Open Journal of
Clinical & Medical
Case Reports

Volume 8 (2022)
Issue 20

Clinical Image ISSN: 2379-1039

A case of non-functioning paraganglioma – A rare phenomena

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Abstract

Non-functioning paragangliomas are rare tumors. I describe a case of a 74-year-old woman who presented with worsening neck mass and subsequently discovered to a non-functioning paraganglioma after surgical resection. These tumors are associated with paraneoplastic diseases including MEN Syndromes, Neurofibromatosis and Von Hippel Lindau and patients should be referred for genetic counseling.

Keywords

Paraganglioma; Rare; Genetics; Men syndrome.

Introduction

Paragangliomas, also known as non-epithelial neuroendocrine tumors, are rare. They are further characterized as sympathetic (functioning) or parasympathetic (non-functioning). It is estimated that the overall incidence is 0.66 cases per 100,000 people per year.

Case Description

74-year-old female with history of multiple thyroid nodules presented with worsening swelling of her left neck. Ultrasound revealed hyper vascular mass at the left carotid bifurcation. A follow up CT Neck with IV contrast showed 2 x 2.3 x 3.5 cm hyper enhancing mass. On admission, her vital signs were unremarkable, and she did not report of weight loss, diarrhea, constipation, chest pain, palpitation, flushing, episodic headache, chest pain or shortness of breath. She subsequently underwent carotid body tumor resection with final pathology revealing paraganglioma.

Conclusion and Discussion

Non-functioning paragangliomas are rare diseases. Most tumors are located in the head & neck mostly in the skull base in the distribution of the IX and X cranial nerves. Patients are asymptomatic and do not exhibit sequalae of pheochromocytoma. It has been reported that most paragangliomas are sporadic however 30% of all paragangliomas are hereditary including the paraganglioma syndromes 1, 3 and 4 subtypes. Other forms may be associated with multiple endocrine neoplasia 2A and 2B (MEN), neurofibromatosis 1 and Von Hippel Lindau (VHL) disease. A thorough History and Physical is of valuable importance however surgical biopsy and resection is warranted. Patients should be closely monitored for possible malignant transformation and associated MEN syndromes based on clinical suspicion. All patients with paragangliomas should be screened for germline mutations. Our patient was referred to the Genetic Counseling team for further assessment.

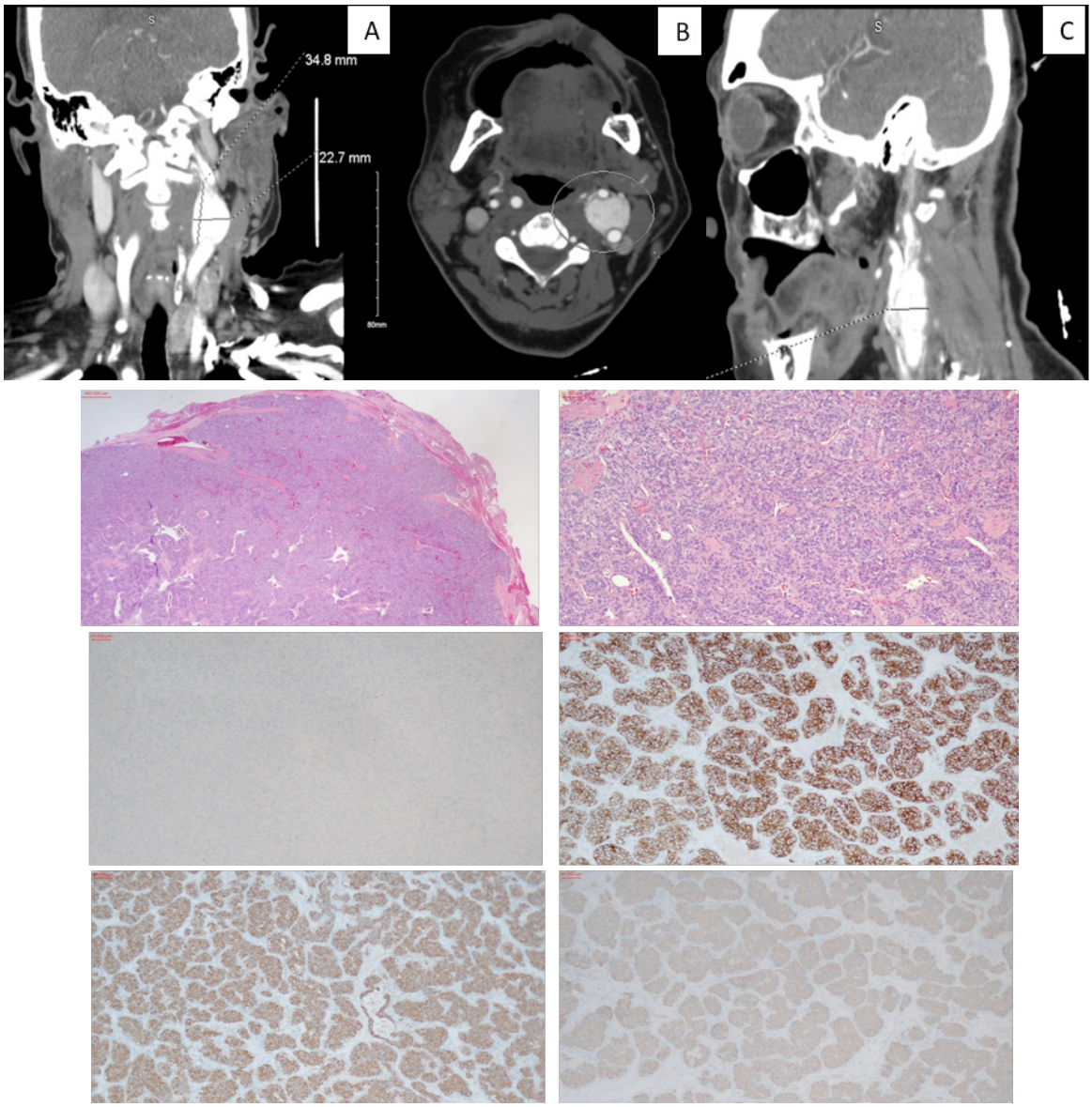

Figure 1: (A,B & C) 2 x 2.3 x 3.5 cm hyperenhancing mass splaying the LEFT carotid bifurcation **(D):** 2X magnification; **(E):** 10X magnification**; (F):** AE1/AE3 staining**; (G):** CD56 staining**; (H):** Chromogranin staining**; (I):** Synaptophysin staining.

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Manuscript Information: Received: October 14, 2022; Accepted: November 07, 2022; Published: November 10, 2022

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Citation: Agyei O, Ali Z, Zhang Y. A case of non - functioning paraganglioma - A rare phenomena. Open J Clin Med Case Rep. 2022; 1933.