Thyroid mass from a malignant glomus tumor: Case report and literature review

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Background and Objective
Glomus tumors, also known as glomangiomias, are rare tumors that arise from the dermal glomus body. Glomus bodies are specialized arteriovenous anastomoses surrounded by nervous tissue that function in temperature regulation via shunting of blood. Glomus tumors typically occur in areas rich in glomus bodies including the subungual region of the digits, the dermis or the subcutis of the palm, wrist, forearm or foot.1 They present as single or multiple painful and thermosensitive nodules in these areas.2 Malignant glomus tumors are exceptionally rare. Here we describe the first reported case of a malignant glomus tumor that spread to the thyroid gland.

Methods
A 40-year-old male with a history of glomus tumor of the dorsal aspect of his left foot status-post wide local excision with metastatic disease that was well controlled for four years with chemotherapy and radiation presents with a slowly enlarging left thyroid mass. The mass was first identified on surveillance CT Chest and enlarged from 1.3 centimeters to 2.4 centimeters over the course of six months (Figure 1).

Fine-needle aspiration of the mass demonstrated atypical epithelioid cells with conspicuous nuclei and scatter mitosis with immunostains positive for SMA, focally positive for calponin and negative for PAX-8 consistent with metastatic malignant glomus tumor.

Figure 1. Pre-operative axial CT chest with contrast showing a 2.4 cm hypodense nodule in the left lobe of the thyroid gland (arrow).

Results
The patient was treated with preoperative radiation to his left calcaneous and left shoulder with 20 grey to palliate pain at these sites. He then underwent total thyroidectomy to resect this mass and palliate any potential airway compressive symptoms (Figure 2). Pathology showed malignant glomus tumor diffusely involving the thyroid parenchyma and invading the perithyroidal soft tissue with a 4.9 focus in the left thyroid lobe and second 1.9 cm area in the right thyroid lobe (Figure 3).

His post-operative course was unremarkable and he was started on temozolomide therapy for palliation of his pain.

Discussion
Glomus tumors, which classically present as subungual nodules in the nail bed or dermal or subcuticular nodules elsewhere in the upper or lower extremity, are uncommon and constitute 1.6% of all soft tissue tumors.2 Glomus tumors are typically benign and localized but rare cases are malignant. Due to the rarity, the diagnosis of malignant glomus tumor can easily be delayed or missed and clinicians should be alert to this possibility.

While glomus tumors typically occur in cutaneous locations in the extremities, extracutaneous presentations tend to occur but are less frequent as glomus bodies are either sporadic or absent throughout much of the rest of the body. Malignant glomus tumors are very rare with a reported rate of malignant transformation of only 1%.3,5 They are typically locally aggressive and rarely metastasize despite histologic features of malignancy.1 The disease is typically fatal when metastases are present, however this is based on only on scattered case reports.3,4,6 Metastatic spread of malignant glomus tumors to the brain, bone, lung, liver, mediastinum, small intestine and lymph nodes have been reported,6 however we did not identify any previous cases of malignant glomus tumors with spread to the thyroid.

We identified one previous case report of a glomus tumor that presented in the thyroid gland, which after initial presentation metastasized to the brain and lung ultimately leading to the patient’s death.7 The authors believed that this was an extracutaneous primary site originating in the thyroid gland. Malignant glomus tumor of the larynx that required total laryngectomy has also been reported.

The primary treatment is cases with or without metastatic disease is surgical resection. In our case, in order to palliate potential morbidity from regional disease in the neck and airway obstruction the patient elected to remove his thyroid mass.

Conclusions
• We present the first reported case of a malignant glomus tumor metastasizing to the thyroid gland. Clinicians treating thyroid disorders should be aware that glomus tumor can metastasize to the thyroid gland.

• Glomus tumors should not be confused with paragangliomas, which were formerly also called glomus tumors. Paragangliomas derive from glomus cells of the sympathetic nervous system that function as chemoreceptors along blood vessels which accounts for the confusing terminology however paragangliomas were reclassified and are no longer called glomus tumors.

References