Carotid Body Tumor in a case of Carcinoma Gingivo Buccal Sulcus

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Citation

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Abstract

We report a case of Carotid Body Tumor in a case of Carcinoma Gingivo Buccal Sulcus. Carotid Body Tumors have a variety of imaging characteristics obviating the need for a pathological diagnosis.

INTRODUCTION

Carotid body tumor is a common neck paraganglionoma which are neuro-endocrine tumors derived from the extra adrenal paraganglia of the autonomic nervous system. On imaging it has an array of characteristics which provide an accurate diagnosis.

CASE HISTORY

A 55 year old woman, chronic betel nut chewer for the past 30 years presented to the ENT OPD with complains of an ulcer in the mouth. On examination a fungating ulcer 2x2cms size was found in the left Gingivo Buccal Sulcus which on pre and post – operative cytology turned out to be a well differentiated squamous cell carcinoma.

On physical examination, another well defined globular swelling, 3x2.5cms in size and hard in consistency was found in the anterior triangle of the neck on the right side and on further questioning said that the swelling is present from the past 8yrs, not painful and not increasing in size.

All other clinical and lab parameters including blood pressure were within normal limits.

The patient was subjected to further radiological examinations for characterization of the neck lesion and to rule out if the neck swelling was a metastatic lesion.

Initially a neck ultrasound was done which showed a well defined echogenic mass lesion at the bifurcation of the right common carotid artery and on Doppler turned out be a highly vascular lesion giving a possible diagnosis of carotid body tumour.

Figure 1

Figure 1- USG image showing an echogenic lesion with splaying of ICA and ECA with internal colour flow.

Next the patient was subjected to a CT contrast study of the neck which again showed a well defined, irregular, circular, heterogenously enhancing mass lesion measuring 4.2x4x6cms (LxBxH) at the carotid artery bifurcation and seen engulfing the ICA. The angle at the bifurcation was splayed and there was no associated lymphadenopathy. These findings increased the possibility of a carotid body tumor.

Figure 2

Figure 2 - CECT axial ,a hypervascular oval mass splaying ICA anteriorly and ECA posteriorly

On MRI, a hyperintense circular lesion of the same size with internal signal voids giving the classical representation of “Salt and Pepper” appearance of a carotid body tumor on T2WI which confirmed our previous diagnosis. MRI also helped in classifying the tumor as grade II Shamblin classification based on the degree of encompassment of the carotid artery.

Figure 3

Figure 3 - T2WFS Coronal images showing right carotid body tumor splaying the ICA and ECA. Note: the characteristic Â“Salt-and-PepperÂ” appearance, with areas of haemorrhage(Salt) and flow voids(Pepper) and a small left parotid gland cyst.

Since the neck lesion was proved radiologically to be a benign carotid body tumor and on the absence of any lymphadenopathy in the neck, the patient was subjected to left hemi-maxillectomy operation for the carcinoma GBS.

Figure 4

Figure 4A – CECT axial views showing post-op left hemi maxillectomy with recurrence of Ca GBS

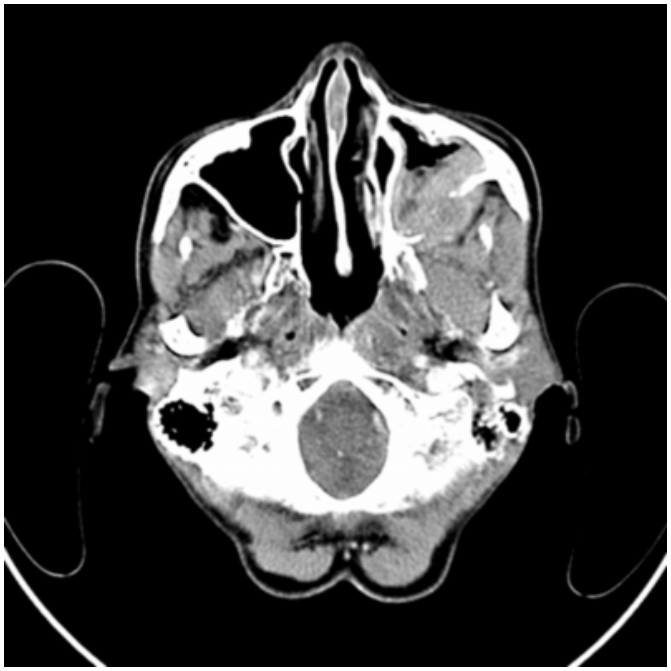
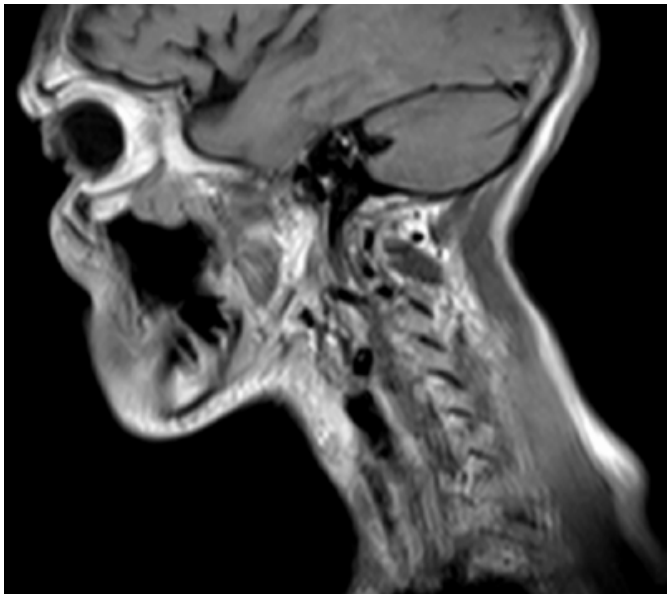


Figure 5

Figure 4B–MRI sagittal views showing post-op left hemi maxillectomy with recurrence of Ca GBS



DISCUSSION

CAROTID BODY TUMOR

Carotid Body Tumor or Glomus Caroticum is derived from the extra adrenal paraganglia of the autonomic nervous system. They are the second most common para gangliomas in head and neck. It was first described by Von Haller in 1743 and the term para ganglioma was coined by Kohn in 1903[1].

The tumor is located on the postero-medial wall of the internal carotid artery at its bifurcation and is attached by Mayer's ligament through which the feeding vessels run. They are ovoid or pear shaped. The normal carotid body measures 3 to 5mm in diameter.

Grossly they are dark tan to purple in color and usually fairly circumscribed and can extend along the internal carotid artery upto skull base.

Sporadic form is more common than inherited and tends to occur slightly more in women. It is multicentric in 10% with bilateral lesions more common. Malignancy occurs in 6 to 12.5%. Hereditary form occurs in 7 to 9% and is more frequently multicentric.

The tumor presents as slowly enlarging (~5mm per year), non tender anterior triangle of neck and as the tumor enlarges symptoms of Dysphagia, odynophagia, hoarseness and cranial nerve (IX to XII) deficits appear.

Compensatory hypertrophy of only the carotid body is seen in patients with prolonged hypoxia and hypercapnia[2] and populations living at high altitudes who are chronically exposed to hypoxic conditions and hence have a ten fold increased prevalence of the tumor[3].

ON DIAGNOSTIC IMAGING

On ultrasonography the lesion appears as a well defined hyperechoic, hypervascular mass showing arterial flow in the vessels within on Doppler. The lesion also demonstrates the relationship to the carotid artery and delineates any intrinsic carotid artery disease.

Figure 6

Figure 5A - USG image showing an echogenic lesion with splaying of ICA and ECA with flow in the vessels within

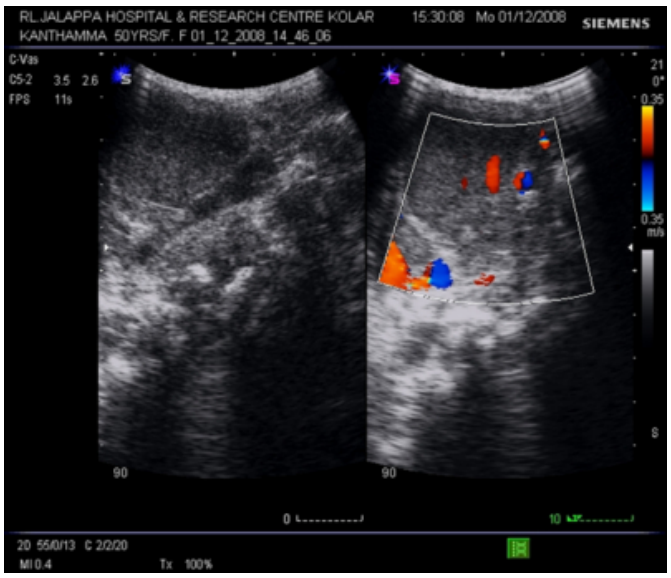


Figure 7

Figure 5B- USG image showing an echogenic lesion with arterial flow in the vessels within on Doppler

On CT and MR these hypervascular tumors show significant contrast enhancement and delay should be avoided as the contrast washes out early.

Dynamic scanning will reveal a hypervascular curve with a sharp filling peak and a rapid washout face[4,5,6]. Such a curve differentiates this vascular tumor from a hypo vascular lesion such as a schwannoma, which enhances because of the slow extra vascular accumulation of contrast in the tumor bed.

Small tumors enhance homogenously whereas larger ones are usually non homogenous with areas of both necrosis and hemorrhage.

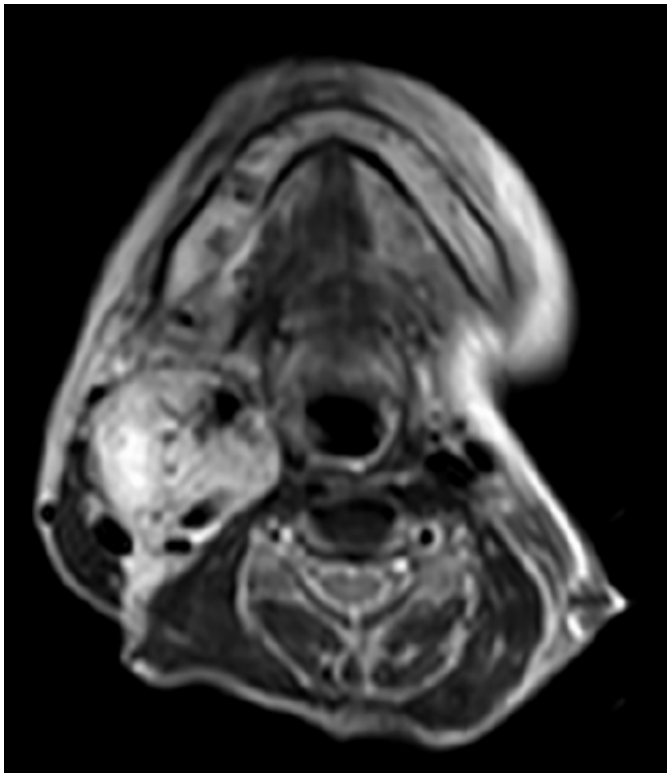
MR imaging is the most useful imaging modality in these cases because of its triplanar view and superior soft tissue contrast. They have an overall intermediate signal intensity background matrix on all imaging sequences usually with some “brightening” on T2 WI. Scattered sites of high T2 WI signal intensity can be seen, which help give the tumor “Salt & Pepper” appearance with areas of haemorrhage accounting for the salt appearance and flow voids the pepper appearance

Figure 8

Figure 6A- T2WI coronal image showing right Carotid Body Tumor splaying the ICA and ECA. Note: the characteristic Â“Salt-and-PepperÂ” appearance, with areas of haemorrhage(Salt) and flow voids(Pepper) and a small left parotid gland cyst.

Figure 9

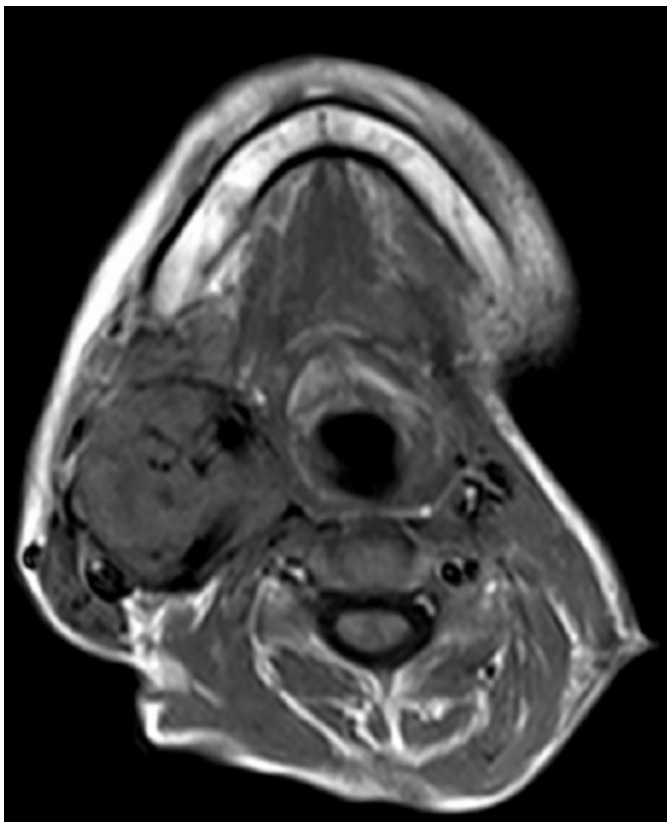
Figure 6B- T2WI axial image showing right Carotid Body Tumor splaying the ICA and ECA. Note: the characteristic "Salt-and-Pepper" appearance, with areas of haemorrhage(Salt) and flow voids(Pepper) and a small left parotid



In larger tumors a characteristic MR finding is serpentine, channel like areas of signal void within the tumor matrix which is present on all imaging sequences, which represents the large caliber, rapid flow dominant vessels of the tumor[7,8,9].

Figure 10

Figure 7 –T1WI non contrast axial, Carotid Body Tumour with areas of serpentine flow voids.



It has been suggested that MR contrast is unnecessary in studying para gangliomas and that the diagnosis is most reliably made on T2-weighted images. This study also confirmed that flow voids are more likely to be found in tumors greater than 2cm in diameter and on T2-weighted images[10].

Figure 11

Figure 8A: T1 Post contrast sagittal, right Carotid Body Tumour, characteristic splaying of the ECA and ICA.

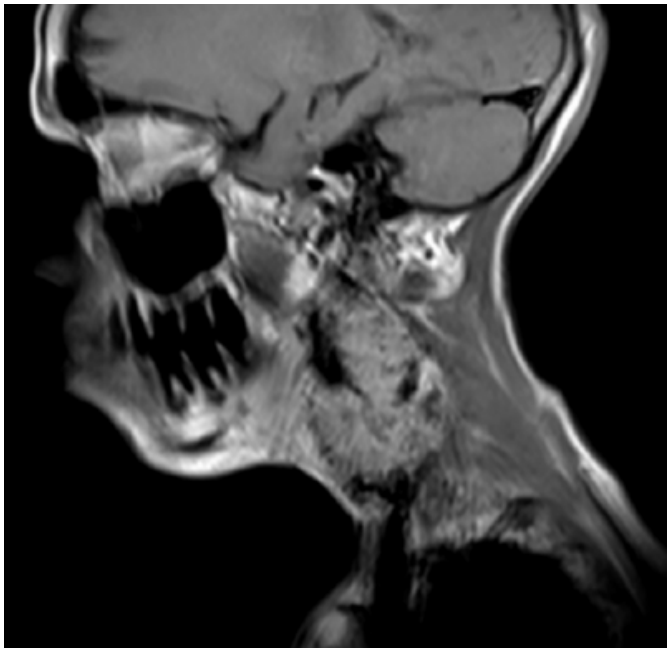
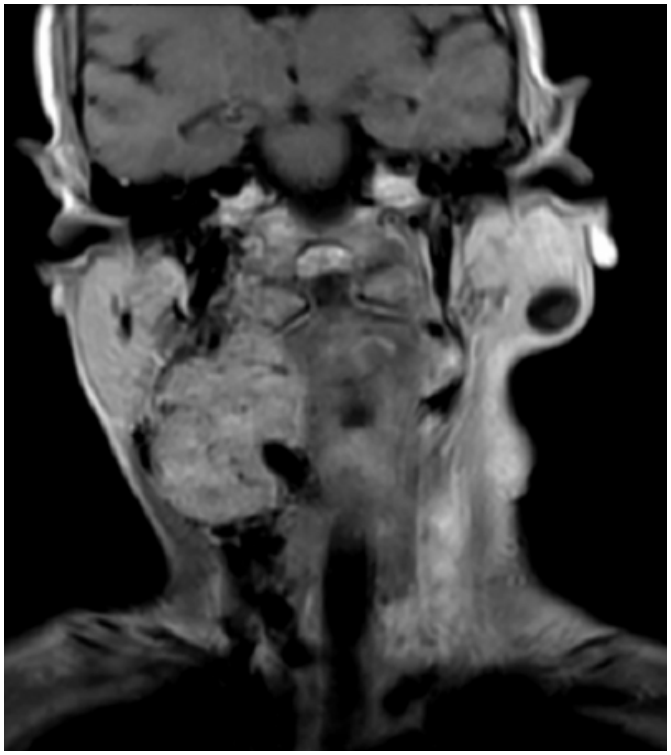


Figure 12

Figure 8B: T1 Post contrast coronal, right Carotid Body Tumour, characteristic splaying of the ECA and ICA.



Both glomus vagale and glomus jugular tumors arise around the vagus nerve dorsal to the ICA and tend to displace the ICA anteriorly. The carotid body tumors also usually displace the internal carotid artery anteriorly; however, splaying of the internal and external carotid arteries is a more constant feature[8].

This splaying causes a lyre-shaped widening of the carotid bifurcation, a distinctive imaging feature well seen on CT, MR, and conventional angiography.

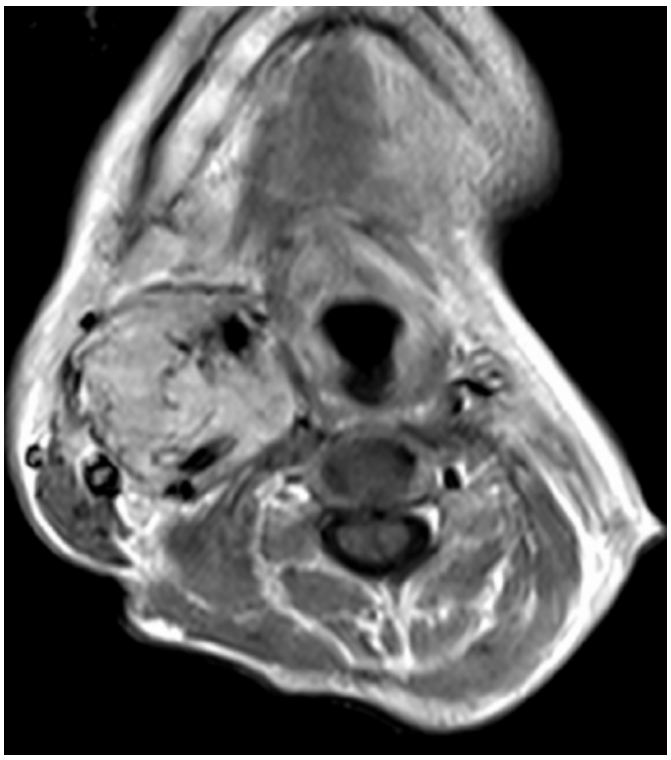
Multislice 3D TOF MRA is superior to 3D Phase contrast and 2D TOF MRA for identifying the first and second order vessels in the neck, the sensitivity of MRA is too low to replace conventional angiography, especially for carotid body tumors[11].

MRI imaging findings usually obviates the need for a definitive diagnosis by angiography.

MRI also has a high specificity (100%) in the Shamblin classification of the tumor to grade the difficulty of resection of the tumor[12]. Group I tumors are defined as small, localized & minimally attached to the carotid vessels, surgical excision was described as carried out without difficulty in this group. Group II included tumors adherent to or partially surrounding the vessels, with moderate arterial attachment and this group tumors were amenable to careful surgical excision. Group III tumors completely encompassed the carotid and careful approachment to these tumors with consideration for vessel replacement is advised[13].

Figure 13

Figure 9 – T1 Post contrast axial view, partial encasement of the right ICA, Shamblin group II.



Carotid angiography is useful in pre-operative embolization & in rare instances where malignant carotid body tumor is suspected. Additionally, angiography with temporary balloon occlusion using clinical and electroencephalographic monitoring, combined with xenon cerebral blood flow scanning can provide specificity as to the tolerance of collateral cerebral circulation across in the circle of Willis in selected cases

MIBG and Octreotide scanning can be used due to the high density of somatostatin receptors in paragangliomas and have been recommended as a possible screening test for familial paragangliomas for patients at risk[14].

Surgery remains the mainstay of the treatment and adjuvant radiation therapy may be considered after surgery for malignant carotid body tumors for locoregional control.

However, no case has been reported till date of a carotid body tumor in a case of carcinoma Gingivo Buccal Sulcus and no association between the above two has been postulated.

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