



RESEARCH ARTICLE

MANAGEMENT OF NEURILEMMOMAS OF HEAD AND NECK A SINGLE CENTRE
EXPERIENCE

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ABSTRACT

Introduction: The neurilemmoma is not a common tumor but it is by no means rare. These are benign encapsulated tumors which are resectable and surgically curable but may be confused with malignant lesions

Material and methods: Four cases of neurilemmoma of head and neck region have been described which were managed in the Department of ENT, Government Medical College Jammu.

Conclusion: Neurilemmomas are less common solitary tumors of head and neck. Most are initially asymptomatic and usually present with swelling in the pharynx, externally or both. Benign schwannoma should be considered in the differential diagnosis of unusual intranasal lesions. An experienced cytopathologist is required to reach at a preoperative diagnosis and prior biopsy is confirmatory in nasal and paranasal lesions.

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INTRODUCTION

The neurilemmoma is not a common tumor but it is by no means rare. These are benign encapsulated tumors which are surgically curable but may be confused with malignant lesions and patient may be subjected unnecessarily to mutilating surgical procedures or may be refused treatment for a lesion which is completely curable.

Neurolemmal tumors arise from neurilemmal sheath of the peripheral, cranial and dsympathetic nerves and are widely distributed in the body. About 25% to 45%¹ of Neurilemmomas reported are found in the head and neck region, the majority being cervical in origin,² where the cranial nerves and sympathetic system with cervical plexiform are abundant. Although intranasal schwannomas are rare and only comprise 4% of head and neck neural tumors, such lesions should be considered for unusual appearing nasal masses.

Parapharyngeal lesions are usually deep seated and they present great difficulty to the clinician for their pre-operative assessment and specific technique of surgery.³ Because of rarity and interesting clinical and radiological findings, four cases of neurilemmoma of head and neck are described here.

MATERIAL AND METHODS

Four cases of neurilemmoma which were managed in Department of ENT, Government Medical College, and Jammu are described. A detailed history and thorough clinical examination of these patients was done. These patients were then subjected to routine investigations and radiological imaging. Also fine needle cytology was done if needle to establish the diagnosis. Once diagnosed all of the patients were subjected to surgery. All of them recovered well and are under our follow up.

RESULTS

Case no 1

A 45 year old male patient presented with a swelling on the left side of the neck and hoarseness of voice. The patient had undergone surgery on the swelling at another hospital. The surgeon did not remove the swelling but took a biopsy from the swelling which on histopathology turned out to be a normal lymph node. The patient developed hoarseness of voice immediately after the surgery.

Local examination revealed a solitary rounded, firm swelling 5 cm x 5 cm with diffused margins involving left submandibular

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region extending upto angle of mandible. The skin overlying the swelling had a scar mark. There were no visible pulsations.

On palpation, the swelling was firm freely mobile, non-tender, extending from anterior border of sternomastoid to the middle of ramus of mandible. The upper limit of the mass could not be palpated. No pulsations were felt over the swelling. Oral examination did not reveal a bulge.

Examination of nose and nasopharynx did not show any abnormality. Indirect laryngoscopy revealed left vocal cord palsy. There was no other cranial nerve involvement.

The routine blood and urine examination were within normal limits. X-ray of the chest and base of the skull did not show any abnormality. Fine needle aspiration cytology was non-diagnostic.

A CT scan of neck with contrast enhancement revealed a moderately enhancing left Parapharyngeal mass.

The patient was operated under general anesthesia through a left inframandibular cervical incision. No connection of the tumor with any cranial nerve could be demonstrated. The patient made an uneventful recovery. The histopathology of tumor mass showed it to be neurilemmoma. The follow-up more than one year did not show any evidence of tumor recurrence.

Case no 2

A 20 year old male was referred with history of painless lump on the right side of the neck for the last two years. This had increased in size during this time. The patient denied dysphagia, dyspnoea, cough or syncope.

On examination, vital signs were found to be normal. His voice was normal. There was a smooth, firm, non-tender, non-pulsatile, irreducible swelling in the right side of neck extending from anterior border of upper third of sternomastoid to 1 cm from midline. The lower end of the swelling was at the level of upper border of thyroid cartilage. The upper border of the swelling was not palpable as it was extending upwards under the ramus of the mandible. The skin over the swelling was normal. One large vessel was palpable on the lateral surface of the swelling and another along its antero-medial border. There was no bruit over the swelling.

Examination of the pharynx revealed a bulge on right lateral wall of pharynx just behind the right posterior pillar, extending from the level of vallecula to above the level of hard palate. Mucosa over the swelling was intact and freely mobile. The swelling was bimanually palpable.

Carotid angiography showed splaying of carotid fork with internal carotid artery displaced laterally and external carotid artery displaced anteromedially. The tumor vascularity was demonstrated.

CT scan with contrast enhancement demonstrated a well – encapsulated irregularity enhancing right Parapharyngeal space mass with central necrosis.

With a tentative diagnosis of neurilemmoma the mass was explored through a curved right upper cervical incision made at the level of hyoid bone extending from mastoid tip up to the

midline. The bifurcation of carotid was exposed, and the encapsulated tumor mass was found. Hypoglossal nerve was lying on the surface of the tumor from which it was separated. The tumor was pushing internal carotid artery laterally and external carotid artery anteromedially. The tumor was separate from both the arteries and carotid fork. The tumor was removed intact patient developed right-sides hypoglossal palsy postoperatively. There is no tumor recurrence one year after the surgery. But the hypoglossal palsy persists.

Case no 3

A 28 year old male presented with chief complaints of mass on the left side of throat and change in voice. There was no history of dysphagia or dyspnoea.

Local examination revealed a bulge on the left pharyngeal wall extending from behind the posterior pillar to almost midline. This was extending from the level of floor of vallecula to above the level of hard palate. The mass was 5 cm x 3 cm, firm, non-tender, non-pulsatile. Mucosa over the mass was intact and freely mobile. There was no external swelling.

Examination of nose, ear and larynx did not reveal any abnormality. There was no cranial nerve palsy.

Trans- oral fine needle aspiration cytology was non-confirmatory. A CT scan of neck with enhancement revealed a moderately enhancing well- encapsulated left parapharyngeal mass.

Using an external approach the patient was explored under general anesthesia. The left submandibular gland was removed to gain a wide access to the upper pole of the tumor. A well-encapsulated mass was dissected out. No connection with any of the cranial nerves was demonstrated. The patient developed Horner's syndrome in the postoperative period. There occurred a bluish discoloration and oedema of pharyngeal mucosa on the operated side which disappeared in a few days. There is no evidence of tumor recurrence on one year of follow-up but the Horner's syndrome persists.

Case 4: A 38 year old male presented to our department with a history of right nasal obstruction for one year, but in recent months it had been slowly progressive with a little left side obstruction also. He denied any history of rhinorrhoea, epistaxis, headache, postnasal discharge or pain. There was no response to multiple antibiotics, nasal steroid sprays, antihistamines and decongestant.

Anterior rhinoscopy revealed a whitish grey, homogenous, large polypoidal mass in the right nasal cavity, also seen on posterior rhinoscopy with left sided deviation of septum. The mass was tough and painless. Rest of the otolaryngologic and physical examination was normal.

Non-contrast enhanced computed tomography (CT) showed a well defined large soft tissue mass in the right nasal cavity with pushing of nasal septum towards left side compromising the left nasal cavity also. There was also thinning of turbinates, medial wall of orbit with extension into preseptal region. Superiorly the mass was extending into the ethmoid air cells and posteriorly into the nasopharynx. Right maxillary sinus was obliterated by the mass. There was no intracranial extension.

The benign clinical course and radiological appearance of the lesion urged the surgical team to take the biopsy which showed a benign schwannoma. The schwannoma was completely removed via a transnasal route which was confirmed endoscopically and the patient experienced an uneventful recovery. The histological diagnosis, confirmed by immunohistochemical studies was positive for S-100 protein.

Complications: We had one complication in case no 2 where the hypoglossal nerve palsy persisted. Case number 3 also had Horner's syndrome, but none of our patient had recurrence of tumor

DISCUSSION

Schwannomas are, in the overwhelming majority of cases, benign slowly growing tumors that characteristically expand the bony confines of the cavities and foramina in which they arise. The nomenclature of these encapsulated neurogenic tumors is varied, resulting in confusion. Neurilemmomas were first established as a pathological entity in 1910 by Verocay who called them "Neurinomas".¹ In 1935, Stout coined the term "Neurilemmoma" believing that the tumor arose from the nerve sheath of Schwann or neurilemma cells. Among other names employed for this tumor are schwannoma, encapsulated neurofibroma, perineural fibroblastoma, periglioma and neurilemmoblastoma etc.² The neural origin of Neurilemmomas is considered to be peripheral motor, sensory, sympathetic and cranial nerves. The optic and olfactory cranial nerves are not potential sites of the origin since they lack sheaths that contain schwann cells.⁴ In the Parapharyngeal space the tumor are diagnosed when they are of considerable size because of deep-seated location. Most of the neurilemmoma are initially entirely asymptomatic. Most patients usually present with a swelling in the pharynx or externally or both. The Parapharyngeal space is a potential space with three rigid walls so that the growth of a tumor in the region proceeds either medially or inferiorly or both. The pattern of growth accounts for distinctive clinical appearance.⁵ Pain is uncommon, dyspnoea, dysphagia, a vague discomfort or a sensation of pressure occurs as a late symptom with large tumor.⁶

The origin of nasal neurilemmomas is presumed to be the-

1. Sheath of the ophthalmic and maxillary branches of trigeminal nerve.
2. Autonomic fibers (parasympathetic) from the sphenopalantine ganglion.
3. Autonomic fibers (sympathetic) derived from the carotid plexus.⁴

Neurogenic tumors of paranasal sinuses are rare. Kragh *et al.*⁷ and Arora *et al.*⁸ have reported 1 and 4 cases, respectively.

In a review of non-epithelial tumors of the nasal cavity, paranasal sinuses and nasopharynx over a 70 year period, Perzin *et al.*⁹ found only 11 Schwann cell tumors among 430,000 cases researched.

Nasal neurilemmoma usually take a benign clinical course, but intracranial extension has also been reported. Bony erosion is not a criterion for malignancy because benign schwannoma can erode bone by pressure as was seen in our case. Inverted

papillomas, polyps and some malignancies also show bone destruction and remodeling.²

Probable histological nature of the tumor in this area is the most important criteria for the particular surgical approach in parapharyngeal tumors³ and for diagnosis in paranasal sinus tumors.⁴ Although aspiration biopsy has been suggested to be a convenient and expedient way of resolving the problem of histological diagnosis by Daly and Roesler¹, fine needle aspiration in our cases was not helpful in reaching the histological diagnosis of Parapharyngeal space tumors.

Because of long list of differential diagnosis of nasal and paranasal lesions biopsy is usually confirmatory. Pathologically, benign schwannoma consist of uniformly shaped spindle cells with variable cellularity. Neurilemmoma tend to be solitary, well-circumscribed tumors with an oval or round shape and encapsulated. They do not entrap the nerve axons but rather push them; therefore nerve may be surgically preserved. They typical exhibits a biphasic histologic pattern of Antoni A and Antoni B. Antoni A areas are regions of high cellularity with spindle shaped cells often arranged in bundles, palisade or whorls. Groups of compact parallel nuclei are also seen and are known as VEROCAY bodies. Antoni B area is less cellular and forms no distinctive pattern. Additionally, Neurilemmomas usually show intense immunostaining for S-100 (particularly Antoni A areas) which help to distinguish from other tumors.

In contrast to neurilemmoma neurofibroma are composed of streams of spindle cells with serpentine nuclei and are often encapsulated, multiple and often entrap the nerve axons which made it difficult to preserve the nerve. Although there is a significant risk of malignant transformation with neurofibromatosis, neurilemmoma very rarely takes this course.

In all the three cases the tumors were removed through an external approach after making an upper cervical incision made at the level of hyoid bone. Excision of submandibular gland in Case-III gave a good visual access to the upper pole of the tumor. No connection of the tumor with cervical sympathetic chain or any other cranial nerve could be demonstrated. Horner's syndrome developed in one case and hypoglossal palsy was observed in another case. The patient developed hypoglossal palsy on 7th postoperative day which could have been due to the 12th nerve involvement in healing and fibrosis. On regular follow up till date there is no evidence of tumor recurrence. Recurrence is not to be expected even if a part of capsule is left behind.⁷

Determining the location and extension of the tumors with preoperative imaging help to choose the approach for nasal tumors. In the present study, after confirmation with prior biopsy the nasal schwannoma was resected completely transnasally with endoscopic confirmation which is preferred by many surgeons in schwannomas without intracranial extension.⁴

CONCLUSION

Neurilemmomas are less common solitary tumors of head and neck. Most are initially asymptomatic and usually present with swelling in the pharynx, externally or both. Benign schwannoma should be considered in the differential diagnosis of unusual intranasal lesions. an experienced cytopathologist is

required to reach at a preoperative diagnosis and prior biopsy is confirmatory in nasal and paranasal lesions.

It was the advent of CT scan that allowed a more systematic preoperative evaluation to determine which tumor requires preoperative angiography to find the size and extent of tumor and to differentiate between parotid and extra parotid masses. MRI can be quite useful for the determination of possible intracranial extension and can better evaluate the causes of opacified sinus structure.

An external approach is recommended for removal of Parapharyngeal space Neurilemmomas because of better visualization and even intracapsular excision is not associated with increased risk of reoccurrence. If confined to nasal and paranasal sinuses prognosis is excellent with regular endoscopic re-examination.

Fine needle aspiration in our cases was not helpful in reaching the histological diagnosis of Parapharyngeal space tumors.

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