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REVIEW

A Multicenter Review of Carotid Body Tumour Management☆

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Objective. *Carotid body tumour (CBT) is a rare but the most common form of head and neck paraganglioma (PGL). We present the biggest ever series on CBT in UK/EU discussing diagnostic challenges, surgical treatment and complications of surgical intervention.*
Method. *A detailed proforma was designed and sent to all members of Joint Vascular Reasearch Group (JVRG). Data of 95 patients was collected. Generic terms including carotid body tumour/s, or paraganglioma/s were used to search a variety of electronic database in order to get latest informations available in literature.*
Results. *A total of 95 patients were recorded in our data from 1979 to 2005. Mean age of presentation was 55 years. Incidence was higher in females. CBT was more common on right side (58%). 18% tumours were bilateral. Neck lump (98%) and pressure symptoms including cranial nerve deficits and pain were main presenting complaints. About 18% of tumours were familial. Only 4.2% were malignant. Duplex scan is the best investigation for diagnosis, though MRI, DSA and CT scan are important for preoperative assessment. Surgery is the treatment of choice. Stroke and cranial nerve injury constitute postoperative morbidity (35%) and mortality (1%). Incidence of postoperative cranial nerve deficit was about 19%. Combined ipsilateral and contralateral recurrence rate was 4.2%.*
Conclusion. *CBT is a rare condition which needs surgical excision by experienced vascular surgeon. Surgical resection is associated with significant morbidity of 35% and mortality of 1%. Mostly CBT is benign but malignant forms are not uncommon.*

Keywords: CBT; Carotid body tumour; PGL; Paraganglioma.

Introduction

Although the most common form of head and neck paraganglioma, Carotid body tumours (CBT) remain rare and most vascular surgeons will encounter only a very few during their career. Publications on CBT management tend to arise from single centre studies.[1] In an attempt to determine more accurately the modes of presentation, investigation and treatment as well as outcome across a broader spectrum of surgeons, the retrospective experience of the 33 vascular units of the Joint Vascular Research group (JVRG) over several years has been pooled. . The Joint vascular research group consists of surgeons from both large tertiary referral units as well as smaller units.

Methods

A detailed proforma was designed and ethical committee approval gained to retrospectively review patient notes . This questionnaire included demographic details of patient, possible symptomatolgy, diagnostic techniques used, type of treatment provided, type of surgeon involved, post surgery complications, recurrence rate and follow up methodology. This questionnaire was emailed to all members of JVRG. Data from a JVRG member from Netherlands was also included.

A literature search was also undertaken in order to compare our data with others. Studies or case reports published between January 1970 and September 2005 were identified through the MEDLINE, EMBASE,

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CINAHL and COCHRANE LIBRARY databases with a search in all languages, through review of Current Contents. Generic terms carotid body tumour, carotid paraganglioma, head and neck paraganglioma, chemodectoma and glomus tumour were used to search the electronic database. Hand searching and looking through references quoted in useful articles were also carried out and yielded few more articles.

Results

11 vascular units did not have any experience of CBT management. 10 major vascular centres (Table 1) had treated these tumours and submitted proformas. 12 members did not submit data. So overall this data represents the experience of 21 out of 33 vascular units (68%). The data of 95 patients of CBT was reviewed to generate summative outcome. We assume that only vascular units with a fair number of cases and with better results submitted their data for combined analysis, therefore the quality of the results we are presenting may be biased by the selection criteria. The Mean age at presentation was 55 years but it varied from 18–94 years. The male to female ratio was 1: 1.9 (63:32). 55 (57%) tumours were found on right side, 24 (25%) on left side and 16 (17%) were bilateral. In patients where there was a family history (16) of CBTs, 4 tumours had had previous surgery and were considered recurrences. The dimensions and grade of each tumour (i.e. relationship and involvement of the carotid arterial/jugular venous system) was difficult to assess due to insufficient data and lack of common consensus on classification system of Shamblin. In this series only one tumour was functional and one was associated with paraganglioma else where, 4 (4.2%) were found to be malignant There was no consensus on the definition of malignant CBT among the members of JVRG but all tumours which were locally invasive and with the evidence of metastasis, were considered as malignant CBT. The Shamblin classification system was not used for every case in all vascular units and detailed information about this classification was not available in patient's history notes. That is why we could not analyze our data to assess the outcome according to Shamblin classification of CBT. The exact cause of higher female preponderance is unknown but possibly hormonal changes due to menstruation and pregnancy might well be a factor.

Almost all tumours presented as a neck lump with or without pressure symptoms like neck pain, dysphonia, hoarseness, stridor, dysphagia, odynophagia, cranial nerve palsy, jaw stiffness and sore throat . About 22% of patients presented with cranial nerve deficit. The functioning (Fig. 1). CBT which was associated with a pheochromocytoma (1) presented with palpitations, tachycardia and hypertension.

Duplex ultrasound scanning was considered primary diagnostic investigation among JVRG members. It depicts growth at carotid bifurcation causing splaying and helps to define hypervascularity of growth. Angiography (DSA), CT, CTA, MRI and MRA were used for preoperative assessment (Fig. 2). DSA was not used very often (only 20%) by JVRG members for the diagnosis of CBT or preoperative assessment.

Surgical resection of CBT was offered to all patients, though only one patient had preoperative radiotherapy for tumour destaging. Surgery was performed by specialist vascular surgeon alone (80%) or by ENT and vascular surgeon together (20%).

Surgical resection of CBT was associated with 1% mortality and 33% morbidity. Cranial nerve damage, stroke, TIA, wound haematoma; cutaneous nerve injury and Horner syndrome were main complications

Table 1. Participating JVRG members

No	Trust/Hospital name	Number of cases
1	Royal Free Hospital London	7
2	St Mary Hospital London	36
3	Mr Hero Van Urk (Rotterdam, Holland)	17
4	Nottingham	3
5	Royal Cornwall Hospitals	4
6	Norfolk and Norwich University Hospital	10
7	Freeman Hospital Newcastle	9
8	South Tees Hospital Middlesbrough	1
9	Birmingham Heartlands Hospitals	1
10	Royal Bournemouth	7
Total		95

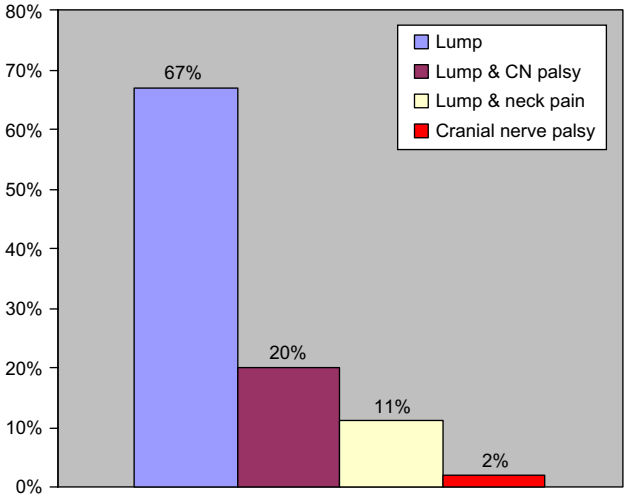

Fig. 1. Presentation of CBT.

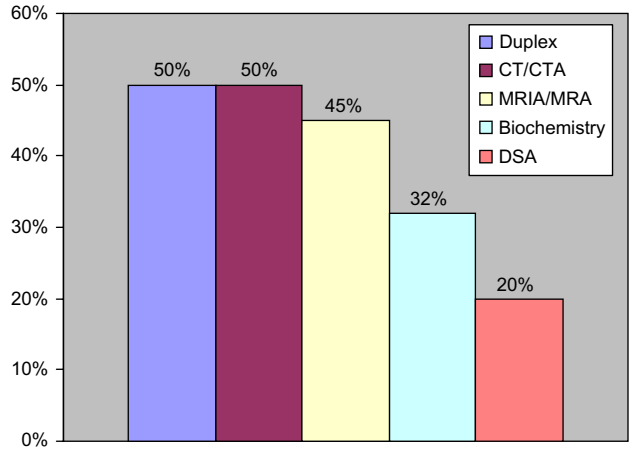

Fig. 2. Investigations used for CBT diagnosis.

after surgery (Fig. 3). The incidence of cranial nerve deficit on first post-operative day was 19%, which reduced to 3% after 6 months and less than 1% at mean follow-up. There was only one case of permanent cranial nerve damage (hoarseness of voice due to vagus nerve injury; 1%) where tumour size was 20 cm, malignant in nature and it was of type IV according to Shamblin classification.

About 95.8% of tumours were benign and 4.2% were malignant. Combined ipsilateral and contralateral recurrence rate was 4.2%. There was no fixed protocol for follow up in any of JVRG centre. Average follow up time was one year. Since this retrospective analysis encompasses 95 patients of CBT over a time

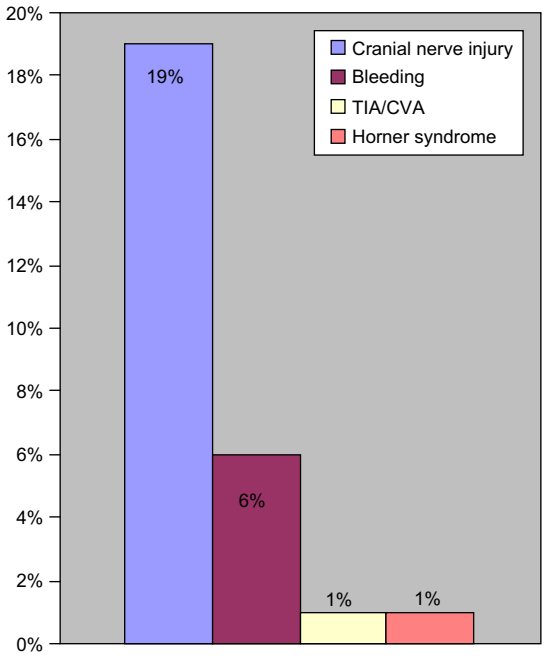

Fig. 3. Complications of CBT surgery.

period of 26 years, with mean follow up time of one year only, it is very difficult to explain the exact recurrence rate. There were 17 patients who lost follow-up and several more cases without follow-up record. Therefore, there is a strong possibility of sample contamination and biased record of CBT patients. Based on this fact, the recurrence rate in our data may be of limited value and same applies on follow-up data.

Since the experience of the management of CBT in different vascular centres of JVRG varies greatly (from 1 patient to 36 patients), it was difficult to compare and contrast the outcome of these units.

Discussion

CBTs are invariably benign but malignant forms are seen occasionally. Like other paragangliomas, CBT originates from cells which develop from neural crest. CBT is a non functioning tumour but sometimes histamine, serotonin, epinephrine and norepinephrine secreting tumours are also seen. Incidence of CBT is about 1 in 30,000. There are 3 distinct types; familial, sporadic and hyperplastic forms. Hyperplastic type is very common in patients of COPD, patients of congenital cyanotic heart disease and in the areas which are more than 5000 feet above sea level like New Mexico, Peru and Colorado. Chronic exposure to hypoxia is responsible for higher incidence of hyperplastic CBT in these patients. Familial type mainly involved younger patients, usually associated with head and neck paragangliomas and was more likely to be malignant.

Von Haller first described the carotid body in 1743. In 1762, Heller introduced the term glomus tumour and this was re-established by Gild in 1953. Reigner attempted excision of a CBT in 1880, but the patient died. Maydl also attempted CBT excision in 1886 but the patient developed a stroke. The first successful CBT excision was performed by Albert in 1889. Kohn first introduced the term paraganglioma (PGL) in 1903. Based on the work of Glenner and Grimley the term paraganglioma is now the most widely used to classify CBT.[2]

As mentioned before surgical resection is the only treatment of choice for CBT, though preoperative chemo-embolization and radiotherapy have been mentioned in literature. Surgical resection is not risk free and it is associated with significant morbidity and mortality.[3] The Shamblin's classification has been widely used for risk stratification before surgical intervention. According to this classification , bigger is the CBT, higher is the risk of cranial nerve injury and vascular injury. If the size of CBT is more than 5 cm, mortality is 1%–3% after surgical intervention.[3]

Neurological morbidity is not best described by this system and need for modified Shamblin's classification has been suggested in literature.[3] Whenever there is need for vascular reconstruction during resection of CBT, the risk of cranial nerve injury increases dramatically. This neurological deficit is more likely related to locally advanced tumour and pathology of the tumour rather than surgical technique.[4]

Mini Literature Review (Table 2)

Surgery is the only treatment of choice for CBT. There is very little if any role of embolization or radiotherapy in the management of CBT. On mini literature review, outcome of surgical intervention in JVRG series is comparable to international data. We compared our three parameters; cranial nerve injury rate, stroke rate and mortality rate with international studies on CBT management.

Luna-Ortiz *et al.* recently published an article quoting 49% incidence of cranial nerve deficit after CBT excision as compared to 19% in our article. However in literature the incidence of cranial nerve deficit after CBT surgery varies from 11% to 49%. Most of these cranial nerve injuries are temporary. The incidence of permanent cranial nerve deficit has been quoted less than 1% in international literature and it is comparable with JVRG data.

TIA/ Stroke rate after CBT excision varies from 0 to 8% in international studies while in our study it is 1%. Torres *et al.* published data of 96 patients of CBT and the incidence of TIA/Stroke after surgical intervention was 0% indicating a great variation in this complication. The difference about the incidence of TIA/Stroke among different publications may be due to experience of operating surgeon. Yang *et al.* published data of 27 patients and mortality in this series was 7.4% while it varies from 0–7.4% in international literature. Mortality in JVRG series was only 1%.

Table 2. Mini-literature Review

Studies (CBT surgery)	Patients	Cranial palsy	Stroke	Mortality
Luna-Ortiz *et al.*[5]	46	49%	0	1.5%
Patetsios *et al.*[6]	29	17%	0	3%
Dardik *et al.*[7]	25	33%	4%	0
Huang *et al.*[8]	30	36%	0	0
Plukker *et al.*[9]	35	11%	8%	0
Wang *et al.*[10]	29	41%	0	0
Westerband *et al.*[11]	31	19%	0	0
Torres *et al.*[12]	96	23%	0	1%
Yang *et al.*[13]	27	30%	2%	7.4%
Gaylis *et al.*[14]	44	18%	2.5%	4.5%
Dickinson *et al.*[15]	32	19%	4%	0

Conclusion

- Carotid body tumours are rare, but they are still the commonest of the head and neck paraganglioms.
- There are distinct hyperplastic, sporadic and familial forms.
- Duplex scan is effective in making the diagnosis, but angiography and CT or MRI is helpful to plan surgery.
- Excision is indicated for most tumours, but the surgery is not risk free and the best results are achieved by specialist team.

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