Introduction

Tumors of the parapharyngeal space (PPS) are rare, accounting for 0.5% of all head and neck tumors, of which approximately 80% are benign. The salivary gland neoplasms represent the commonest tumors, followed by neurogenic tumors and paragangliomas. Rare lesions include metastatic lymph nodes, lymphoma, lipoma, rhabdomyoma, metastatic thyroid cancer and branchial cleft cysts. During the last one and half years, we have encountered four exceptional tumors of the parapharyngeal space in our Institute. Presented herein is a summary of these four cases and review of literature.

Materials and Methods

This was a retrospective analysis of four unusual parapharyngeal tumors operated upon by the principal author (NKP) in the department of Otolaryngology, Postgraduate Institute of Medical Education and Research, Chandigarh between August 2001 and May 2002.

Results

The clinical presentation and radiological findings on CT and MRI scan are discussed in Table 1 while Cytopathology; histopathology, treatment and outcome of the four cases are summarized in Table 2.
Unusual Malignant Tumours Of The Parapharyngeal Space-A Diagnostic Dilemma

Table 1: Clinical and Radiological features of Parapharyngeal tumor patients.

<table>
<thead>
<tr>
<th>S No</th>
<th>Diagnosis</th>
<th>Age</th>
<th>Sex</th>
<th>Symptoms with Duration</th>
<th>Signs</th>
<th>CT/MRI Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Squamous cell carcinoma</td>
<td>55E</td>
<td></td>
<td>Occipitomental mass in Lt. tonsil cavity for 6 months</td>
<td>Headache, 2 cm x 2 cm subcutaneous mass in Lt. tonsil region</td>
<td>1 x 1 x 1 cm mass extending to infratemporal fossa, involvement of right hypoglossal nerve</td>
</tr>
<tr>
<td>2</td>
<td>Embryonal rhabdomyosarcoma</td>
<td>26M</td>
<td></td>
<td>Right parotid mass for 8 months</td>
<td>Meningeal invasion, mass effect on brainstem</td>
<td>1 x 1 x 1 cm mass with extension to right temporal fossa, involvement of right trigeminal nerve</td>
</tr>
<tr>
<td>3</td>
<td>Undifferentiated Carcinoma</td>
<td>53M</td>
<td></td>
<td>Painful mass in Left submandibular fossa, painless for 2 months</td>
<td>Neck mass, fixed to adjacent structures</td>
<td>2 x 2 x 2 cm mass with extracapsular spread, involvement of left vagus nerve</td>
</tr>
<tr>
<td>4</td>
<td>Fibrosarcoma</td>
<td>45M</td>
<td></td>
<td>Progressive, tender submandibular mass</td>
<td>Neck mass, fixed to adjacent structures</td>
<td>3 x 3 x 3 cm mass with infiltration of adjacent structures</td>
</tr>
</tbody>
</table>

Abbreviations: Sma submandibular; Rt. Right; Lt. Left;

Table 2: Cytological, Histopathological, treatment and outcome of parapharyngeal tumour patients.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>FNAC</th>
<th>Surgery</th>
<th>Histopathology</th>
<th>RT</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Squamous cell carcinoma</td>
<td>Cystic, inflammatory</td>
<td>Transcervical, submandibular gland excision</td>
<td>Osseous, myxoid material with odontoid and chondroid areas</td>
<td>XRT, postop</td>
<td>NED 2 yrs</td>
</tr>
<tr>
<td>Embryonal sarcoma</td>
<td>Anaplastic sarcoma</td>
<td>Radical resection with PPS clearance</td>
<td>Osseous, undifferentiated sarcoma</td>
<td>XRT, postop</td>
<td>NED 10 yrs</td>
</tr>
<tr>
<td>Undifferentiated Carcinoma</td>
<td>Adenocarcinoma</td>
<td>Radical resection with PPS clearance</td>
<td>Osseous, poorly differentiated spindle cells</td>
<td>XRT, postop</td>
<td>NED 5 yrs</td>
</tr>
</tbody>
</table>

Abbreviations: XRT External Radiotherapy; NED No evidence of disease

EPIDEMIOLOGY

The male to female ratio was 3:1. The age of the patients ranged from 28 to 52 years with a mean age of 38.8 years.

CLINICAL PRESENTATION

Tumors of the parapharyngeal space PPS remain clinically silent for a long time till they attain a sufficiently large size so as to produce a palpable or visible lump in the oral cavity and neck. The presenting symptoms also depend on the...
extent of the swelling. The first and second cases presented with a palato-facial bulge and an additional swelling in the submandibular region owing to large size (Fig 1). Parotid involvement in the third and fourth cases leads to additional preauricular bulge (Fig 2). Extension to infratemporal fossa (ITF) in the first and fourth cases leads to trismus in them. Mass effect on the palate, oropharynx and supraglottis produced symptoms of voice muffling, dysphagia and odynophagia. Severe localized pain due to malignant infiltration was a prominent feature in the fourth case. The second case presented with paresthesia over the mandibular nerve territory and referred otalgia, owing to invasion of foramen ovale. Metastatic cervical lymphadenopathy was seen in the third case alone. Noteworthy is the absence of lower four cranial neuropathy or Horner’s syndrome in any of the cases.

Figure 5
Figure 1: Clinical photograph of case 2 (synovial sarcoma) showing submandibular swelling.

RADIOLOGICAL FEATURES
Computerized tomography in four cases and MRI in one case was done to determine the nature, site, extent, vascularity, relation to great vessels, adjacent bony infiltration and retropharyngeal or cervical lymphadenopathy (Fig 3,4,5). The findings are tabulated in Table 1.
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Figure 7
Figure 3: Contrast CT scan (axial section) showing a discrete, capsulated hypo dense lesion in the parapharyngeal space in case 2 (synovial sarcoma).

Figure 8
Figure 4: Contrast CT scan (axial section) showing heterogeneous, calcified tumor in the parapharyngeal space extending to the infratemporal fossa eroding the mandibular ramus. (Fibrosarcoma).

Figure 9
Figure 5: MRI scans T1 weighted with contrast (coronal section) showing the vertical extent of the above lesion (Squamous cell carcinoma).

CYTOPATHOLOGY
The second vital investigation performed was fine needle aspiration cytology (FNAC) of the lesion by transoral route in one case and transcervical in three cases. FNAC corroborated with the final histopathological report in one case (synovial sarcoma). FNAC was not contributory to diagnosis in the remaining three cases.

SURGERY
All patients were taken up for surgical excision by either trans-cervical approach (2/4 cases) or trans-parotid approach (2/4 cases) with added neck dissection in the third case having metastatic neck secondaries. Resection of the involved vertical ramus of mandible was performed in two cases (second and fourth) and the involved facial nerve was sacrificed with the tumour in two cases (third and fourth). There were no minor or major complications in any of the cases. The fourth case underwent reconstruction of the post-operative soft tissue defect by pedicled latissimus dorsi flap after one year of disease-free interval.

HISTOPATHOLOGY
The post-operative histological reports of the excised specimens are summarized in Table 2. All four neoplasms were malignant namely squamous cell carcinoma, undifferentiated carcinoma, fibrosarcoma and synovial sarcoma (Fig 6).
DISCUSSION

This study comprises four unusual parapharyngeal malignancies encountered in a single institute over a one and half year period. The mean age of the patients was 38.8 years and this value was comparable with the mean age of 40 years in the Stanley series of PPS tumours. The commonest clinical presentation was a submandibular lump followed by dysphagia and alteration in voice. In a study of parapharyngeal tumors by Pang et al, 60% had painless lump, 12% had globus and 6% had aural symptoms. In the Carrau et al series, 53% of the 51 patients presented with a neck mass, 51% had pharyngeal mass, 9% had parotid mass, 4% trismus, 11% vocal cord palsy, 2% each had mandibular nerve, accessory nerve and hypoglossal nerve palsy.

CT scan and MRI were employed to assess the extent of the lesion and plan surgical approach. MRI is currently considered as a superior modality owing to advantages like specific signal intensity characteristics, delineation of fat planes and ability to reveal carotid attenuation by the tumour.

The second investigation i.e. fine needle aspiration cytology yielded an accurate diagnosis in only one case (25%) contrary to 77% accurate diagnosis reported by Pang et al.s Though fine needle aspiration cytology has been reported to have a high sensitivity, it cannot be relied upon entirely particularly in cases where suspicion of the tumour being malignant is high. It is therefore suggested that in such cases it is prudent to undertake complete excision and histopathological confirmation.

Surgery is the mainstay of treatment in parapharyngeal tumors. The surgical approach chosen should facilitate complete tumour extirpation with minimal morbidity. The conventional surgical approaches described for the parapharyngeal tumours are transoral, trans-parotid, transcervical with or without mandibulectomy and midline transmandibular-oropharyngeal approaches. Of them transcervical and trans-parotid approaches are commonly used. The later approach can lead to temporary facial weakness and Frey’s syndrome. The trans-cervical approach is associated with risks of permanent lower cranial nerve and cervical sympathetic neuropathies as seen in 17% cases in Carrau series and 1/13 in the Pang series. The other fatal complications are mainly related to paraganglioma excision namely neurovascular injury, cerebro-vascular accident and mortality.

Squamous cell carcinoma (case 1) in the parapharyngeal space is usually seen as a metastatic nodal disease or less commonly as an extension of deep lobe parotid tumor into the parapharyngeal space. Som et al and Maran et al have reported an incidence of 14% and 17% respectively. Rarely, primary PPS tumours can arise from lateral pharyngeal wall or minor salivary gland tissue of PPS (SCC comprises 1.2% of minor salivary gland epithelial neoplasms). The tumour in our case probably originated de novo from minor salivary glands of the parapharyngeal space or from lateral pharyngeal wall.

Synovial sarcoma (case 2) comprising 8-10% of all sarcomas predominantly involves the extremities. The commonest site in the head and neck is hypopharynx, followed by prevertebral, maxillofacial, pharyngeal and laryngeal spaces. This high grade tumour has a male preponderance and usually occurs in the third decade. Histologically, this tumour may be biphasic, comprising both epithelial and sarcomatoid cells or monophasic with predominant spindle cells (as in our case) or predominant epithelial cells. t (x; 18) analysis could not be done as this facility does not exist. Metastasis to regional lymph nodes occurs in 12.5% cases. A marked tendency for local recurrence and pulmonary metastasis lowers survival to 40% at 5 years and 25% at 10 years. Wide resection with a cuff of normal tissue is indicated in all cases. Post operative radiotherapy is required in case of residual, microscopic disease.

The third case was an undifferentiated carcinoma of the deep lobe of parotid extending to the parapharyngeal space. This is a rare salivary gland tumor, comprising 3.2% of primary
epithelial parotid tumors and 10.2% of malignant parotid tumors. It is seen predominantly in the middle age. There are two varieties; the small cell type being twice as common as the large cell type. This tumor is very aggressive and requires radical surgery followed by postoperative radiotherapy. The five- year survival rate is 20-30%.

Fibrosarcoma (case 4) is an uncommon tumor of the infratemporal fossa and parapharyngeal space. In a study by Carrau et al., 11 out of 54 PPS tumors (20%) were found to be malignant, of which one was fibrosarcoma (9%). In a study by Shaheen, 3 out of 10 cases of primary infratemporal fossa tumors proved to be fibrosarcoma. This tumor is also enlisted among the seven rare primary malignant neoplasms of the PPS encountered in medical literature. In our case Masson's trichrome staining was done. It showed delicate collagen fibrils along the spindle cells and occasionally within the cells. Extensive sampling did not reveal any squamous cell carcinoma component or any chondrosarcomatous or osteoid component to suggest this to a spindle cell variant of squamous cell carcinoma or a de differentiated primary bone tumor. As regards tumor involving the bone, the tumor arose primarily in the infratemporal fossa eventually extending to involve the ramus of the mandible.

CONCLUSIONS

To conclude, parapharyngeal space tumors are a rare entity. The above four neoplasms are even more rarely encountered in the parapharyngeal space. Diagnosis of these unusual malignancies may pose a tough challenge to the clinician, radiologist and pathologist alike. When the diagnosis is not clear on fine needle aspiration cytology, complete surgical excision and histopathological examination is recommended for optimum management.

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