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Head and Neck Cancer

Surgical Management of Parapharyngeal Tumors: Our Experience

Poonam Joshi[1] Kamal Deep Joshi[1] Sudhir Nair[1] Muddasir Bhati[1] Deepa Nair[1] Munita Bal[2] Amit Joshi[3] Naveen Mummudi[4] Vidisha Tuljapurkar[1] Devendra A. Chaukar[1] Pankaj Chaturvedi[1]

[1]Department of Head and Neck Surgery, Tata Memorial Hospital, Homi Bhabha National Institute, Mumbai, Maharashtra, India
[2]Department of Pathology, Tata Memorial Hospital, Homi Bhabha National Institute, Mumbai, Maharashtra, India
[3]Department of Medical Oncology, Tata Memorial Hospital, Homi Bhabha National Institute, Mumbai, Maharashtra, India
[4]Department of Radiation Oncology, Tata Memorial Hospital, Homi Bhabha National Institute, Mumbai, Maharashtra, India

Address for correspondence Poonam Joshi, MS,Department of Head and Neck Surgery, ACTREC, Tata Memorial Hospital, Homi Bhabha National Institute, Mumbai, Maharashtra, 410210, India (e-mail: drpoonamjoshi80@gmail.com).

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Abstract

Poonam Joshi

Context Tumors of parapharyngeal space (PPS) are rare and histologically diverse. The management of these tumors requires diligent assessment and planning with due consideration of various anatomical and pathological aspects of the lesion.

Aims This retrospective study aims to present our experiences in the clinical and pathological aspects of PPS tumors with a critical evaluation of management.

Settings and Design Retrospective analytical study.

Methods and Material The electronic medical records of 60 cases of PPS tumors, managed surgically from 2007 to 2017, were reviewed and analyzed using SPSS 22 software. The mean follow-up duration was 44 months.

Results The mean age was 45 years with a male-to-female ratio of 1.7 (38:22). The majority of the tumors were benign (71.7%) and the most common presentation being upper neck mass or oropharyngeal mass. Histologically, neurogenic tumors were most common (43.3%) PPS tumors, followed by tumors of salivary gland origin. Magnetic resonance imaging was used as a diagnostic modality in 70% of cases, and computed tomography scan and positron emission tomography/CT were used in 26.7 and 3.3% of cases, respectively. In our study, the diagnostic accuracy of fine-needle aspiration cytology was 71% for benign and 47% for malignant lesions. The most common approach for surgery used was transcervical (72%).

Conclusion The study reveals that cranial nerve palsy is the most common complication associated with PPS tumors. Completely resected, malignant tumors originating within PPS have a good prognosis, as compared with tumors extending or metastasized to PPS.

Keywords
- parapharyngeal space tumors
- surgical approach
- cranial nerve palsies
- deep neck space tumors
- schwannoma

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Introduction

Parapharyngeal space (PPS) tumors are rare, constituting approximately 0.5% of all head and neck tumors with intriguing diverse histology.[1-3] It is also interesting to note that many of the tumors occurring in PPS are not primary PPS pathology, but are metastatic or directly spreading tumors from adjacent structures. The primary PPS tumor can originate in the prestyloid compartment or the poststyloid compartment, which are separated by fascia between the styloid process and the tensor veli palatini muscle.[4] The common tumors of the prestyloid origin are salivary gland tumors, lipomas, lymphomas, and rarely neurogenic tumors. In contrast, most of the tumors of the poststyloid compartment are neurogenic as this compartment contains the internal jugular vein, carotid artery, vagus nerve, and sympathetic plexus.[5] Most of the PPS tumors are benign and only 10 to 20% of these are malignant.[6] The management of tumors of PPS is challenging. Presently, surgery with an appropriately chosen approach is the primary modality for the treatment and radiotherapy is utilized for the management of unresectable tumors, as an adjuvant treatment, or in patients with a high risk of injury to cranial nerves during surgery.[7]

There is paucity of adequate literature on this disease entity. The rarity of the diseases precluded conducting of prospective studies with high level of evidence. The present study was done with an aim to present our experiences in various clinical and pathological aspects of PPS tumors with a critical evaluation of management.

Subjects and Methods

The present study is a retrospective analysis of 60 cases of PPS tumors managed surgically from 2007 to 2017. After obtaining ethical clearance from the institutional review board, the complete medical records were obtained from the electronic medical record system of the institute. The demographical, clinical, and pathological details and management with surgical details were recorded. Any preoperative and postoperative complications during follow-up were also recorded. Analysis of the collected data was done using SPSS 22 software.

Results

A total of 60 patients who underwent surgical resection of PPS tumors from the year 2007 to 2017 were included in the study. The demographical details are given in ►Table 1.

The most common presentation was swelling/mass in the parotid or upper neck region in 36 (60%) cases. Twenty-four (40%) patients presented with oropharyngeal (tonsillar) or soft palate bulge. Dysphagia was the presenting symptom in 14 (23.3%) patients, while 6 (10%) patients had a change in voice. Pain was the chief complaint in 5 (8.3%) patients. Other rare presentations were trismus (2 cases), foreign body sensation, and hearing loss (1 case each).

The primary imaging modality utilized for evaluation was magnetic resonance imaging (MRI) with contrast in 42 (70%) cases, followed by contrast-enhanced computed tomography (CECT) scan in 16 (26.7%) cases and positron emission tomography/CT (PET/CT) scan in 2 (3.3%) cases. CT angiography was done for two patients along with MRI for suspected vascular tumor.

Table 1 Demographic details

	Number	Percentage
Total number of cases	60	
Gender		
Male	38	63.33%
Female	22	36.67%
Male: Female ratio	1.7:1	
Mean age in years (range)	45.22 (22–82)	
Treatment naive	53	88.3%
Recurrent cases	7	11.66%
Location		
Prestyloid	39	65%
Poststyloid	21	35%
Mean follow-up duration in months (range)	44 (4–161)	
Deaths during follow-up	2	3.3%

A total of 42 patients underwent fine-needle aspiration cytology (FNAC) for histological evaluation. The diagnostic accuracy of FNAC was 71% for benign lesions and 47% for malignant lesions. FNAC was nondiagnostic in 13 (30.9%) patients, and the majority of them were schwannomas (53.8%). On the final histopathological assessment, 43 (71.7%) were benign and 17 (28.3%) were malignant. The histological distribution of various tumors of PPS is given in ►Table 2.

Among the tumors of neurogenic origin, schwannoma was the most common tumor. Pleomorphic adenoma was the most common tumor of salivary gland origin and the majority was present in the prestyloid compartment. Other histopathologies with their distribution are given in ►Table 2.

Surgery was the treatment modality in all the cases. The most common surgical approach was transcervical, which was used in 43 patients. The combination of transcervical and transparotid approach was used in eight patients, a combination of transmandibular and cervical approach in six patients, and transoral robotic surgery (TORS) was performed in three patients.

Adjuvant radiotherapy/concurrent chemoradiotherapy was advised to 11 patients out of 16 patients having malignant tumors. A total of nine patients could complete the treatment and two deaths were reported during the treatment. At the end of the follow-up duration, seven patients were disease-free and two patients had developed nonsalvageable recurrence after adjuvant therapy. None of the patients with the benign histology had disease recurrence during follow-up period.

Table 2 Tumors of PPS: histological distribution

Type (number, percentage)		Benign (number)	Malignant (number)
Neurogenic origin (26, 43.3%)	Prestyloid	Nil	Nil
	Poststyloid	Schwannoma (22) Neurofibroma (1) Vagal paraganglioma (2) Meningothelial meningioma (1)	
Salivary origin (24, 40%)	Prestyloid	Pleomorphic adenoma (13)	Mucoepidermoid carcinoma (2) Myoepithelial carcinoma (1) Carcinoma ex pleomorphic (1) Synovial sarcoma (1) Adenoid cystic carcinoma (1)
	Poststyloid	Pleomorphic adenoma (1)	Mammary analogue secretory carcinoma (1) Myoepithelial carcinoma (2) Salivary duct carcinoma (1)
Miscellaneous (10, 16.7%)	Prestyloid	Fibromatosis (1) Lymphovascular malformations (1)	Metastasis from papillary carcinoma (3) Metastasis from medullary thyroid carcinoma (1)
	Poststyloid	Solitary fibrous tumor (1)	Metastasis from papillary carcinoma (1) Metastasis from medullary thyroid carcinoma (1) Metastatic squamous cell carcinoma (1)
Total		43 (71.7%)	17 (28.3%)

Abbreviation: PPS, parapharyngeal space.

Cranial nerve palsies/paresis were the most dreaded complication of PPS tumors. A total of 6 patients who presented with cranial nerve palsies in the preoperative period, did not show any recovery during follow-up. Cranial nerve palsies were present in 16 patients in the postoperative period. The most commonly involved cranial nerves were facial, lower cranial nerves (IX, X, XI, XII), and sympathetic chain. The most common neurological complication was marginal mandibular nerve paresis, seen in eight cases. One-half of these cases (4/8) improved with physiotherapy. A list of cranial palsies in the preoperative and postoperative period in the present study is given in ►Table 3.

Discussion

The PPS is described as an inverted pyramid, with the base at the skull base and apex oriented toward the hyoid bone. This anatomical space is seated deeply, surrounded by other anatomically described spaces. The anatomy of PPS is complex and is neither easily accessible for clinical assessment, nor a direct surgical approach is possible. The tumors arising from PPS are most challenging for head and neck surgeons due to their rare occurrence, late presentation, difficulty in establishing a diagnosis, varied histopathology, and finally due to their complex surgical management.

The present retrospective analysis was done to share the clinical experience of these rare tumors managed at Tata Memorial Hospital, Mumbai, Maharashtra, India. In our experience, the mean age of presentation and sex distribution is comparable to preexisting literature.[5]

PPS tumors have an indolent course and remain asymptomatic for long periods. Riffat et al have given a comprehensive analysis of presenting symptoms in their systemic review of PPS tumors. The most common presentation is the cervical or oropharyngeal mass lesion.[5,6] In our study, the cervical mass was the most common presenting symptom followed by the tonsil and soft palate submucosal bulge. Other presenting symptoms noticed in our study were attributed to mass effect or cranial nerve deficits.

Imaging has an important role in diagnosis and guiding the management plan for the PPS tumors. The various imaging modalities available for PPS tumors include MRI, CECT scan, and angiography and these are complementary to each other. The origin of the lesion can have an implication on the choice of surgical approach. Imaging can be helpful in determining the origin of the tumor by assessing the displacement pattern of fat in PPS.[7] There are few advantages of MRI, which is considered superior to CT scan in providing better soft tissue contrast resolution between PPS masses and surrounding fat, while the spatial resolution of CT scan delineates the vital structures well. The MRI enables a better assessment of margins, stage of the disease, and fat infiltration. It is also useful in cases with an equivocal CT scan and to assess skull base or perineural spread.[8]

Angiography is particularly useful for vascular and enhancing lesions. The role of PET/CT is limited as a diagnostic modality; however, it can be used to rule out distant metastasis in malignant tumors.[9]

In our study, primary modality, MRI was done in 42 patients, while CECT was done in 16 patients. PET/CT was done in only two patients presenting with metastases.

FNAC may have an important role in guiding the surgeon especially in malignant cases for complete extirpation of tumor and can aid in selecting the approach as well. This can lead to preoperative and postoperative planning, prognostication, and counseling of the patients. In our study,

Table 3 Preoperative and postoperative nerve palsies

Serial no.	Histopathology	Cranial nerve palsy			Remarks
		No. of patient	Preoperative	Postoperative	
1	Carcinoma ex pleomorphic	1	VII	IX	Lower branches of VII nerve and IX nerve sacrificed
2	Metastasis from papillary carcinoma	1	X	XII	Left superior laryngeal nerve and left hypoglossal nerve sacrificed
3	Mucoepidermoid carcinoma	1		VII	Facial nerve - lower trunk sacrificed
4	Adenoid cystic carcinoma	1		VII	Marginal mandibular nerve paresis
5	Pleomorphic adenoma	4		VII	Marginal mandibular nerve paresis
7	Salivary duct carcinoma	1	VII		Facial nerve trunk involved by the tumor and sacrificed
8	Vagal paraganglioma	1	X		Left vocal cord palsy
9	Vagal paraganglioma	1		X, Horner's syndrome, XII	X nerve palsy
10	Schwannoma	1		IX, Horner's syndrome, VII	Originating from IX nerve and hence excised, marginal mandibular nerve paresis
10	Schwannoma	1	Horner's syndrome		Sympathetic trunk origin and engulfed by tumor and sacrificed
11	Schwannoma	1		Horner's syndrome	Sympathetic trunk origin and engulfed by tumor and sacrificed
12	Schwannoma	1		X	Left vocal cord palsy
13	Schwannoma	1	Horner's syndrome	X	Recurrent schwannoma sympathetic trunk
14	Schwannoma	1		VII	Recurrent schwannoma marginal mandibular nerve paresis
15	Schwannoma	1		VII	Marginal mandibular nerve paresis
16	Schwannoma	1		VII	Facial nerve sacrificed
	Total patients	19	6	16	

the diagnostic accuracy of FNAC was 71% for benign PPS tumors and 47% for malignant PPS tumor. The diagnostic accuracy of FNAC for benign PPS tumors was higher compared with diagnostic accuracy for malignant tumors. It is to be noted that nondiagnostic cases were not included in statistical analysis for measuring diagnostic accuracy. In our study, the higher nondiagnostic results for benign pathologies can be attributed to the FNAC being hemorrhagic in paragangliomas and hypocellular in schwannomas.

On the final histopathological assessment, 43 (71.7%) cases were benign and 17 (28.3%) were malignant. In our series, schwannoma was the most common pathology (22 cases), followed by pleomorphic adenoma (14 cases). In a systematic review, Riffat et al have shown that among the 70 different histologies reported in 1,118 primary cases, 82% were benign and 18% were malignant. The most common pathology reported was pleomorphic adenoma.[6]

The proximity of PPS tumors to neurovascular structure often leads to menacing neurological complications, which can be seen in the preoperative or postoperative period. It is observed that preoperative nerve palsies are less symptomatic as compared with postoperative nerve palsies, most probably due to slow onset and activation of rehabilitative mechanisms. In malignant pathologies, usually, there is early involvement of nerve due to infiltration, whereas in benign pathologies, tumor attains large size before palsy is even apparent. In our series, patients presenting with preoperative cranial palsies did not improve after surgery. A list of preoperative and postoperative cranial nerve palsies is given in ►Table 3.

Various surgical approaches that can be used for excision of the parapharyngeal tumors are transcervical, combined transcervical-transparotid, transmandibular, transoral, and transoral robotic approach. The choice of approach is guided by the location, size, histology, and relationship with various anatomical structures.[1,3,10,11] Various facets of these approaches are described in the subsequent paragraphs.

In our study, the transcervical approach was used in the majority (72%) of the patients. However, this approach can be used for tumors with limited cranial extension and not involving the parotid gland. The combined transcervical-transparotid approach was used in lesions arising from the parotid gland. This approach has the advantage of better exposure of the facial nerve, but cannot be used for tumors greater than 4 cm in size.[6] The transcervical-transmandibular approach was used in cases where the tumor was reaching up to the skull base in the region of the foramen ovale or was in close proximity to the styloid process.

The transoral approach is minimally invasive but offers the least exposure and used in none of the patients in the present study. The endoscopic-assisted transoral approach can increase the visibility, provide better hemostatic control, and decrease complications.[12] The TORS has the advantage of being minimally invasive like the transoral approach but provides better exposure with augmented reach.[11] Boyce et al have concluded that the TORS is safe and effective, but has the limitation of reach in far lateral and superior areas of PPS.[13] In the present study, robotic approach was used for two cases of pleomorphic adenoma that had submucosal palatal swellings and one case of schwannoma arising from the posterior pharyngeal wall.

A treatment algorithm by Chu et al, suggest that the TORS can be an approach of choice in a well circumscribed tumor with a clear plane of cleavage from neurovascular bundle and has displaced internal carotid artery laterally. It can safely accomplish the excision if uncontrolled bleeding or tumor spillage is not present. However, it should not be utilized if preoperatively trismus, infiltration of surrounding tissue, or medial displacement of the internal carotid artery is detected.[14]

In conclusion, PPS tumors arise from an anatomically complex area and have heterogeneous histopathology. Most of these tumors are benign, indolent in nature, and usually become symptomatic when they attain large dimensions. Multiple cranial nerve palsies are often associated with tumors of neurogenic origin arising from the poststyloid space. The MRI is the best imaging modality to assist in the diagnosis; however, the CT scan and angiography are complementary. The FNAC has low accuracy for malignant lesions in our series. Surgery is the primary modality of treatment and the majority of these tumors are accessible through transcervical approach. The aim of surgery should be complete resection of the tumor with minimum morbidity. Malignant tumors originating from PPS when resected completely have a better prognosis but those with metastasis to PPS have a poorer prognosis.

Key Messages

The parapharyngeal space (PPS) tumors are histologically diverse and rare tumors. The article presents a single-institutional experience on clinical aspects and management challenges of PPS tumor in one of the largest case series (60 cases).

Conflict of Interest

None declared.

Acknowledgment

None.

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