinus surgery with Microdebrider	Synechia, Hyposmia	3-12 months - No recurrence

6	Nasopharynx	Endoscopic excision	No	4 months - No recurrence
7	Vagus Nerve	Endoscopic with intraoral	No	4-8 months - No recurrence
8	Ansa cervicalis	Transcervical	No	6 months - No recurrence
9	Brachial Plexus	Transcervical	No	6-12 months - No recurrence
10	Posterior pharyngeal wall	Transcervical	Mild neurological deficit	6-12 months - No recurrence
11	Parapharyngeal space	Rigid laryngoscopy with bipolar cauterization	No	3-6 months - No recurrence
12	Retropharyngeal space	Intraoral	Post-op pain	4 months - No recurrence

DISCUSSION

Peripheral nerve sheath tumors include both neurofibroma & schwannoma. Neurofibromas are more common but with less malignant potential than schwannomas.[5] Schwannomas are usually solitary sporadic lesions in comparison to neurofibromas. There are certain conditions like type-2 neurofibromatosis (Von Recklinghausen's disease) & schwannomatosis which are associated with

unilateral or bilateral vestibular schwannomas. Incidence of extra-cranial schwannomas is common in the cervical region and peripheral extremities.[6] Only a few handful of cases occur in the sino-nasal region, nasopharynx, oral cavity, and posterior pharyngeal wall.

Schwannoma equally affects both sexes and has an average age incidence of 20-50 years.[7] These tumors take origin from Schwann cells of peripheral nerves, and cranial nerves except olfactory & optic nerves and spinal & autonomic nerves. The most common sensory nerve affected is the 8th cranial nerve and the 7th nerve is the motor nerve involved. [4,8] The incidence of schwannomas in the head and neck is around 25-50% of cases.[2] In the oral cavity and nasal cavity, it accounts for 1% and <4% of head and neck schwannomas respectively.

The most common location of schwannoma in the head and neck area is the para-pharyngeal space and the vagus nerve is frequently involved.[9] Usually, the patient is asymptomatic and may present a painless oral cavity or a cervical mass. The most common symptom in oral cavity schwannomas is a painless swelling involving the tongue, palate, buccal mucosa, lips, floor of mouth, or mandible. [2,10] Some of the differential diagnoses of lingual schwannoma include traumatic neuroma, mucosal neuroma, ganglioneuroma, lipoma, solitary circumscribed neuroma, pleomorphic adenoma, dermoid or dermoid.[11]

The nasal cavity and paranasal sinuses are very rare locations and may simulate chronic rhino-sinusitis or a nasal polyp. They commonly involve the trigeminal nerve or its branches. Differential diagnoses include fibroma, juvenile nasopharyngeal angiofibroma, hemangiopericytoma, solitary fibrous tumor, rhabdomyosarcoma, meningioma, and malignant peripheral nerve sheath tumor.[12] In our study, Sino-nasal schwannoma shows a well-circumscribed homogenous mass in the nasal cavity extending to the nasopharynx & paranasal sinus without any bone destruction on contrast-enhanced CT scan. The absence of fibrous capsules and predominant hypercellularity on

histopathology in sino-nasal schwannoma differentiates it from other schwannomas. The reason behind this is an absence of epineurium over the nerve. Complete excision of the tumor via endoscopic, trans palatal, lateral rhinotomy, or combined approach is the treatment of choice. They can be resected surgically without damaging the nerve if its origin is from the nerve lining. In a subset of cases, endoscopic assistance was utilized during the surgical procedure. Additionally, a combination of endoscopic and intra-oral approaches was employed in surgeries involving the nose, paranasal sinuses, and nasopharynx. These endoscopic techniques were integrated to enhance visualization and precision during specific steps of the surgical excision process. Their application underscores the evolving role of minimally invasive techniques in the management of schwannomas in the head and neck region, potentially leading to reduced morbidity and improved surgical outcomes.

Posterior pharyngeal wall schwannoma is extremely rare and considered to originate from the peri-pharyngeal nerve plexus. If it is a solitary lesion like in our case, then it can be excised intraorally using a laryngoscope.[13] Brachial plexus schwannomas are rare and account for around 5% of cervical region schwannomas whereas cervical vagal schwannomas constitute about 2-5% of tumors. Vagal schwannomas push the internal jugular vein and internal carotid artery as they grow between them.[14] MRI is the investigation of choice to delineate location, margins, and extension.

Imaging modalities like ultrasonography show a hypoechoic homogenous solid mass. Contrast-enhanced CT scan shows a moderately enhancing homogenous mass lesion. On gadolinium contrast MRI scan, schwannoma appears in isointense or hypointense areas on the T1-weighted image and hyperintense on the T2-weighted image.[15]

On gross examination, the tumor appears as an encapsulated lobulated mass with an overlying smooth surface. Histologically, the tumor cells are organized in 2 different patterns -hypercellular

Antoni A & hypocellular Antoni B regions. Antoni A region consists of compact spindle-shaped cells with a palisading arrangement of the nucleus in an eosinophilic cytoplasm known as verocay bodies. Antoni B region consists of spindle-shaped cells which are arranged in a loose connective tissue matrix.[16] This biphasic appearance differentiates it from neurofibroma where the spindle cells are uniformly distributed throughout the tumor within a collagenous to myxoid stroma. It is not encapsulated & less vascular. None of our cases showed features of malignant transformation like pleomorphism, high mitotic figures (>10/2mm$^{2)}$, features of necrosis, infiltration, and irregular positivity for S-100.[17] Schwannomas show strong S-100 positive on immunohistochemistry.[18] Total excision of the tumor with preservation of nerve function is the definite treatment of choice because of its eccentric location in the epineurium & encapsulation.[19] We used an intra-oral approach with coblation technology for the excision of a base of tongue schwannoma. Recurrence of tumor after excision and complications is uncommon. Sometimes, neurological deficits may occur if nerve fibers are damaged during surgery.[20]

CONCLUSION

Pre-operative diagnosis of schwannoma is difficult. Therefore, a differential diagnosis of schwannoma should be considered for a slow-growing, painless swelling in the head and neck region. Pre-operative contrast-enhanced CT scan, MRI & FNAC helps in diagnosis as well as planning surgical excision.

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