



Outcome of Treatment in Locally Advanced Parapharyngeal Tumors

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Abstract

Parapharyngeal tumors are uncommon and a challenge to the surgeon as they are large and intimately related to carotids and lower cranial nerves at the time of presentation. This was a retrospective study of 36 large parapharyngeal tumors operated at a tertiary care rural hospital. Seventy eight percentage of tumors were benign, most common being schwannoma followed by pleomorphic adenoma. Most common malignant tumor was mucoepidermoid carcinoma (19%). Pre operative imaging and fine needle aspiration biopsy helped in diagnosis and planning of surgical approach. Tumors of neural origin were post-styloid and salivary origin tumors were pre-styloid. Majority of patients presented with large neck swelling and 28% had medial displacement of tonsil compromising the oropharyngeal space. 11% presented with stridor requiring emergency tracheostomy. Cervical with transparotid approach was most common surgical approach followed by only cervical approach. FNAB correlated with histopathology report in 94% patients. Transient paralysis of lower division of facial nerve was the most common complication. Only 2 patients required sacrifice of facial nerve and in one of them nerve was grafted. Only 1 patient in this series had permanent paralysis of vagus and hypoglossal nerve. A 22% of them were malignant salivary tumors and one of them recurred leading to death. Benign tumors did not recur. In our experience these challenging tumors have a good outcome but require meticulous planning and surgery.

Keywords: Parapharyngeal space tumors; Surgical approach; Schwannoma; Neurofibroma; Salivary tumors; Cranial nerve palsy

Introduction

Parapharyngeal tumors are uncommon and contribute to 0.5% to 1% of Head & Neck masses [1]. These include salivary gland tumors, neurofibromas, schwannomas and paragangliomas. Majority of these tumors (80%) are benign [1]. However, patients often present with locally advanced tumors which are intimately associated with important neurovascular structures like carotids, vagus & hypoglossal nerves. These tumors can extend higher into masticator space and can be intimately associated with facial nerve and skull base. They can compress the airway and present with stridor. Some of them can secrete vasoactive amines and can be part of a syndrome involving multiple sites. Some of these tumors may be associated with cranial nerve palsies at the time of presentation [2]. Surgery of these locally advanced tumors is challenging and requires expertise and experienced surgeon and anesthesiologist. Though the outcome of treatment of benign parapharyngeal tumors is good with regard to survival, serious complications and morbidity can result due to treatment. Clinical presentation and imaging guide a surgeon accurately regarding the origin and extensions of the tumor. We would like to share our experience regarding the presentation, histology, approach used and outcome of surgery in 36 locally advanced parapharyngeal tumors operated in our institution.

Objectives of the study

1. To document the various clinical presentations and histologies of parapharyngeal tumors in our region.
2. To correlate the clinical presentation with histology and extent in these tumors.
3. To document the outcome of surgery in these locally advanced tumors.

Materials and Method

This is a retrospective observational study done in a tertiary hospital in a rural area between

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Received Date: 30 Mar 2020

Accepted Date: 08 Apr 2020

Published Date: 10 Apr 2020

Citation:

Gupta A, Azeem Mohiyuddin SM, Sagayaraj A, Prasad CSBR. Outcome of Treatment in Locally Advanced Parapharyngeal Tumors. Clin Surg. 2020; 5: 2801.

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Table 1: Clinical presentation.

Histology	Large Neck Swelling	Displacement Of Tonsil	Stridor	Dysphagia	Change in Voice (Hot Potato Voice)	Dyspnoea	Syncope
Schwanomma	10	0	2	2	2	3	0
Pleomorphic Adenoma	7	6	0	1	0	0	0
Mucoepidermoid Carcinoma	6	4	0	1	0	0	0
Carotid Body Tumor	1	0	0	0	1	1	1
Neurofibroma	3	0	2	1	1	0	0
Adenoid Cystic Carcinoma	1	1	0	0	0	0	0

Table 2: Location.

Histology	Pre Styloid	Post Styloid	Anterior To Ica	Posterior To Ica	Lateral To Ica
Schwanomma	0	12	0	12	0
Pleomorphic Adenoma	6	2	6	2	0
Mucoepidermoid Carcinoma	6	1	6	1	0
Carotid Body Tumor	3	0	0	0	0
Neurofibroma	0	5	0	4	1
Adenoid Cystic Carcinoma	1	0	1	0	0

Table 3: Surgical approach.

Histology	Cervical + Transparotid	Cervical	Cervical + Transparotid + Mandibulotomy
Schwanomma	3	9	0
Pleomorphic Adenoma	8	0	0
Mucoepidermoid Carcinoma	4	0	3
Carotid Body Tumor	0	3	0
Neurofibroma	2	3	0
Adenoid Cystic Carcinoma	0	0	1

January 2012 to January 2019.

Thirty six patients aged between 15 years to 55 years with locally advanced parapharyngeal tumors and operated in our institution and having follow up of minimum 1 year were included in this study. Patients with metastatic lymph node presenting as parapharyngeal mass, primary tumors of masticator space only, recurrent tumors and those having history of radiation to neck were excluded from this study. Following an informed written consent by the patient or patient attendants to be included in this study, the medical case records were analysed regarding symptomatology, mode of presentation, clinical findings, airway compromise and involvement of carotid sheath and cranial nerves. The tumors included schwannoma, neurofibroma, pleomorphic adenoma, deep lobe parotid, and carotid body paragangliomas. All patients had contrast enhanced computed tomography and guided fine needle aspiration biopsy.

Surgery was the main stay of treatment for all the patients and the approach was decided based on location and extent of tumor, relationship to important neuro-vascular structures and suspicion of malignancy on FNAB. All patients with severe compromise of airway in oropharynx underwent elective tracheotomy.

Lymph nodes in the parapharyngeal space were sampled and subjected to frozen section. A modified radical neck dissection was done in patients whose lymph nodes were found to be metastatic on frozen section.

Patients whose tumors were found to be high grade malignancy (Mucoepidermoid carcinoma and Adenoid cystic carcinoma) were

administered post operative radiotherapy.

The airway compromise and adverse events during intraoperative and postoperative period were documented and the complications and interventions to manage them along with the outcome were documented. The data was entered on SPSS excel sheet and a detailed descriptive analysis of the above factors was done. An attempt was made to correlate the clinical location of the tumor, imaging findings and histological diagnosis.

Results

The patients in our series were aged between 15 years to 55 years. However 30 (83.3%) patients were below 40 years of age and included 16 females & 20 males. Majority of tumors of neural origin (12 out of 17) were seen in males and tumors of salivary origin (11 out of 16) patients were females.

Majority of patients 28 out of 36 (77.7%) presented with a unilateral neck swelling. 7 (19.4%) presented with dyspnea. Five (13.8%) patients presented with dysphagia. Four patients (11.1%) presented with stridor and respiratory distress and underwent emergency tracheostomy.

Among patients who presented with neck swelling, the size of swelling was more than 5 cm in diameter. Eleven of them had medial displacement of tonsil and among them 4 had a large mass compromising the oropharyngeal lumen and also had change in voice (hot potato voice). Four patients among these 11 had severe compromise of oropharyngeal cavity (Table 1).

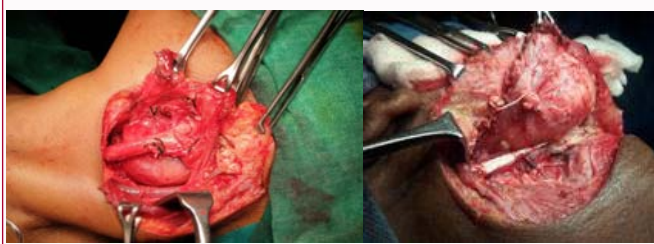
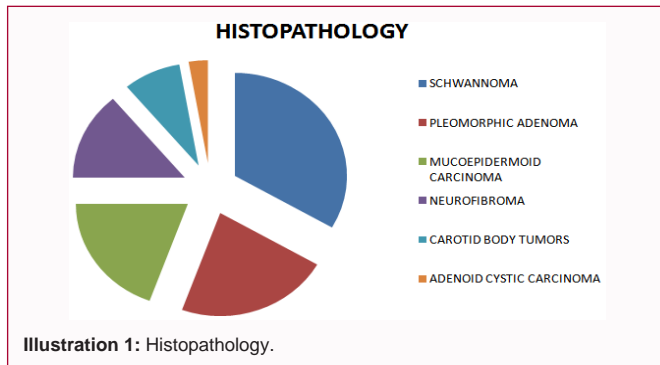


Figure 1: Parapharyngeal tumors accessed by cervical approach, (a) post styloid neurofibroma, (b) pre styloid mucoepidermoid carcinoma.

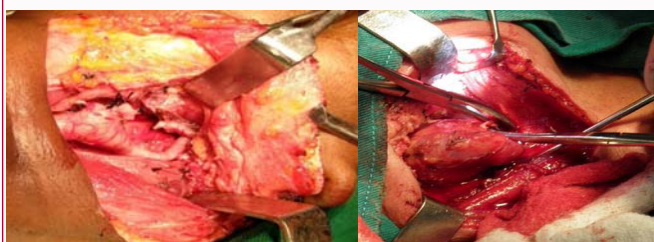


Figure 2a, 2b: Parapharyngeal tumors extending to masticator space accessed by cervical approach.

All patients underwent contrast enhanced computed tomography and guided fine needle aspiration biopsy and preoperative diagnosis was established. On histology 12 patients were found to have schwannoma, 5 had neurofibromas, 8 patients had pleomorphic adenoma, 7 had mucoepidermoid carcinoma, 3 carotid body tumors, and 1 patient had adenoid cystic carcinoma (Illustration 1).

In our series, all patients with schwannomas (33.3%) and neurofibromas (13.8%), and 2 (5.5%) pleomorphic adenomas, 1 (2.7%) mucoepidermoid carcinoma presented in the post-styloid compartment of parapharyngeal space pushing the carotid sheath anterolaterally. None of the patients with paragangliomas in our series were secreting vasoactive amines. Majority of the tumors of salivary gland origin which included 6 (16.6%) pleomorphic adenomas, 6 (16.6%) mucoepidermoid carcinomas, 1 (2.7%) adenoid cystic carcinoma were situated in the pre-styloid compartment. All the 3 carotid body tumors were located at the bifurcation of the common carotid and found to be extending cranially in the parapharyngeal space but the upper extent was well below the angle of mandible (Table 2). All patients in our series were treated surgically. Four patients (11.1%) were found to have large mass severely compromising the airway or causing tracheal shift and underwent elective tracheostomy to secure airway as well as for general anesthesia. The approach was transcervical in 15 patients, transcervical and transparotid in 17 patients and 4 patients had cervical approach with paramedian



Figure 3: Parapharyngeal tumors (a) pre styloid accessed by cervical and transparotid approach, (b) accessed by cervical approach.



Figure 4: (a) Deep lobe parotid tumor accessed by cervical and transparotid and mandibulotomy approach, (b) After removal.

mandibulotomy. Among patients who underwent mandibulotomy, 3 were mucoepidermoid carcinoma and 1 was adenoid cystic carcinoma. All these 4 tumors were extending high into parapharyngeal and masticator spaces (Table 3) (Figure 1-4).

All patients with mucoepidermoid carcinoma underwent ipsilateral modified radical neck dissection. Three of the mucoepidermoid carcinomas were found to be low grade and only had closed follow up postoperatively whereas 4 were found to be high grade and received post operative radiotherapy. One patient with high grade mucoepidermoid carcinoma recurred after 6 years and succumbed to the disease. One patient had neurofibroma of the facial nerve (cervicofacial division), 1 patient had neurofibroma of the cervical sympathetic chain and 1 patient had neurofibroma of vagus nerve. The other 2 neurofibromas took origin from branches of the cervical nerve roots. One patient who was reported as mucoepidermoid carcinoma had facial nerve involvement and facial nerve was intentionally sacrificed and grafted.

In all except 2 patients, the histopathology report confirmed the fine needle aspiration biopsy diagnosis. However, in 2 patients the FNAB had suggested pleomorphic adenoma but histopathology showed low grade mucoepidermoid carcinoma.

The most common complication of surgery was marginal mandibular nerve palsy seen in 8 patients (22.2%). All these patients had transient weakness and recovered after 4 months. Four patients had complete facial palsy. Among them 2 were transient (House Brackman Grade II) and recovered within 2 months. One patient had extensive involvement of facial nerve and hence the nerve was sacrificed. In one patient only the lower division of facial nerve was involved by tumor and hence the nerve was sacrificed and facial nerve grafting was done. This patient improved after 6 months. Two patients (1 schwannoma and 1 neurofibroma from vagus) had vagal palsy. One of these (schwannoma) had a transient palsy and recovered within 6 months. The patient having neurofibroma of vagus also had paralysis of the hypoglossal nerve and required long term tracheostomy with cuff tube and swallowing therapy. All

patients except 1 who had emergency tracheostomy at the time of presentation or elective tracheostomy at the time of surgery were decannulated within the 10th postoperative day. Only the patient who had vagal and hypoglossal palsy (neurofibroma of vagus) continued to be on tracheostomy with cuffed tube. In our series 6 patients underwent postoperative radiotherapy. These included 5 patients with high grade mucoepidermoid carcinoma and 1 patient with adenoid cystic carcinoma.

Discussion

Parapharyngeal tumors are uncommon accounting for about 0.5% to 1% of all head & neck tumors. In our hospital also parapharyngeal tumors accounted for about 0.5% of all head and neck masses. Majority of the patients in our series were in the 3rd decade of life and a few about 50-years-of-age. Other studies in literature also have reported maximum incidence of parapharyngeal tumors in the age group of 40 years to 47 years [1]. The average size of parapharyngeal space tumor was 6 cm ± 3 cm. This was similar to most studies which reported average diameter of 5 cm [2]. Few of our patients presented with tumors of massive size (10 cm × 12 cm). In our series tumors of neural origin were more common in males and tumors of salivary origin and paragangliomas were more common in females. The tumors of neural origin were almost similar in number to tumors of salivary origin. A large systematic review of various series of parapharyngeal tumors showed that the most common tumor was pleomorphic adenoma [1]. However, a large single institution study in United States showed that the most common tumor was schwannoma. This was similar to our study where schwannoma followed pleomorphic adenoma were the most common benign tumors.

In our series only 8% of patients had paraganglioma at the carotid bifurcation. This was in contrast to few western studies which reported higher numbers of paraganglioma both at carotid bifurcation as well as jugular foramen. Unlike the large series reported by Fenglin Sun et al., none of our patients had erosion of skull base by the parapharyngeal tumor [3]. This could be because majority of tumors in our series were benign and low grade malignancies. 78% of the parapharyngeal tumors were benign in our series. This was similar to other studies in literature which have reported that 80% to 85% parapharyngeal tumors are benign [4,5]. 22% tumors in our series were malignant, the majority being mucoepidermoid carcinoma (19.4%). This was in contrast to most other studies which reported adenoid cystic carcinoma to be most common malignancy. One study also reported lymphomas and soft tissue sarcomas in parapharyngeal space. This was not seen in our series. These distributions of benign and malignant tumors were similar to the series reported by Mayo clinic. Other studies have reported lesser percentage of malignant tumors.

In our series, 78% patients presented with unilateral neck swelling which was more than 5 cm on presentation with 27.7% patients having medial displacement of tonsil on clinical examination. Western studies have also shown unilateral neck swelling to be the most common presentation with significant number and having medial displacement of tonsil. However, systematic review done in United Kingdom showed that the most common presentation was intraoral swelling [1]. The second most common presentation being dyspnea in 19.4% followed by dysphagia in 13.8%. Literature from United States showed that majority of the patients were asymptomatic at presentation [2]. In our study we observed that 19.4% patients presented with dyspnea which is a higher in comparison to other

studies. None of our patients had cranial nerve palsy at the time of presentation. In contrast to a study done in Minnesota where 10% patients presented with cranial nerve palsies.

81.25% of the salivary origin tumors were found in the pre-styloid segment and majority were seen anterior to internal carotid artery whereas all the neural tumors were seen in the post-styloid segment displacing the internal carotid artery anteriorly. Other studies have also reported a similar finding [3].

The diagnoses was made in all patients by fine needle aspiration cytology and were conclusive in 94.4% as 2 patients who were diagnosed as pleomorphic adenoma were reported as mucoepidermoid carcinoma after surgery. The role of FNAB in parapharyngeal tumors is controversial as various studies report the accuracy to be between 70% to 100% and it can carry the risk of hemorrhage [1,6]. All the tumors of neural origin were in post-styloid compartment and most of the salivary origin tumors were seen in the pre-styloid compartment. This was similar to observation in other studies.

Most common surgical approach in our series was cervical and transparotid followed by only cervical approach. This was similar to approaches described in the systematic review where cervical with transparotid was the most common approach. However, few large single institution studies had adopted mainly cervical approach. None of the cases in our series had incomplete removal or massive hemorrhage during surgery showing that cervical with transparotid approach gives adequate exposure and working space. Mandibulotomy was reserved for extensive mucoepidermoid carcinoma extending deeply into masticator space (8.3%). A similar approach was adopted by authors in western countries [1-3,7]. None of the patients in our series required transmastoid approach or intraoral approach. This can be explained by the fact that no patient had skull base erosion and most of the tumors were more than 5 cm in diameter. A few western authors have adopted transoral approach for smaller tumors, however this approach is not considered safe by majority of authors. Few studies adopted transmastoid approach in addition to cervical in tumors reaching the jugular foramen [8,9]. This was not required in our study.

A 11% patients underwent an emergency tracheostomy at the time of presentation as all these patient presented with stridor. Elective tracheostomy has been done in 11.1% patients. 90% of the tracheostomized patients had been decannulated within 10 days. Similarly patient who underwent tracheostomy in western countries were also decannulated in 8 days to 10 days [1,2,4]. One patient with neurofibroma of vagus had major aspiration and dysphagia due to hypoglossal and vagal palsy and was not decannulated.

Most common complication of surgery in our series was transient palsy of marginal mandibular nerve and transient palsy of facial nerve in 2 patients. This could have been due to handling or devascularisation of the facial nerve. Similar observation was made by Fenglin Sun et al. [3] who reported that the most common complication was transient facial palsy (House Brackman grade II). However, in 2 patients the facial nerve was sacrificed because of encasement by the tumor. It was grafted in 1 of the patients and showed improvement after 6 months. In these 2 patients the remnant facial palsy was sequelae and not complication. In our series there is low incidence of lower cranial nerve palsy (2 patients - 5.5%) 1 patient with schwannoma who had transient vagal palsy recovered in

3 months and 1 patient with neurofibroma of vagus had permanent vagal and hypoglossal palsy. This is contrast to few western studies which reported higher incidence of lower cranial nerve palsies [10]. This difference in complications between our study and western studies can be explained by the fact that in our series most tumors were benign tumors or low grade malignancies and none of them had eroded skull base.

None of the benign tumors in our series recurred and no radiation was given to benign tumors. This was similar to various other studies in literature. Only 1 patient with high grade mucoepidermoid carcinoma recurred in our series. Literature also quotes very good disease free survival in patients with benign parapharyngeal tumors. However the prognosis in malignant parapharyngeal tumors depends on extent, location and involvement of important neuro-vascular structures.

Conclusion

Locally advanced parapharyngeal space tumors present with large tumors with close proximity to important neurovascular structures and some of them are malignant. Surgery is challenging and the main modality of treatment. Majority of the tumors can be accessed by cervical approach with transparotid approach in few cases. Accurate imaging and meticulous surgery results in good outcome in benign tumors. Majority of complications are transient and nerve grafting holds promise.

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