Chondrosarcoma of the larynx and trachea: A report of two rare cases

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INTRODUCTION

Tumors with cartilaginous differentiation are rarely encountered in the upper airway. Laryngeal chondrosarcomas comprise 0.1% to 2.0% of all neoplasms of the larynx and 3% of all chondrosarcomas. Approximately 250 cases of laryngeal chondrosarcoma have been reported, with most cases revealing low-grade histological and clinical features. Management of the tumors is surgical, with most sources advocating conservative resection. Total laryngectomy is generally reserved for high-grade, recurrent, or extensive disease. Experience with radiation of cartilaginous tumors is limited and chemotherapy is not effective. Clear cell chondrosarcoma of the larynx is an extremely rare subtype, of which only four cases are known. Primary tracheal chondrosarcomas are also rare, with only 17 published reports. We present two rare cases of chondrosarcoma of the upper airway, one in arising in the trachea and another of the clear cell variant originating in the thyroid cartilage.

CASE 1

An 80-year-old male presented to his primary care physician with a complaint of cough and increased sputum production over the past several weeks. CT showed an exophytic tracheal soft tissue mass with internal calcifications measuring 3 x 2 x 2 cm. The mass involved the anterior and left lateral walls of the first through the fourth tracheal rings with extraluminal extension. Fiberoptic bronchoscopy revealed a large exophytic mass 2 cm distal to the glottis with enhanced vascularity and 70% airway obstruction. Biopsy was consistent with chondrosarcoma. The patient was referred to our institution and underwent composite tracheal resection of the first through fourth rings and left thyroid lobectomy with primary tracheal closure. Gross examination of the specimen demonstrated a 2 x 2 x 1.5 cm tan-gray mass on the anterior half of the upper tracheal rings protruding into the tracheal cavity. The mass also passed externally through the tracheal rings to abut against the adjacent thyroid tissue. Histopathological analysis revealed characteristic features for a chondrosarcoma, including lobular growth comprised of chondrocytes with increased cellularity, binucleated cells and nuclear hyperchromasia (Fig. 1). Most of the lesion showed features of a low-grade (Grade I) chondrosarcoma, however some areas showed increased cellularity comprised of more pleomorphic-appearing nuclei representing an intermediate-grade (Grade II) chondrosarcoma (Fig. 1). There was no evidence of angioinvasion or neurotropism. A delphian lymph node was free of metastatic tumor. Recovery was unremarkable. CT scans of the neck 3 and 7 months postoperatively showed no evidence of recurrence. Thirty-six months after surgery the patient remains asymptomatic.

DISCUSSION

Chondrosarcoma of the Trachea
Cartilaginous tumors of the trachea and upper respiratory tract are rare. Only 18 cases of tracheal chondrosarcoma have been reported. Tracheal chondrosarcoma is histologically similar to conventional chondrosarcoma arising in bone. Large, round or polygonal cells with hyperchromatic nuclei and prominent nucleoli are described within a pink or pale blue background containing elements of chondroid tissue. The gross appearance of the tumor is typically submucosal, smooth, pedunculated, and tan or white. Treatment is tracheal resection with end-to-end anastomosis. Radiation may play a role in recurrent or aggressive cases. Chemotherapy is ineffective.

Clear Cell Chondrosarcoma of the Larynx (CCCL)
Only 4 other cases of laryngeal clear cell chondrosarcoma have been reported. Histologically, CCCL appears as rounded cells with enlarged, centrally located, moderately polymorphic nuclei and abundant clear cytoplasm. The average age at diagnosis is 48, unlike other forms of laryngeal chondrosarcoma, which usually present in the 70’s. Locoregional recurrences have been reported in 2 of 5 cases, both of which occurred three times. CCCL may represent a more aggressive variant of laryngeal chondrosarcoma. It occurs in younger patients and is highly likely to recur after conservative resection. Selective neck dissection is indicated for known nodal disease. Organ-sparing surgeries may be attempted as long as adequate margins are attainable. However, in light of the clinical behavior of this lesion, adjuvant radiation warrants further consideration.

REFERENCES