ORIGINAL ARTICLE

HEAD AND NECK PARAGANGLIOMAS: 10 YEARS INSTITUTIONAL EXPERIENCE

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ABSTRACT

Background: Paragangliomas originate from neuroectodermal derived cells. They are very slow growing tumors. Patients usually present with a neck mass or hearing loss and tinnitus in jugulotympanic paragangliomas. In advanced stage cranial nerve palsies are more common. Radiographic studies are important for diagnosis and depending on location and stage of tumor, either surgery alone or post-operative radiation therapy is the mainstay of treatment.

Material & Methods: It was a retrospective review of case records of patients treated at Aga Khan University Hospital, Karachi from October 2000 to October 2010.

Results: Total number of patients was 24; 15 males and 9 females. Average age was 52.3 years. Fourteen (58.34%) were carotid body tumors, 5(20.83%) glomus tympanicum, 3(12.53%) glomus jugulare and 2(8.33%) were glomus vagale. CT scan was done in all patients to confirm the diagnosis and extent of tumor. Preoperative angiography and angioembolization was done in all patients. All patients underwent surgical resection. Complete resection was possible in all except in two cases of glomus jugulare. Recurrence was only in 2 cases of glomus tympanicum. Post-operatively, cranial nerve dysfunction was noted only in glomus vagale, where vagus nerve was scarified in both patients.

Conclusion: Surgery has promising outcome in carotid body tumors but in jugulotympanic paragangliomas complete resection is challenging. In glomus vagale, surgery is always associated with vagus nerve palsy.

Key Words: Paraganglioma, Carotid body tumor, Glomus tympanicum, Gloms jugulare, Glomus vagale.

INTRODUCTION

Paragangliomas are rarely encountered in day to day surgical practice. They arise from the paraganglion system. Paraganglion are the neuroectodermal derived cells, responsible for catecholamine secretion. Most sites of paraganglia disappear after the birth except adrenal medulla and around the autonomic nervous system.[1] In head and neck, there are three main sites where there is concentration of paraganglia, which can form the tumors. They are; in descending order; around the carotid bifurcation, jugulotympanic area and the vagus nerve.

They are usually slow growing tumors with minimal symptoms initially. Carotid body tumors (CBT) and Vagus paragangliomas (VP) usually appear as mass in neck[2] while Jugulotympanic paragangliomas (JP) present initially as tinnitus and hearing loss. They are highly vascular tumors and also known as chemodectomas or Glomus tumors. Most paragangliomas are benign while less than 10% are thought to be malignant.[3] Malignancy can only be diagnosed when there is evidence of lymph node involvement or distant metastasis. Multiple paragangliomas can be found in 10-15% cases and in multicentricity, 50% cases shows familial inheritance.[4] Diagnosis depends on clinical examination and specific imaging characteristics. Biopsy is usually not advocated as they are highly vascular tumors. Main treatment modality is surgical resection with or without preoperative embolization while in advance cases radiation therapy can also be offered. The purpose of this article is to review the clinical presentation, diagnosis and outcomes of offered treatment modality in our patients over the period of 10 years.

MATERIAL AND METHODS

It was a retrospective review of the charts of the patients from October 2000 to October 2010. All the patients who were treated at Aga Khan University Hospital Karachi were included and the patients who were treated initially outside were excluded from the study. The data was analyzed on SPSS version 16. Percentage and mean was used to analyze the results of study.

RESULTS

Total number of the patients were 24; 15(62.5%) were males and 9 (37.5%) were females. Average age was 52.3 years. Fourteen (58.34%) patients had unilateral carotid body tumors, 5 (20.83%) had glomus tympanicum, 3 (12.53%) glomus jugulare and 2(8.33%) patients had glomus vagale. All the carotid body tumors presented with non-tender neck mass, slowly growing in size over the period of years. Among the vagus nerve tumors, 1 patient came with neck mass while another presented with neck mass and also loss of gag reflux. All the glomus tympanicum patients came with on and off complaint of tinnitus and mild hearing loss. Two patients of glomus jugulare came with hearing loss and headache and 1 presented with cranial nerve IX and X palsy.

Detailed history, clinical examination, and different work up was done in different patients. In all the 8 patients of glomus tympanicum and glomus jugulare hearing test, pure tone audiometery were done which were affected depending upon the location and size of tumor. CT scan was done in all the 24 patients. In 5 patients of carotid body tumors, doppler ultrasound was done initially and then CT scan was also done. 24 hours urinary VMA was also done in all the patients and no one was found to have secretory tumor.

Pre-operatively, in all the patients, angiography and angioembolization was done to decrease the risk of bleeding per-operatively. In carotid body tumors, complete surgical resection was achieved in all the 14 patients. In 1 patient, 5 mm part of carotid artery was removed and vascular reconstruction done by using saphenous vein graft. In 3 patients neck nodes were also removed. No patient developed any complication post-operatively.

Two patients of Glomus vagale also underwent surgery and complete resection was achieved but post-operatively both had vagus nerve palsy. Five patients with glomus tympanicum underwent ear exploration and tumors were removed completely but postoperatively 2 patients suffered increased hearing loss. In glomus jugulare, among 3 patients, complete resection was achieved only in 1 patient and in 2 patients there was residual disease post-operatively. These 2 patients then underwent radiation therapy. Post-operatively, final histopathology reports were consistent with paragangliomas. All the nodes which were removed were also negative for any malignancy.

Mean follow-up period was 3.5 years. 1 patient of glomus jugulare died due to the disease. Among 5 patients of glomus tympanicum, recurrence was noted in 2 patients after 18 months and then they were kept under observation with serial CT scans. No increase in size of tumors was noted after 2 years in both patients. No new lesion was seen in any patient during follow up period.

DISCUSSION

Head and neck paraganliomas most commonly occurs along the blood vessels (carotid bifurcation and jugular bulb) or along the nerves such as vagus nerve and tympanic plexus in the middle ear. They are highly vascular and it may be due to angiogenic growth factors.[5] These tumors are very slow growing, painless masses. Carotid body tumors (CBTs) arise from the carotid bifurcation and become noticeable after reaching certain size when they become visible or can be palpated. In our study, neck mass without any cranial nerve dysfunction was the most common finding of CBTs. Carotid body tumors can be classified according to the shamblin classification depending upon the carotid artery involvement.[6] Vagal paragangliomas usually originate just below the skull base at the level of nodose ganglion. Tumor may go unnoticed until vagus or adjacent cranial nerve dysfunction occurred. In our study 1 patient presented with cranial nerve dysfunction while another patient came with neck mass. Glomus jugulare develop in the temporal bone at the level of bulb of internal jugular vein. A distinct mass is usually not the presenting complaint. In our patient, the present complaint was sensoryneural hearing loss. More over involvement of one or more cranial nerve (IX, X, XI, XII) is often noted. Similarly, glomus tympanicum originate in the middle ear from tympanic plexus. Our patients came with history of hearing loss and tinnitus. Glomus tympanicum has sun rise appearance under the microscope.

Another important consideration in paraganliomas is the presence of multiple lesions. Although in our study, none of the patient had multiple paraganliomas but in literature it is up to 22%.[7] Similarly, if family history is positive then there are 78% to 87% chances of multiple tumors.[8] Malignancy in paragangliomas is more common in sporadic and non familial patients[9] but in our study no case of malignancy was found. Presence of tumor in lymph node or systemic metastasis is the only acceptable criteria for malignancy because histological examination of tumor is not diagnostic for its malignant potential.

For the diagnosis of paragangliomas, radiological studies are very important. CT scan is usually done for temporal bone paraganliomas (glomus jugulare and glomus tympanicum) to see the bony erosion. MRI has equally efficacy in diagnosing these vascular tumors in neck areas. Angiography is more sensitive in diagnosing these lesions and especially when there are multiple syn-

chronous paraganliomas.[10] More over angiography provides information to the surgeons regarding the feeding vessels, so that preoperatively necessary strategies should be adopted to decrease the bleeding. In our study, angiography was done in all patients pre-operatively for diagnosis as well as for embolizing the feeding vessels to decrease the risk of bleeding.

Traditionally, the mainstay of treatment is surgical resection but the outcomes depend on many factors. Tumor size, its location, patient age and general conditions help in deciding which treatment should be offered. Radiation therapy is reserved for extensive intracranial or skull base tumors, multiple or bilateral tumors with potential post-operative cranial nerves dysfunction and in poor risk patients.[11] Surgical resection, most often, results in different post operative complications. Lower cranial nerves palsy is the most common post operative complication. In our study, carotid body tumors resection was not associated with any cranial nerve dysfunction but excision of vagal nerve paragangliomas almost always require vagus nerve resection[7] and in our patients vagus nerve was also scarified in the both patients of glomus vagale.

Similarly, glomus jugular are intimately associated with cranial nerve IX, X, XI and XII involvement.[12] In our study, among 3 patients, 1 patient came with IX and X nerve involvement. Although, Post operatively in these 3 patients, no cranial nerve dysfunction was observed due to surgery but complete tumor removal was also not achieved in 2 patients. These 2 patients were offered radiation therapy later on. Complete surgical removal of these tumors is often not possible and optimal treatment consists of surgical resection and radiation therapy.[13]

Glomus tympanicum arises from the tympanic plexus in middle ear. All our 5 patients of glomus tympani underwent surgery and post aural approach was adopted to have a good view of operative field. Complete tumor removal was achieved per-operatively. But 2 patients shown recurrence after 18 months. Both of these patients were observed for another 2 years with serial CT scan and no increase in size was observed. Another 2 patient's developed increased hearing loss just after surgery and no improvement had been shown over the period of one year but these 2 patients did not developed any recurrence.

Surgery remains the first choice of treatment of all paragangliomas and every effort should be made for complete resection.[14,15] Jugulo tympanic tumors are challenging for complete resection. Therefore, radiation should be offered post-operatively if there is residual disease.

CONCLUSION

Head and neck paragangliomas are rare tumors. Detailed clinical examination and rdiographic studies are important for diagnosis. Surgical resection for carotid body tumors has excellent outcome but for vagua nerve tumors, X nerve dysfunction results in major morbidities. Complete resection for jugulotympanic tumors is challenging and radiation therapy should be offered if there is any residual disease.

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