CLINICAL VIGNETTE

Carotid Epithelioid Hemangioendothelioma – A Management Dilemma

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Case Report

A 58-year-old male presented to a local ENT with several months of progressive dysphagia to solid food, odynophagia, and transient painless vision impairment. Physical exam was noticeable for subtle right neck fullness without obvious lymphadenopathy. Neurological exam was normal. Flexible indirect laryngoscopy showed paralysis of right true vocal fold. CT angiography (CTA) of the neck followed by duplex ultrasound demonstrated a mass within the right carotid bifurcation and proximal internal carotid artery concerning for a carotid bifurcation tumor. A cervical MRA demonstrated a mass described as possible thrombosed pseudo-aneurysm at right carotid artery bifurcation. He was placed on dual-antiplatelet therapy, but continued to deteriorate with worsening dysphagia and tongue weakness. He had multiple admissions due to aspiration pneumonia and eventually underwent feeding tube placement. Repeat CTA two months later showed growth of the soft tissue mass at the bifurcation raising concern for a malignant etiology. He underwent radical surgical resection of the mass, and pathology revealed epithelioid hemangioendothelioma (EHE) of the carotid artery with extravascular invasion into the jugular vein, hypoglossal and vagal nerve. Given the extent of his disease, he completed adjuvant concurrent chemo-radiation with weekly carboplatin and paclitaxel as radiation sensitizers. His symptoms gradually improved and he has been disease free for over one year.

Discussion

Epithelioid hemangioendothelioma (EHE) is a rare malignant vascular neoplasm that derives from endothelial cells. It can be variably aggressive with biological behavior between benign hemangioma and malignant hemangiosarcoma. Peak incidences are between the ages 30 to 50 years. The incidence in the US is about 20 cases per year with primary vascular EHE accounting for about 50% of cases. EHE can also occur in soft tissues, bones, liver and lungs. Over 30 vascular EHE cases have been previously reported predominantly from veins rather than arteries.[1] The diagnosis of intravascular EHE remains extremely challenging due to its rarity, and the difficulty in distinguishing it from thrombosis, as their radiographic characteristics are rather nonspecific. There are no specific imaging features or preferred imaging modality to differentiate primary intravascular EHE from atherosclerotic, inflammatory and thrombotic lesions.

Our patient had EHE involving the carotid artery and the tumor mimicked a thrombosed carotid artery aneurysm on serial imaging studies. Despite his presenting symptoms concerning for cranial nerve involvement, the diagnosis was delayed due to imaging challenges. We have found only two published cases of EHE arising from the carotid artery.[2,3] Clinicians should be aware of carotid artery EHE to include in the differentials of carotid body masses. This is especially important when cranial nerve palsies are present. Further efforts are needed for a better radiologic characterization of such intravascular tumors as timely diagnosis and complete surgical intervention remains the primary treatment. The effectiveness of adjuvant chemotherapy and/or radiation therapy remains ambiguous. Adjuvant radiation after surgical resection can be explored for localized EHE in order to control the residual disease given the risk of recurrence. Systemic chemotherapy is primarily for widespread disease.

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