PRIMARY MALIGNANT MELANOMA OF THE TRACHEA

Mohamad Nattout, Nabil Fuleihan,
Omar Sabra, Ibrahim Aburizk,
Abdul-latif Hamdan

Abstract

Tracheal melanomas represent the rarest type of extracutaneous melanomas. The clinical manifestation is similar to other tracheal tumors and ranges from symptoms of airway obstruction such as dyspnea and stridor to other nonspecific symptoms such as hoarseness, cough and hemoptysis. Bronchoscopy is required to draw the origin of the lesion biopsy is needed to establish histologic diagnosis. Treatment consists of either palliative surgery aiming at restoring the airway or tracheal resection and end to end anastmosis. We would like to present here below a rare case of tracheal melanoma and discuss the various diagnostic and therapeutic means.

Key Words: melanoma; airway obstruction; trachea.

Introduction

Primary tumors arising from the trachea are uncommon. The clinical incidence does not exceed 0.2 per 100,000 people per year and the post-mortem prevalence is about 1 per 15,000 autopsies[1]. When present it is important to recognize the association of primary tracheal tumors with other head and neck tumors, with the increased risk being estimated as 11 fold[2]. More than fifty percent of these tumors are malignant and the most frequent histopathologic types are squamous cell carcinoma followed by adenocarcinoma There are only few reports in the literature on primary tracheal melanomas and these represent the rarest type of extracutaneous melanomas[3-6]. This case report describes the relevant radiologic and clinical features of this uncommon malignancy.

* Lecturer.
** Professor.
*** Resident.
**** Associate Professor, Department of Otolaryngology, American University of Beirut Medical Center.
Corresponding author: Abdul-Latif Hamdan, MD, FACS, American University of Beirut, Department of Otolaryngology, P.O. Box: 110236, Tel/Fax: 961-1-746660. E-mail: alhamdan@scvlb.com

Case Presentation

A 28 year old male, previously healthy, presented to the American University of Beirut with history of severe upper airway obstruction. He reported history of progressive dyspnea, shortness of breath and hemoptysis of two month duration. Medical history was negative for asthma or chest pain or other systemic illnesses. His physical exam revealed "stridor" and suprasternal retractions. Fiberoptic laryngeal endoscopy did not show any laryngeal abnormalities with normal mobility of the vocal folds. Computerized tomography of the neck and chest revealed a tracheal mass obstructing almost ninety percent of the tracheal lumen (Fig. 1). Magnetic Resonance imaging did not show extension into the mediastinum. PET scan did not show any metastatic lesion or other primary except the tracheal lesion. Patient underwent tracheostomy under local anesthesia followed by bronchoscopy. Bronchoscopy revealed a tracheal mass with a nodular surface arising from the right posterolateral wall at the level of the fifth tracheal ring and extending to the seventh tracheal ring. The lesion was occupying most of the tracheal lumen. Biopsy was taken and sent for histopathological examination. The following immunohistochemical stains were performed: HMB45, S-100, Vimentin and Cytokeratin AE1/AE3. All the results were positive except for cytokeratin AE1/AE3 which was negative. The diagnosis was consistent with a primary malignant melanoma of the trachea (Fig. 2).

Fig. 1
Computerized Tomography of the neck showing a tracheal tumor arising from the right posterolateral wall of the trachea and occluding almost all the tracheal lumen

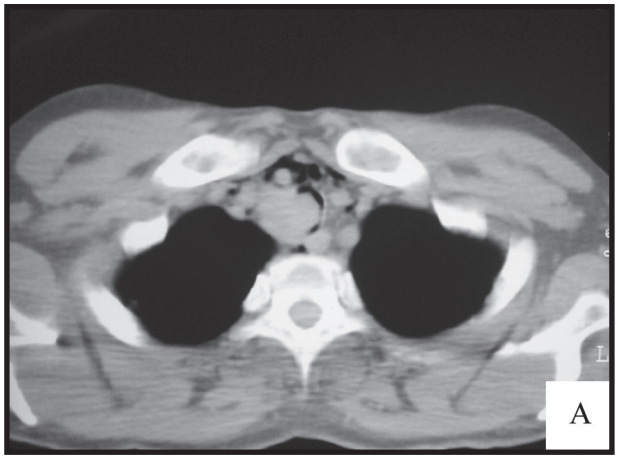
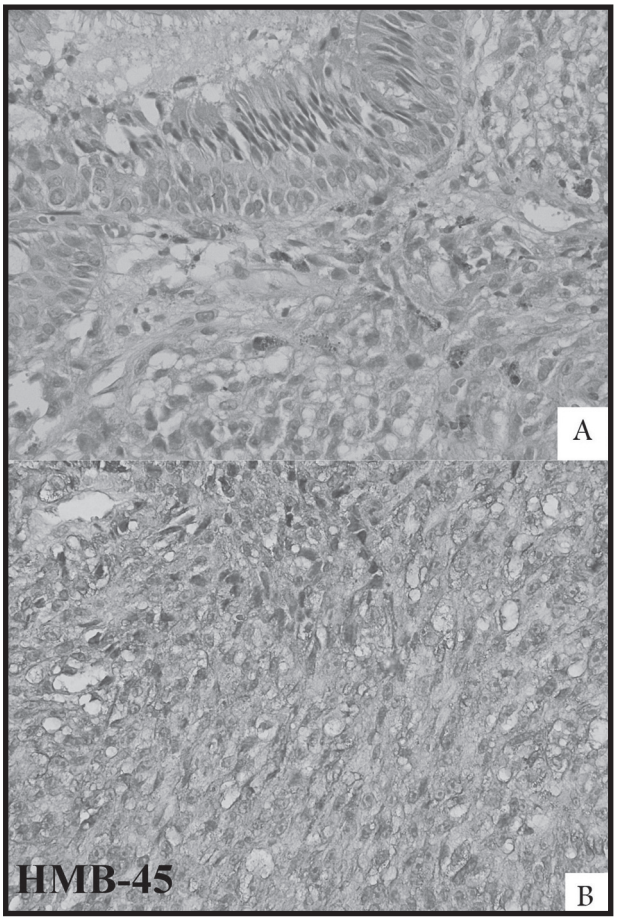


Fig. 2
A: H & E stain: Tumor composed of interlacing bundles of spindle cells, with vesicular nuclei containing prominent nucleoli, occasional cytoplasmic melanin is noted. The tumor fills and expands the subepithelial stroma. B: HMB-45 positive cells are seen



Discussion

Primary tracheal melanoma is extremely uncommon despite the increase in the incidence of malignant melanoma of the skin and the few reports of intratracheal metastasis[3]. In a retrospective review of 360 primary tracheal tumors, only one patient had tracheal melanoma[7]. The clinical manifestation of primary tracheal melanomas is similar to other tracheal tumors and ranges from symptoms of airway obstruction such as dyspnea, stridor wheezes and cyanosis, to other nonspecific symptoms such as hoarseness, cough, hemoptysis, weight loss and chest pain. Invasion into adjacent structures may present as a neck mass with the presence of dysphagia and odynophagia.[2,5]. Tracheal tumors are rarely discovered by chest or neck radiographs especially in the early stages. Not more than 50% are identified based on

these means as reported by a radiologic and clinical study conducted by Li et al. Barium esophagogram is helpful in detecting esophageal invasion when the mass is arising from the posterior tracheal wall. Computerized tomography is the most important imaging modality with an accuracy that exceeds tomography. It provides a good cross sectional image of the trachea and the endoluminal tumor extent, however it may underestimate the vertical length of the tumor in view of the volume averaging[5,6]. Bronchoscopy is required to draw the origin of the lesion and to map the extent of the tumor in relation to the vocal folds. Biopsy remains the hallmark to establish histologic diagnosis. There are strict criteria for the microscopic diagnosis of primarily melanoma of the trachea and these include the presence of a solitary tracheal tumor containing melanin histologically in the absence of tumor outside the trachea, junctional changes in the mucosa between melanoma cells and normal lining and invasion beyond the epithelium and submucosa[8,9]. Immunostains for HMB-45 and S-100 are usually positive in these tumors. These were the findings in our reported case.

There were several attempts to explain the oncogenesis of mucosal melanomas in general and to postulate the histogenesis of lower respiratory tract melanomas in specific. Shivas and MacLennan have suggested the term "melanogenic metaplasia", where mucus-secreting cells are histologically altered into melanin- producing cells. Reid and Metha and Walsh reported that squamous metaplasia is the origin of mucosal melanomas. Others have speculated that melanocytes migrate during embryogenesis to the developing lower respiratory tract or arise from a neuroendocrine cell precursor (Kul-chitsky cell)[5-9].

Treatment consists of either palliative surgery aiming at restoring the airway by means of laser usage or therapeutic with tracheal resection and end to end anastmosis[10]. For laser usage, various therapeutic options are available and these include the carbon dioxide CO_2 laser, the argon, the neodymium:yttrium-aluminum-garnet (Nd: YAG) and the new argon plasma coagulation (APC) technique under flexible bronchoscopy. Radiation therapy or endobronchial brachytherapy may be added especially in cases of incomplete resection. Chemotherapy is recommended however the prognosis remains poor irrespective of the modality of treatment.

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