Egyptian Journal of Surgery Vol. (19), No. (2), April, 2000

CAROTID BODY TUMOURS: SURGICAL MANAGEMENT AND PREDICTORS OF OPERATIVE RISK

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In spite of a low but constant incidence of neurologic complications, resection of carotid body tumours of all sizes in appropriate surgical candidates has been advocated as the sole line of treatment of these rare lesions. This study summarizes our experience in the diagnosis and treatment of these lesions trying to identify the factors that predict the magnitude of operative risk.22 patients with 23 non-familial carotid body tumours presenting to the Cairo and Alexandria University Hospitals over a 4-year period were reviewed. There were 10 females and 12 males and the mean age was 42 years. A preoperative diagnosis, of being carotid body tumour, was not reached in 7 patients. In the remaining 16 tumours, duplex scanning, angiography, CT scanning and MRA clenched the diagnosis preoperatively, one tumour had FNAB and another open biopsy. 21 tumours were resected and the remaining 2 were irradiated. Internal carotid artery reconstruction employing great saphenous vein was required in 7 cases, end to end anastomosis in one case and lateral repair in another. A temporary shunt was used in 5 instances. Three patients suffered a transient hemiparesis while another 3 suffered a transient hypoglossal nerve palsy.It was concluded that surgical resection remains an effective form of treatment but is not without risk. The predictors of operative difficulty and hence increased risk include lack of preoperative diagnosis, mid and large sized tumours, Shamblin classes 2 and 3, use of FNAB as well as previous surgery

Keywords: Carotid body tumour, chemodectoma, carotid artery injury, postoperative neurologic complications, paraganglioma

INTRODUCTION

Since first reported by Marchand [1] in 1891 carotid body tumours (CBT, chemodectoma, carotid paraganglioma) have been relatively rare and posed a difficult surgical problem because of their vascularity, proximity and possible infiltration of the carotid bifurcation, compression of cranial nerves in the neck and extension to the skull base [2].

Other factors incriminated include the substantial size reached before' definitive diagnosis [3] as well as the lack of apreoperative diagnosis in up to 30% of patients [4].

Early reports were associated with morbidity and mortality rates of 40% and 30% respectively leading some surgeons to consider non operative treatment [5,6]. However, because of the locally invasive nature and uncertainty about their natural history. resection of chemodectomas is still highly recommended soon afier diagnosis [7,8,9] particularly that advances in preoperative evaluation, surgical technique and intraoperative monitoring reduced the mortality to zero.

Unfortunately, the incidence of neurologic complications remained between 10-40%[10, 11].

This study summarizes our experience in the diagnosis and management of these lesions trying to identify the factors that predict an increased operative risk.

MATERIALS AND METHODS

This study included patients who were treated for carotid body tumours at Cairo and Alexandria University Hospitals between 1995 and 1998. Patients' demographics,

mode of presentation, preoperative evaluation, tumour size, treatment modalities, Shamblin classes, neurologic complications (stroke, cranial nerve injuries), histopathology, metastasis and recurrence were all noted.

Preoperative evaluation included duplex scanning, angiography, CT scanning and MRA. FNAB was done in one case and open biopsy in another. Tumours were arbitrarily classified into small (less than 4 cms), mid sized (4 to 5 cms), and large tumours (more than 5 cms).

21 tumours were managed by resection. The remaining 2 received irradiation, because they were judged too extensive for resection. Preoperative embolization was not used in this study. The Shamblin classification was used to assess the difficulty of resection. Class 1 consisted of tumours easily isolated and dissected from the carotid vessels, class 2 tumours were more adherent to the adventitial layer and partially encircled the vessels at the bifurcation, and class 3 tumours were densely adherent to the carotid vessels and completely encircling the carotid bifurcation.

The factors associated with increased operative difficulty and hence risk were identified

RESULTS

Over the 4 year period of this study, 22 patients with a total of 23 non-familial CBTs were reviewed. Of these patients 10 were women and 12 were men and the mean age was 42 years (range 18 - 78). One male patient had bilateral tumours. None of the cases had a relationship to high altitude. Co-morbid conditions are listed in (Table 1), of particular note is the lack of cerebrovascular disease in all 22 patients.

Table (1)*: Co-morbid conditions*

Condition	*N (%)*
Smoking	12 (54.54%)
Hypertension	7 (3 1.8 1%)
Peripheral vascular disease	4 (1 8.1 8%)
Coronary artery disease	2 (9.09%)
Diabetes mellitus	4(18.18%)

All patients presented with a mass just below the angle of the mandible. A preoperative diagnosis was not reached in *7(30.4%)* carotid body tumours which were confused with enlarged cervical lymph nodes. No specific imaging modalities were used in these 7 tumours.

In the remaining 16, duplex scanning (Fig. 1) was done 8 times, angiography 8 times, CT twice (Fig. 2), and MRA only once (Fig. 3).

One asymptomatic tumour was detected during duplex scanning done for contralateral tumour. One tumour had FNAB and another had open biopsy.

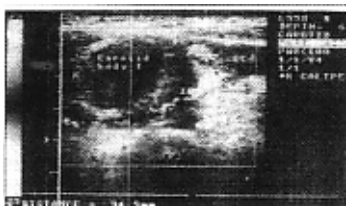

Fig. (1): Duplex scan showing a carotid body tumour

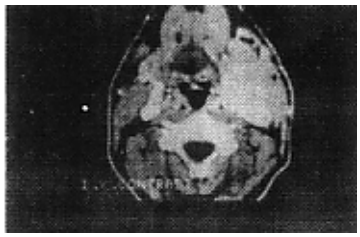

Fig. (2): CT scan showing a carotid body tumour

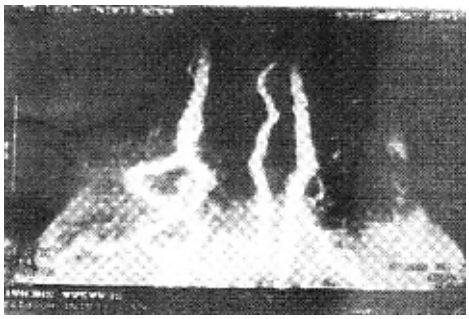

Fig. (3): MRA showing characteristic splaying of the carotid bifurcation

There were 8 small (up to 4 cms), 11 (4-5 cms) mid

sized tumours and large tumours (more than 5 cms). 21 carotid body tumours were managed by resection while the remaining two were irradiated (and not resected) owing to the extent. Of the 21 resected tumours, two were resected after 2 previous unsuccessful attempts, 1 was resected after one prior attempt while in 3 tumours, the authors were called because of bleeding encountered during the excision of what was thought to be a lymph node. The technique used involved subadventitial (sometimes periadventitial resection, cutting into the tumour was sometimes resorted to in the advanced Shamblin classes to free the carotids of the encircling tumour and finally securing of feeders. Resection entailed internal carotid artery reconstruction employing non- reversed great saphenous vein in 7 cases (one leg was prepared in all cases with a preoperative diagnosis of chemodectoma), end end anastomosis in one case and lateral repair in another. Temporary carotid shunts were used in 5 cases. Out of the remaining 4 (of the 9 patients who required some form of carotid reconstruction), in one it was not required during lateral repair and in the other 3, it was not used because these were the instances in which the authors were called in. The external carotid artery was ligated in 7 instances.

According to the Shamblin classification, there were 7 (30.4%), Class 1 tumours, 12 (52.1%) class 2 tumours, and 4 (1 7.3%) class 3 tumours.

All class 1 -tumours were small sized (< 4 cms), while class 2 tumours belonged to the small and mid-sized tumours (4-5 cms), all class 3 lesions were large (>5 cms).

Of the 9 tumours requiring some form of carotid artery reconstruction, 5 were not diagnosed preoperatively, (2 of these were subjected to more than 2 attempts at resection), 2 were Shamblin class 1 (but then again not diagnosed preoperatively), 5 were mid-sized Shamblin class 2, 2 were large Shamblin class 3 and one had FNAB.

Three (14.28%) patients suffered a transient hemiparesis and another 3 (14.28%) a transient hypoglossal nerve palsy. All 3 strokes occurred in patients who had been subjected to an attempt at excision and another surgeon had to be called in because of bleeding as a result of carotid injury (i.e. they were all among the 9 patients who required carotid artery reconstruction) and no shunt had been used. All 3 hypoglossal nerve injuries occurred in patients with midsized and large Shamblin class 2 and 3 tumours.

Only one patient in this study required a transfusion of 2 units of blood. There were no instances of baroreceptor failure. Histopathology confirmed the benign nature of all resected carotid body tumours (Fig. 4). There was no operative mortality in this series. There were neither tumour recurrence nor evidence of metastasis over a postoperative follow up of 19 months, range (8-24 months).

Fig. (4): Microscopic appearance typical of CBT

(well defined nests and groups of cuboidal cells (zellballen) with central rounded nuclei separated by thin vascularized fibrous septa (H&E, x 40).

DISCUSSION

Carotid body tumours are uncommon lesions; approximately 1000 cases had been reported by the year 1990 [12]. Most carotid body tumours appear to be sporadic (70-80 %), the remaining show familial tendency and bilateraty, either svnchronous or metaclironous [13]. Males and females. in the fifth decade of life, are affected with the same frequency, however, tumours occurring at high altitudes have a marked predilection for female patients [4].

Patients evaluated in this study conformed to these features except that all cases were sporadic and none was apparently related to hypoxaemic stimuli (i.e., significant cyanotic heart disease or severe chronic obstructive pulmonary disease), which have been found by some investigators [14] to be associated with both hyperplasia and neoplasia of the carotid body.

Only one patient had bilateral tumours and then again sporadic (as proven by screening of the family).

Lack of preoperative diagnosis has been reported in up to 30% of patients [4] with these tumours. Some are confused with enlarged lymph nodes and other neck lumps and consequently subjected to unwise attempts at biopsy or exploratory surgery.

Approximately one third (7/23) of patients in this study were not diagnosed as having CBTs preoperatively and were subjected to attempts at excisional biopsy. In one small Shamblin [15] class 1 lesion, the procedure was uneventful and in effect curative, in another three the authors were called because of brisk hemorrhage and in the remaining 3, the procedure was deferred to another sitting.

The advent of new non-invasive imaging technologies potentially allows the detection of CBTs at an earlier stage[16]. Duplex scanning currently appears to be the most useful test for confirmation of the presence of CBTs and the assessment of Shamblin classes[16]. It also allows the detection of smaller sized lesions as well as very small impalpable ones. Its role in follow up of these lesions and screening for familial tumours cannot be overlooked. It also serves to identify coexistent cerebrovascular disease.

On the other hand, CT may be a better means of delineating the relationship of these tumours to nerves and vessels and detecting multiple lesions, thus, it is more than a screening tool; it is a thorough evaluative method [13].

Although conventional angiography yields a characteristic appearance its routine use may be limited in the future, owing to its invasiveness to those patients with indeterminate findings on the other non invasive tests. Apart from identifying individual tumour feeders, it provides access for preoperative embolization.

Cases in this series were subjected to conventional angiography in the early phase of the study but findings in the later part of the study have confirmed value of duplex scanning as the standard, sensitive and specific preoperative test In our series it could detect a silent contralateral tumour and it has also been use to screen relatives of the patient with bilateral CBTs.

The invasiveness of angiography has undoubtedly contributed to the reluctance of requesting this investigation in the preoperative evaluation of neck lumps. Now that duplex is widely available, we recommend its use in evaluation of all solitary lumps suspicious of being CBTs.

The rationale of aggressive surgical excision of CBTs was initially based earlier data that about 50% are malignant[17]. However, subsequent studies which restricted the term malignant to metastasizing tumours have shown that only 5-10% are malignant [18,19]. This fact led some authors to adopt non operative treatment with careful follow up to avoid the risk of carotid artery and cranial nerve injury during excision of CBTs [19].

More recent reports advocate resection of all CBTs once diagnosed to avoid the risk whatever small, of malignancy[20,21], and before difficulty in excision arises. The increasing use of newer imaging modalities particularly duplex scanning may allow earlier detection and hence safer operation [22]. Discovery of such very small tumours creates a dilemma, should such tumours be resected In one study [16], one such tumour in a 77 year old patient was treated conservatively with annual duplex surveillance.

We have resected 21 carotid body tumours none of which seemed a candidate for conservative treatment. Although carotid artery injury with subsequent repair was required in 9 instances in this study only 3 suffered transient hemiparesis. Also 3 suffered transient hypoglossal nerve injury.

Carotid artery repair was required in 5 out of 7 tumours with no preoperative diagnosis and in only 4 out of 16 with a confirmed preoperative diagnosis. In all except 2 the size of the tumour exceeded 4 cms and belonged to the Shamblin classes 2 and 3. Still in the 2 Shamblin class 1 tumours. the preoperative diagnosis was lacking and the tumours were towards the 4 cm size. Also, 2 were subjected to more than 2 attempts at resection and one had FNAB which was particularly difficult to resect.

All 3 transient strokes occurred in instances when another surgeon was called in and no shunt was used. All 3 transient hypoglossal nerve injuries occurred in patients with mid sized and large Shamblin class 2 and 3**.** These results agree with other reports (21, 23) suggesting that small tumour size permits easier removal and a lower incidence of operative complications.

This study also confirmed the work of Davidge Pitts and Pantanowitz [19] who showed that there is a positive correlation between the size of the tumour and the Shamblin classification determined intraoperatively.

It seems reasonable to classify the risks associated with operating on a CBT into vascular and non-vascular complications. The former represent carotid artery injury, the forerunner of stroke and the latter representing cranial nerve injury Although stroke is not inevitable after carotid injury it is more prone to occur if preoperative diagnosis is lacking. Still, although the larger, Shamblin class 2 and 3 tumours are more risky, it is class 1 that is prone to misdiagnosis and then a undiagnosed Shamblin class 1 tumour becomes risky.

Further analysis of the results of this study has also shown that most complications were associated with more than one risk predictor and that combination of predictors would point to a vascular rather than a non-vascular complication.

Two unresolved therapeutic issues are the use of preoperative angioembolization and the use of radiotherapy. Angioembolization has be recommended before the resection of large tumours because it may decrease the vascularity reducing intraoperative blood loss and transfusion requirements[7]. On the other hand, this technology is associated with potential risk of reflux particulate matter into the ophthalmic or cerebral circulation[13].

In one study [16] this adjunctive procedure was successfully performed in 5 of 6 cases, the remaining patient sustained an ipsilateral procedure- related stroke. There was no objective evidence of diminished intraoperative blood loss.

In yet another study [24], there was no difference between embolized and non-embolized tumours particularly in the treatment of mid-sized tumours.

Preoperative embolization was not used in this series and only one patient required the transfusion of 2 units of blood.

Finally, controversy exists as to the role of radiotherapy in treating CBT Because recurrence has been observed after initial control [8] it is usually reserved for metastatic lesions or lesions presumed to be malignant.

Radiotherapy has been used to treat 2 tumours in this study, both judged too extensive as shown by CT. Both developed recurrence during the follow up period.

CONCLUSION

Carotid body tumour is a rare lesion, which is benign in the vast majority cases. Although resection of carotid body tumours of all sizes in appropriate surgical candidates remains the standard of care, it is not without risk of carotid arterv and cranial nerve damage.

The predictors of risk include the lack of a preoperative diagnosis, mid-sized and large tumours, Shamblin classes 2 and 3 the use of FNAB, and previous operative intervention. Invariably one or more of these predictors were present in patients who required carotid artery reconstruction as opposed to simple subadventitial resection. Preoperative embolization of these lesions does not appear to be standard practice. Also biopsy, whether FNAB or open is not recommended.

The wide spread use of duplex scanning will allow more and more suspicious lesions of the neck to be properly diagnosed preoperatively as well as smaller sized tumours to be detected without resorting to invasive angiography.

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