CME ARTICLE

# Update in Carotid chemodectomas

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## ABSTRACT

Carotid chemodectomas or carotid body tumors are rare tumors, generally benign, slow-growing and generally asymptomatic, and represent the most common paraganglioma of the head and neck. Sometimes there is a compression syndrome or carotid sinus syndrome. Malignancy occurs in 6 - 12.5% of cases. The diagnosis of the carotid body tumor is based on clinical features (site, size, and consistency) and non-invasive tests such as duplex scan or computed tomography (CT) scanning. Imaging is necessary for diagnosis when there is a palpable lateral neck mass. Ultrasounds allow to detect a solid mass within the carotid bifurcation; color flow duplex scanning shows typical tumor hypervascularity. Tumor blush and splaying the carotid bifurcation are the typical features of these tumors.

CT and magnetic resonance imaging (MRA) angiographies are the preferred pre-operative diagnostic imaging investigations and can demonstrate other tumors sites, so they are very useful in combination with ultrasound for treatment planning of the mass. Angiography was in the past the gold standard before surgery to see the vascularity of the tumor. Actually, the treatment of choice is surgical excision preceded by embolization of feeding arteries 24 hours before the surgery. Embolization can decrease blood loss, improve tumor excision and preserve the internal carotid artery flow but may cause fibrosis making dissection difficult. When surgical risk is high, radiation therapy can be performed. Follow-up is important is important because sometimes carotid body tumors can produce metastases.

**Keywords:** *Chemodectomas, carotid body tumors, Extra-Adrenal Paragangliomas,diagnosis, treatment*

## INTRODUCTION

Chemodectoma is one of possible non atherosclerotic pathologies of the neck that include vessel thrombo-embolism, dissecting aneurysms, fibromuscular dysplasia, Takayasu's arteritis, pseudoaneurysms and arteriovenous fistulae. The prevalence of nonatherosclerotic pathologies was calculated as 0.65% in a previous study of Labropoulos done in 11480 ultrasound tests held from 1997 to 2003.[1]

Carotid chemodectomas are called paragangliomas or carotid body tumors (Fig 1). Carotid body was first described in 1743. Kohn first used the term *paraganglion* to describe the carotid body.[2] Paragangliomas are highly vascular and slow growing tumors (5 mm annually) deriving from the paraganglia, which are specialized tissue of the extra-adrenal neuroendocrine system.[2] They are formed by cells divided by vascular channels and fibrosis matrix (Fig 2). Usually they are localized in the neck anterior to the sternocleidomastoid muscle at the level of the hyoid bone. Glennes and Grimley[3] proposed a classification of these tumors based on location, innervation and microscopic appearance, and helped distinguish the adrenal paragangliomas from the extra-adrenal paragangliomas (Table 1).

Paragangliomas are uncommon: they have an incidence of 0.6% of head and neck tumors and 0.03% of all tumors, can be bilateral in 10% of cases, familial in 30% with autosomal dominant hereditary transmission. Malignancy is more common in the non-familial than in the hereditary form. Paragangliomas can be seen in adult 30-60 years old, generally women.[4]

Carotid body tumors are composed by two cells types: the type I cells are the chief or paraganglionic cells; the type II cells are the sustentacular cells. Chief cells are arranged in a characteristic pesudoalveolar pattern described as Zellballen (which means cells balls). These cells contain eosinophilic cytoplasmatic granules and predominate in case of paraganglioma.[5] These tumors are functionally active only in 1-3% of cases. Usually sporadic and unilateral, they can be familiar in 10% of cases with autosomic dominant transmission:the latter is bilateral up to 30%. Generally benign, malignant transformation may occur in up to 10% of the cases. Metastases are found in less than 5% and multiple tumors in 5%.[6] Malignancy is usually more common in the sporadic form and it is not related with tumor size. The malignant type of chemodectoma cannot be predicted by the initial clinical presentation or by the histologic appearance. Nuclear pleomorphism, atypia or

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