Auris Nasus Larynx xxx (2016) xxx–xxx

Contents lists available at ScienceDirect

Auris Nasus Larynx

journal homepage: www.elsevier.com/locate/anl

Case report

A case of desmoid tumor co-existing with recurrent squamous cell carcinoma in the larynx

Shogo Shinohara [a,*], Atsushi Suehiro [b], Masahiro Kikuchi [a], Hiroyuki Harada [a], Ippei Kishimoto [b], Yukihiro Imai [c]

[a] Department of Otolaryngology-Head and Neck Surgery, Kobe City Medical Center General Hospital, Minatojima-minamimachi 2-1-1, Chuo-ku, Kobe 650-0047, Japan
[b] Department of Otolaryngology-Head and Neck Surgery, Graduate School of Medicine, Kyoto University, Sakyo-Ku, Kyoto 606-8507, Japan
[c] Department of Pathology, Kobe City Medical Center General Hospital, Minatojima-minamimachi 2-1-1, Chuo-ku, Kobe 650-0047, Japan

ARTICLE INFO

Article history:
Received 23 April 2016
Accepted 12 July 2016
Available online xxx

Keywords:
Desmoid tumor
Aggressive fibromatosis
Larynx
Near-total laryngectomy
Squamous cell carcinoma

ABSTRACT

Extra-abdominal desmoid tumor, also known as aggressive fibromatosis, has aggressive behavior with local infiltration and tendency for recurrence. Though head and neck is reported to be one of the most common sites, a desmoid tumor in the larynx is extremely rare. A 67-year-old male visited our hospital with prolonged hoarseness and received laryngo-microsurgery with the diagnosis of laryngeal polyp. After the operation, he eventually developed a laryngeal squamous cell carcinoma with papilloma, confirmed by second laryngo-microsurgery and received radiation therapy. After the third laryngo-microsurgery to remove residual papilloma, white irregular mass appeared on the right vocal cord and grew rapidly beneath the glottis, causing dyspnea. After 2 additional laryngo-microsurgeries, he was diagnosed having the dermoid tumor co-existing with recurrent squamous cell carcinoma. He underwent near-total laryngectomy and is currently alive without disease, speaking using a vocal shunt. Only five cases of the desmoid tumors arising in the adult larynx have been reported in the English literature. In this case, repeated surgery and radiation were suspected as the causes. Also, the present report is the first to describe desmoid tumor co-existing with recurrent squamous cell carcinoma in the larynx.

1. Introduction

Desmoid tumor (DT), which is also known as desmoid type fibromatosis or aggressive fibromatosis, represents a group of non-metastasizing, clonal, infiltrative spindle cell proliferation with fibrosis. This rare tumor has an annual incidence of 2–4 cases per million [1] and some of the DTs were reported to be associated with hormonal factors, e.g. estrogen [2], genetic factors, e.g. familial adenomatous polyposis (FAP) gene and preceding local invasion, e.g. surgery or trauma [2,3]. DT has aggressive clinical behavior with local infiltration and tendency for recurrence, which makes surgical treatment challenging. Approximately 37–50% of desmoid tumors arise in the abdominal region while the most common extra-abdominal sites are reported to be the shoulder, chest wall, upper arm, thigh and head/neck [4]. Within the head and neck, the largest percentage of DT arises in the neck, followed by the face, the oral cavity, scalp, paranasal sinuses, and orbital areas [5]. DTs of the larynx were extremely rare. To date, we are aware of only 5 adult cases of DT involving the larynx published in the English literature [4,6–9]. This is the 6th case of DT of the larynx arising in an adult and the first case co-existing with squamous cell carcinoma of the larynx.

* Corresponding author. Fax: +81 78 302 7537.
E-mail address: sinosino@kcho.jp (S. Shinohara).

http://dx.doi.org/10.1016/j.anl.2016.07.006
0385-8146/

2. Case report

A 67-year-old male visited our hospital with prolonged hoarseness and received laryngo-microsurgery (LMS) with the diagnosis of laryngeal polyp on the left vocal cord in the summer of 2008. One year later, he received the second LMS for biopsy of the tumor on the posterior commissure of the right vocal cord and was diagnosed pathologically as having laryngeal squamous cell carcinoma (SCC) with papilloma (Fig. 1A). Conventional radiation therapy (2 Gy/fr/day, 30 fr) to the larynx was done under the diagnosis of glottic SCC, clinical T1aN0M0 in the summer of 2009. The third LMS surgery for the estimation of the response of radiation revealed the rest of the papilloma in the intraoperative frozen section and the papilloma was removed. Four months after the third LMS, white irregular mass appeared on the right vocal cord and grew so rapidly that it caused dyspnea in a month (Fig. 2). An emergent tracheostomy followed by the fourth LMS was performed in June 2010. Pathological diagnosis at that time was granuloma (Fig. 1B). He was moved to the outpatient clinic under close observation with remaining tracheostoma. The white irregular mass recurred soon and grew month by month in spite of the 2 times of intratumoral injections of triamcinolone acetonide (Fig. 2). The fifth LMS was performed for pathology and he was diagnosed having laryngeal desmoid tumor with the recurrence of SCC.

Magnetic resonance imaging (MRI) before the fifth LMS demonstrated a subglottic mass, heterogeneous in T2 weighed images and low intensity and well enhanced with Gadolinium in T1 weighted images, on the right side of the vocal cord (Fig. 3). The tumor extended from the right posterior commissure to the anterior commissure of the glottis but did not involve the posterior half of the left vocal cord in MRI (Fig. 3). The Pearson's near-total laryngectomy [10] with right lateral neck dissection was performed aiming at both of the curative resection and the partial preservation of vocal cord function in December 2010. Shortly, the larynx was excised vertically on the half of the left ala of the thyroid cartilage and the center of the back of the cricoid cartilage. After the confirmation of cancer free on surgical margins by intraoperative frozen sections, a mucosal flap was developed from left pyriform sinus, rotated downward and sutured vertically to the cut margin of laryngeal mucosa in order to produce a vocal shunt.

The tumor occupied most of the subglottic space and extended 3.5 cm downward, close to the tracheal cut end (Fig. 1C). Histologically, the entire tumor was made of the same component; the deeper area of the tumor consisted of uniform proliferation of spindle cells with bland elongated nuclei of fine bright chromatin and interlacing collagenous matrix. Superficial area consisted of same kind of spindle cell proliferation and covering and infiltrating squamous cell carcinoma (Fig. 1D and E). Although lymphatic invasion of SCC was also observed deep near to cartilage, no lymphnode metastasis was observed in neck dissection specimen (left II–IV). Immunostaining of the spindle cells revealed positivity for smooth muscle actin (SMA) and vimentin, and negativity for desmin, CD34, S100, D2-40, WT1, cKit, c-myc, cyclin D, p16, p53, β-catenin and ALK1. All available epithelial markers keratin-side, CK-MNF116, CAM 5.2, AE1/AE3, CK7, CK20, EMA, e-Cadherin were also negative for spindle cells. Ki67 proliferative index of spindle cells was 15% including infiltrating inflammatory cells, which demonstrated less aggressive nature of this tumor than the other malignancy, such as spindle cell carcinoma. These pathological

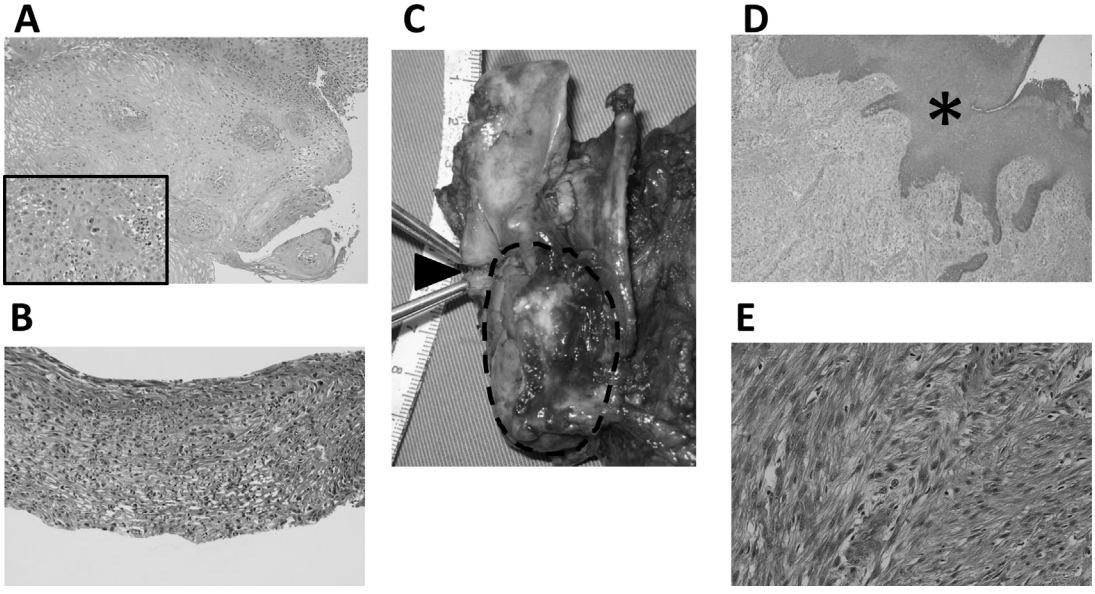

Fig. 1. (A) The biopsied specimen at the second laryngo-microsurgery showed the hyperplastic squamous mucosa on fibrovascular cores. This appearance was compatible with squamous papilloma. In some areas, nuclear enlargement, pleomorphism, and anaplasia of squamous epithelium were observed and this led to the diagnosis of squamous cell carcinoma (inset). (B) Only the granulation tissue was seen in the specimen of the forth laryngo-microsurgery. (C) The surgical specimen. A macroscopic view showed that a tumor occupied most of the subglottic space and extended to supraglottic area (black dotted line). The level of the vocal cord was indicated with a black arrow head. (D) The tumor consisted of round- or spindle-shaped nucleus and boundless, eosinophilic cytoplasm below the cancerized epithelium (*). (E) High power view revealed uniform bland spindle cell proliferation with interlacing collagenous stroma. (All the pathological findings were obtained with hematoxylin and eosin staining.)

Please cite this article in press as: Shinohara S, et al. A case of desmoid tumor co-existing with recurrent squamous cell carcinoma in the larynx. Auris Nasus Larynx (2016), http://dx.doi.org/10.1016/j.anl.2016.07.006