Case Report

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Pathology Section

Parapharyngeal Schwannoma- A Rare Case Report

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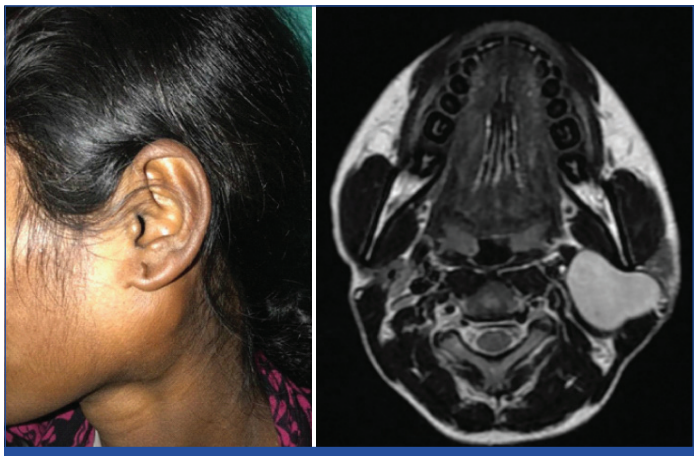
ABSTRACT

The incidence of schwannomas is 25-45% in the head and neck region but less than 0.5% when restricted to the parapharyngeal space. The clinical presentation of this lesion varies depending on the anatomical location of its occurrence and thus does the treatment modality. Present report is a case of a 20-year-old female who reported to the Department of Otorhinolaryngology with swelling in the lateral part of the neck which was subsequently diagnosed as schwannoma and appropriate surgical excision was done. Histopathology of the excision lesion confirmed the diagnosis. It becomes crucial to report and revise the tumours of the parapharyngeal space because of the complexity of the area.

Keywords: Antoni cells, Neurilemmoma, Parapharyngeal space, Schwann cell, Tumours of neck

CASE REPORT

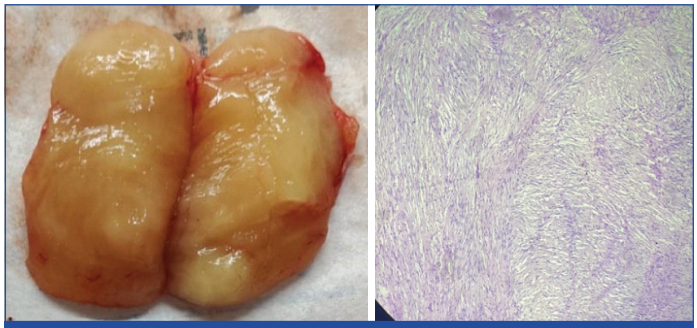
A 20-year-old female patient presented to the Department of Otorhinolaryngology in a tertiary care centre in central India with complaints of swelling on the left side of the neck since three months, which was of a slow-growing, indolent nature [Table/Fig-1]. On examination, the swelling was a solitary, well-defined, oval lesion which was firm in consistency, freely mobile and not fixed to the underlying structures. The overlying skin was normal and pinchable. There was obvious lymph node enlargement on palpating the surrounding area. The patient had no fever or any other symptomatic abnormality. The lesion was further assessed radiologically using Computed Tomography (CT) scan which suggested a well-defined, oval, hypodense cystic lesion measuring 5.3×3.0×2.3 cm in the left parotid space, abutting the parotid gland laterally, pterygoid muscle, and submandibular gland anteriorly, paravertebral muscles medially and sternocleidomastoid posteriorly. It was extending inferiorly till the level of C4. Magnetic Resonance Imaging (MRI) confirmed the CT findings and gave a probable diagnosis as schwannoma [Table/Fig-2].



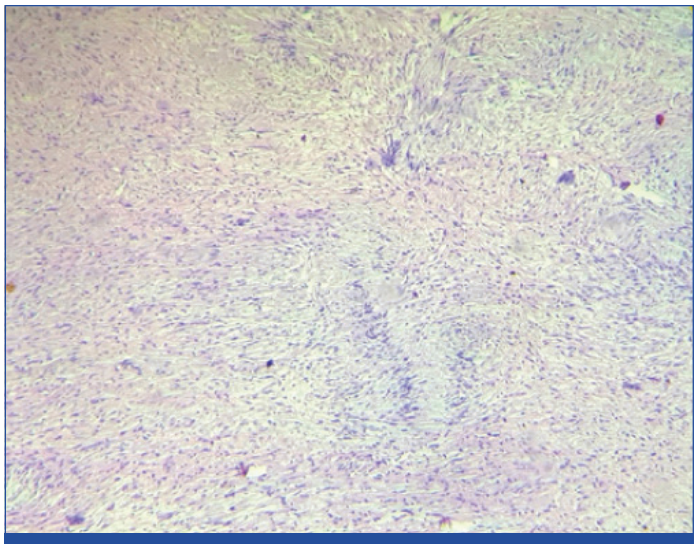
[Table/Fig-1]: Slow-growing swelling on the left side of the neck since 1 year. **[Table/Fig-2]:** T2/STIR image showing hyperintense, well-circumscribed lesion in the left parotid space suggestive of Vagal Schwannoma. (Images from left to right)

Surgical excision of the lesion was performed under aseptic conditions using the transcervical approach and the specimen was sent for histopathological examination. On gross examination, the specimen was a single, globular, whitish-yellow encapsulated mass measuring 5×5×1 cm in dimensions. The cut surface was solid, firm, and yellowish-white homogenous areas were identified [Table/Fig-3].

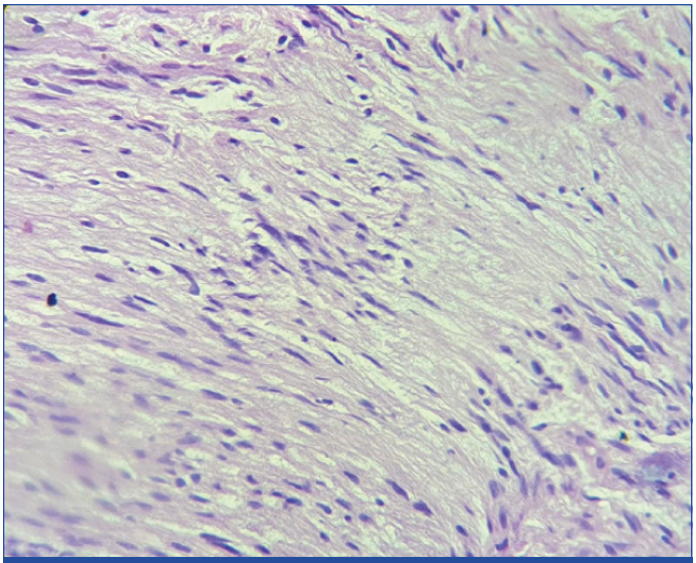
Multiple sections were taken which showed predominantly cellular areas of spindle cells with pulled-up nuclei. Microscopic examination also showed biphasic morphology with hypercellular Antoni A and hypocellular Antoni B areas. Antoni A areas are cellular areas with spindle cells arranged in a palisading fashion [Table/Fig-4,5]. Few places show an organoid arrangement of cells forming Verocay Bodies [Table/Fig-6], whereas in Antoni B areas the tumour cells are separated by abundant edematous fluid which forms cystic spaces. Histopathological features were suggestive of schwannoma.



[Table/Fig-3]: Grossly, the lesion was a single, globular, encapsulated mass with pale yellow areas identified on the cut surface; **[Table/Fig-4]:** Section showing a dimorphic pattern of cellular (Antoni A) and loose myxoid (Antoni B) areas (40x magnification) H&E stained. (Images from left to right)



[Table/Fig-5]: Section showing parallel rows of nuclei known as nuclear palisading (100x magnification) H&E stained.



[Table/Fig-6]: Section showing eosinophilic cell bodies nearly encircled by rows of nuclei called as Verocay bodies (400x magnification) H&E stained.

The patient came for routine follow-up on an outpatient basis, one month and then three months later and there were no complaints or symptoms. The benign nature of this lesion translates into a good, but guarded prognosis as the occurrence of this lesion in the parapharyngeal space might create a surgical bias towards preservation of the nearby structures rather than a just surgical enucleation.

DISCUSSION

The neoplasms occurring within the structures of the parapharyngeal space are predominantly of salivary origin and/or neurogenic. They can either be benign accounting for 70-80% of the lesions or malignant (20%) accounting for the rest of them [1]. A schwannoma, also known as neurilemmoma is an uncommon, slow-growing, indolent benign lesion having its origin from the peripheral neural sheath [2]. Out of all the peripheral nerve sheath tumours, one-fourth of them occur in the head and neck region and amongst these, less than 1% are found in the parapharyngeal space. Schwannomas are mostly asymptomatic [3].

Out of all the cases reported of Schwannoma, upto 45% occur in the head and neck region specifically in the intracranial cavity, scalp, face, medial and lateral regions of the neck, mastoid, orbit, nasal, oral cavities, and even parapharyngeal space. Other sites being the flexor surfaces of extremities, mediastinum, and retroperitoneum [4], it's occurrence in the parapharyngeal space is relatively very rare. It is known to occur more commonly in young adults and middle ages and there is no gender predominance. Half of the schwannomas reported in the parapharyngeal space are known to originate from the vagus nerve {Cranial Nerve (CN) X} and its branches and the remaining from the cervical sympathetic chain [5].

Parapharyngeal space or lateral pharyngeal space is a pyramidal potential space placed in the lateral aspect of the pharynx. Bounded by the carotid sheath posteriorly, parotid gland on the posterolateral aspect, medial pterygoid plate on the anterolateral aspect, and medially by the pharynx, this space is divided into two compartments by the styloid process and the stylopharyngeal fascia, the prestyloid or masseteric space and the poststyloid or carotid space. The prestyloid space contains the maxillary artery, ascending pharyngeal artery, and the parotid deep lobe. The post-styloid space contains the internal carotid artery, internal jugular vein, sympathetic chain, lymph nodes, carotid bodies, and the last four cranial nerves i.e., the glossopharyngeal nerve (CN IX), vagus nerve (CN X), accessory nerve (CN XI) and the hypoglossal nerve (CN XII) [6]. Considering the anatomical aspects, and the multitude of surgical differentials occurring for the surgeon from this region, like vagal benign or malignant paraganglioma or a carotid body tumour, the applied aspect primarily focuses on restricted surgical modalities to preserve the salient structures around, which could further possibly result in incomplete excision and maybe even recurrence [7].

A Schwann cell or a fibroblast supporting the nerve usually forms the nevus for the origin of this neoplasm. Schwannomas are truly encapsulated, white to yellowish-white and mucoid on cutting surface, and sometimes show cystic degeneration, calcification, and haemorrhage. The characteristic microscopic feature on low power is alternate Antoni A and Antoni B areas which are areas of hyper and hypocellularity, respectively. The cellular areas consist of organised spindle cells with nuclear palisading and may often contain Verocay bodies. The fewer cellular areas contain oval to spindle cells arranged more haphazardly in a loose myxoid matrix. Many cases have prominent thickened hyalinised vessels. Changes of degeneration like cystic change, calcification, hyalinisation, haemosiderin deposition, haemorrhage, and foamy histiocytes can also be present [8]. Thus, the gross examination and microscopic examination are mandatory to confirm the diagnosis and differentiate it from other more frequent tumours originating from the parapharyngeal space.

As the masses found in this space could originate from either the salivary gland, connective tissue, lymphoid lesions, cystic lesions, or neurogenic tissue, Confirmation using immunohistochemistry should be performed for which schwannoma will give positivity for S100 protein and SOX10 (Sry-related HMG Box gene 10), while it is negative for Endomysial Antibody (EMA), CD34, and claudin-1. In addition to the conventional schwannoma, there are various variants like cellular schwannoma, epithelioid schwannoma, plexiform schwannoma, and microcystic/reticular schwannoma. Cellular schwannomas are more commonly found in the mediastinum, retroperitoneum, and pelvis and lack Antoni B hypocellular areas. Epithelioid schwannomas will have mainly epithelioid cells in sheets, cords, and clusters with or without the cells of the conventional schwannoma. Plexiform schwannoma is seen in relatively younger patients and has extensive intraneural growth creating a plexiform pattern. Lastly, the microcystic/reticular occur mainly in the gastrointestinal tract consisting of anastomosing spindle cells forming a microcystic or reticular pattern [9].

The well-circumscribed and encapsulated nature of the neoplasm makes the prognosis favourable, and mere surgical excision is a safe modality of management. But when the patient starts presenting with symptoms like hoarseness of voice and dysphagia, the clinician needs to start suspecting a malignant transformation [10].

CONCLUSION(S)

A confirmed diagnosis of schwannoma can be made on histopathological and subsequent immunohistochemical examination if the histopathological diagnosis has not given satisfactory confirmation. Though the tumour is benign, it is site-sensitive, and as in this case, there could be a possibility of incomplete surgical resection because of the critical surrounding structures. One should be aware of the differentials of the tumour of the parapharyngeal space and emphasise the likelihood of recurrence in this region, therefore taking utmost care that there is a timely and regular follow-up of the patients.

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