Case Report

Endotracheobronchial lymphoma: Two unusual case reports and review of article

Trilok Chand, Avdhesh Bansal, Harsh Dua[1], Kapil Sharma

Departments of Respiratory, Critical Care and Sleep Medicine and [1]Medical Oncology, Indraprastha Apollo Hospitals, New Delhi, India

ABSTRACT

The tracheobronchial origin of non-Hodgkin's lymphoma (NHL) is a very rare presentation, and there are only a few case reports of primary tracheal or endobronchial NHL. We have two cases of primary tracheobronchial NHL; one case was incidentally diagnosed as anaplastic large cell lymphoma of endobronchial origin when a comprehensive workup and surgery were carried out for an endobronchial aspergilloma which was actually sitting on top of lymphoma. The second patient was a case of myelodysplastic syndrome who presented with acute respiratory distress; on thorough workup, he was found to have endotracheal B-cell lymphoma. Both cases were responding well with standard chemotherapy. The mortality in these kinds of patients is due to disease progression or airway compromise and treatment complications.

KEY WORDS: Aspergilloma, chemotherapy, endobronchial lymphoma, non-Hodgkin's lymphoma, tracheal tumor

Address for correspondence: Dr. Trilok Chand, B-241, Sarita Vihar, New Delhi - 110 076, India. E-mail: rao272@gmail.com

INTRODUCTION

The trachea is an uncommon site of primary lung malignancy, then endobronchial origin of malignancy. The primary neoplasm of the trachea is only 2% of all malignancies[1] and representing <0.1% of cancer death. The most common tracheal tumor is squamous cell carcinomas followed by adenoid cystic carcinomas.[2] The tracheal lymphoma is again a very rare presentation, accounting <3% of all tracheal tumors. Endobronchial lymphoma is also a rare presentation, and more common is Hodgkin's lymphoma as compared to non-Hodgkin's lymphoma (NHL). Anaplastic large cell lymphoma accounts for 2% of NHL, and its endobronchial presentation is extremely rare. We present a series of two case reports of which one appeared to be endobronchial aspergilloma (another rare entity) initially on bronchoscopic biopsy but was proven to be endobronchial anaplastic large cell lymphoma (EALCL) on surgical excision and the second one was a case of myelodysplastic syndrome, came with shortness of breath, which was later diagnosed with a case of endotracheal B-cell lymphoma. Both cases were successfully treated by surgery and chemotherapy.

Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/0970-2113.192873

CASE REPORTS

Case 1

A 24-year-old male, nonsmoker, presented with a 1-month history of shortness of breath, dry cough, and fever for 2 weeks. On examination, air entry was diminished on the right side of the chest, and breath sounds were absent in infrascapular area on the right side.

The patient's routine blood investigations were within normal limits, but his chest X-ray (CXR) showed the right lower lobe (RLL) collapse and contrast-enhanced computed tomography (CECT) of the thorax revealed a soft-tissue mass

How to cite this article: Chand T, Bansal A, Dua H, Sharma K. Endotracheobronchial lymphoma: Two unusual case reports and review of article. Lung India 2016;33:653-6.

in the right main bronchus with partially opened upper lobe and intermediate bronchus with the collapse of a posterior basal segment of RLL [Figure 1]. The CECT of the chest did not show any mediastinal or hilar lymphadenopathy. Fiberoptic bronchoscopy revealed a yellowish glistening endobronchial mass in the right main bronchus almost totally occluding it [Figure 2]. No other segment could be visualized on the right side. As the CXR and computed tomography (CT) had shown only lower lobe collapse, the mass was presumed to be originating from one of the lower lobe segments. The mass could be made to move while taking multiple biopsies proving that it had a stalk.

Histopathology revealed acute necrotizing inflammation due to *Aspergillus*. In view of the large size of *Aspergillus* mass, thoracotomy with right lower lobectomy and extraction of fungal mass from the right main bronchus was done. The mass was found attached to the posterior wall of RLL bronchus.

Histopathology of the mass showed anaplastic large cell lymphoma in the stalk (positive staining with LCA, CD30, and Alk-1) [Figure 2], and the head portion of the mass was again found to have *Aspergillus* in necrotic material which was initially reported on bronchoscopic biopsy. There was no involvement of the lung tissue by lymphoma.

Case 2

A 65-year-old female, nonsmoker, presented with cough, breathlessness, and generalized weakness for 1 month. On examination, the patient was conscious, afebrile; vitals were stable except oxygen saturation at room air was 91%. On chest auscultation, bilateral air entry was slightly sluggish.

She was a known case of psoriasis and on ayurvedic medication for the previous 3 years of presenting here. She was also diagnosed myelodysplastic syndrome 1 year back before presented here.

The patient's routine blood investigations were within normal limit except low Hb (8.6 g%) and low total leukocyte count (3400/cmm) due to myelodysplasia. Her CXR showed increased bronchovascular markings, but high-resolution computed tomography of the chest showed intratracheal growth and old calcified nodules with peribronchovascular distribution but no intrathoracic lymphadenopathy [Figure 3]. Two-dimensional echocardiography revealed global hypokinesia with ejection fraction - 30% and mild concentric left ventricular hypertrophy. Her bronchoscopy showed a large rounded mass lesion just below the vocal cords on the posterior tracheal wall and two small masses on anterior aspect of the vocal cords [Figure 4]. Furthermore, there were multiple small nodules seen on the mucosa of the trachea till carina.

A biopsy was taken from a large mass lesion and histopathology confirmed the lymphoproliferative B-cell

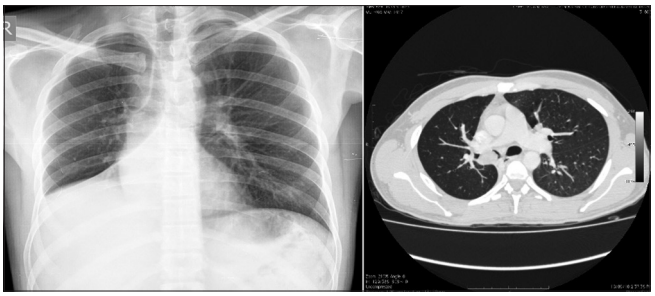

Figure 1: Chest X-ray shows the right lower lobe collapse, and computed tomography section shows mass in right main bronchus

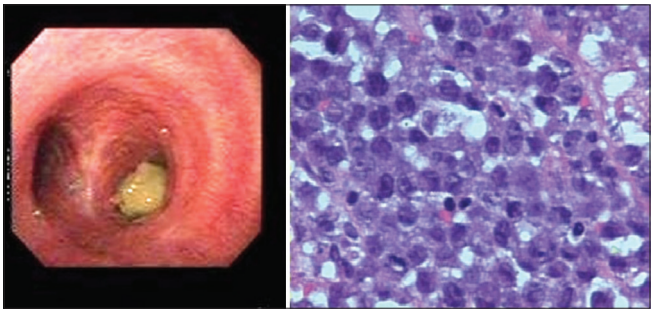

Figure 2: Fiberoptic bronchoscopy shows glistening mass in right main bronchus, and histopathology slide shows anaplastic large cell lymphoma

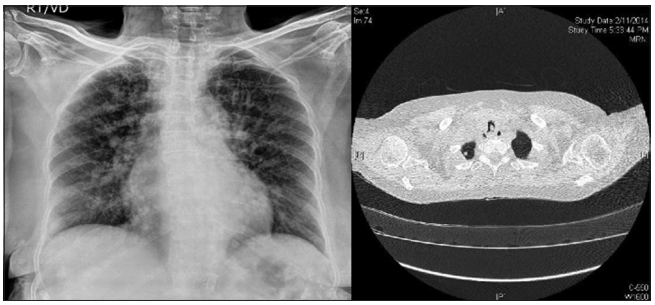

Figure 3: Chest X-ray shows prominent bronchovascular marking, and computed tomography section shows growth in the upper trachea

lymphoma (NHL) with the positivity of CD20, CD3, CD138 and limited CK positivity to surface epithelium. The patient was started on standard chemotherapy, and she was kept under close observation in hospital for a few days because of risk of complete tracheal obstruction by tumor mass. After a few weeks, her check bronchoscopy was done to evaluate the mass which revealed regression of the mass with patent trachea.

DISCUSSION

The majorities of tracheal tumors are malignant and mostly are metastases from other sites such as lung, thyroid, esophagus, or larynx. The primary tracheal tumor is uncommon, and lymphoma of tracheal or endobronchial origin is a rare instance. The NHL of tracheobronchial tree is less common than Hodgkin's lymphoma. Primary tracheal NHL accounts for only 0.2–3% of all tracheal tumors.[3-6] The most comprehensive

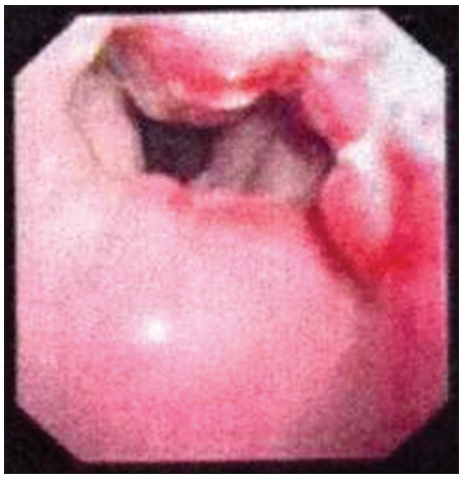

Figure 4: Fiberoptic bronchoscopy shows multiple intraluminal growths in the upper part of trachea

review of the literature identified only 28 primary tracheal lymphomas from 1973 to 2000.[5] Extranodal lymphoma itself is not uncommon, but patients with extranodal lymphoma only have a 3.6% rate of tracheobronchial involvement.[7]

The first case of endobronchial NHL was described in 1955 by Dawe *et al*.[8] In an autopsy study of 55 patients of NHL, none of the cases showed endobronchial lymphoma.[9] In another autopsy study, only one patient had endobronchial lesion out of 93 patients with pulmonary lymphoma.[10] Our both patients found to have NHL which is less common than Hodgkin's lymphoma, and one patient has anaplastic large cell type of Hodgkin's lymphoma which is again rare. The endobronchial anaplastic large cell lymphoma comprises 2–7% of NHLs.[11]

The median age of patients with primary tracheal lymphoma is 44 years, with a range of 4–81 years.[5] The clinical presentations of primary tumors of the trachea are usually insidious in onset, with signs and symptoms of upper airway obstruction. In a series of 329 patients with primary tracheal malignancies, dyspnea was the most frequent symptom (71%) followed by cough (40%), hemoptysis (34%), asthma (19.5%), and stridor (17.5%).[12] Although hemoptysis is uncommon in tracheal lymphoma because of its submucosal involvement of the tracheal wall, blood-tinged sputum can occur in endobronchial lymphoma.

The less common symptoms are hoarseness of voice and dysphagia, due to involvement of adjacent structures, but rarely, the patient can also present with acute respiratory distress due to compromised airways. Retention of secretions may lead to symptoms of persistent cough, wheezing, or stridor.[7]

The diagnosis is based on clinical history and radiological findings. The plain CXR sometimes appears normal, and the diagnosis is usually delayed or misinterpreted as asthma. The diagnosis may also be delayed in adults because of the large functional reserve of the tracheal lumen, which often requires at least 50–75% occlusion before symptoms present.[5,7]

The CT scan of the chest is the most useful method to assess tracheal or endobronchial tumors radiologically because it allows assessment of tumor extent and relationship to adjacent structures.[7] CT scan can also demonstrate coexisting mediastinal or hilar lymphadenopathy though in our both cases CT of the chest did not reveal any mediastinal or hilar lymphadenopathy.

The treatment of tracheal lymphoma includes chemotherapy, radiotherapy, or surgery alone or in combination. NHLs are known to be highly radiosensitive. Radiotherapy alone is a standard treatment for Stage I to II gastric and nongastric mucosa-associated lymphoid tissue lymphomas.[13] In patients with symptomatic tracheal stenosis, temporary stenting followed by chemotherapy, radiotherapy, or both has been suggested.[14]

The treatment of choice for endobronchial lymphoma is chemotherapy though our patient was undergoing surgical removal of the mass due to its large size and initial diagnosis of endobronchial aspergilloma. EALCL requires surgical treatment only when they cause respiratory embarrassment due to obstruction, and it is mainly stenting or laser removal through the rigid bronchoscope. Our case represents a rare EALCL whose head was covered with *Aspergillus* probably from inhaled environmental exposure. The treatment and prognosis of tracheobronchial lymphoma depend on the histopathologic subtype and extent of tumor in surrounding tissues. The overall outcome of tracheobronchial lymphoma is favorable though patients with tracheal cancers had a poorer prognosis compared with lung cancer.

CONCLUSION

We conclude that lymphoma of tracheobronchial tree is a rare tumor and non-Hodgkin's type of lymphoma is a very rare presentation. Patients of tracheal lymphoma are more symptomatic and have acute presentation because of compromised conduit access to the lungs. Diagnosis was confirmed by biopsy and histopathologic examination and had a favorable response to chemotherapy and/or surgery.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Nuwal P, Dixit R, Singhal AK. Primary adenoid cystic carcinoma of trachea presenting as midline neck swelling and mimicking thyroid tumor: A case report and review of literature. Lung India 2010;27:167-9.

2. Choudhury BK, Barman G, Singh S, Ahmed K. Adenoid cystic carcinoma of the upper trachea: A rare neoplasm. J Clin Imaging Sci 2013;3:39.
3. Fidias P, Wright C, Harris NL, Urba W, Grossbard ML. Primary tracheal non-Hodgkin's lymphoma. A case report and review of the literature. Cancer 1996;77:2332-8.
4. Yang KY, Chen YM, Huang MH, Perng RP. Revisit of primary malignant neoplasms of the trachea: Clinical characteristics and survival analysis. Jpn J Clin Oncol 1997;27:305-9.
5. Takami A, Okumura H, Maeda Y, Kumano Y, Asakura H, Oda M, *et al*. Primary tracheal lymphoma: Case report and literature review. Int J Hematol 2005;82:338-42.
6. Kaira K, Ishizuka T, Tanaka H, Tanaka Y, Ishizuka T, Yanagitani N, *et al*. B-cell non-Hodgkin lymphoma presenting as an endobronchial polypoid mass. J Thorac Oncol 2008;3:530-1.
7. Gasparetto TD, Azevedo FB, Toledo A, Zanetti G, Marchiori E. Primary tracheal non-Hodgkin lymphoma: Case report with an emphasis on computed tomography findings. J Thorac Imaging 2010;25:W24-6.
8. Dawe CJ, Woolner LB, Parkhill EM, McDonald JR. Cytologic studies of sputum, secretions and serous fluids in malignant lymphoma. Am J Clin Pathol 1955;25:480-8.
9. Vieta JO, Cravor LF. Intrathoracic manifestations of the lymphomatoid diseases. Radiology 1941;37:138-59.
10. Papaioannou AN, Watson WL. Primary lymphoma of the lung: An appraisal of its natural history and a comparison with other localized lymphomas. J Thorac Cardiovasc Surg 1965;49:373-87.
11. Solomonov A, Zuckerman T, Goralnik L, Ben-Arieh Y, Rowe JM, Yigla M. Non-Hodgkin's lymphoma presenting as an endobronchial tumor: Report of eight cases and literature review. Am J Hematol 2008;83:416-9.
12. Velez Jo ET, Morehead RS. Hemoptysis and dyspnea in a 67-year-old man with a normal chest radiograph. Chest 1999;116:803-7.
13. Chung HT, Shia SL, Schechter N. Non-Hodgkin's lymphoma. In: Hansen EK, Roach M, editors. Handbook of Evidence-Based Radiation Oncology. 2nd ed. New York: Springer; 2010. p. 583-92.
14. Dannenberg C, Haupt R, Mantovani L, Skuballa A, Körholz D. Primary high-grade non-Hodgkin lymphoma of the trachea in an adolescent. Pediatr Hematol Oncol 2003;20:399-402.