American Journal of Medical Case Reports, 2023, Vol. 11, No. 2, 31-32
Available online at http://pubs.sciepub.com/ajmcr/11/2/6
Published by Science and Education Publishing
DOI:10.12691/ajmcr-11-2-6

A Case of Non-functioning Paraganglioma – A Rare Phenomena

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Received January 05, 2023; Revised February 11, 2023; Accepted February 19, 2023

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 have a non-functioning paraganglioma after surgical resection. These tumors are associated with paraneoplastic diseases including MEN Syndromes, Neurofibromatosis and Von Hippel Lindau. Patients should be referred for genetic counseling once diagnosis is established.

Keywords: *Paraganglioma, rare, genetics, MEN syndrome*

Cite This Article: Obed Agyei, Zonera Ali, and Yanmin Zhang, "A Case of Non-functioning Paraganglioma – A Rare Phenomena." *American Journal of Medical Case Reports*, vol. 11, no. 2 (2023): 31-32. doi: 10.12691/ajmcr-11-2-6.

1. Case Report

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: 74-year-old female with history of multiple thyroid nodules presented with worsening swelling of her left neck. Ultrasound revealed hypervascular mass at the left carotid bifurcation. A follow up CT Neck with IV contrast showed 2 x 2.3 x 3.5 cm hyperenhancing 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. Patient was subsequently referred to Genetics for further testing.

2. Conclusion and Discussion

Non-functioning paragangliomas are rare diseases. Most tumors are located in the head & neck mostly in the skull base at the distribution of the IX and X cranial nerves. Patients are typically 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 are 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.

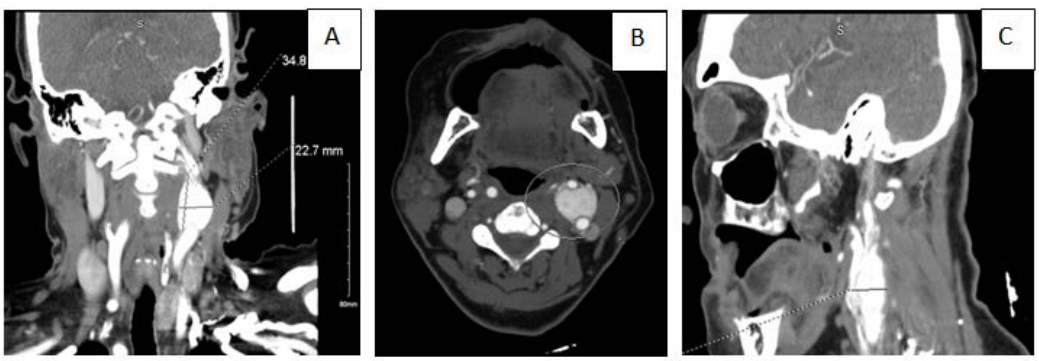

Figure A, B and C. 2 x 2.3 x 3.5 cm hyperenhancing mass splaying the LEFT carotid bifurcation

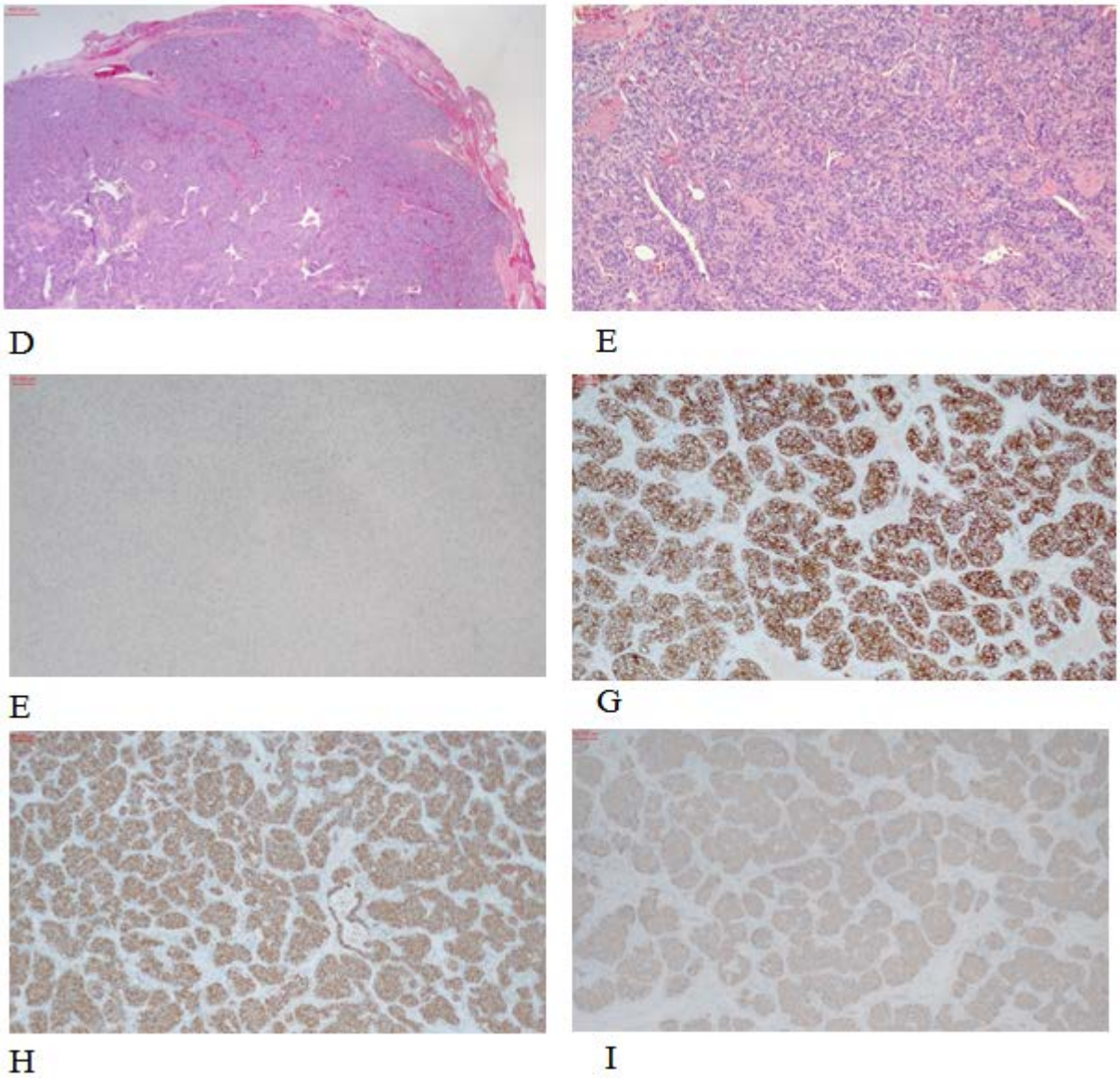

Figure D. 2X magnification; **Figure E.** 10X magnification; **Figure F.** AE1/AE3 staining; **Figure G.** CD56 staining; **Figure H.** Chromogranin staining; **Figure I.** Synaptophysin staining

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