Presentation and Management of Carotid Body Tumors

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Abstract

Objective: To review the presentation, management and outcome of carotid body tumors at a University Hospital in Karachi.

Setting: The Aga Khan University Hospital, Karachi - a tertiary care hospital in Pakistan.

Methods: The medical records of all the patients who were admitted between 1st January1991 to 1st January 2002, with a diagnosis of carotid body tumor and underwent surgical resection, were identified using the ICD-9-CM coding system. The variables reviewed included demographic features, clinical presentation, diagnosis, management and outcome. The data was analyzed using the student t-test for continuous variables and the Fischer Exact test for categorical data, with computer-based SPSS version 10 software package.

Results: A total of 8 patients with 8 carotid body tumors were treated surgically during the study period of 11 years. The study group consisted of 6 males and 2 females with a median age of 46 years (range: 26-68 years). All the eight patients presented with a history of neck mass which was progressively increasing in size. Physical examination revealed well-circumscribed firm masses at the angle of jaw in 6 patients. The other two patients had large tumors extending to the base of skull. The mainstay of final diagnosis was angiography, which was performed in all 8 patients. Pre-operative angio-embolization was done in three patients; followed by complete surgical resection in all 8 patients. There was no peri-operative mortality or stroke in these patients. Transient nerve palsies involving 7th and 12th cranial nerves were noted in three patients. One patient had permanent unilateral vocal cord palsy resulting in hoarseness of voice. Median follow up was 1 year with range from 8 weeks to 3 years; none of the patients had any recurrence during the follow-up period.

Conclusions: Surgical resection of the carotid body tumors can be performed in suitable patients without major morbidity and mortality. There is a small but definite risk of neurologic complications, which should be clearly explained to the patients. Pre-operative angio-embolization facilitates surgery and reduces operative complications (JPMA 53:306;2003).

Introduction

The carotid bodies are reddish brown, ellipsoid structures, lying embedded in the adventitia of the carotid artery bifurcation.[1,2] Physiologically, they are known to be involved in the reflex control of heart rate, blood pressure and respiration via the chemical composition of blood and its temperature.[1] Described first by Haller in 1743, they are derived from the epitheloid cells of neuroectodermal origin.[3] Tumors of this tissue were originally described as chemodectomas by Mulligan; now they are considered as a part of the widely described group of tumors known as paragangliomas.[1]

Carotid body tumors are very rare neoplasms constituting less than 0.5% of all body tumors.[4] An incidence of 0.012% of all surgical specimens has been reported by a hospital based study describing the paragangliomas of the head and neck region.[5] Exact etiology of these tumors is not known, but they are noted to be more common in people living at high altitudes.[6] Sporadic forms of carotid body tumors are more frequent, while familial forms account for about 10% of the cases in most series.[7] The tumors are bilateral in 30% of the familial, but only 5% of the sporadic cases.[8]

Carotid body tumors are usually benign. Symptoms are generally due to local involvement of the nerves and vessels. The malignant potential with possible metastasis has been estimated to be around 2% to 9%; but as for most other neuroendocrine tumors, the usual histologic criteria for malignancy i.e., nuclear atypia and nuclear to cytoplasmic ratio, do not apply; and the likely clinical behavior cannot be predicted from the routinely stained tissues.[9,10] The true proof of malignancy is the presence of lymph node or distant metastases, which may not become evident even years after the original resection. Familial, bilateral or multiple paragangliomas in young patients are more prone to aggressive behavior, and these tumors can metastasize to the lungs, liver and bones.[4] Some researchers suggest that all carotid body tumors be considered malignant because of their progressive involvement of local neurovascular structures.[11]

Most of the carotid body tumors are asymptomatic in the early clinical phase. Eventually, at least 75% of the patients develop symptoms such as enlarging neck mass, neck pain, hoarseness or syncope.[4] Rarely the patients may exhibit other symptoms when the tumors are endocrinologically active. The physical examination usually reveals a pulsatile mass below the angle of mandible which can be moved laterally but not vertically (Fontaine's sign).[4]

A number of diagnostic and therapeutic approaches have been suggested for the management of carotid body tumors, but they continue to pose diagnostic, investigative and therapeutic challenges. The diagnostic modalities used for the evaluation of carotid body tumors include, color-flow duplex ultrasonography, computed tomography and magnetic resonance imaging. Angiography is conventionally considered to be the gold standard. The treatment options include surgery, radiotherapy and angio-embolization.[12] The decision to operate depends upon the age of the patient, co-morbid conditions, symptoms, rate of growth and the size of the tumor.[12] As it is a relatively slow growing tumor, a select group of patients in the advanced age group or with significant co-morbids may be carefully observed with possible use of radiotherapy, resulting in a reasonable survival.[12] In young patients who are otherwise low risk for anesthesia, surgery remains the treatment option of choice, as it is the only available curative therapy.

The basic objective of our study was to review the presentation, management and outcome of carotid body tumors at our hospital.

Patients and Methods

The medical records of all the patients who were admitted at The Aga Khan University Hospital (AKUH), Karachi, between January 1991 to January 2002, with a diagnosis of carotid body tumor were identified and retrieved using the ICD-9-CM coding system. The data was collected for patients who underwent surgical resection, and the variables reviewed included demographic features, clinical presentation, diagnosis, management and outcome. A special note was made for the type of therapeutic intervention used including the use of pre-operative angio-embolization and surgical procedure. The morbidity and mortality related to each procedure was also identified. The data was analyzed by using the student t-test for continuous variables and the Fischer Exact test for categorical data with computer-based SPSS version 10 software package.

Results

A total of 8 patients with 8 carotid body tumors were treated surgically at the AKUH during the study period of 11 years. The study group comprised of 6 males and 2 females with a median age of 46 years (range 26-68 years). One patient had a family history of carotid body tumors, while another had an extra cervical ganglioma (abdomen) resected previously. All the eight patients presented with history of a neck mass, which was progressively increasing in size. In 6 patients the mass was asymptomatic, while two patients complained of mild neck pain. None of the patients had symptoms suggestive of endocrinological activity. Physical examination revealed well-circumscribed firm

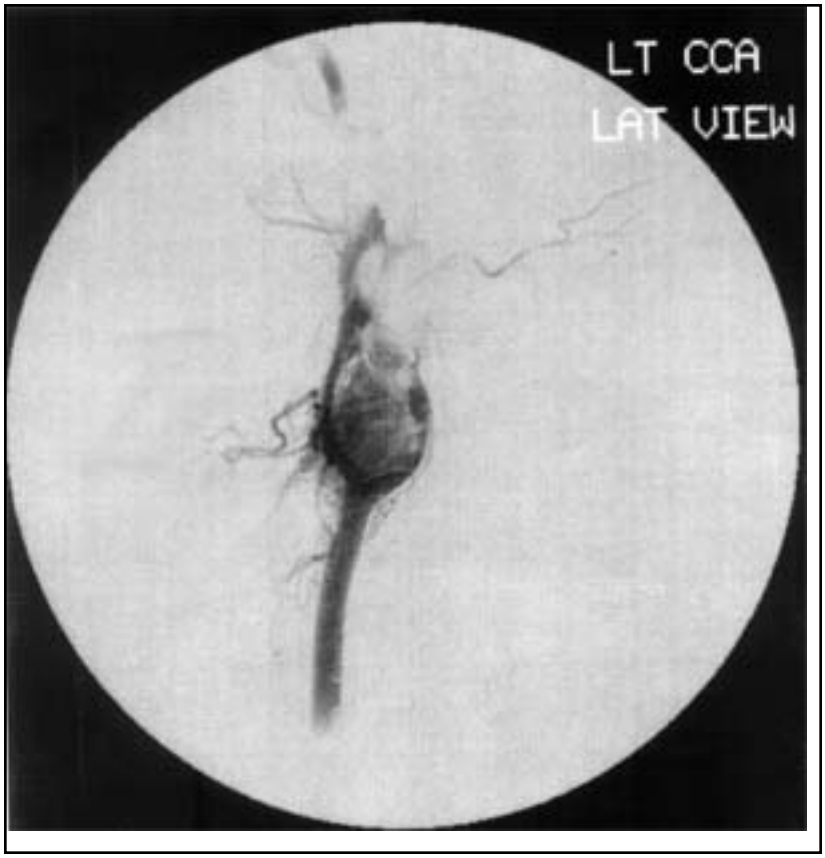

Figure 1. Angiogram displaying typical highly vascularized carotid body tumor with splaying of the internal and external carotid vessels.

Figure 2. Post embolization angiogram showing loss of tumor blush.

masses at the angle of jaw in 6 patients, which were mobile in the transverse plane. Two patients had large tumors extending to the base of the skull. There was no palpable bruit over any of the tumors. None of the patients had clinical evidence of a cranial nerve involvement.

The mainstay of final diagnosis was angiography, which was performed in all 8 patients. It showed a typical highly vascularized mass causing splaying of the internal and external carotid arteries. (Figure 1). Duplex ultrasound was also performed in 4 patients before angiography. It showed typical features of a hypoechoic vascular mass with low resistance arterial flow suggestive of the diagnosis. In two patients CT scan of neck (base of skull) was performed to evaluate the extent of tumor. FNA was done in two patients, one out side AKUH and the other at AKUH; and at both occasions it was inconclusive. Tumor size reported in these patients with investigations ranged from 2x3 to 8x5 cms.

Pre-operative angio-embolization was done in three patients. The selection of the patients for embolization was random, and there were no definite criteria for this selection. Subjectively, pre-operative embolization was found to be very effective in reducing the vascularity of the tumor, and facilitated surgery. (Figure 2). Objectively, it was not possible to compare the estimated blood loss during the surgery in patients who underwent pre-operative embolization as compared to those who underwent surgery without embolization, as this was a retrospective study and the relevant data was missing from the medical records. Surgery was performed within 24 hours of embolization in all three cases. No complication of the embolization was observed in any case.

Complete surgical resection was done in all 8 patients, using vertical incisions along the anterior border of sternocleidomastoid muscle in 7 cases and transverse incision in one case. Proximal and distal control of large vessels was achieved by using vascular slings. The hypoglossal and vagus nerves were identified and preserved. Peri-adventitial plane (Gordon Taylor's plane) was then identified and resection of the tumor was done. In 4 patients, the tumors were large and encircling the major vessels. Out of these 4, the external carotid arteries were ligated and divided in three cases, while in one case a segment of the internal carotid artery was excised and then reconstructed with a graft. In 3 patients enlarged lymph nodes were found per-operatively and these were removed. Histopathology revealed only reactive changes in all the three cases. Closed suction drains were placed in all the patients at the end of the procedure. Only one patient required blood transfusion per-operatively.

There were no peri-operative strokes or death in these patients. Transient nerve palsies involving 7th and 12th cranial nerves were noted in three patients, but all recovered spontaneously. One patient had permanent unilateral vocal cord palsy resulting in hoarseness of voice. All the patients were discharged home between 48 to 72 hours after the surgery. Median follow-up duration was 1 year with a range from 8 weeks to 3 years. Although it would be meaningless to draw any conclusions regarding the recurrence of the tumor due to such a short follow up, nevertheless, none of the patients had any recurrence during this period.

Discussion

Carotid body tumors are uncommon lesions and there have been a number of sporadic reports in the literature. Approximately 1000 cases had been reported in the literature till 1990s.[13] The Mayo Clinic reported one of the largest series to-date, describing 153 cases over a period of 50 years.[14]

Carotid body tumors can be a diagnostic challenge for the clinician and lack of pre-operative diagnosis has been reported in up to 30% of the cases in different series.[6,15] The usual presentation is a slow growing mass at the angle of mandible. Sometimes these tumors are confused with enlarged lymph nodes, branchial cysts, salivary glands, carotid aneurysms, or neurofibromas. The patients are consequently subjected to unwise attempts at biopsy or explorative surgery. In our study, none of the patients underwent a preoperative open biopsy, although two patients had an attempted FNA that was inconclusive. The possibility of a gradually enlarging mass in the anterior triangle of neck should alert the clinician to the possibility of a carotid body tumor. Additional investigations are mandatory before a surgical exploration is attempted. Exploration and biopsy can be disastrous and should be avoided in the management of carotid body tumors[7,12], while fine needle aspiration (FNA) cytology is usually inconclusive.[1]

Color-flow duplex ultrasonography and angiography are the mainstay of diagnosis. The color-flow duplex has high specificity and sensitivity and can be used for the diagnosis, follow-up and also for screening of high-risk patients.[6,7] It is a non-invasive and easily performed procedure, and recent studies have delineated typical sonographic features pathognomic of carotid body tumors. In our study, color-flow duplex ultrasound was performed in 4 patients and was diagnostic in all cases. Angiography shows a highly vascularized mass causing typical splaying of the external and internal carotid arteries. Angiography should be performed on both sides, as in 5 to 10% cases the tumors are bilateral. Although angiography remains the gold standard for diagnosis,[7,16,17] its routine use has been questioned recently due to the risk of associated complications and comparable results with non-invasive imaging modalities.[6] CT and MRI are usually performed to evaluate the intracranial extent, and involvement of the cranial nerves.[6,12,18,19] In our study, CT scan was performed in 2 patients to assess the extent of the tumor. Catecholamine screening can be done if endocrinological activity in the tumor is suspected on clinical grounds.

Although resection of carotid body tumors had long been considered a surgical challenge, surgery is the treatment of choice for these tumors. Initial attempts at surgical resection yielded devastating results with very high rates of mortality, cerebrovascular accidents and cranial nerve palsies. First surgical excision was reported by Maydl in 1886, but unfortunately the patient became hemiplegic and aphasic after the operation.[1] First successful excision of carotid body

tumor without ligation of carotid vessels was done by Albert in 1889.[1] The persistently high morbidity and mortality associated with the resection of carotid body tumors led Martin et al[20] to state in 1957 that these tumors should be subjected to biopsy only.

With the advancement in the fields of vascular surgery, peri-operative monitoring and good pre-operative evaluation the scenario has changed reducing the mortality to near zero and stroke to 1-3%. Cranial nerve palsies still remain one of the major complications, which can occur in 10-40% cases.[6] William et al[16] summarized the results for resection of carotid body tumors of major studies from 1986 to 1990 (177 patients) and found a mortality of 0.6%, stroke in 2.3% and postoperative peripheral nerve dysfunction in 29% of patients. The major series reported in literature from 1991 to 1995 show further reduction of the over all incidence of morbidity (stroke in 1.9% and nerve palsy in 22.3% of patients) without any mortality.[7] These figures are well matched with our results; in our study there was no mortality or stroke, but 3 patients (37%) had transient and one patient (12.5%) had permanent nerve dysfunction.

The routine use of preoperative angio-embolization for carotid body tumors has been questioned because of the potential neurologic complications associated with the accidental reflux of particulate matter into the ophthalmic or cerebral circulation.[6] Some authors advocate its use before the resection of large tumors because it decreases the vascularity of the tumor, reducing intra-operative blood loss and transfusion requirements.[6,21] In our series, this adjunctive procedure was successfully performed in 3 of 8 patients and it was felt that it is effective in reducing the vascularity. There were no neurologic complications associated with angio-embolization in our series, and overall only one patient required peri-operative transfusion. We believe that the benefits of angio-embolization should be weighed against the risk of stroke in an individual patient. Another possible role of angio-embolization might be palliative use for unresectable tumors.[1]

Another unresolved therapeutic issue is the benefit of radiotherapy in these tumors. Radiotherapy has been used in patients with metastatic lesions or tumors presumed to be malignant in certain series. Although some reports have indicated that such tumors were radioresistant,[20,22] others have shown that radiotherapy can result in regression in some instances.[12] However, after initial control, recurrence has been observed.[1] Overall, there is little evidence to suggest that carotid body tumors are radio-sensitive and it is generally believed that radiotherapy has very little to offer in the way of curative treatment.[12]

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As it is very slow growing tumor with the growth rate of less than 0.5 cm per year, small tumors in old patients with significant risk factors for surgical intervention can be managed by observation alone. Martin et al reported the natural history of 28 patients with carotid body tumors, who received no treatment and were observed over a mean period of 11 years[7]; developed lymph node metastases and 2 died of distant metastases.[20] It is recommended that in older patients with asymptomatic and slow-growing tumors, the situation must be carefully discussed with the patient before a final decision is made in favor of surgery.[12]

It is concluded that despite many controversies in the management, surgical resection of the carotid body tumors can be performed in suitable patients without major morbidity and mortality. There is a small but definite

risk of neurologic complications which should be clearly explained to the patients. Pre-operative angio-embolization may be performed in large tumors, as it facilitates surgery and reduces operative complications.

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