# Paraganglioma with radiographic dye: an interesting histopatholological finding

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#### Abstract

Extraadrenal paraganglioma makes up a dispersed neuroendocrine system which extends from the base of the skull down to the pelvic floor. Paraganglioma have been found in particularly every site in which normal paraganglia are known to occur. Herein, we present a typical case of carotid body paraganglioma according to both histological and immunohistochemical findings. The presence of a large ribbon-like amphophilic to basophilic amorphous material which, according to the history of angiography, must result from the precipitation of angiographic dye makes this entity unique and the first to be reported.

Keywords: Paraganglioma; Radiographic dye

#### Introduction

Extraadrenal paraganglia make up a dispersed neuroendocrine system having a centripetal and roughly symmetrical distribution which extends from the base of the skull down to the pelvic floor.<sup>1</sup> These constitute paraganglia in the head and neck including aorticopulmonary paraganglioma which have a close alignment with the parasympathetic nervous system and paraganglia of the sympathoadrenal neuroendocrine system.

A wide variety of hormones and regulatory neuropeptides have been identified in normal paraganglia and their corresponding tumors. Some of these substances have an endocrine function through an interaction with receptors at distant sites, whereas others such as those with paracrine function mediate a regulatory or even perhaps an autoregulatory (autocrine function) effects on neighboring cells. Paraganglia in the head and neck region are closely aligned with the parasympathetic nervous system and often have a close spatial relationship with neural or vascular structures.<sup>2</sup> Paraganglioma have been found in par-

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ticularly every site in which normal paraganglia are known to occur.<sup>2,3</sup> Paraganglioma of the head and neck region are categorized into 5 groups according to their locations and comprise Carotid body (Chemodectoma), Jugulotympanic, Vagal, laryngeal and Aorticopulmonary paraganglioma Immunostaining for neuron specific enolase has been most consistently positive, but it may also be nonspecific. Immunoreactivity for chromogranin and synaptophysin has been reported in virtually all cases, although some were also positive for serotonin and other hormones such as ACTH and VIP etc.

#### **Case Report**

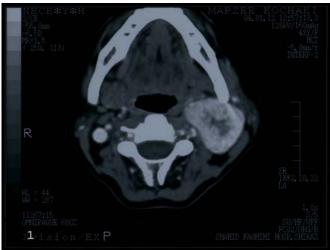
The case presented herein was a 40 years-old female with a slow -growing painless mass in lateral side of the neck since 3 years previously. Computed tomography showed a mass that is attached to carotid artery (Fig. 1). The tumor was completely excised after the patient had undergone arteriography, which confirmed the presence of a vascular tumor.

Grossly the tumor was well demarcated rubbery firm with tan appearance in cut section and measuring about  $3 \times 3 \times 1$  cm. Microscopically the tumor cells arranged as organoid pattern referred to as "zellballen".

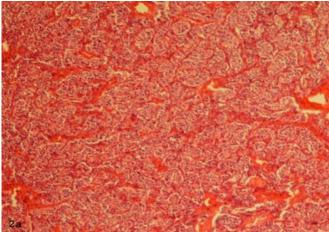
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The tumor cells had eosinophilic and faintly granular cytoplasm with indistinct borders (Fig. 2). Congested blood vessels and hemorrhage were also present within the tumors. The tumor exhibited focal nuclear hyperchromacia and pleomorphism which was peculiar to neuroendocrine tumors and of no prognostic significance.



**Figure 1:** Computed tomography of the lesion, show the location of the tumor near carotid blood vessel.



**Figure 2:** Typical organoid clustering of neoplastic cells (Zelballen) are separated by highly vascularized fibrous septa (H&E, ×40).

In one area of the tumor some ribbon-like amorphous amphophilic materials were present that seemed to be due to deposition of angiographic dyes (Fig. 3A and 3B). The sections were immunostained by streptavidin–biotin method whereby the individual tumor cells were intensely positive for Chromogranin (Fig. 4), synaptophysin and neuron specific enolase using prediluted corresponding antimouse antibodies (DAKO) with S100 positive sustentancular cells in fibrovascular septa.

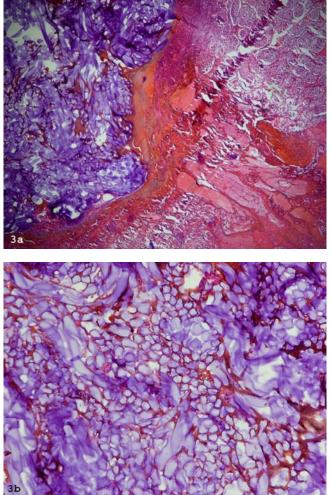
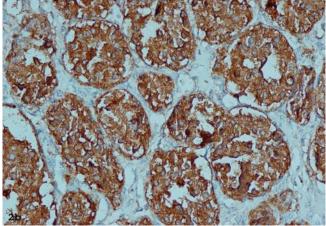


Figure 3A, B: low power and high power view of ribbon like amorphous materials (H & E).



**Figure 4:** Tumor cells are positive for chromogranin. (Immunohistochemistry, ×40).

## Discussion

The first chemodectoma was reported by Marchand in a patient who had undergone operation for a painless mass in 1891.<sup>2</sup> Carotid body and aorthopulmonary paraganglia seems to have a chemoreceptor role, with modulation of respiratory and cardiovascular function in response to changes in arterial PO2, PCO2 and pH.<sup>4</sup>

There is no evidence, however, that any of these tumors has a functional role in chemosensation.<sup>2</sup> Paraganglioma of the head and neck, especially carotid body tumors, are about ten times more frequent in individuals living in high altitude than in those who live at sea level.<sup>3</sup> These have been exaggerated examples of hyperplastic changes due to exposure to prolonged and sever hypoxic stimulation.<sup>3</sup> The occurrence of this tumor as a component of multiple endocrine neoplasm (MEN)<sup>5</sup> or in Von Hipple Lindue disease have already been reported.<sup>2,3,5</sup>

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The carotid body paraganglioma presented herein, is a typical case according to both histology and immunohistochemical staining. This is also unique and the first to be reported, in that a large ribbon-like amphophilic to basophilic amorphous material was present which according to history of angiography, it must be due to the precipitation in tissue of angiographic dye. These consisted of materials which were neither crystals nor light reflecting particles. Substances such as phosphoglyceride<sup>6</sup> or calcium pyrophosphate dehydrate or urate crystals in different shapes were reported in tissue sections.<sup>7</sup>

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