ABSTRACT

Purpose: The purpose of this study is to evaluate parapharyngeal space (PPS) tumors as regards clinicopathological features, preoperative assessment, different surgical approaches, perioperative complications, patterns of recurrence and the role of non-surgical treatment.

Materials and Methods: This study included twenty five patients with (PPS) tumors presented to the NCI, Cairo University, from October 2001 to March 2003. The data of each patient included age, sex, presenting symptoms and signs, provisional diagnosis, preoperative investigations, operative data, histopathological examination, non-surgical treatment and state at follow up. All were collected and analyzed.

Results: This study included 12 males and 13 females. The mean age was 37.1 years. The main presenting symptom and sign was neck swelling. All patients were subjected to CT scan, while 9 patients had MRI. Nineteen patients underwent fine needle aspiration cytology (FNAC) which was conclusive in only 16 patients. Benign lesions were found in 12 patients (48%) and malignant lesions in 13 patients (52%). Parotid gland tumors (40%) and neurogenic tumors (16%) were the commonest. Surgical excision was done in 22 cases. There was no postoperative mortality and overall postoperative morbidity was 9% (2/22). Eight patients received postoperative radiotherapy. Three patients with lymphoma were treated with chemotherapy and two of them received involved field radiotherapy to the Waldey er's ring region. On follow up to 12-30 months, there were only one local and two distant recurrences in the malignant group.

Conclusion: Surgery is the mainstay treatment for tumors of the (PPS). The addition of postoperative radiotherapy in certain indications in malignant tumors of the (PPS) will improve the local control.

Key Words: Parapharyngeal space (PPS) tumors - Management.

INTRODUCTION

The parapharyngeal space (PPS) is one of the potential fascial planes of the head and neck, that may become involved in various pathological processes: inflammatory and neoplastic. The latter represents less than 1% of all head and neck tumors [1]. The parapharyngeal space resembles an inverted pyramid with concave faces. The base is the skull base and the apex is the greater cornu of the hyoid bone, whereas the space is further divided into prestyloid and poststyloid compartments by the styloid process and its attached muscles and fascia [2]. Neoplasms of this space, although vary in origin tend to have similar clinical manifestations. These tumors usually present by an asymptomatic mass in 50% of the patients [3]. The purpose of this study is to evaluate (PPS) tumors treated at the NCI, Cairo University, as regards clinicopathological features, preoperative assessment, different surgical approaches, perioperative complications, patterns of recurrence and the role of non-surgical treatment.

PATIENTS AND METHODS

This is a prospective study conducted at the National Cancer Institute, Cairo University during the period from October 2001 to March 2003. It included twenty five patients presenting with PPS tumors. Patients were subjected preoperatively to history and physical examination with special attention to presenting symptom(s) and clinical examination evaluating the primary lesion and nodal status. CT scan of the neck was done for all patients. MRI of the neck was done in 9 patients to delineate more details about the soft tissues. Magnetic resonance angiography (MRA) was done in 3 cases. Fine needle aspiration cytology (FNAC) was tried whenever possible. Other investigations, as
chest X-ray as a part of metastatic work up and routine laboratory work up, were done for every patient.

Surgical approaches in our series depended on the size of the tumor, the location, the relationship of the tumor to great vessels and the suspicion of malignancy. We used the following surgical approaches in our series:

1- Transcervical approach [4] which provides access to the PPS through the neck with or without excision of the submandibular gland. The former is called (Transcervical - submandibular) approach.

2- Transparotid-transcervical approach [5] which provides exposure for facial nerve dissection, performing a superficial parotidectomy and incising the stylo-mandibular ligament. This allows for anterior dislocation of the mandible to provide an adequate exposure to remove many tumors especially those of the deep lobe of the parotid gland.

3- The transcervical-transmandibular approach [6] utilizing midline or lateral mandibulotomy. In this procedure, with the hemimandible reflected laterally, wide exposure is afforded to approach extraparotid or post-styloid space lesions.

Operative details regarding the surgical approach used for the resection of the primary lesion and management of cervical lymph nodes had been reported and evaluated. Gross and histopathological examination of the surgical specimens were carefully reported and reviewed with particular attention to safety margins and nodal involvement. Postoperative data concerning early outcome including morbidity, mortality and hospital stay and late outcome including functional results and patterns of recurrence were reported and evaluated through follow up of patients for 12-30 months. Patients were referred to chemotherapy and/or radiotherapy departments when indicated.

RESULTS

The total number of patients was twenty five. They were (12) males and (13) females. In male patients, (6) had benign lesions and (6) had malignant lesions. In female patients, (6) presented with benign lesions and the remaining (7) presented with malignant lesions. The mean age for all patients was 37.1 years.

Table (1) showed the main presenting symptom(s), while (Table 2) shows the main presenting sign(s). The main presenting symptoms and signs were neck or parotid swellings. In two cases, the main presentation was an oral swelling (Fig. 1).

All patients were subjected to neck CT scan (Figs. 2-4), while 9 patients had MRI studies to reveal the exact nature of the tumor (Figs. 5-6). MRA was done in 3 cases with carotid body tumors (Fig. 5 B).

Nineteen patients underwent FNAC, 3 patients were previously biopsied and pathologically diagnosed at first presentation to the NCI and 3 patients had no preoperative biopsy with surgical exploration based on MRI data suggesting carotid body tumors. The patients who underwent FNAC showed that the test was conclusive in 16 patients and the results were as follows: benign mixed salivary gland tumors in 4 patients, parotid carcinoma in 4 patients and neurofibroma in 3 patients. Chondrosarcoma, spindle cell sarcoma, benign schwannoma, malignant branchial cyst and infected epidermal cyst all were revealed in one patient individually. In 3 patients, FNAC was inconclusive, so biopsy and marker study were needed to reveal the pathology which proved to be large cell lymphoma in 2 patients and undifferentiated carcinoma of the parotid in one patient.

Surgery is the mainstay treatment for tumors of the PPS. Surgical excision was done in 22 cases. Table (3) shows the different surgical approaches used in our study. In our study, the transparotid-transcervical approach was the most common approach used in 10 patients (45.5%), as the tumors were related to the parotid gland. Fig. 7 (A-D) showed the transparotid-transcervical approach for a case of pleomorphic adenoma of the deep lobe of the right parotid. The trans-cervical approach was used in 8 patients (36.3%) for benign lesions, lesions in the prestyloid compartment and tumors in the lower part of the PPS extending in the neck. Fig. (8) shows the transcervical approach for a case of paraganglioma of the right PPS and Fig. (9) shows the transcervical approach for a case of plexiform neurofibroma of the left PPS. The transcervical-submandibular approach was used in 2 patients (9.1%) for lesions that were large in size and that needed an excision of the submandibular gland for better extirpation of the
tumor. The transcervical-transmandibular approach was used in two patients (9.1%) with malignant lesions (fibrosarcoma and chondrosarcoma).

Neck dissection was performed in 7 patients (4 patients with mucoepidermoid carcinoma of the parotid, 2 patients with undifferentiated carcinoma of the parotid and one patient with malignant branchial cyst).

Primary closure was done in all patients, except 2 cases. A trapezius myocutaneous flap was used in one patient, while a deltopectoral flap was used in the other.

There was no postoperative mortality. Postoperative complications occurred in 2 patients, in the form of superficial sloughing of the edges of the wound which was treated conservatively by repeated dressings and sloughing of trapezius myocutaneous flap, which was replaced by a pectoralis major myocutaneous flap.

Postoperative histopathological results are represented in Table (4). Parotid gland tumors and neurogenic tumors were the commonest.

Postoperative radiotherapy was given to all patients with malignant salivary gland tumors (4 mucoepidermoid and 2 undifferentiated carcinomas). The indications were high grade, positive neck nodes and/or inadequate surgical margin. The patients received 50-60 Gy/25-30 fractions to the primary site with its extension in the parapharyngeal space and ipsilateral neck nodes. Two patients with fibrosarcoma and chondrosarcoma received also postoperative radiotherapy because of inadequate surgical margin. The chondrosarcoma patient received 70 Gy/35 fractions, while the fibrosarcoma patient received only 42 Gy/21 fractions out of 70 Gy/35 fractions due to the patient’s non-compliance. The radiation treatment was given to the primary site only. The patient with squamous cell carcinoma of the branchial cyst had positive neck nodes and received 56 Gy/28 fractions to the whole neck. The 3 patients with parapharyngeal lymphoma received chemotherapy in the form of 6-8 courses of CHOP (Cyclophosphamide, Adriablastine, Vincristine and Prednisone). Two patients of them received involved field radiotherapy, 40 Gy/20 fractions, to the Waldyer’s ring region.

The follow-up period for all patients ranged from 12 to 30 months. One local failure and two distant failures were observed. The single patient with local failure had fibrosarcoma with positive surgical margin and he did not complete his postoperative radiotherapy (42 Gy). The two distant failures were observed in one patient with undifferentiated carcinoma of the parotid and another patient with non-Hodgkin’s lymphoma.
Fig. (4-A): Axial CT scan showing malignant lymphoma of the left PPS.

Fig. (4-B): Coronal CT scan of the same patient.

Fig. (5-A): Sagittal MRI showing right carotid body tumor splaying the carotid branches.

Fig. (5-B): MRA verifying the vascular nature of the mass by virtue of splaying the carotid bifurcation.

Fig. (6-A): Pre contrast axial T1WI showing prestyloid left PPS tumor (Plexiform neurofibroma).

Fig. (6-B): Post contrast axial T1WI of the same patient.
Fig. (7-A): Operative view after superficial parotidectomy showing pleomorphic adenoma of the deep lobe of the right parotid.

Fig. (7-B): Deep lobe of the parotid is excised preserving facial nerve branches.

Fig. (7-C): Operative bed after excision of the tumor.

Fig. (7-D): Postoperative specimen.

Fig. (8): Operative view showing paraganglioma of the right PPS.

Fig. (9): Operative view showing plexiform neurofibroma of the left PPS.
DISCUSSION

This study included 25 patients with PPS tumors. All were primary cases except one which was a recurrent one. The male to female ratio was 1:1.1. In other series it was 2:3 (3), 3:5 (7), 5:6 (8).

In our study the mean age of the female patients was 45.3 years, and for the male patients it was 29.6 years. In a large series of 172 cases [8] the mean age of the female patients was 53.4 years and for the male patients it was 46.7 years.

In our study, 13 out of 25 patients (52%) had malignant tumors. In other series, malignant tumors of PPS represented only 20% [3,8], 27% [9] and 30% [1].

A review of several large series revealed that salivary gland tumors (primarily of parotid origin), neurogenic tumors, paragangliomas and lymphomas comprise nearly 80% of PPS masses. The remainder includes a wide variety of lesions, including lipoma, liposarcoma, haemangioma, haemangiopericytoma, haemangioidothelioma, meningioma, lymphangioma, branchial cleft cyst, fibrosarcoma, malignant fibrous histiocytoma, rhabdomyosarcoma, leiomyoma, chordoma and metastatic lesions [10].

In our study, 10 patients (40%) had salivary gland tumors, 4 patients had benign (mixed salivary gland) tumors and 6 had malignant tumors (mucoepidermoid carcinoma in 4 cases and undifferentiated carcinoma in 2 cases). Four patients had neurogenic tumors (16%), 3 patients had paragangliomas (12%), 3 patients had lymphomas (12%), while miscellaneous tumors were found in 5 patients (20%) (Table 4).

Pitman et al. [11] reported that 50% of the neoplasms were of deep lobe of parotid or minor salivary gland origin, while 20% were of neurogenic origin. Batsakis and Sneige [1] found that neoplasms of salivary gland origin accounted for 40-50% of PPS lesions and were located in the prestyloid compartment of the PPS. The most common prestyloid PPS lesion was pleomorphic adenoma, which represented 80-90% of salivary neoplasms in the PPS. Carcinoma ex-pleomorphic adenoma and adenoid cystic carcinoma were the most common salivary malignancies of the PPS. Approximately, 20% of all salivary lesions in the PPS were malignant. Johnson et al. [12] collected data from 4 studies
representing 213 patients. They found that neurogenic tumors might account for more PPS lesions than thought. In their study, schwannomas accounted for 20% of all PPS tumors and were the most common enhancing extraparotid tumors. Carrau et al. [3] reported that neurogenic tumors comprised 31 out of 54 patients. Twenty-six patients (83%) had benign lesions, while 5 patients (17%) had malignant neoplasms. Our study included 4 patients with tumors of neurogenic origin and all tumors were benign (100%).

Three patients with benign carotid body tumors were reported in our study, Sniezek et al. [13] reported 23 patients of paragangliomas of the PPS, 16 out of which were carotid body tumors (70%) and 7 patients were glomus vagale tumors (30%). Only 2 patients had malignant tumors (8.7%).

Batsakis and Sniege [1], found that carcinoma ex-pleomorphic adenoma and adenoid cystic carcinomas were the most frequently reported PPS parotid malignancies. In our study, mucoepidermoid carcinoma (4 cases) and undifferentiated carcinoma (2 cases) were the two types encountered.

In our study, lymphomas represented 3 patients out of 13 patients with malignant tumors (23%). They were large cell Non-Hodgkin lymphomas. This is higher than other series, e.g. (1/11) 10% (3) and (3/35) 8.6% (8).

In our study, patients presented with a wide variety of symptoms and signs. The most common complaint was a mass in the neck. Parotid swelling and otalgia were the next common presenting symptoms, while parotid swelling and facial palsy were the next common signs. Kenneth et al. [8] in Mayo Clinic, found that the most common symptom initially seen was awareness of an intraoral or a neck mass (84%). They found that most patients had only vague symptoms, ear pressure or pain (36%), dysphagia (12.8%), hearing loss (11%), hoarseness (10.5%) and facial or jaw pain (6.4%) of the whole 172 patients included in their study. Trismus, although considered a common finding by Clarke and Hebert [14], was not encountered in our cases.

Lang [15], found that IX, X and XI cranial nerve palsies were common in the paragangliomas and malignant tumors, but not in other tumors and the cranial nerve most commonly involved was the vagus. They found that IX, X and XI cranial nerve palsies resulting in paralysis of the stylopharyngeus muscle, loss of taste on the posterior 1/3 of the tongue and disturbances in swallowing and speech (Vernet's syndrome) and the tumors involving the hypoglossal nerve (Collet's syndrome) and cervical sympathetic chain (Horner's syndrome) were common.

In our study, imaging modalities in combination with fine needle aspiration cytology were diagnostic in most of our patients. FNAC was done in 19 patients. The test gave conclusive results in 16 patients. Thus FNAC is a safe and valuable test to diagnose PPS tumors and this is similar to that reported by Keiner et al. [16].

Evaluation of PPS tumors is mostly radiographic with CT scanning and MRI offering useful diagnostic information [7]. Appropriate diagnosis can be reached radiographically in 95 % of patients without tissue biopsy [3]. Becker [17] reported that CT scan could be considered as a reliable technique to evaluate and localize suprahypoid neck lesions. Mondal and Gupta [18] mentioned that MRI was able to differentiate tumors from normal salivary gland tissue better than CT, especially using T2 weighted images. Histological differentiation could be demonstrated between different tumor types based on the MRI. Neural tumors are usually homogenous in appearance and often have a multilobulated surface. MRI currently provides the most useful preoperative information about tumor extent and its relationship to surrounding structures. The addition of MRA provides the preoperative information necessary to determine the optimal surgical procedure for tumor removal. Salivary neoplasms arising in the prestyloid compartment will tend to displace the carotid artery posteriorly. Neural tumors are poststyloid and will displace the carotid artery anteriorly. Carotid body tumors will exhibit the characteristic bowing between the internal and external carotid arteries (Lyre sign).

Surgical extirpation remains the treatment of choice for PPS tumors [4]. External beam irradiation has shown only limited utility in a small subset of cases [19]. Thus, surgery is the mainstay treatment for tumors of the PPS.

The choice of the surgical approach depends on the size of the tumor, the location, the rela-
tionship of the tumor to great vessels and the suspicion of malignancy. In our study, the transparotid-transcervical approach was the most common approach used in 10 patients followed by the trans-cervical approach which was used in 8 patients. Besides the four surgical approaches used in our study (Table 3), there are 2 other approaches described in the literature. They include the transoral approach [20] and the infratemporal approach [21]. The transoral approach is no longer recommended due to inadequate exposure offered, leading to high rate of vascular injury, tumor spillage and local recurrence. The infratemporal approach is recommended for proximal vagal lesions with both extracranial and intracranial components such as glomus jugulare paragangliomas.

In our study, there was no postoperative mortality. Postoperative complications were encountered in 2 patients who were managed successfully. Injury to cranial nerves IX, X, XI, XII and cervical sympathetic chain may result from surgery of PPS tumors. The incidence of post operative cranial nerve deficits ranges from 0-57% with higher incidence seen in malignancies or neurogenic lesions [22,23]. In our study the facial nerve was completely sacrificed in 2 cases with malignant parotid tumors and the upper trunk was sacrificed in 2 other cases with malignant parotid tumors due to evident involvement of the tumor by preoperative examination. The cervical sympathetic chain was affected by the tumor, leading to Horner’s syndrome observed in preoperative examination in one patient.

Kenneth et al. [8] studied 172 patients with a median follow-up of 6.1 years. The status of all patients at the time of the report was “alive without disease” (63%), “alive with disease” (8.5%), “dead of disease” (26.15%), “dead of other causes” (14.8%) and “lost to follow-up” (14.8%). Of the 137 patients with benign lesions, 12 (9%) had recurrences (5 paragangliomas, 3 pleomorphic adenomas, 1 neurofibroma, 1 angioblastic, 1 desmoid tumor and 1 meningioma). Among the 35 patients with malignant neoplasms of the PPS, recurrent tumors were noted in 27 and by time 22 of them had died of the disease. In our study, the follow-up period for all patients ranged from 12 to 30 months. One local failure and two distant failures were observed. All occurred in the malignant group.

We conclude that PPS tumors, when suspected, should be referred to a specialized center, where meticulous examination, assessment and better investigations are done. Surgery is the mainstay treatment and surgeons of head and neck should be acquainted by the different surgical approaches.

REFERENCES


