



Myxoid chondrosarcoma of the cricoid cartilage

Sir,

Primary cartilaginous bone tumors of the laryngeal region are extremely rare. Laryngeal chondrosarcomas account for 0.07–2% of all laryngeal neoplasms and less than 1% of all sarcomas.^[1] A 95% of all laryngeal chondrosarcomas are low-grade and are often underdiagnosed as benign chondromata. Biopsy is the cornerstone of the diagnosis. For definitive treatment there is a general consensus that conservative surgery is the most appropriate method.^[2] These tumors are known for their indolent course, low metastatic potential and high percentage of recurrence. The clinical outcome was the worse for patients with myxoid type of tumors, regardless of tumor grade or applied form of therapy.^[3]

A 65-year-old man with no family history of malignant or hereditary diseases and a 50-year history of smoking 20 cigarettes a day presented with a 7-month history of breathy dysphonia.

The initial evaluation in a local clinic, 2 months after the onset of his symptoms, yielded no distinctive diagnosis. After further 5 months of uninterrupted symptoms the patient came for an examination in our university hospital. Laryngoscopy revealed a 3cm large swelling on the front wall of the larynx. Computed tomography (CT) of the neck demonstrated a mass lesion of variable density in the area of the left cricoid cartilage with extralaryngeal growth extended beyond the outer circumference of the cartilage and into the adjacent posterior soft tissue of the neck [Figure 1]. Pathological lymph nodes were absent. A probatory biopsy of the mass was done. Pathologic examination revealed a tumor to be a differentiated cricoid chondrosarcoma with a myxoid component [Figure 2]. The patient refused irradiation of the tumor and strongly demanded total removal of the larynx. However the entire lesion could not be removed due to the proximity of the carotid and the vagus nerve. Total laryngectomy was done. Definite histology showed cartilaginous tumor with minimal nuclear atypia, however together with myxoid changes of the interstitium and focal invasion of the preexisting cartilage. Histological and clinical findings match the diagnosis of myxoid chondrosarcoma. Due to positive margins and residual tumor adjuvant radiation therapy was applied. The postoperative course was uneventful. Two years after the surgery the patient is free of recurrence and metastasis.

Laryngeal chondrosarcomas most commonly affects posterior cricoid cartilage lamina.^[1,3] The etiology of laryngeal chondrosarcoma is still unresolved. The disordered ossification, ischemic change in a chondroma, radiation therapy, smoking have all been linked to development of the disease, but specific etiologic link has not been definitively established.^[4] Typically it occurs more frequently in men with a peak incidence in the sixth decade.^[3,5] Hoarseness is the most common presenting symptom, followed by dyspnea, dysphagia, dysphonia, voice changes, cough, neck mass, airway obstruction (stridor), and pain. The symptoms are frequently present for a long duration supporting the notion of an indolent tumor. The pathology of laryngeal chondrosarcomas is well described showing that low-grade tumors are the most common variety observed.^[1,3,4] Moderate and high-grade tumors are less common. It is believed that myxoid change in these tumors is of prognostic importance, automatically places tumor into grade 2 lesion for which is more likely to have an adverse clinical outcome and metastatic spread.^[3] While conservative surgery is the primary treatment of choice for low-grade tumors, the specific modality and technique performed have been the subject of many discussions. The role of radiotherapy still is a matter of debate, generally it

seems to be ineffective in the management of laryngeal chondrosarcoma.^[3] On the contrary there have been few isolated reports of effective radiation therapy and encouraging reports of fractionated proton radiation therapy for low-grade chondrosarcoma.^[5] There have been no reports of effective adjuvant chemotherapy although high-grade chondrosarcomas warrant consideration of adjuvant chemotherapy. The rarity of myxoid laryngeal chondrosarcomas, specific presentation with

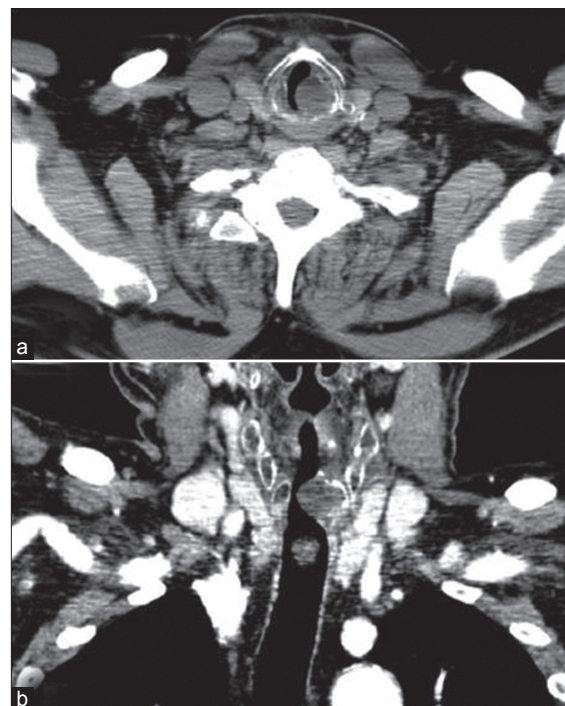


Figure 1: CT images of our patient diagnosed with cricoid chondrosarcoma. (a) Axial section showing a heterogeneous mass with a diameter of 3cm destroying the left half of the cricoid ring. (b) Anterior radiograph of the neck showing presence of a mass entering the airway

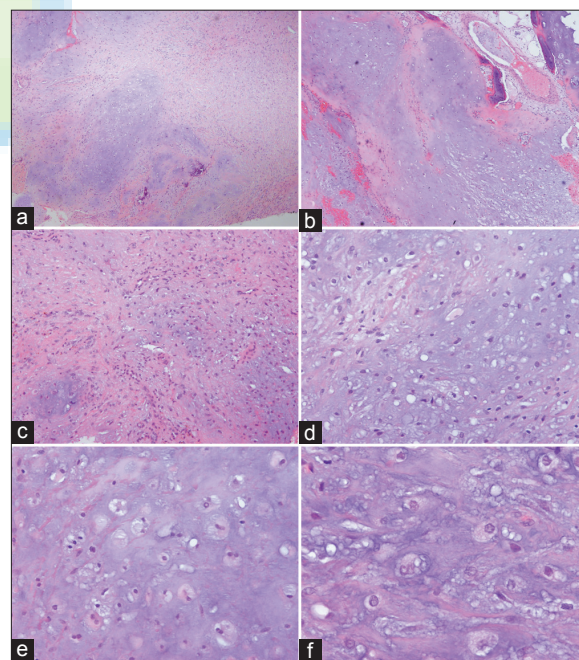


Figure 2: Microscopic features of the resected tumor, Hematoxylin and eosin staining (a-f). Photomicrographs showing tumor tissue composed predominantly of hypo or slightly cellular lobules with myxochondroid appearance (a and b, $\times 40$) which are separated by fibroblast-like spindle cells (c, $\times 100$). The tumor cytomorphology: atypical neoplastic chondrocytes with small hyperchromatic nuclei surrounded by abundant cytoplasm (d, $\times 100$ and e, $\times 200$). Enlarged, binucleated atypical cells with an increased nuclear-to-cytoplasmic ratio and nuclear chromatin distribution irregularities (f, $\times 400$)

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