Amyloidoma of the Carotid Body: A Rare Presentation of Head and Neck Amyloidosis

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ABSTRACT

Amyloid deposition in the head and neck most commonly occurs in acquired immunoglobulin light chain (AL) amyloidosis and occurs in approximately 20% of cases. It is typically isolated to the larynx, usually the glottis, but has been seen in the thyroid [1-3]. Of note, Amyloidoma of the larynx accounts for less than 1% of all benign laryngeal tumors [1]. Systemic (AA) Amyloidosis can present with macroglossia and periorbital purpura in approximately one-third of the affected population. Other locations included scattered submucosal deposits within the nasal cavity, soft palate and supraglottic mucosa. Presentation of Amyloidosis as a lateral neck mass is exceedingly rare. To the best of our knowledge, there are no previous reports in the literature describing an Amyloidoma on the carotid artery.

CASE PRESENTATION

59F smoker with past history of COPD and hypothyroidism presents with a 6 month history of a right neck mass. The mass was nontender and had a 20% increase in size over this time frame. She complained of referred otalgia and occasional temporal headaches, but was otherwise asymptomatic, with no dysphagia, odynophagia, hoarseness, dyspnea or weight loss. On exam she was noted to have a firm 2.5-3 cm nontender mass at the right carotid bifurcation. The remainder of her exam was negative. CTA of the neck demonstrated a 2.9 x 1.5 cm mass in the right carotid bifurcation with central tortuous vessels, and coarse calcifications. Additional study includes an MRA that demonstrated a T1 isointense soft tissue signal 1.3x1 cm immediately above the carotid bifurcation on the right side. The differential diagnosis at that time was peripheral nerve sheath tumor versus carotid body paraganglioma. The patient's imaging findings were atypical for carotid body tumor; however, this did not affect the surgical approach and she was consented for surgery.

Intraoperatively the vagus nerve and internal jugular vein were preserved. Dissection on the medial aspect of the proximal internal carotid artery demonstrated a gritty, hard consistency that was difficult to establish a surgical plane on. Superiorly, it was possible to circumferentially dissect on the internal carotid artery approximately 1 cm away from the bifurcation. On the external carotid artery, a similar approach was noted as the external carotid artery was dissected towards the bifurcation. Unfortunately bleeding occurred from transluminal compromise of the vessel by the overlying disease process. Suture placement with 6-0 Vicryl was not capable of obtaining adequate hemostasis due to transmural spread of the disease, which was extending from the carotid bifurcation to the proximal internal and external carotid artery. At this point in the case, Vascular Surgery was asked to assist. Given the extent of tumor involvement, it was necessary to resect the common and internal and external carotid artery and reconstruct with a right great saphenous vein interposition graft.

Several hours after recovery in the ICU, she was taken back to the OR for an expanding neck hematoma. There were several venous sources noted, however the arterial reconstruction was hemostatic. The patient ultimately recovered and was discharged home in stable condition. Congo red staining of the pathology specimen was consistent with Amyloidosis. The patient was then referred to Hematology/Oncology for management of AL Amyloidosis.

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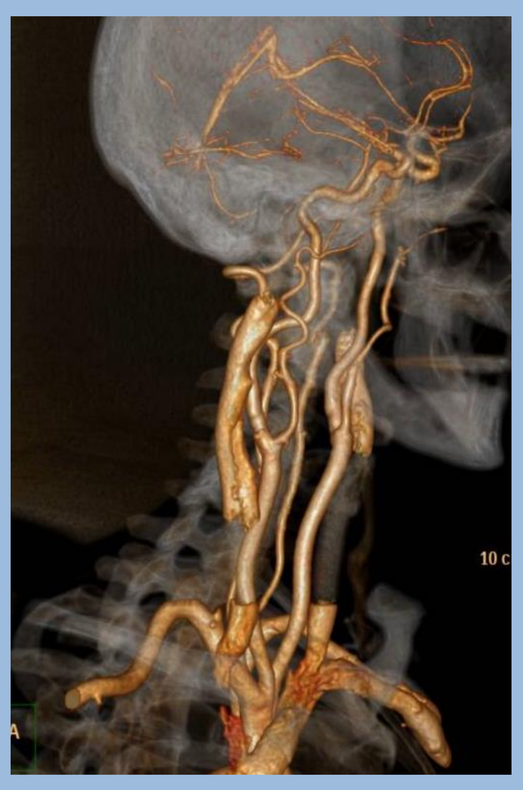
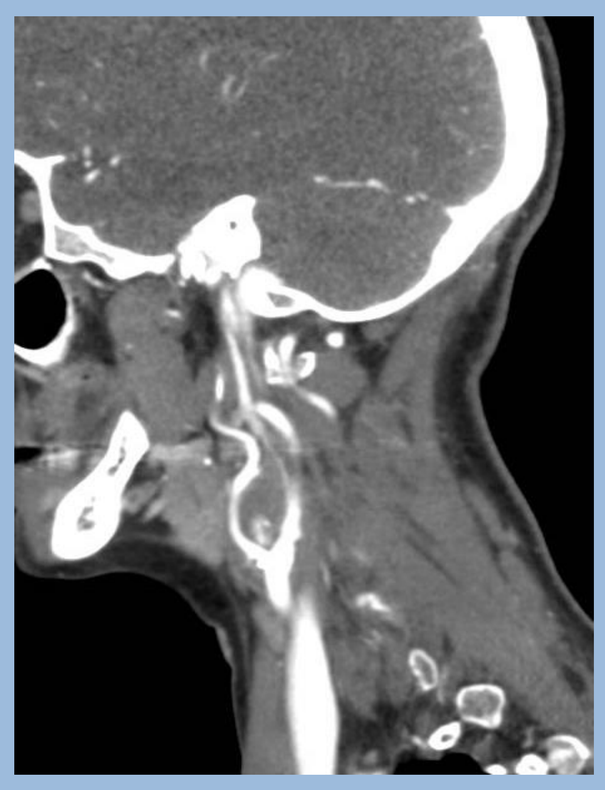
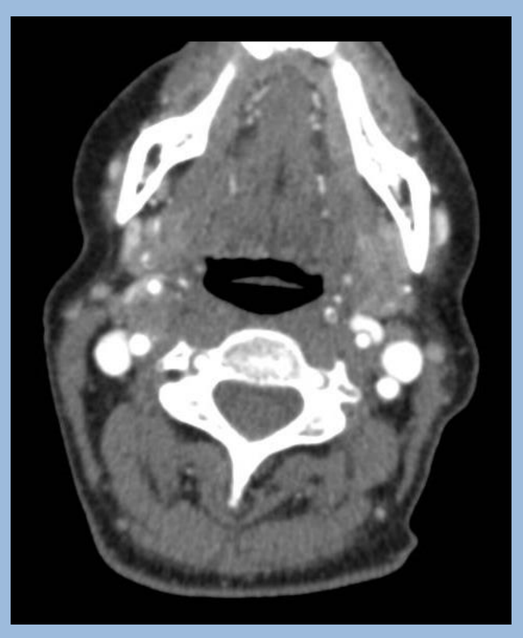
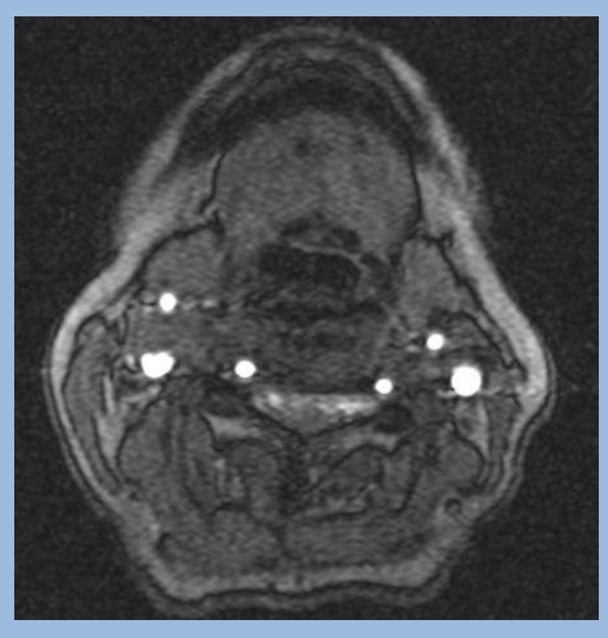
IMAGING









DISCUSSION

Masses in the lateral neck at the carotid artery bifurcation are most commonly carotid body tumors. These tumors are hypervascular benign neoplasms of the tissue that houses chemoreceptors and monitors changes in arterial oxygen, carbon dioxide and pH. These tumors are slow-growing masses with potential to cause local neck discomfort, headache, dizziness, dysphagia, and voice changes. Three distinct forms are described: sporadic, familial, and hyperplastic, which occurs in individuals with COPD or those who live at high altitude. CT scans and/or MRI with angiography are the initial diagnostic tools to assess size and location; carotid angiography is the gold standard. Carotid body tumors are treated with surgical excision, but the surgery carries considerable mortality risk and risk of hemorrhage, stroke, and damage to vagus and other cranial nerves [4-6]. Amyloidoma is rarely on the differential for carotid body tumors, as the typical presentation for head and neck amyloidosis is within the larynx or scattered in the submucosa of the upper aerodigestive tract. Though there was splaying of the internal and external carotids, the calcifications were not typical of a carotid body tumor.

CONCLUSION

This case illustrates an interesting initial presentation of Amyloidosis. Not only was the patient's first lesion in the head and neck, but it was located at the carotid body, a never before reported location. Carotid body tumors or paragangliomas, are the most common tumor of the bifurcation, but this case highlights an atypical lesion. Careful intra-operative resection and close collaboration with Vascular Surgery colleagues were paramount to this patient's care.

REFERENCES

1. Gouvea, A.F., Prado Riberio, A.C., Leon, J.E., Carlos, R., Pais de Almeida, O., Lopes, M.A., "Head and neck amyloidosis: clinicopathological features and immunohistochemical analysis of 14 cases," J Oral Pathol Med (2012), 41:178-185.
2. Penner, C.R., Müller, S., "Head and neck amyloidosis: A clinicopathologic study of 15 cases," Oral Oncology (2006), 42:421-429.
3. Kerner, M.M., Wang, M.W., Angier, G., Calcaterra, T.C., Ward, P.H., "Amyloidosis of the head and neck: A clinicopathologic study of the UCLA experience, 1955-1991," Arch Otolaryngol Head Neck Surg (1995), 121:778-782.
4. Metheetrairut, C., Chotikavanich, C., Kekool, P., Suphaphongs, N., "Carotid body tumor: a 25-year experience," Eur Arch Otorhinolaryngol (2016), 273: 2171-2179.
5. Warshawski, S.J., de Souza, F.M., "The carotid body tumor," J Otolaryngol (1989), 18:306-310.
6. Makeieff, M., Raingeard, I., Alric, P., Bonafe, A., Guerrier, B., Marty-Ane, C., "Surgical management of carotid body tumors," Ann Srg Oncol (2008), 15:2180-2186.