Case Series

A Report of 4 Laryngeal Sarcoma Patients

Ömer Bayır¹, Tuğba Karagöz*, Ünsal Han², Güleser Saylam¹, Emel Çadallı Tatar¹, Ali Özdek¹, and Mehmet Hakan Korkmaz¹-³

¹Department of Otorhinolaryngology and Head and Neck Surgery, Ministry of Health, Dışkapı Yıldırım Beyazıt Research and Training Hospital, Turkey
²Department of Pathology, Ministry of Health, Dışkapı Yıldırım Beyazıt Research and Training Hospital, Turkey
³Department of Otorhinolaryngology and Head and Neck Surgery, Yıldırım Beyazıt University, Turkey

Abstract

Sarcomas account for a very small percentage of the head and neck malignancies in adults. Laryngeal sarcomas represent 1% of all laryngeal malignancies. Chondrosarcoma is the most common laryngeal sarcoma, and originates mostly from the cricoid cartilage. Less than 3% of all cervicocephalic rhabdomyosarcomas is located in the larynx and laryngeal rhabdomyosarcoma is characterized by less aggressive spread than other rhabdomyosarcomas. Pleomorphic variant of rhabdomyosarcoma is usually encountered in adults. Laryngeal sarcomas have different prognosis and treatment approaches than squamous cell cancers. In this presentation, we discussed the clinical and histopathological features of four laryngeal sarcoma cases; which 2 were diagnosed as chondrosarcoma of arytenoid and cricoid cartilage, and 1 was as spindle cell rhabdomyosarcoma originating from aryepiglottic fold, and 1 pleomorphic variant of rhabdomyosarcoma.

INTRODUCTION

Sarcomas derived from mesodermal tissue components represent less than 5% of the head and neck malignancies. Sarcomas are even rare in larynx and usually present as a submucosal mass without any specific symptoms. Chondrosarcomas are reported to be 2-5% of head and neck neoplasms especially those arising from maxilla. Although the incidence for larynx cancers is 0.07-0.2%, it is still the most common non-epithelial malignant laryngeal tumor [1]. Posterior lamina of cricoid cartilage is observed to be the most commonly involved area which can be attributed to the insertion of muscles and ossification of the cartilages and subsequent mechanical influence of the contracting muscles [2]. It frequently occurs in the sixth or seventh decades of life in men. Dysphonia is an initial complaint while dysphagia and dyspnea occurs in the advanced stages. Surgery is the primary treatment and can vary from endoscopic excision to total laryngectomy depending on the size and histologic grade of lesion. Recurrences and metastases are rare after surgical excision [1]. The next sarcoma type is rhabdomyosarcoma and less than 3% of all rhabdomyosarcomas in the cervicocephalic region is localized in the larynx. Laryngeal rhabdomyosarcoma has less metastatic potential than other rhabdomyosarcomas but needs multimodal treatment approach [3]. In this paper, we present rarely encountered chondrosarcomas, rhabdomyosarcoma originating from larynx, and we discuss their management of treatment in the light of literature.

CASE PRESENTATION

Between the years 2006 and 2015, (321+4) 325 laryngeal cancers were treated in our clinic.

Case 1

A 58 year old man, smoking 30 p/y, presented with dysphonia for one year duration. Preoperatively, a laryngeal mass with a smooth surface extending from the left arytenoid over left vocal cord was seen on endoscopic examination. The left vocal cord was fixed and there was no evidence of cervical lymph node metastases by clinical examination. Preoperative magnetic resonance imaging (MRI) scans revealed 33x29 mm high density mass in the arytenoid cartilage (Figure 1). A 4 cm horizontal neck incision was made caudad to the cricoid cartilage. Then the thyroid cartilage was explored by division of sternocleidomastoid muscles in the midline. The mass was identified at the cricoarytenoid area. Curettage excision of the 2x2 cm involving the posterior cricoid lamina mass was applied (Figure 2). The curettage of the mass was thoroughly completed and the laryngeal mucosa was preserved. Histopathological examination revealed 'low grade chondrosarcoma' of cricoid cartilage. There was no recurrence and metastases after 2 years of follow up.
Case 2

Sixty-five year old man was referred to our clinic with a complaint of progressive dyspnea. In his medical history, he had 3 previous excisions (1 endolaryngeal, 2 external) of chondrosarcoma of cricoid cartilage elsewhere in a period of 7 years. In our institution he had 3 more external excisions in a period of 5 years with the same histopathologic diagnosis. During these years he had sometimes tracheostomy and sometimes decannulated but he denied total laryngectomy. Three years ago along with his dyspnea he started to have dysphagia. His physical examination proved postcricoid area was covered by mass (Figure 3). The CT scan revealed a mass extending to upper trachea and pushing the cervical esophagus. Finally the patient accepted total laryngectomy (Figure 4). His operation was challenging as the mass was extending to the upper mediastinum. The operation and the postoperative period was uneventful. Histopathological examination confirmed low grade chondrosarcoma (Figure 5). Tracheoesophageal puncture was made after 1 year from the surgery for the voice restoration. No recurrence and complications were observed after 3 years of follow up.

Case 3

47 year old woman was admitted to our clinic with the complaint of dysphagia and dyspnea. Laryngeal examination revealed fullness at the right aryepiglottic fold and ventricular fold. The mass was removed by carbon dioxide fiber laser (CO₂ laser), and was reported as neurofibroma without any other physical findings of the disease. After 6 months a recurrence was determined at the same localization, it was again excised using a transoral approach with diode laser. This time histopathological examination was reported as ‘spindle cell variant of rhabdomyosarcoma’ (Figure 6). The patient received chemoradiation; of 4100 cGy and chemotherapy as an treatment. Because of swallowing and aspiration problems in the postoperative period, swallowing exercise therapy applied done by Swallowing and Speech Therapy team in our clinic. The swallowing problem resolved in 1 month. After 2 year follow-up period, she had no evidence of recurrence radiologically and endoscopically (Figure 7).

Case 4

34 year old male patient presented with a 1-year history of progressive dysphonia. Laryngeal endoscopic examination revealed an ulcerated mass in the right vocal cord with vocal fold immobility. Biopsy of the mass under direct laryngoscopy revealed rhabdomyosarcoma. MRI scan showed heterogeneously enhancing lesion measuring 21x8 mm in the right glottic area. Using vertical hemilaryngectomy, lesion was excised completely (Figure 8) and pleomorphic variant rhabdomyosarcoma was
diagnosed (Figure 9,10). Peri and postoperative periods were uneventful. He received chemotherapy. This patient was followed up regularly for about 4 years and was completely asymptomatic.

**DISCUSSION**

Being derived from the embryonal mesodermal components of the body sarcomas have heterogeneous histopathological variants. Laryngeal sarcomas similarly have different histological types with a wide clinical spectrum ranging from relatively slow-growing lesions to locally and regionally aggressive destructive lesions. Although laryngeal sarcomas are rare, the most common sarcoma arising in the larynx is chondrosarcoma. Chondrosarcoma as an uncommonly encountered cartilaginous tumor was first described in larynx by New in 1935 [4]. Low grade chondrosarcoma has low incidence as all the cartilaginous laryngeal tumors were misdiagnosed as chondroma in the previous reports. Today diagnosis of chondroma is less common than previously thought. Although both pathologies are different, it is difficult to distinguish them by gross examination or even by histology. Histologically, chondromas show a characteristic well-defined lobular pattern with benign looking and evenly distributed chondrocytes that lack nuclear pleomorphism and mitotic activity. Chondromas are smaller than chondrosarcomas and less cellular, with less pleomorphism, lacking mitoses and necrosis. Both can be synchronous in the same lesion or malignant transformation of chondroma can lead to chondrosarcoma (metachronous) [5].

Chondrosarcoma of the larynx generally occurs in the age group between the sixth and seventh decade of life, especially in men and originates mostly from the posterior cricoid cartilage followed by the thyroid cartilage (9–20%), and the arytenoid [6]. Notably this tumor does not develop in the elastic cartilages (epiglottis). Despite unknown etiology smoking, radiotherapy, teflon injections can be risk factors [7].

Clinically patients present with hoarseness, slowly progressing dyspnea, dysphagia and neck mass. On physical
While primary treatment is conservative surgery, endoscopic debulking, laryngofissure, partial laryngectomy can be performed according to the localization of the lesion. Surgical margin is the most important factor for recurrences. Lymph node metastasis is very rare therefore neck dissection is not always performed. Aggressive treatment should be considered for recurrent disease. Radiotherapy can be a choice for palliative therapy of the residual disease [12]. Chemotherapy can be used in high-grade chondrosarcomas, clinically aggressive tumors with rapid local recurrence and cases with a high probability of metastasis [13]. Our both cases underwent only surgery with the diagnosis of low grade histopathology and no recurrence was detected during their follow up.

Five year survival %70-80 [12] and prognostic factors include disease resectability, grade, stage and localization. Death from disease is very uncommon and is usually the result of uncontrolled local growth into vital structures of the neck.

Laryngeal rhabdomyosarcoma is extremely rare. Unlike other laryngeal sarcomas rhabdomyosarcomas (RMS) are more aggressive and have a tendency to metastasize [3]. Three histological subtypes are described: embryonal, alveolar and pleomorphic. Pleomorphic variant is usually encountered in adults, especially in men but its incidence is the least of all. In RMS a multimodality approach is needed comprising surgery followed by chemo and/or radiotherapy [3]. Spindle cell sarcomatoid carcinoma is recognized as a morphologically biphasic neoplasm containing an epithelioid and spindle-shaped neoplastic proliferation. Five year survival is about 80% and most patients have a low stage [14]. Our third case had been diagnosed as neurofibroma in the initial excisional biopsies. However total reexcision of the recurrent lesion was compatible with spindle cell variant of rhabdomyosarcoma. Finally the patient was diagnosed to have spindle cell variant rhabdomyosarcoma in addition to neurofibromatosis. The other patient with pleomorphic rhabdomyosarcoma was also given adjuvant therapy. There was no recurrence during follow up.

Sarcomas of larynx are very rare as was delineated in our laryngeal cancer patients series (4 out of 325 patients diagnosed between 2006-2014). Laryngeal sarcomas are treated according to histologic type, grade and localization.

REFERENCES


