Background Information:
Paraganglia are neural crest derived cells, and as such, they are found along the embryologic migration routes of neural crest tissues from the base of the skull to the lower pelvis, not including the area of the extremities. They are most closely associated with blood vessels, where they act as chemoreceptors, such as in the carotid body. Paragangliomas are neoplasms that arise from these paraganglia, usually as a benign tumor. These extra adrenal paragangliomas are histochemically non-chromaffin, and tend to be located in the head and neck region, superior mediastium, and retroperitoneum. Specifically, the most common locations are the carotid body, jugular bulb, Jacobson's tympanic plexus in the middle ear, and the vagal nerve. Overall, paraganglioma of the head and neck region are incredibly rare and constitute only 0.012% of tumors in that area, and due to their strong association with blood vessels, an overwhelming 80% of these paragangliomas arise from the carotid body. Thyroid associated paraganglioma is especially uncommon as it is not an area that should contain neural derived tissue. There are only 28 recorded cases of primary thyroid paraganglioma in the English-based publication, and their origin is still not fully understood.

Although it is not known for sure how it forms, it is thought that the paraganglioma arises from the inferior laryngeal paraganglia, which are being displaced downwards to the lateral aspects of the thyroid gland, or that the paraganglioma is formed in the thyroid capsule itself.

Case History:
The patient was a 68 year old female with a history of a solitary thyroid nodule in the right lobe. She was diagnosed with hypertension and sarcoidosis previously, but was currently in remission. There were no changes in her voice or dysphagia, and she was biochemically euthyroid. CT scans showed low grade uptake in an enlarged right thyroid lobe with mass. She underwent a total thyroidectomy to remove the thyroid and impacted nodes. The thyroid mass was firm and immobile, and showed significant vascularity that posed tremendous issues for the patient due to the size of the tumor. Patient then underwent adjuvant radiation therapy and was directed to follow up for adjunct PET/CT scans to see if any additional systemic therapy is necessary.

Results:
Final diagnosis was a malignant paraganglioma of the thyroid due to the presence of metastasis to the paratracheal lymph node. There was also extrathyroidal extension, although this could be due to the size of the tumor. Patient then underwent adjuvant radiation therapy and was directed to follow up for adjunct PET/CT scans to see if any additional systemic therapy is necessary.

Conclusion:
Distinction should also be made between benign and malignant paragangliomas of the thyroid, although malignant types are incredibly rare. Pathologists are not able to use IHC and H&E to distinguish between benign and malignant. Therefore the only way to diagnose a paraganglioma of the thyroid as malignant is if it metastasizes to non-neuroendocrine tissue, usually the cervical lymph nodes. Origin for the paraganglioma is also very important, as they can be familial or sporadic. Sporadic makes up the majority of cases at about 90%, but familial is not to be overlooked. In those who have familial paragangliomas, and in 10% of sporadic paragangliomas, there is a 30% risk of mutations in the genes encoding for mitochondrial complex II (SDH-B, SDH-C, SDH-D). Individuals with this are at risk of multiple tumors. Because of these potential mutations, or in the case of malignant paraganglioma, follow up is crucial in treatment.

Because they are so rare, paraganglioma is not commonly listed in differential diagnoses for thyroid gland lesions. Usually they are discovered through signs and symptoms, mass effect indications, as an incidental finding on a imaging scan of the thyroid, or during a familial screen for hereditary paraganglioma. 4 When the growth is discovered it is initially misdiagnosed as a more common thyroid nodule, which occur in 5% of women and 1% of men, making it far more likely than paraganglioma. 5 Despite this fact, modern imaging and staining indications have enabled set of guidelines to help distinguish between paraganglioma and its more frequent thyroid malignancy lookalikes, most commonly lymphanizing trabecular adenoma of the thyroid and medullary carcinoma of the thyroid.