Granular Cell Tumor of the Cervical Esophagus: Case Report and Literature Review

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Granular cell tumors (GCT) are a rare, benign neoplasm of neural derivation, thought to arise from Schwann cells. Most commonly found in subcutaneous tissue, other sites of origin include the oral cavity, breast, larynx, bronchus, and gastrointestinal (GI) tract, with approximately 45-65% of tumors occurring in the head and neck. GCT’s of the cervical portion of the esophagus are an especially isolated occurrence. In reviews of GI tract GCT’s, the incidence of cervical esophageal lesions has been found to be 1.3% of all GI GCT’s and only 4.2-5.0% of all esophageal GCT’s. Due to the rarity of this condition, treatment protocols have not been elucidated, but in general, observation has been recommended for lesions less than 1 centimeter in size. In an effort to better characterize these uncommon lesions, we present a case of a cervical esophageal GCT and review the literature to determine trends in therapy and provide management recommendations.

INTRODUCTION

Granular cell tumors are uncommon lesions. They are found at any age and may present with symptoms such as dysphagia, hematemesis, or the sensation of a lump in the throat. Histologically, they are characterized by a storiform pattern of eosinophilic cells with clear cytoplasm and oval nuclei. Treatment generally consists of complete excision, but in cases of large or loculated tumors, resection may be necessary. A review of the literature reveals less than 20 reported cases of cervical esophageal GCT’s.

METHODS AND MATERIALS

Case report and review of the literature. A MEDLINE search was performed through the United States National Library of Medicine’s “PubMed” online database. Using the search terms “esophagus” and “granular cell tumor” with results limited to the English language, over 83 papers were obtained. Case reports, series and reviews were identified and their citations examined for further resources. Eighteen resources were identified, comprising a total of nineteen patients (Table 1). These publications were reviewed to extract pertinent information.

RESULTS

Case Report:
A 55 year old female with a greater than 10 year history of progressive dysphagia without weight loss, neck masses, otalgia, or voice changes. CT scan revealed a cervical esophageal tumor. Endoscopy confirmed the presence of a 2.7 cm mass in the cervical esophagus. The patient was referred to our institution for further evaluation.

LITERATURE REVIEW

While esophageal GCT’s are a rare entity, accounting for only 1.7% of all GCT’s and 1.2% of all esophageal masses, GCT’s of the cervical portion of the esophagus are even less common. Characterized histologically by large cells with abundant granular cytoplasm, indistinct nuclei, and bland nuclei that stain positive with Periodic Acid-Schiff and S100, these lesions are benign in nature, but may be incorrectly diagnosed as squamous tumors due to a high percentage demonstrating pseudoepitheliomatous hyperplasia on superficial biopsy and an infiltrative growth pattern.

Treatment of GCT’s has generally consisted of complete excision, especially in those greater than 1 cm in size, in order to prevent growth and eliminate the risk of malignancy. Transformation estimated to occur in 1-2% of cases. Though up to 25% of lesions are asymptomatic, morbidity related to excision of cervical esophageal GCT’s often requires sacrifice of contiguous esophageal musculature due to an infiltrative growth pattern. Resection of larger lesions is especially problematic due to close proximity to vital structures such as the recurrent laryngeal nerve, larynx, and pharynx. In fact, 40% of symptomatic lesions reviewed required segmental esophageal resections, with 20% also requiring local pedicled or free flap reconstructions. While 20% of asymptomatic patients also required segmental esophageal resection, the smaller size of these tumors affords less risk to adjacent structures. As 35% of cases reviewed had concurrent GCT’s, all of which were in the upper aerodigestive tract and stomach, definitive management evaluation of cervical esophageal GCT’s should necessarily include upper endoscopy.

CONCLUSIONS

Cervical esophageal granular cell tumors, although benign, are difficult to manage lesions due to infiltrative growth and often large on presentation. Tumors, if detected early, should be treated aggressively to prevent the risk of further growth posing risks to nearby structures and malignant potential. Upper endoscopy is recommended to rule out second primary lesions due to a high rate of concurrent lesions.

REFERENCES