Case Report

A case report of primary anaplastic large cell lymphoma arising from the trachea

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Abstract: Anaplastic large cell lymphoma (ALCL) is an aggressive non-Hodgkin's lymphoma. Its presentation as an isolated primary lesion in the airway is extremely rare and not often considered in the differential diagnosis of airway lesions. Thus, it is important to be aware of its presenting manifestations, imaging features and treatment complications. Here, we report a case of pathologically confirmed tracheal-based ALCL. We especially focus on presenting the salient imaging features of this entity and the associated clinical and pathological findings of this rare large airway disease.

Keywords: Anaplastic large cell lymphoma (ALCL); trachea; computed tomography (CT); imaging

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Introduction

Primary malignant tumors of the trachea are rare, accounting for about 0.1–0.4% of all malignancies (1-3). The most common pathology is squamous cell carcinoma, followed by cystadenoma (*Table 1*) (1,4). Although extranodal lymphoma is commonly encountered, lymphoma presenting in the large airways are incredibly rare (5), constituting less than 3% of tracheal tumors. Less than 10 cases have been reported in the literature to date (6,7). Presenting symptoms can be mistaken for other entities, and imaging can play a vital role in diagnosis and management. Here, we present a case of anaplastic large cell lymphoma (ALCL) of the trachea with an emphasis on the imaging features of this entity and a discussion of treatment complications.

Case presentation

A 31-year-old man presented to our institution complaining of a 3-month history of productive cough and 1 week of shortness of breath. The cough was associated with chest pain and hemoptysis. The patient denied fevers, fatigue or loss of appetite. A physical exam was notable in that the patient exhibited a high-pitched wheeze in the throat and upper chest.

High resolution computed tomography (CT) was performed, which demonstrated a 2.0 cm × 2.0 cm × 1.6 cm endotracheal nodule extending from the lower right tracheal wall (*Figure 1*). The lesion was homogeneous with mild contrast enhancement. It caused near-complete occlusion of the trachea. In addition, there was a right paratracheal lesion adjacent to the endotracheal mass with extension into the medial right upper lobe.

Resection of the tracheal lesion and sampling of the right paratracheal mass was performed via bronchofiberoscopy. Histology was consistent with ALCL (*Figure 2*). Anaplastic lymphoma kinase (ALK) staining was positive. Other immunohistochemistry stains included: CD30(+), CD8(–), CD4(–), CD5(–), Granzyme B(+), Perforin(+), TIA-1(+), CD2(–), CD43(–), CD68(–), EMA(+), PLAP(–), Myogenin(–), Des(–), MyoD1(weak +), CD99(+), Myoglobin(+), Vim(+), LCA(+), and Ki67(90%+).

A bronchial stent was subsequently placed after mass resection, followed by autologous stem cell transplantation, chemotherapy and radiotherapy (*Figure 3*). Patient's dyspnea

Table 1 The imaging features of different tracheal tumors

Features	Adenoid cystic carcinoma	Carcinoid	Mucoepidermoid carcinoma	Lymphoma
Predilection site	Trachea or main bronchus; the main body is in the lateral posterior wall	Main bronchus	Lobar or segmental bronchus, rare in trachea or main bronchus	Trachea or main bronchus
Growth characteristic	Mass encircling the trachea with wall thickening; usually involve more than 180° of the airways circumferences; both intra and extra the lumen	Nodule within the lumen and mass in the external lumen	Smooth circular or round like nodule attached the wall, with broad base. The long axis is parallel to the lumen	Mass within the tracheobronchial lumen
Density	Homogeneous	Homogeneous; flake necrosis can be seen in atypical type	Mostly homogeneous; lower than muscle	Homogeneous; necrosis is rare
Calcification	Rare	Characteristic manifestation: diffuse, eccentric; gravel, nodular, popcorn	Punctate or nodular	Rare
Enhancement	Mild	Obvious	Mild heterogeneous enhancement	Mild homogeneous enhancement

markedly improved post-operatively. The right paratracheal mass also significantly decreased in size. The patient was subsequently admitted multiple times over the course of 4 years for recurring anhelation. CT imaging from his first post-operative hospitalization raised concern for tumor recurrence, given the increased tracheal wall thickening that resulted in re-stenosis (*Figure 4*). Trans-bronchial biopsy performed at that time demonstrated granulomatous changes without evidence of lymphoma. This tissue was resected, and the stent exchanged. The patient has required further stent exchanges due to recurrence of granulomatous tissue, but no evidence of tumor recurrence has been noted to date.

Discussion

ALCL is a sub-type of non-Hodgkin lymphoma (NHL) (8) and can be categorized as ALK-positive and ALK negative. The distinction of them is important and essential because of clinical and prognostic differences. Namely, ALK-positive ALCL is more common in younger patients who generally have superior outcomes when treated with standard chemotherapy as compared to patients with ALK-negative ALCL. ALK-positive ALCL patients typically have 5-year overall survival rates of more than 70%, whereas patients with ALK-negative ALCL have less than 50% (8,9). Those with ALK-negative ALCL are often middle-aged patients (8). There is a strong (6:1) male predilection.

Clinical manifestations of tracheal ALCL, including dyspnea, stridor and wheezing due to obstruction (8,10), can rapidly progress to respiratory failure and require emergency management. They can often be initially misdiagnosed as chronic obstructive pulmonary disease. Thus, if the above symptoms are present, it is important to consider the presence of a large airway tumor on the differential diagnosis.

Airway lesions are difficult to diagnose on chest radiography and CT is very helpful for imaging evaluation. ALCL mainly occur in the trachea or main bronchus, manifesting as a mass within the tracheobronchial lumen. Pharyngeal lymphoma does occur but is also a rare entity (11). These masses are generally not necrotic, unlike other tracheal lesions (*Table 1*). Most of the cases present locally, but involvement of the adjacent mediastinum and lung can sometimes occur. Because of patients' airway symptoms, patients are usually diagnosed prior to significant mediastinal or parenchymal involvement. Diffuse lymphadenopathy is not common in primary tracheal ALCL.

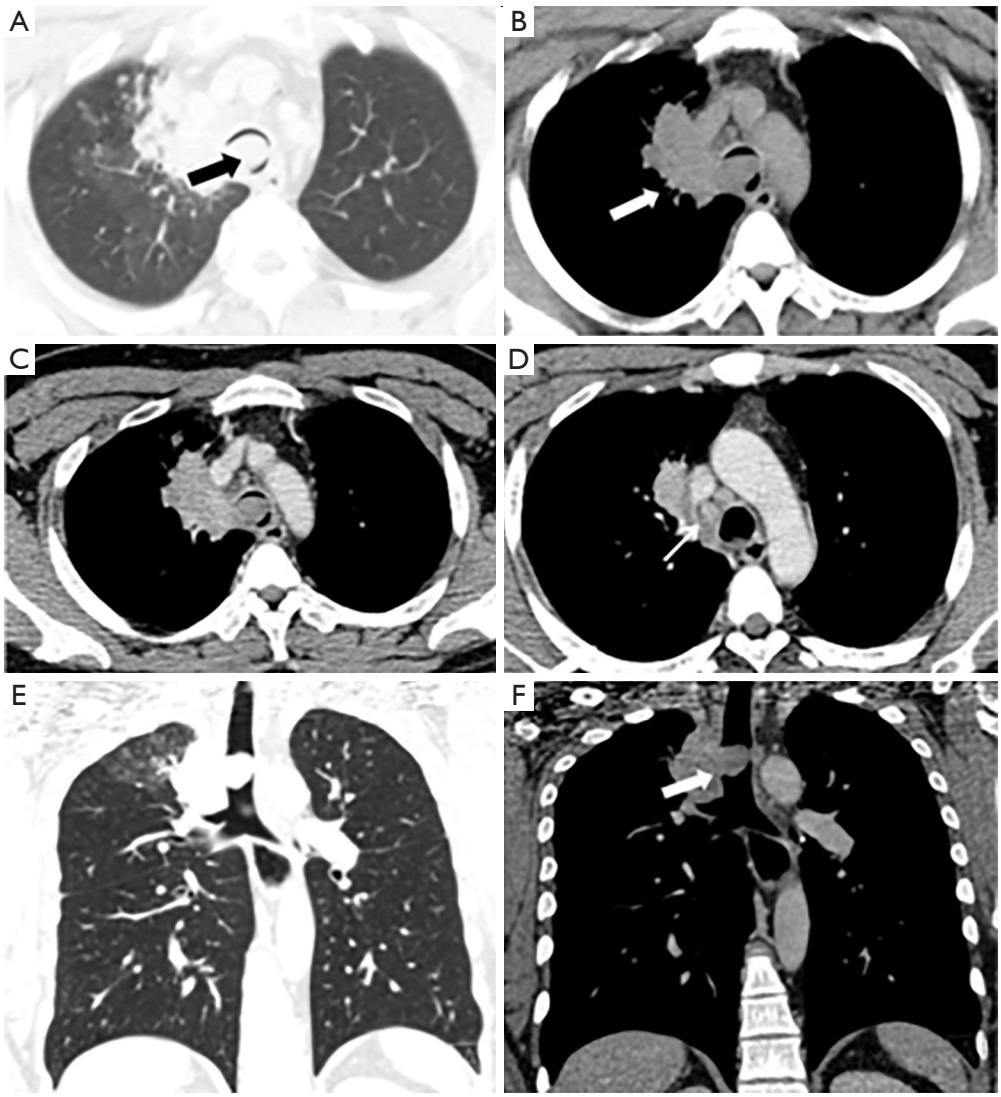

Figure 1 CT manifestation of the lesion. (A) CT demonstrates nodular lesion (arrow) within the lumen of the trachea, occupying about two thirds of the lumen. The margin of the protruding part of the lesion is smooth; there is a broad base connection with the tracheal wall; (B) a consolidative lesion is present adjacent to the tracheal mass in the medial right upper lobe that extends into the mediastinum (arrow). This mass appears to be contiguous with the endotracheal lesion and without a definite margin identified. The endotracheal and right paramediastinal mass enhances homogeneously without evidence of necrosis (C,D). Adjacent paratracheal lymphadenopathy was also present (arrow); (E,F) coronal view of the endotracheal lesion shows near complete occlusion of the lower trachea by the lesion. CT, computed tomography.

Treatment strategies for patients with primary tracheal lymphoma are controversial and may depend on pathology subtype. Surgery, chemotherapy, and radiation therapy usually are used either alone or in combination. Positron emission tomography (PET)/CT maybe useful to search for systemic disease and for treatment response evaluation. The International Prognostic Index (IPI) is important for evaluating NHL, which was determined using prognostic factors (the age at diagnosis, performance status, lactate dehydrogenase (LDH), stage and number of extranodal sites). This patient had a low-risk IPI, with 0 risk factors. For patients whose LDH was unknown, it was presumed to be normal for calculating the IPI.

Local resection is usually performed for symptom

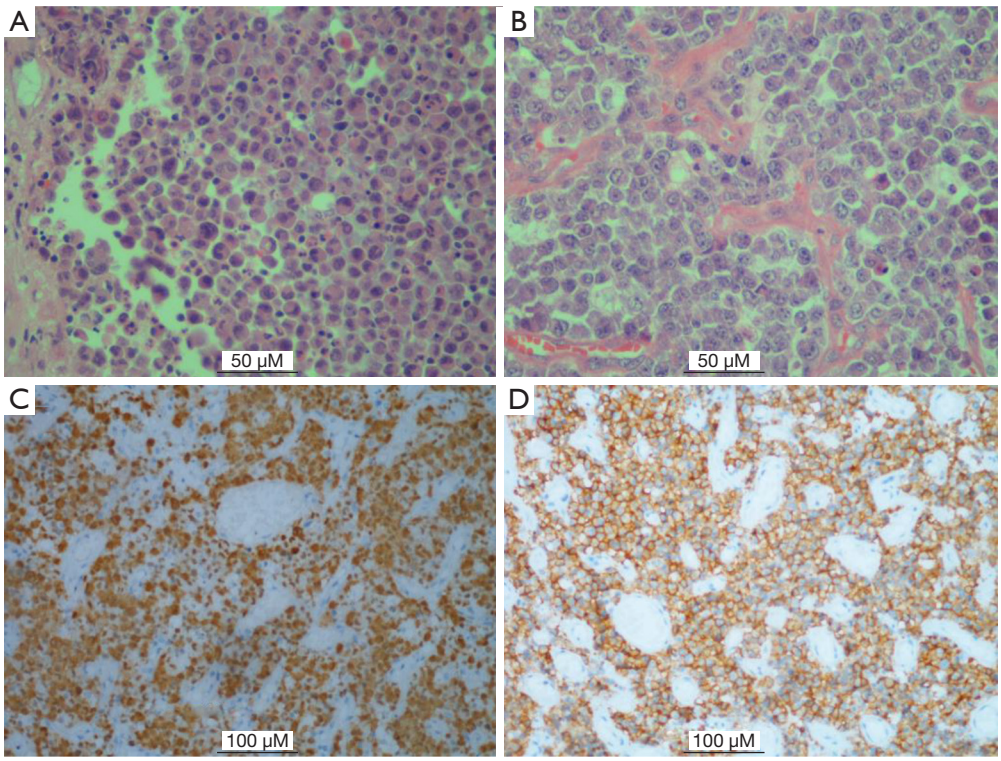

Figure 2 Histology and immunohistochemistry of the endotracheal mass. (A,B) ALK-positive ALCL. The hallmark cell of ALCL is a large cell with an eccentrically placed embryo-like or reniform nucleus, a distinct eosinophilic Golgi zone in the ample amphophilic cytoplasm (HE ×400); (C) ALCL. All malignant cells are strongly positive for ALK (IHC ×200); (D) ALCL. All malignant cells are strongly positive for CD30 (IHC ×200). ALK, anaplastic lymphoma kinase; ALCL, anaplastic large cell lymphoma; HE, hematoxylin and eosin; IHC, immunohistochemistry.

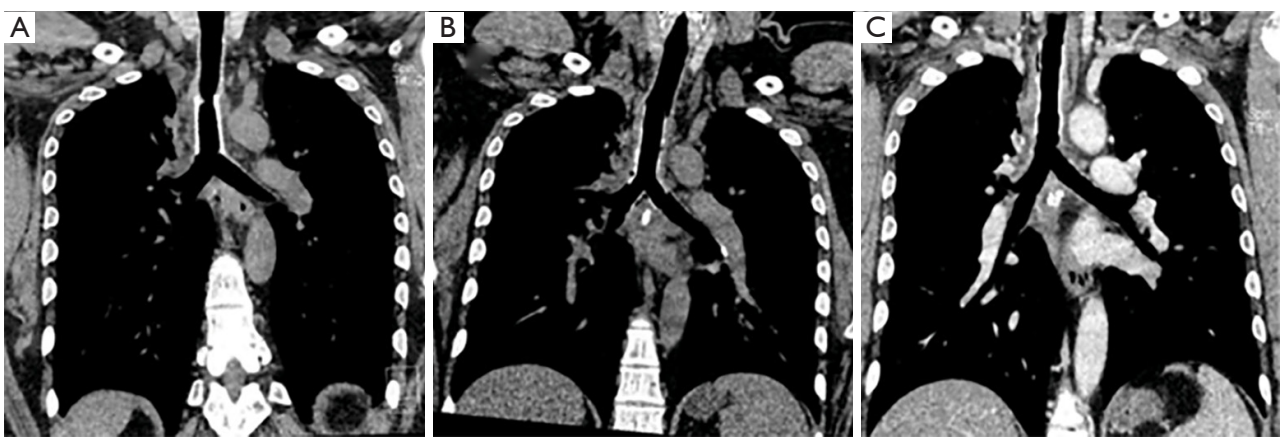

Figure 3 Follow-up chest CTs. Follow up chest CTs in (A) November 2015, (B) September 2016, and (C) October 2017 showing bronchial stent in place. No tumor recurrence has been detected for 4 years post treatment. CT, computed tomography.

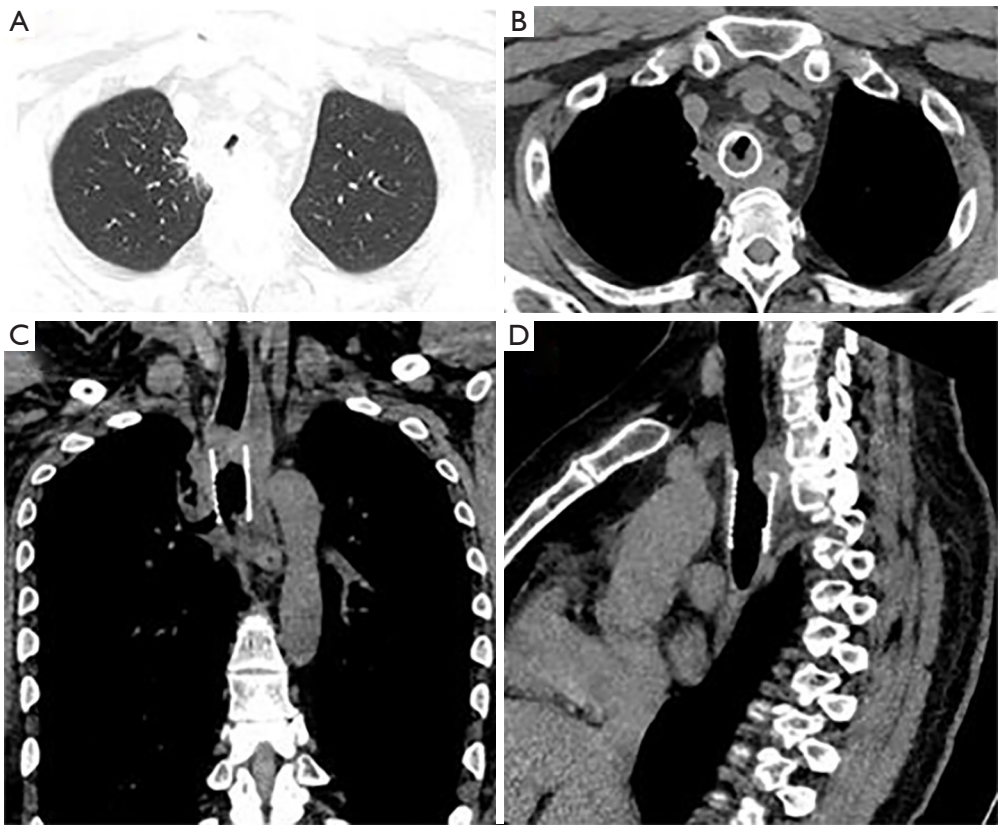

Figure 4 Postoperative CT findings. Two years after operation, CT imaging raised concern for tumor recurrence, given the increased tracheal wall thickening resulting in re-stenosis. Trans-bronchial biopsied performed at that time demonstrated granulomatous changes without evidence of lymphoma. CT, computed tomography.

relief. Relapsing disease is very uncommon, but has been known to occur. Thus, imaging follow-up is important in these patients. Even without relapse, complications from local benign granulomatous growths can lead to recurrent clinical symptoms (12). For our patient, the increased soft tissue around the endobronchial stent required multiple debridement and stent exchanges to maintain airway patency. Quantitative evaluation of the airway prior to stent placement, which can be provided by imaging, may be useful to reduce this complication.

In conclusion, primary ALCL is a rare endotracheal tumor with presenting symptoms that can often be misdiagnosed as chronic obstructive pulmonary disease. Taken together with patient's clinical history, imaging features on CT can help guide the clinician to consider this diagnosis. Follow-up imaging post treatment is important to monitor treatment complications and, in rare cases, disease recurrence.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at http://dx.doi.org/10.21037/tcr.2019.02.05). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related

to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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