Print ISSN: 2321-6379
Online ISSN: 2321-595X
DOI: 10.17354/ijss/2016/519

Original Article

# Accuracy of Pre-operative Imaging Predictors of Shamblin Grades in Carotid Body Tumors

**Sudha Kiran Das[1], T P Suman[2], Kamal Kumar Sen[3], Shivananda Siddappa[4]**

[1]Assistant Professor, Department of Radiodiagnosis and Imaging, JSS Medical College and Hospital, JSS University, Mysore, Karnataka, India, [2]Resident, Department of Radiodiagnosis and Imaging, JSS Medical College and Hospital, JSS University, Mysore, Karnataka, India, [3]Professor, Department of Radiodiagnosis and Imaging, JSS Medical College and Hospital, JSS University, Mysore, Karnataka, India, [4]Associate Professor, Department of Vascular Surgery, JSS Medical College and Hospital, JSS University, Mysore, Karnataka, India

## Abstract

**Background and Purpose:** Magnetic resonance imaging (MRI) is both sensitive and specific imaging modality in the diagnosis and surgical planning of carotid body tumors (CBTs). Radiological criteria to predict Shamblin grades has been described based on MRI, however, literature validating the same is sparse. The purpose of this study was to evaluate the accuracy of radiological criteria used to predict Shamblin grades in correlation with pre-operative grading.

**Materials and Methods:** MRI images of 11 patients diagnosed to have CBTs during April 2014 to July 2016 at our institution were correlated with per-operative Shamblin grade. Radiologically, the tumors were classified into 3 types based on the arc of vascular contact with the internal carotid artery (ICA); contact ≤180° was categorized as Type 1, Type II tumors had more than 180° and <270°, and Type III tumors had a maximum circumference of contact of 270° or more.

**Results:** Of the 11 patients, two patients were excluded from our study, one had bilateral carotid tumors on imaging and was operated elsewhere, and the other two, histology of one was vagal schwannoma, and the others were a nodal metastatic adenocarcinoma. Of the eight, six were Type II, one was Type 1, and one was Type III. Pre-operative prediction of Shamblin grades correlated accurately with per-operative Shamblin group in all the 8 operated tumors.

**Conclusions:** MRI can accurately predict Shamblin group preoperatively based on the degree of circumferential contact of the CBT with the ICA on axial images.

**Key words:** Carotid Body Tumors, Shamblin grade, MRI

## INTRODUCTION

Carotid body tumors (CBT's) are slow-growing hypervascular tumors classically sited at the bifurcation of the common carotid artery and accounts for more than 50% of head and neck paraganglioma. These tumors are also known as glomus caroticum or chemodectoma; they arise from the carotid body, which is located in the adventitia of carotid bifurcation. Histologically, the carotid body comprises of both ectoderm and mesodermal tissue and has two types of glomus cells, namely Type I (chief or paraganglion cells) and Type II (sustentacular cells), physiologically it functions as a vascular chemoreceptor and is sensitive to hypoxia, hypercapnia, and acidosis.[1,2]

CBTs are mostly benign in etiology and run an indolent course and present anywhere between the third and fifth decade. The exact etiology of CBT is unknown, although its higher incidence among people living at high altitudes or those with chronic obstructive pulmonary disease suggests role of chronic hypoxia.[13] Mostly unilateral in occurrence, can be bilateral in 5% of sporadic cases, and in 33% of familial cases. Familial cases have been shown to be caused by germline mutations in three of the four succinate dehydrogenase subunit genes. Incidence of malignancy varies between 5% and 7% and usually in familial cases.[3,11] Mostly nonfunctional, however, occasionally they secrete catecholamines and can present with hypertension and tachycardia.

Access this article online

www.ijss-sn.com

Month of Submission : 08-2016
Month of Peer Review : 08-2016
Month of Acceptance : 09-2016
Month of Publishing : 10-2016

**Corresponding Author:** Dr. Sudha Kiran Das, Department of Radiodiagnosis and Imaging, JSS Medical College and Hospital, JSS University, M G Road, Mysore - 570 004, Karnataka, India. Tel.: +91-9902063135. E-mail: sudha7492@gmail.com

Surgical excision is the mainstay of treatment for CBT. Surgical morbidity and mortality is secondary to injury to the internal carotid artery (ICA) or the nerves at the time of resection. Meticulous pre-operative planning and careful patients' selection is essential for a successful surgical outcome. Shamblin *et al.*[4] graded these tumors based on clinicopathological studies to predict vascular morbidity. Arya *et al.*[5] proposed criteria based on arc of contact with ICA on axial magnetic resonance imaging (MRI) sequences for preoperatively prediction of Shamblin grades. Involvement of external carotid artery or the common carotid artery is not assessed, as morbidity is mainly related to ICA encasement. The purpose of our study is to validate the accuracy of these imaging criteria in correlation with per-operative Shamblin grade.

## MATERIALS AND METHODS

MRI images of 11 patients diagnosed to have CBT during April 2014 - July 2016 on imaging were included for the study. Patients with no operative records or with a histological diagnosis other than CBT were excluded from the study as all tumors histologically confirmed.

Computed tomography (CT) was done in two patients and MRI was done in 10 patients. The surgical records were blinded to the radiologists while they reviewed the MRI. Radiologically, the tumors were classified into 3 types based on the arc of vascular contact with the ICA; contact ≤180° was categorized as Type 1, Type II tumors had more than 180° and <270°, and Type III tumors had a maximum circumference of contact of 270° or more.

### Imaging Protocol

Contrast enhanced CT angiography was performed on 128 slice CT with bolus tracking, helical acquisitions of 0.625 mm from the level of aortic arch to the level of frontal sinus was done following injection of iodinated contrast media. Iohexol (Omnipaque 300 mg/ml) was injected at a rate of 3.5 ml/sec followed by saline chase using the dual head CT pressure injector. Axial images were reconstructed at 1 mm thickness with coronal and sagittal reformations.

Multiplanar and multiple sequence pre- and post-gadolinium contrast enhanced MRI was done on Philips 3 tesla MRI using a dedicated neck coil with three-dimensional (3D) time of flight angiography MR angiography.

### Imaging Criteria as Proposed by Arya *et al.*[5]

Radiological classification was based on the arc of contact; the angle suspended between two intersecting lines passing from the center of the ICA to the tumor. The degree of circumferential contact of the ICA with the tumor (Figure 1) was noted. Large tumors, wherein arc of contact with ICA exceeded 180°, the angle was assessed on the rest of the tumor, circumferential contact was inferred following subtraction by 360°. Based on this methodology, the tumor was radiologically classified into 3 types, Type I, ≤180°; Type II, >180° and <270°; Type III, ≥270° (Figure 1).

Angle measurements were made on T2-weighted axial images and correlated with post-contrast T1-weighted sequences using the inbuilt angle measurement tool available on Philips 3 Tesla MRI workstation. Two patients, one underwent a routine CT neck protocol and another was planned for CT angiography. Measurements could not be obtained on the former as the enhancement of ICA and CBT equaled in the delayed arterial phase. However, accurate depiction of tumor and ICA was possible on the angiography protocol, in this patient the same principle of measurement were used. This patient could not undergo MRI as she was claustrophobic.

### Per-operative Shamblin Grades

According to the currently applied Shamblin's classification[4] CBTs, based on the relationship between the mass and the carotid artery wall, are classified into three types: Type I, referring to those without encasement of the vessel wall, tumor size <5 cm, no widened carotid bifurcation, and easy for surgical removal; Type II, referring to those attached to the blood wall, but without encasement; Type III, referring to those located inside the blood vessel with encasement of the blood wall, tumor size larger than 5 cm with widened carotid bifurcation.[4] The Shamblin group of the tumor was classified based on at least 2 of the following criteria: (1) The extent of circumferential encasement of the carotid vessels by the tumor as seen intraoperatively; (2) the feasibility of obtaining a plane of dissection between tumor and the vessel; (3) the presence of adventitial infiltration on gross examination.

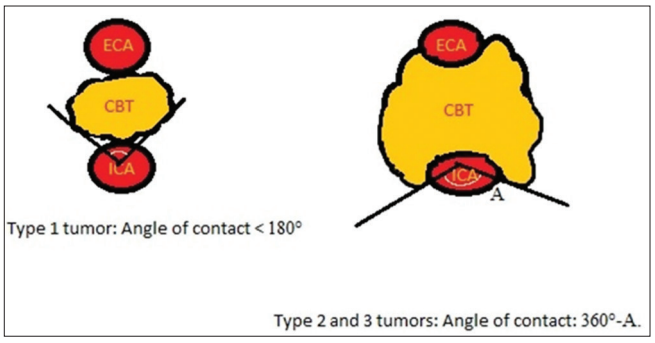


**Figure 1: Schematic representation: Measuring circumference of tumoral contact with internal carotid artery**

## RESULTS

Of the 11 patients, one lady bilateral carotid tumors on imaging and was operated elsewhere, and the others, one was diagnosed to be a vagal schwannoma and the other as metastatic nodal adenocarcinoma; three were excluded from the study

Of the eight, seven were female and one a male. The youngest was 18 years old and the oldest was 80 years. In this study, there was a definite female predominance as opposed to literature that documents male to female ratio of 2.5:1 was observed. Three tumors were on the right and five on the left. However, few other studies state that females are more frequently affected as seen in our study. All were sporadic cases, no predisposing etiology was observed.

In all the 8 patients, imaging appearances were classical for CBT. T2-weighted fat suppressed sequences depicted all of the tumors as hyperintense elliptical masses splaying the CCA bifurcation and with punctuate flow voids with the classical "lyre sign" (Figure 2). On CT, intensely enhancing hypervascular tumor splaying the CCA was seen. Five tumors splayed the CCA in the mediolateral direction, and three in the anteroposterior direction. The size of the tumor varied between 3 cm and 5 cm in the axial dimension.

Based on imaging criteria, six were Type II, one was Type 1, and one was Type III, this was concordant with per-operative grading. Per-operative grading Shamblin I tumor encased the vessels partially and could be easily dissected from the vessels, and did not show adventitial infiltration. Shamblin II tumors were difficult to dissect from the vessels, which had partial or focal adventitial infiltration. Shamblin III tumors encased the vessels almost completely, and despite meticulous dissection, plane between the tumor and vessels could not be obtained due to adventitial infiltration leading to incomplete resection (Figure 3).

It was possible to preserve, the ICA in all the cases and none of the patients we evaluated had post-operative neurological deficits or nerve injury. The radiological classification proposed by Arya *et al.*, was found to be accurate in all the cases with 100% positive predictive value.

## DISCUSSION

Our small case series reiterate the accuracy of pre-operative imaging prediction of Shamblin grades based on the criteria proposed by Arya *et al.* In this small series of

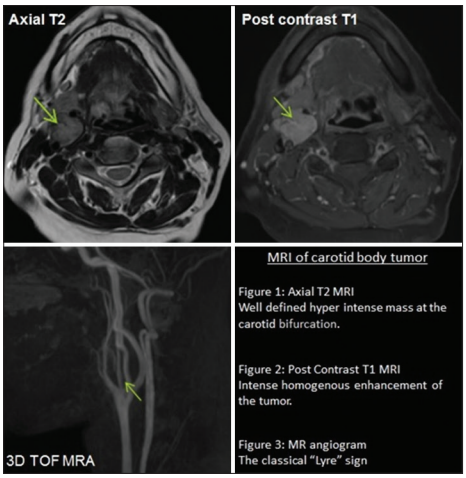


**Figure 2: Magnetic resonance imaging of carotid body tumor**

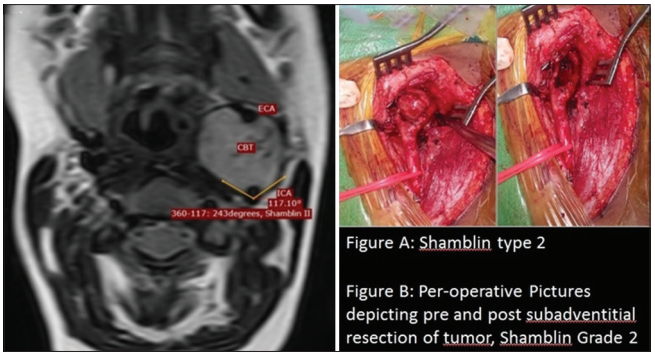


**Figure 3: Shamblin Grade II: Axial T2-weighted magnetic resonance imaging versus per-operative picture**

CBTs are mostly benign in etiology and run an indolent course. Usually unilateral, bilateral occurrence is rare, varies between 5% and 10% and is usually seen in the familial type. Incidence of malignancy varies between 5% and 7%. The incidence of CBTs is <1 in 30,000.[1,11]

Lushka[1] first described tumors arising from the carotid body in 1862. Etiopathogenesis of the tumor is largely unknown. The tumor mostly occurs in sporadic form while a minority of patients had a familial type which is thought to be related to genetic factors. They can occur at any age but are usually more common in the third to sixth decade of life. A higher incidence has been reported in those living higher altitudes typified by relative hypoxia.[6] Recent biogenetic discoveries reveal that mutations in oxygen sensing genes may also be responsible, accounting for approximately 35% of cases, these two etiologies are probably additive.[6,7] The tumor is usually nonfunctional but occasionally tumors capable of catecholamine secretion are diagnosed with symptoms similar to pheochromocytoma such as hypertension and tachycardia.

Carotid arterial angiography in decades past was the mainstay for diagnosis of these tumors. It aided in detecting

the tumor size, vascularity, blood supply, and presence of multiple tumors. The main vascular supply of the tumor is from bifurcation and external carotid artery, through the ligament of Meyer but may have contributory blood supply from ICA, vertebral artery, and thyrocervical trunk. A biopsy is contraindicated owing to tumoral vascularity.[7,8]

Color Doppler study, panoramic imaging with 3D sonography, CT angiography, and MR angiography are now the modalities for pre-operative imaging. Color Doppler study highlights vascularity of the tumor at the level of carotid artery bifurcation and is the first diagnostic tool, and this can be coupled with panoramic imaging as well as 3D sonography for optimal delineation of the tumor.[9,12] CT or MR angiography is imperative prior surgery as they aid in surgical planning.[14,15,17]

Rapid evolution of imaging techniques has replaced angiography in the present decade. Invasive angiography is resorted to only in patients planned for pre-operative embolization to reduce tumor vascularity; however, this too is not preferred as inflammatory response followed by fibrosis impedes sub-adventitial dissection peroperatively resulting in incomplete resection.[18,19]

The tumor is a surgical challenge due to its location, high vascularity and potential morbidity, and mortality secondary to injury to ICA or nerve injury. Novel surgical techniques have evolved over the years, but meticulous sub-adventitial dissection described by Gordon Taylor as the "white line" has stood the test of time. Although surgical removal of the tumor is the treatment of choice, if patient is elderly and unfit for surgery, radiotherapy can also be tried.[15,16]

Surgical classification by Shamblin *et al.* continues to be a predictor of vascular morbidity. Shamblin *et al.* classified these tumors into 3 groups based on the operative notes and gross specimen examination and established that the risk of surgical intervention depends mainly on the relationship of the tumor with the carotid vessels. According to Makeieff *et al.*,[20,21] The rate of serious complications, i.e., permanent nerve palsy and vascular complications was 2.3% for Shamblin Class I/II tumors and 35.7% for Shamblin Class III tumors ($P < 0.001$) and O'Neill *et al.*[10] found in his series that cranial nerve injury was more likely following the removal of larger tumors.

The importance of Shamblin classification increases significantly if pre-operative cross-sectional imaging can accurately predict the Shamblin group. CT and MRI is a frequently used imaging method in the diagnosis and pre-operative workup, however, most reports on the diagnosis and surgical management of CBTs do not mention specific or consistent imaging criteria to predict this classification. Van der Mey *et al.* have stressed the need for a uniform classification system for these tumors so that the communication in the literature could be consistent.

Our study highlights the accuracy of radiological classification based on arc of contact of tumor to the ICA which can be easily inferred on the axial imaging, be it CT or MRI. We propose the use of this simplistic radiological classification to be incorporated in all reports of CBT so as to aid the surgeon in pre-operative planning.

## ACKNOWLEDGMENTS

Department of Vascular and ENT Surgery, JSS Medical College and Hospital, Mysuru.

## REFERENCES

1. Peterson EW, Meeker LH. Tumors of the carotid body. Ann Surg 1936;103:554-71.
2. McPherson GA, Halliday AW, Mansfield AO. Carotid body tumours and other cervical paragangliomas: Diagnosis and management in 25 patients. Br J Surg 1989;76:33-6.
3. Fruhmann J, Geigl JB, Konstantiniuk P, Cohnert TU. Paraganglioma of the carotid body: Treatment strategy and SDH-gene mutations. Eur J Vasc Endovasc Surg 2013;45:431-6.
4. Shamblin WR, ReMine WH, Sheps SG, Harrison EG Jr. Carotid body tumor (chemodectoma). Clinicopathologic analysis of ninety cases. Am J Surg 1971;122:732-9.
5. Arya S, Rao V, Juvekar S, Dcruz AK. Carotid body tumors: Objective criteria to predict the Shamblin group on MR imaging. AJNR Am J Neuroradiol 2008;29:1349-54.
6. Atefi S, Nikeghbalian S, Yarmohammadi H, Assadi-Sabet A. Surgical management of carotid body tumours: A 24-year surgical experience. ANZ J Surg 2006;76:214-7.
7. Ward PH, Jenkins HA, Hanafee WN. Diagnosis and treatment of carotid body tumors. Ann Otol Rhinol Laryngol 1978;87:614-21.
8. Knight TT Jr, Gonzalez JA, Rary JM, Rush DS. Current concepts for the surgical management of carotid body tumor. Am J Surg 2006;191:104-10.
9. Derchi LE, Serafini G, Rabbia C, De Albertis P, Solbiati L, Candiani F, *et al.* Carotid body tumors: US evaluation. Radiology 1992;182:457-9.
10. O'Neill S, O'Donnell M, Harkin D, Loughrey M, Lee B, Blair P. A 22-year Northern Irish experience of carotid body tumours. Ulster Med J 2011;80:133-40.
11. Rush BF Jr. Familial bilateral carotid body tumours. Ann Surg 1963;157:633-6.
12. Shibuyaa Y, Umedab M, Yoshikawab T, Komori T. Carotid body tumour: Case report. Oral Oncol 2002;38:313-7.
13. Rodriguez-Cuevas H, Lau I, Rodriguez HP. High-altitude paragangliomas diagnostic and therapeutic considerations. Cancer 1986;57:672-6.
14. Persky MS, Setton A, Niimi Y, Hartman J, Frank D, Berenstein A. Combined endovascular and surgical treatment of head and neck paragangliomas – A team approach. Head Neck 2002;24:423-31.
15. Luna-Ortiz K, Rascon-Ortiz M, Villavicencio-Valencia V, Granados-Garcia M, Herrera-Gomez A. Carotid body tumors: Review of a 20-year experience. Oral Oncol 2005;41:56-61.
16. Suárez C, Rodrigo JP, Mendenhall WM, Hamoir M, Silver CE, Grégoire V, *et al.* Carotid body paragangliomas: A systematic study on management with surgery and radiotherapy. Eur Arch Otorhinolaryngol 2014;271:23-34.
17. van den Berg R. Imaging and management of head and neck paragangliomas. Eur Radiol 2005;15:1310-8.
18. Jansen JC, Baatenburg de Jong RJ, Schipper J, van der Mey AG,

van Gils AP. Color Doppler imaging of paragangliomas in the neck. J Clin Ultrasound 1997;25:481-5.
19. Netterville JL, Reilly KM, Robertson D, Reiber ME, Armstrong WB, Childs P. Carotid body tumors: A review of 30 patients with 46 tumors. Laryngoscope 1995;105:115-26.
20. Litle VR, Reilly LM, Ramos TK. Preoperative embolization of carotid body tumors: When is it appropriate? Ann Vasc Surg 1996;10:464-8.
21. Makeieff M, Raingeard I, Alric P, Bonafe A, Guerrier B, Marty-Ane C. Surgical management of carotid body tumors. Ann Surg Oncol 2008;15:2180-6.

**How to cite this article:** Das SK, Suman TP, Sen KK, Siddappa S. Accuracy of Pre-operative Imaging Predictors of Shamblin Grades in Carotid Body Tumors. Int J Sci Stud 2016;4(7):24-28.

**Source of Support:** Nil, **Conflict of Interest:** None declared.