Parapharyngeal Space Tumors: Our Experience In A Tertiary Hospital In Andhra Pradesh, India
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Citation

Abstract
Introduction
Parapharyngeal space tumors are rare, accounting for 0.5% of all head and neck tumors; 80% of them are benign. According to the site of tissue origin, parapharyngeal space tumors are mainly grouped into four varieties i.e., salivary tumors, neurogenic tumors, lymphoid masses and paragangliomas.

Material and methods
Our study period was 5 years. During this time, 13 patients were operated for parapharyngeal space tumors. The age ranged from 14-70 years with the mean age being 42 years. Male-to-female ratio was 1.16:1. All patients presented with neck mass. Clinical diagnosis was uncertain in cases of neural origin. Routine investigations and fine-needle aspiration cytology were done for all cases. Imaging studies were done when the fine-needle aspiration cytology report was inconclusive.

Results
Out of 13 cases operated, 5 were salivary neoplasms, 3 neurogenic tumors and 5 lymph node masses. Out of the 5 salivary neoplasms, 2 were reported as pleomorphic adenomas, 1 was a duct cell carcinoma and 1 a schwannoma. Four cases were arising from the parotid gland; one case was reported as plexiform neurofibroma which was arising from the submandibular salivary gland. Out of 3 neurogenic tumors, 2 were reported as neurofibromas and the remaining one was a ganglioneuroma arising from the vagus nerve. Out of 5 lymph node masses, 2 were metastatic masses from papillary carcinoma and 1 was a metastatic mass from medullary carcinoma of the thyroid gland. The remaining 2 were metastatic masses from squamous cell carcinoma. Four cases presented with occult primary while the medullary carcinoma of the thyroid presented with a small nodule in the right lobe. Fine-needle aspiration cytology was diagnostic in pleomorphic adenomas and secondary lymph node masses. It was inconclusive in diagnosing neurogenic tumors. Computerized tomography was done in complicated cases when fine-needle aspiration cytology was inconclusive. It helped in locating the site and extension of the tumor and in delineating its plane. Ultrasonography proved helpful in locating the tumor in the thyroid gland.

Conclusion
Parapharyngeal space tumors are rare accounting for only 0.5% of all head and neck tumors. There are difficulties in clinical assessment as they are deep seated. Fine-needle aspiration cytology has limited value in neural tumors. Radiological evaluation is essential to minimize the intraoperative risk. Excision biopsy is advisable in view of the complex anatomy of the space. It is diagnostic and curative.

INTRODUCTION
Parapharyngeal space tumors are rare, accounting for 0.5% of all head and neck tumors (1). They are deep seated masses on the lateral wall of the pharynx; 80% of them are benign tumors, difficult to diagnose clinically, and biopsy is also hazardous because of the complex anatomy. Excision biopsy is both diagnostic and curative.

The parapharyngeal space is a cone-shaped space extending from the jugular foramina at the base of the skull to the hyoid bone below. Medially the space is bounded by buccopharyngeal fascia and superior constrictor muscle. Laterally, it is formed by the ramus of the mandible and posteriorly by vertebrae and prevertebral muscles. The parapharyngeal space is further divided into prestyloid and poststyloid spaces by the tensor-styloid fascia which contains the vascular bundle of the ascending palatine artery.

Anatomically, the space is very important and potentially dangerous because it is very narrow and contains carotid
vessels, jugular vein, 9th, 10th, 11th and 12th cranial nerve and the cervical sympathetic chain. Optimal care in preoperative evaluation is essential to avoid injury to the above structures during the surgical procedure.

Parapharyngeal space tumors are mainly grouped into four types as per tissue of origin i.e., salivary tumors, neurogenic tumors, lymphoid masses and paragangliomas. Tumors of salivary origin occupy the prestyloid region and comprise 40-50% of the parapharyngeal tumors (2). They include parotid, submandibular and minor salivary glands.

Neurogenic tumors occupy the poststyloid region and comprise 25-30% of the parapharyngeal tumors. These include schwannomas, neurofibromas, ganglioneuromas and paragangliomas.

Paragangliomas are benign neoplasms arising from paraganglion or extraadrenal neural crest tissue. Two per cent of paragangliomas secrete catecholamines. Lymph node masses are mostly secondary metastatic lesions. Metastasis is either from thyroid tumors or from malignancies of the upper aerodigestive system.

MATERIAL AND METHODS

A retrospective study was carried out in the department of General Surgery, Kamineni Institute of Medical Sciences, Narketpally-Nalgonda District of Andhra Pradesh, from April 2006 to June 2011. The study includes 13 patients, who were diagnosed to have tumors arising from the parapharyngeal space. All patients were examined clinically to evaluate the tumor location, extension, plane and intraoral extension. Neurological examination was also carried out for the involvement of cranial nerves and cervical sympathetic chain. Apart from routine haematological and urine investigations, fine-needle aspiration cytology was done in all cases. Computerized tomography was done in 2 complicated cases where fine-needle aspiration cytology failed to confirm the diagnosis. It confirmed the location, plane of the swelling and relation to great vessels. (Figure 1)

RESULTS

In this study, patients were within the age group of 15-70 years with the mean age being 42 years. Three patients were in the 2nd decade (25%), 3 patients were in the 3rd decade (25%), 3 patients were in the 4th decade (25%), 1 patient each in the 5th & 6th decade (8.34%) and 2 patients in the 7th decade (16.66%). (Table 1)

Table 1: Age Incidence

<table>
<thead>
<tr>
<th>Age (Years)</th>
<th>No of Cases</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>10 – 20</td>
<td>3</td>
<td>23.07 %</td>
</tr>
<tr>
<td>21 – 30</td>
<td>3</td>
<td>23.07 %</td>
</tr>
<tr>
<td>31 – 40</td>
<td>3</td>
<td>23.07 %</td>
</tr>
<tr>
<td>41 – 50</td>
<td>1</td>
<td>7.69 %</td>
</tr>
<tr>
<td>51 – 60</td>
<td>1</td>
<td>7.69 %</td>
</tr>
<tr>
<td>61 – 70</td>
<td>2</td>
<td>15.38 %</td>
</tr>
</tbody>
</table>
The sex incidence was 7 male and 6 female patients, with a male-to-female ratio of 1.16:1. (Table 2)

**Figure 3**
Table 2: Sex Incidence

<table>
<thead>
<tr>
<th>Sex</th>
<th>No. of Cases</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>7 Cases</td>
<td>53.84%</td>
</tr>
<tr>
<td>Female</td>
<td>6 Cases</td>
<td>46.16%</td>
</tr>
</tbody>
</table>

Neck swelling was the most common presentation (100%). Dysphagia and pain were present in 2 cases (16.66%) of metastatic lymph node masses from squamous cell carcinoma from an occult primary. Another case also presented with pain in the bone metastases from medullary carcinoma of the thyroid. No malignant lesion was found in the oral cavity and no swelling was extending intraorally.

Out of 13 cases, 8 cases (61.53%) were having benign swellings and 5 cases (38.48%) were having malignant tumors. Out of five malignant tumors, two cases were reported as papillary thyroid carcinoma, one case as medullary carcinoma of the thyroid and the remaining two were of squamous cell origin.

Of the 13 cases, 5 were of salivary origin (38.46%), 3 of neurogenic origin (23%) and 5 were having secondary metastatic lymph node deposits (38.46%). Ten (77%) cases were located in the prestyloid parapharyngeal space and 3 (23%) in the poststyloid space. (Table 3)

**Figure 4**
Table 3: Anatomical location and tissue origin of parapharyngeal space tumours [n = 13]

<table>
<thead>
<tr>
<th>Anatomical location</th>
<th>Tissue origin of tumors</th>
<th>Diagnosis</th>
<th>Number of cases*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parotid gland</td>
<td>a) Plasmacytoma of parotid gland</td>
<td>4 (30.75%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>b) Pleomorphic adenoma of parotid gland</td>
<td>1 (7.69%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>c) Dysplasia of parotid gland</td>
<td>1 (7.69%)</td>
<td></td>
</tr>
<tr>
<td>Submandibular salivary gland</td>
<td>a) Plasmacytoma of submandibular salivary gland</td>
<td>1 (7.69%)</td>
<td></td>
</tr>
<tr>
<td>Prestyloid tumors</td>
<td>a) Secondary in neck (primary – follicular carcinoma of thyroid)</td>
<td>5 (38.46%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>b) Secondary in neck (primary – medullary carcinoma of thyroid)</td>
<td>5 (38.46%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>c) Secondary in neck (primary – lymphoma)</td>
<td>5 (38.46%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>d) Secondary in neck (primary – undetermined)</td>
<td>5 (38.46%)</td>
<td></td>
</tr>
<tr>
<td>Lymph nodes</td>
<td>a) Secondary in neck (primary – lymphoma)</td>
<td>5 (38.46%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>b) Secondary in neck (primary – lymphoma)</td>
<td>5 (38.46%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>c) Secondary in neck (primary – lymphoma)</td>
<td>5 (38.46%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>d) Secondary in neck (primary – lymphoma)</td>
<td>5 (38.46%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>e) Secondary in neck (primary – lymphoma)</td>
<td>5 (38.46%)</td>
<td></td>
</tr>
<tr>
<td>Nerves</td>
<td>a) Neurofibroma</td>
<td>3 (23.07%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>b) Neurofibroma with cystic degeneration</td>
<td>3 (23.07%)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>c) Ganglioneurofibroma</td>
<td>3 (23.07%)</td>
<td></td>
</tr>
</tbody>
</table>

All the cases except 2 were operated. Among the 11 operated cases, 8 cases were operated by transcervical approach, 3 by transparotid approach and 2 cases of secondary deposits of squamous cell origin were treated by radiation.

Among the salivary neoplasms reported in our study, 2 were pleomorphic adenomas, 1 was a duct cell carcinoma (Figure 2) with early recurrence, 2 were neural tumors, 1 was a schwannoma arising from the parotid gland and another one a plexiform neurofibroma of the submandibular gland. (Figure 3)

**Figure 5**
Figure 2: Cut section of duct cell carcinoma of the parotid gland
Among the neural tumors in the poststyloid region, two cases of neurofibromas and one case of ganglioneuroma arising from the vagus nerve were reported. One of the two cases of neurofibroma presented with cystic degeneration. (Figure 4)

**Figure 5**
Gross specimen of vagal ganglioneuroma with the vagus nerve

Wound infection was present in 2 cases (16.66%). All patients were followed for a year and recurrence was reported within three months in one patient with duct cell carcinoma of the parotid gland.

**DISCUSSION**
Parapharyngeal space tumors are rare, accounting for only 0.5% of all head and neck tumors (1). As per tissue of origin, four main groups of parapharyngeal space tumors are identified. They are tumors of salivary origin, neurogenic tumors, lymphoid masses and paragangliomas.

The parapharyngeal space occupies the lateral wall of the pharynx. It is further divided into prestyloid and poststyloid spaces. Tumors arising from the prestyloid region make up 70-80% (in our study 75%), whereas 20-30% (in our study 25%) of the tumors arise from the poststyloid region (2).

Parapharyngeal space tumors are mostly benign [73 % (in our study 61.53 %)] (3). The most common parapharyngeal tumors are salivary in origin (pleomorphic adenomas). Tumors arising from lymph nodes are secondary metastatic deposits from upper aerodigestive and thyroid malignancies. The most common age group is the 4th and 5th decade (4). In our study, the presenting age group is the 2nd, 3rd and 4th decade. Neurogenic tumors present at an early age; three cases were reported in our study with an age below 18 years. Secondary deposits from papillary carcinoma of the thyroid present in an early age, whereas the secondary deposits of squamous origin, follicular and medullary carcinomas of thyroid present in late age.

Neck swelling is the most common presenting feature of...
Parapharyngeal space tumors (such as in our study) [4,5]. In benign lesions, the tumor can produce pressure effects on adjacent structures resulting in dysphagia and difficulty in breathing. The malignant lesions can present with pain, cranial nerve palsy, trismus, hoarseness of the voice and otalgia.

Taking biopsy is challenging because of the complex anatomy of the space, and surrounding vital structures. Obtaining preoperative tissue biopsy is not crucial, since 70-80% of the tumors are benign (5). Appropriate radiological evaluation by computerized tomography is the cornerstone for diagnostic evaluation (1). Computerized tomography will show a fat plane between tumor and parotid gland. Prestyloid tumors displace carotid vessels laterally while poststyloid tumors displace the carotid vessels anteromedially (5).

Fine-needle aspiration cytology is an essential part of the diagnosis with an accuracy rate of 95% (6). It has a 100%-accuracy in case of secondary deposits and epithelial tumors of the parotid swellings, but not in neurogenic tumors. In our study, none of the neural tumors was diagnosed by fine-needle aspiration cytology. Computerized-tomography-guided fine-needle aspiration cytology is very useful in case of clinically non-detectable swellings (7). Open biopsy is reserved for non-resectable tumors but one has to be careful not to injure neurovascular structures.

Computerized tomography with contrast is very useful. It helps to know the origin, site, extension and plane of the swelling. Magnetic resonance imaging has advantages over computerized tomography because of use of non-ionizing radiation. Multiple planes are taken without tilting the patient’s position. Superior tissue resolution is obtained. Vessels along with the feeder vessel can be identified with magnetic resonance angiogram.

The patient must be informed well in advance about possible surgical complications like hoarseness of the voice, Horner’s syndrome, dysphagia, excessive hemorrhage and stroke. Surgical excision is diagnostic and curative; it is the treatment of choice in benign tumors. Multiple approaches have been described in the literature. The three basic approaches are transcervical, transparotid and mandibular swing. The surgical approach is considered according to the size and site of the tumor. Most surgeons prefer the transcervical approach. The mandibular swing operation is used for big and vascular tumors which require maximum exposure at the base of the skull. Poststyloid lesions are better approached by transcervical incisions. We excised all three neural lesions by transcervical incision. Parotid swellings can be excised by transparotid incision. Cranial nerve palsy can present if the tumor arises from cranial nerves. Vocal cord palsy may occur if the recurrent laryngeal nerve is paralyzed and Horner’s syndrome results if the sympathetic chain is involved.

Squamous cell carcinomas extending from the nasopharyngeal area respond well to radiotherapy. Techniques such as intensity modulated regimen, hyper fractioned regimen and stereotactic radio-surgery have offered exciting advances in this field. Thyroid metastases require lymph node dissection along with total thyroidectomy followed by radioactive iodine as an adjuvant therapy. Chemo-radiation is the treatment of choice for primary malignant parapharyngeal malignancies. Doxorubicin is indicated for glandular neoplasm and for squamous cell carcinomas cisplatin and 5-fluouracil are given (8). In our study, total thyroidectomy with neck lymph node dissection was carried out in thyroid secondaries.

All patients were followed for a year and no recurrences were reported except one. The patient with duct cell carcinoma of the parotid gland had a recurrence within three months.

CONCLUSION

Parapharyngeal space tumors are located deep in the upper part of the neck. The location makes clinical evaluation difficult because of the limited space. Fine-needle aspiration cytology has limited value in neural tumors. Radiological evaluation is essential to minimize the intraoperative risk. Considering the critical anatomical site, excision biopsy is both diagnostic and curative. Preoperative diagnosis can be obtained by incisional biopsy in malignant cases. If malignancy is proved, further treatment by chemo-radiation is instituted.

References

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