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ORIGINAL ARTICLE

A Study of Primary Parapharyngeal Space Tumors in a Tertiary Care Center

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ABSTRACT

Objectives: This study was conducted to evaluate the demographic profile, clinicopathological features, and surgical approaches used to treat primary parapharyngeal space tumors.

Materials and methods: This was a retrospective study of the primary parapharyngeal space tumors treated surgically from April 2012 to March 2015 in a tertiary care teaching center. The study population included 16 cases. The inflammatory lesions and secondary metastasis in parapharyngeal space were excluded. The clinicopathological features, surgical management, and outcome of all the cases were analyzed.

Observations: The gender distribution was 56.2% males and 43.7% females. The median age was 38 years with range from 16 to 62 years. The most common presenting symptom was neck swelling in 87.5% cases, and oropharyngeal bulge was the most common examination finding in 100% cases. Magnetic resonance image was done in 87.5% cases and contrast computed tomography (CT) scan in 31.25% cases, while 25% cases underwent both. The tumor was in the pre-styloid compartment in 62.5% cases and post-styloid in 31.25% cases. In 75% cases, cytology was done directly, while in 25% cases, CT–guided cytology was performed. Cytological diagnosis was histologically correlated in 81.2% cases while changed in 18.7% cases. On histology, 87.5% cases were benign and 12.5% were malignant. The most common histological variant was pleomorphic adenoma in 56.25% cases. The most common surgical approach used was transcervical in 75% cases. Mean tumor size was 7 cm. Complications occurred in 12.5% cases.

Conclusion: Imaging modalities in combination with fine needle aspiration cytology are a very good diagnostic tool before planning for intervention. The transcervical approach is an excellent technique to deal with small to moderate-size tumors and even for large well-defined tumors.

Keywords: Parapharyngeal space tumor, Pleomorphic adenoma, Transcervical approach.

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INTRODUCTION

Primary parapharyngeal space (PPS) tumors are rare neoplasms representing only 0.5% of all the head and neck neoplasms.[1] Approximately 80% of PPS tumors are benign and the remaining 20% are malignant. They often pose therapeutic and diagnostic problems due to variable nonspecific symptoms and complex anatomy of the region. Most of the lesions of this region are salivary gland neoplasm followed by neurogenic tumors, paraganglioma, and a variety of uncommon lesions.[2-4] A literature review by Riffat et al,[5] which included 1,143 cases of PPS tumors, found that most frequently these lesions presented as a cervical or an intraoral mass (50% and 47% respectively). Pleomorphic adenomas were the most common primary lesion (34%). Because PPS tumors are located deep within the neck, clinical examination and palpation are limited and unreliable.[6] With the advent of cross-sectional imaging studies incorporated along with guided fine needle aspiration cytology (FNAC), interpretation of the nature of these PPS lesions have definitely proved to be of much help.[7] Clinical examination of PPS is difficult, and therefore, computed tomography (CT) and magnetic resonance image (MRI) are essential to delineate the tumor extent, spread, intracranial involvement, and relationship to the adjacent vital structures. Computed tomography scanning and MRI investigations are complementary, and both studies should be performed for the evaluation of PPS. Imaging also helps in planning the surgical procedure. Although histopathology remains the gold standard, owing to the difficult surgical approach of tumors in this region, guided FNAC has proved to be useful both in terms of morphological analysis and efficacy with regard to cost and time. When lesions are readily accessible or palpable in the retromandibular or cervical area or transorally, FNAC may aid in surgical and postoperative planning.[8] Biopsy of the PPS mass should be performed after obtaining the results

of radiologic studies. Complete surgical excision is the mainstay of treatment for tumors of the PPS.

MATERIALS AND METHODS

This retrospective study was carried out in the tertiary care teaching hospital. It comprised of 16 patients with primary PPS tumor treated successfully from April 2012 to March 2015. Only those patients who presented with mass primarily in PPS and treated surgically were included. The inflammatory lesions and secondary metastasis in PPS were excluded in the present study. The clinical features, histological and surgical management, and outcome of all the 16 patients were analyzed in a retrospective manner. Radiological imaging in the form of either CT or MRI was performed in all patients. And FNAC was done directly from the retromandibular or transcervical area or transorally in readily accessible lesions, and CT-guided FNAC was performed in the four cases and a cytologic diagnosis was made. Confirmation was done by histopathology in all the cases. Depending on the location of the tumor in PPS, the cases were classified as pre-styloid and post-styloid space tumors. All patients underwent surgical excision, and postoperatively, patients were followed up till 6 months. All operated cases were approached externally. The study also emphasizes the approach used in accessing the tumor and its complications.

RESULTS

The total numbers of patients were 16. Out of these, 9 (56.2%) cases were males and 7 (43.7%) were females. There was a male predominance. Age ranged from 16 to 62 years, and the average mean age was 38 years. In our study, maximum patients fell within the age group 40 to 50 years. The most common presenting symptom was neck swelling in 14 (87.5%) cases followed by oropharyngeal swelling in 10 (62.5%) cases, and the most common examination finding was oropharyngeal bulge seen in all the 16 (100%) cases followed by cervical swelling in 12 (75%) cases (Fig. 1). Table 1 shows the presenting symptoms and examination findings. One case presented late with acute respiratory obstruction. An MRI (Fig. 2) was done in 14 (87.5%) cases and contrast CT scan in 6 (31.25%) cases. Four (25%) cases underwent both CT and MRI scan. In 10 (62.5%) cases, the tumor was in the pre-styloid compartment and in 6 (31.25%) cases in the post-styloid compartment. No adjacent bony erosion and intracranial extension were found in all the cases. In 12 (75%) cases, FNAC was done directly from readily accessible lesions, while in 4 (25%) cases, CT-guided FNAC was performed and a cytologic diagnosis was made. Table 2 shows the correlation between the cytologic and histologic diagnosis. In our study, out of 16 cases, 14 (87.5%) were benign, while 2 (12.5%) were malignant on histology. Out of 16 cases, cytological diagnosis was histologically confirmed in 13 (81.2%) cases, while cytological diagnosis was changed in 3 (18.7%) cases. Two cases diagnosed as adenoid cystic carcinoma on cytology later on turned out as pleomorphic adenoma and carcinoma ex-pleomorphic adenoma on histopathology. One case

Fig. 1: Clinical photograph showing left cervical swelling

Table 1: Presenting symptoms and clinical signs

Sl. no.	*Symptoms*	*No. of cases (%)*	*Clinical signs*	*No. of cases (%)*
1	Neck swelling	14 (87.5)	Oropharyngeal swelling/bulge	16 (100)
2	Oropharyngeal swelling	10 (62.5)	Cervical swelling	12 (75)
3	Muffled voice	5 (31.2)	Parotid swelling	5 (31.2)
4	Dysphagia	5 (31.2)	Bimanual ballottement	2 (12.5)
5	Acute airway obstruction	1 (6.2)	Decreased mouth opening	1 (6.2)

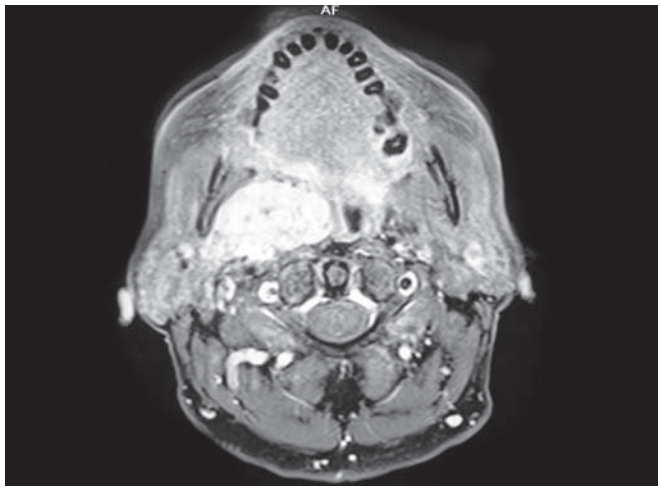

Fig. 2: T1-weight MRI axial section showing mass in right parapharyngeal space

Table 2: Correlation of cytology with histopathology and types of tumor

Sl. no.	*Types of parapharyngeal tumor*	*Cytology (FNAC)*	*Histopathology*	*Percentage*
1	Pleomorphic adenoma	8	9	56.25
2	Schwannoma	4	5	31.25
3	Mucoepidermoid carcinoma	1	1	6.25
4	Adenoid cystic carcinoma	2	0	0
5	Benign spindle cell neoplasm	1	0	0
6	Carcinoma ex-pleomorphic adenoma	0	1	6.25

diagnosed as benign spindle cell neoplasm on cytology turned out to be schwannoma on histopathology. All the patients underwent surgical excision by different external surgical approaches (Table 3). The most common surgical approach that was done was transcervical in 12 (75%) cases (Fig. 3) followed by the transcervical-transparotid approach in 3 (18.7%) cases, while in 1 (6.2%) case, a large PPS tumor was excised by transcervical with mandibulotomy. One case underwent emergency tracheostomy for large tumor presenting as upper airway obstruction,

Table 3: Type of surgical approach used

Sl. no.	*Surgical approach*	*No. of cases operated (%)*	*Complications*	*Recurrence on follow-up*
1	Transcervical	12 (75)	Nil	Nil
2	Transcervical and transparotid	3 (18.7)	Pharyngeal abscess – 1 Facial weakness – 1	Nil
3	Transcervical with mandibulotomy	1 (6.2)	Nil	Nil

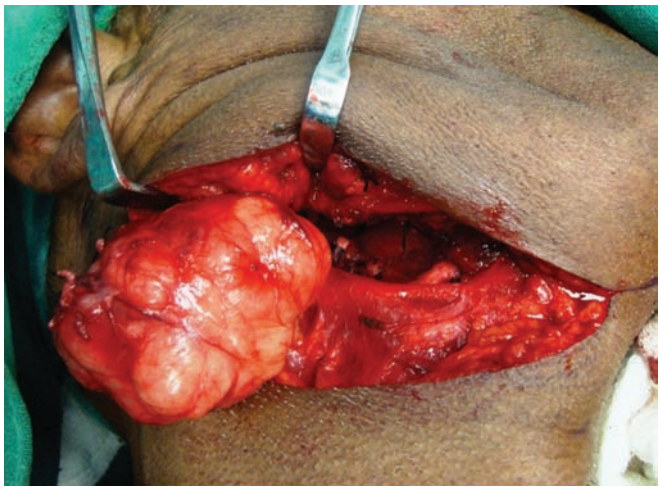

Fig. 3: Tumor being delivered by transcervical approach

Fig. 4: Excised tumor specimen

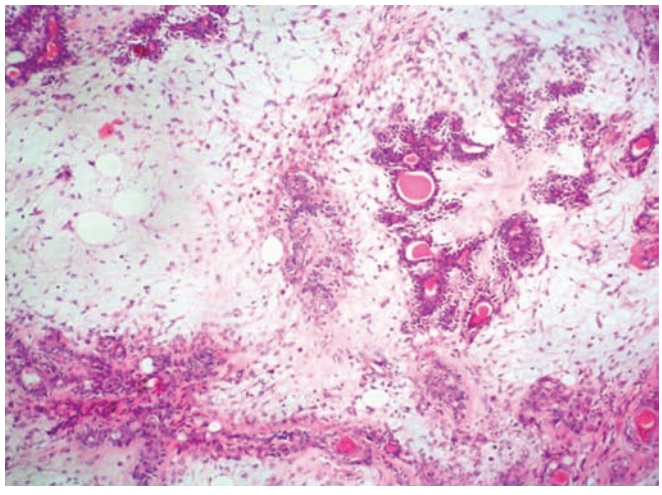

Fig. 5: Microphotography (H&E × 40) showing pleomorphic adenoma

and after successful excision of the tumor, the patient was decannulated on 7th postoperative day. The mean tumor size in our study was 7 cm (4–12 cm) (Fig. 4). The most common histological variant that we found in our study was pleomorphic adenoma in nine (56.25%) cases (Fig. 5) followed by schwannoma in five (31.25%) cases. Complications occurred in two (12.5%) patients. One patient developed wound infection and presented as pharyngeal abscess after 1 week of surgery. Facial weakness of lower trunk was present in one case due to tumor involvement, which had to be sacrificed during surgery as the cytological diagnosis of patient was mucoepidermoid carcinoma. One (6.25%) patient received adjuvant radiotherapy that was diagnosed as mucoepidermoid carcinoma. On follow-up, no recurrence was reported till the last follow-up.

DISCUSSION

Primary parapharyngeal space tumors are rare, and clinical detection of these tumors in the initial stage is difficult. The tumor has to grow to 2.5 to 3.0 cm to be detected

clinically, and generally, these tumors tend to present late, attaining an impressive size. Meticulous intraoral and cervical examination including bimanual palpation give the clinician a rough impression of the size of the tumor. Generally, it presents as an asymptomatic mass causing mild bulging in the soft palate or tonsillar region, or fullness near the angle of the mandible. The ensuing symptoms depend on the affected site. The pre-styloid lesion can present as serous otitis media, voice change, nasal obstruction, aspiration, or dyspnea. The post-styloid lesion can compress the lower four cranial nerves and cause hoarseness, dysphagia, dysarthria, or Horner's syndrome. Cranial nerve palsy, pain, and trismus often suggest malignancy. Trismus indicates infiltration of pterygoid muscles in malignancy or mechanical obstruction to the coronoid process of the mandible. The most common age group of presentation as documented in various literatures is 4th to 5th decade.[9,10] In our study, the most common age group involved was 40 to 50 years. In our study, majority of the cases presented with neck swelling (87%) followed by oropharyngeal swelling (62.5%). On examination, oropharyngeal bulge was seen in all the cases in our study as the tumor size in our study was more than 4 cm in all the cases. This correlates with various published studies in the literatures. The FNAC is an important and essential investigation with a 92% accuracy rate. It can be improved even further to 95% by combining with CT, which is essential for lesions that are not directly detectable without imaging techniques.[11] In our study, FNAC overall correctly detected the diagnosis in 87.5% cases, which was was comparatively lower than various reports in the literature; probably, the reason may be limited exposure of the cytologist to these rare lesions. Imaging studies, such as CT, MRI, and angiogram are important modalities to support the diagnosis of parapharyngeal tumors and distinguish pre-styloid tumors from those in the post-styloid space. They help to know the site of origin, size, and extent of the tumor and involvement of the surrounding structures by the tumor. An MRI delineates soft tissue density better. The relationship between the tumor and the carotid artery can be more clearly seen with MRI. Evaluation of PPS tumors is mostly radiographic, with CT scanning and MRI offering useful diagnostic information.[10] Appropriate diagnosis can be reached radiographically in 95% of patients without tissue biopsy.[12] In our study, imaging modalities in combination with FNAC were diagnostic in 81.2% of the cases. In 18.7% cases, where these failed, histopathology confirmed the final diagnosis. Hughes[13] reviewed 172 patients with parapharyngeal space neoplasm and found that pleomorphic adenoma was the most common neoplasm (40%), followed by paraganglioma (20%) and neurogenic tumor (14%) respectively. This result is consistent with most reports. In our study, a different pattern of tumor distribution was observed probably due to less number of cases. However, there are few literatures reporting neurogenic tumors as the most frequent entities. In one of the most recently published study on primary parapharyngeal tumors, Luna-Ortiz et al[14] reported that neurogenic tumors represented 57% of all tumors in his study population. Pleomorphic adenoma was the most common salivary gland tumor of the PPS observed in our study. The frequency of malignant parapharyngeal salivary neoplasm varied greatly in the literature. In our study, mucoepidermoid carcinoma and carcinoma ex-pleomorphic adenoma were two malignant tumors. In most of the reports, neurogenic tumor is the 2nd most common tumor in the PPS.[5] Schwannoma is the most common type of neurogenic tumor.[5] Similar observations were made in our study. In our study, paraganglioma and neurofibroma were not reported probably due to less number of cases. Definitive surgical treatment will be considered first as this is the most common treatment modality for maximum primary PPS lesions. It may be essential that the planned approach must be adequate for a full oncological resection as 2nd look surgery in this space is difficult. The goal of parapharyngeal surgery is to provide adequate tumor visualization to achieve complete tumor removal, while preserving the surrounding nerves and vessels and controlling any hemorrhage. Many surgical approaches have been reported in the literature. Overall, transcervical and transparotid are the two main approaches. They have been reported to be used alone or in combination with each other. They have also been used with mandibulotomy to increase exposure. Transcervical approach is preferred in the post-styloid space tumors. This approach frequently involves blind finger dissection in the PPS and does not provide enough exposure for larger benign lesions extending cranially or those with a more aggressive growth pattern. Exposure in this approach is limited superiorly by the angle of mandible, and this approach is suitable only for small to moderate tumors. Tumors extending near the base of the skull cannot be managed by this approach due to limited exposure.[15,16] The transcervical approach can be combined with mandibulotomy for larger tumors. Olsen[17] recommended mandibulotomy for vascular tumors extending into the superior PPS, solid tumors that are confined to the superior aspect of the PPS. The transparotid approach is commonly used for parotid deep lobe tumor and pre-styloid tumors. Many surgeons prefer the transcervical-transparotid approach. Olsen[17] reported using this technique in 80% of his case series. He recommended this approach for all deep-lobe parotid tumors, extraparotid salivary tumors, and most of the post-styloid neurogenic tumors. Hughes[17] also

reported the transcervical-transparotid approach as the preferred procedure for removal of most parapharyngeal tumors in his review of 172 patients with primary parapharyngeal tumors. The transpalatal approach was considered only for small benign tumors, which are extra-parotid and non-vascular since exposure is very limited and adequate control of great vessels is quite difficult. The transoral approach provides poor access and poor visualization and is not a recommended approach. Combined infratemporal fossa and transfacial approach is a new alternative approach for massive tumors within the infratemporal fossa and pterygopalatine fossa. This provides excellent control of internal carotid artery without leaving facial scars. In our study, all the patients underwent surgical excision by different external surgical approaches. The most common surgical approach used was transcervical in 75% cases followed by the transcervical-transparotid approach in 18.7% cases, while in 6.2% cases, a large tumor was excised by the transcervical with mandibulotomy approach. While surgery is the mainstay of the treatment for parapharyngeal tumors, radiation therapy should be considered in elderly patients with paragangliomas. In our study, one case underwent adjuvant radiotherapy for malignant tumor. Shahab R et al recently published their review of 114 parapharyngeal tumors for over 27 years of experience, the 2nd largest study in the literature. The 5- and 10-year survival for benign parapharyngeal tumor was 100%. For malignancies, the 5-year survival was 93%, but reduced to 57% at 10 years.[18] This study showed that a patient is highly unlikely to die of a benign PPS tumor; therefore, careful consideration of surgical treatment and discussion with the patient are crucial. In our study, all cases were doing well and on follow-up, no recurrence was reported. The follow-up period was ranged from 6 months to 2 years.

CONCLUSION

Imaging modalities in combination with FNAC are a very good diagnostic tool before planning for surgical intervention. Surgery is the best way to treat PPS tumors. The choice of surgical approach is dictated by the size of the tumor, its location, and its relationship to the great vessels. The transcervical approach is a very good technique to deal with small to moderate-size tumors and even for large well-defined tumors. A majority of the tumors are benign, with pleomorphic adenoma being the most common tumor followed by schwannoma.

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