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CAROTID BODY TUMOR: OUR EXPERIENCE

Clinical Research

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ABSTRACT

Carotid body tumors are paraganglia arising at carotid bifurcation and commonly presents as painless slow growing mass with characteristic MRI finding – salt and pepper appearance and lyre sign – splaying of external and internal carotid artery. Surgical management is treatment of choice and radiotherapy reserved for inoperable case.

KEYWORDS

Paraganglioma, carotid body tumor. salt and pepper appearance.

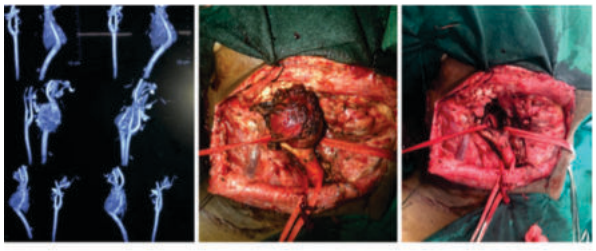
INTRODUCTION

Carotid body tumors are Paragangliomas that arise from neural crest cells, >90% occur in the adrenal paraganglia – termed as pheochromocytoma and rest occur outside of the adrenal gland - abdomen, thorax, and 3% in the head and neck [1]. The most common paraganglioma tumor subsite within the head and neck is carotid bifurcation – termed as Carotid body tumor with rare sites of occurance are jugulotympanic region, vagus nerve, sympathetic trunk, laryngeal paraganglia, ciliary ganglion, nasal cavity, and paranasal sinuses [2, 3, 4].

Head and neck paragangliomas are parasympathetic in origin and non-secreting (non-chromaffin paragangliomas) [5]. Paragangliomas can arise as hereditary or sporadic. When hereditary, gene mutations are usually found in the succinate dehydrogenase or mitochondrial complex II genes [3]. Most sporadic paragangliomas present as a single tumor, with 10–20% multifocality. However, familial paragangliomas are multifocal in 80% of cases [7].

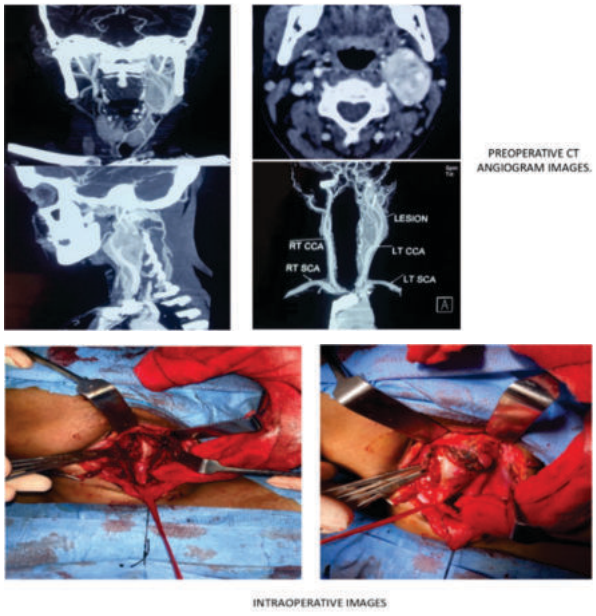
Here we are demonstrating our experience of last 5 year of carotid body tumors at Kidwai memorial institute of oncology, Bengaluru, India. We have operated 5 cases of carotid body tumors, out of which we are describing two cases in details.

Case 1:
27 years old female presented with left side carotid body tumor-shamblin type-3 on preoperative angiogram and tumor removed without any complication.



A- pre operative ct angiogram showing splaying of carotid bifurcation.
B- intraoperative shamblin 3 tumor.
C- intraoperative after tumor removal.

Case 2-
31 years old female presented with left side neck swelling since last 3 to 4 years but was ignored. On evaluation with CT as there was enlargement and cosmetic concerns showing space occupying lesion at bifurcation of common carotid artery. On reference to our institute, CT with angiogram done which was suggestive of 5X5 cm lesion in left carotid space with encasement of distal portion of left common carotid artery, proximal portion of left internal and external carotid artery. The lesion was extending from C2 to C5 vertebra. Prophylactically as advanced grade – balloon occlusion test was done which was suggestive of good cross blood flow with no venous lag or clinical neuro deficits. Patient was taken for surgery and there was no possibility of separating tumor from carotid vessels and vagus nerve. Intraoperative decision was taken to ligate common carotid and branches with scarification of vagus and hypolglossal nerve. Post operatively patient was having vocal cord plasy left side, tongue deviation on left side with protrusion and without any neurological deficits.



PREOPERATIVE CT ANGIOGRAM IMAGES.

INTRAOPERATIVE IMAGES

DISCUSSION

Carotid body tumors present as painless, slow growing lateral neck masses. Rarely, they secrete catecholamines leading excessive sweating, headache, flushing, palpitations, and chest or abdominal pain. On physical examination there is a lateral neck mass that is mobile in an anterior-posterior plane but immobile in a cranio-caudal plane. Cranial nerve palsy of X-XII and Horner's syndrome may be found. Magnetic resonance imaging with contrast of Head and neck is recommended for diagnosis and to rule out multiple or bilateral lesions and it shows hyperintensity on T2-weighted and salt and pepper pattern on T1-weighted contrast-enhanced imaging due to flow voids [7]. Carotid body tumors can splay the internal and external carotid artery and produce “lyre sign”. If a genetic predisposition is suspected or confirmed, [18F]-DOPA positron emission tomography (PET) is recommended to detect potential multiple paraganglioma tumor sites with high levels of both sensitivity and specificity [8, 9]. If a secreting paraganglioma is suspected, urinary or plasma fractionated metanephrines are obtained. Fractionated metanephrines are more sensitive and preferred over catecholamine concentration measurements [10]. Computed tomography or magnetic resonance imaging is used to follow tumor growth in small tumor and patient is not candidate for operability. Interestingly, mean growth rate was 2.0 mm/year.

The useful classification system to decide operability and risk of surgery is the Shamblin grade, which separates carotid body tumors into three groups [14]. Group 1 tumors do not encase the internal or external carotid arteries, group 2 tumors partially encase the carotid arteries, and group 3 tumors completely encase the carotid arteries. For patients undergoing surgical excision, preoperative tumor embolization may be considered. Possible complications of embolization include pain, postembolization fever, transient ischemia attack, and cerebrovascular accident [15]. So, tumors larger than 3 cm and Shamblin class II or III lesions only should be considered for preoperative embolization but that no clear evidence or guideline indicates when embolization is most appropriate.

Surgery is the gold standard of treatment for resectable carotid body tumor due to the risk of adjacent neurovascular compression with progressive tumor growth and risk of malignancy [2]. However, surgery can carry a high risk of perioperative morbidity. So, an angiographic should be obtained to confirm the diagnosis, evaluate the contralateral carotid system, assess the collateral intracerebral vasculature, and embolize the tumor, if indicated. For poor surgical candidates and with extensive tumor involvement around the carotid artery or multiple tumor sites, observation and radiotherapy are viable treatment alternatives. Successful surgical outcome is absence of tumor recurrence after complete resection. In young, healthy patients local control rates are between 94% and 100% [11-13]. Radiotherapy of carotid body tumors is generally reserved for tumors having unacceptable risk of severe perioperative complications; however, similar rate of tumor control as compared with surgical resection can be found [16].

CONCLUSION

Carotid body tumors are vascular tumor with high peri-oprative morbidity requiring expectant surgical management in experienced hands.

There Is No Coflict Of Interest Among Authors.

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