

CASE REPORT

Tracheobronchopathia osteochondroplastica: awareness is the key for diagnosis and management

K Devaraja,¹ Prem Sagar,² Amit Singh Chirom²

¹Department of Otorhinolaryngology, Kasturba Medical College, Manipal, Karnataka, India

²Department of Otorhinolaryngology, All India Institute of Medical Sciences, New Delhi, Delhi, India

Correspondence to

Dr K Devaraja,
deardr@gmail.com

Accepted 21 July 2017

SUMMARY

Tracheobronchopathia osteochondroplastica is a rare airway disease of unknown aetiology. Due to overlapping symptomatology and lack of awareness, the condition is often missed resulting in unnecessary medical or surgical treatment. A male patient presented with a long-standing history of hoarseness and had earlier received treatment for bronchial asthma and tuberculosis. On evaluation, he had typical submucosal calcified nodules distributed throughout the trachea sparing the posterior membranous part. Although the biopsy confirmed the diagnosis of tracheobronchopathia osteochondroplastica in our case, histopathological examination is not always needed to make this diagnosis. Our patient has been kept under conservative management and is having non-progression of disease at 1-year follow-up. After having reviewed the literature related to pathophysiology and management of tracheobronchopathia osteochondroplastica, we emphasise on the fact that the treating physicians' awareness about this condition is the key to its diagnosis and management.

BACKGROUND

Tracheobronchopathia osteochondroplastica (TO) is a rare idiopathic disease of the tracheobronchial framework. Most often it is diagnosed incidentally and managed conservatively because of its self-limiting course. However, when symptomatic, it may mimic airway diseases like asthma, chronic obstructive pulmonary disease, sarcoidosis and so on.¹ The non-specific symptomatology and lack of awareness might lead to misdiagnosis of TO as some other condition resulting in unnecessary treatment.² Through this case report of TO we are readdressing the classical clinico-radiological features of the condition, which are often diagnostic. Extensive but relevant literature has been presented in a systematic manner to create awareness about the condition.

CASE PRESENTATION

A 56-year-old male patient came to us with recent worsening of hoarseness which was there with last 10 years. He had no history of aspiration or swallowing difficulty. The patient was on multidose inhaler therapy for bronchial asthma and had taken antitubercular treatment 27 years back for pulmonary tuberculosis. On examination, the supraglottis and the glottis were normal, but there was some nodular thickening in subglottis, as shown in [figure 1A](#). The tracheoscopy also showed similar

irregular nodular lesions distributed along the entire length of the trachea, except the posterior wall, as in [figure 1B](#). Cross-section imaging revealed numerous irregular deposits of calcified nodules along the mucosal surface of the trachea, with no significant lumen narrowing, as shown in [figure 2](#). A biopsy was taken from these lesions to rule out any neoplastic or chronic granulomatous aetiology. However, the histopathological examination was suggestive of TO, as shown in [figure 3](#).

OUTCOME AND FOLLOW-UP

The patient was kept under conservative management with the close observation and has a non-progressive course at 1-year follow-up.

DISCUSSION

The reported incidence of TO is 0.1%.³ Although the aetiopathology of this condition is elusive, it is characterised by inflammatory osseocartilaginous deposits underneath the mucosa of the tracheobronchial tree. The infection has been proposed as probable aetiology responsible for TO, as the affected individuals usually have a history of recurrent respiratory infections.⁴ Reported cases of TO associated with the IgA deficiency also support this theory.⁵ Certain bacteria has also been isolated from bronchoalveolar lavage of the affected individual.⁶ TO mostly affects elderly in the sixth decade and has a male predilection.^{3 7 8} However, there are few reports of TO affecting children.^{4 6} In the clear majority of the affected individuals, this condition is diagnosed incidentally.^{9 10} However, symptomatic patients may present with a long-standing history of an unresolved non-productive cough, change in voice and/or breathing difficulty.¹¹⁻¹³ Cough is the most common complaint, seen in 66% of the symptomatic patients.¹⁴ Due to non-specific and implicit symptoms, TO may mimic laryngeal amyloidosis or sarcoidosis clinically. There are reports of TO being misdiagnosed as asthma or chronic obstructive pulmonary disease.^{1 6 15} Our patient has also received treatment for bronchial asthma and tuberculosis in the past, which could have been avoided by appropriate diagnosis.

The key in identifying the patient with TO is 'the awareness' about the condition. All these disease entities have similar and overlapping symptomatology, but the signs of TO are quite peculiar and diagnostic. The fibre-optic bronchoscopy (FOB) reveals characteristic whitish submucosal hard nodules along the walls of the trachea, with pathognomonic sparing of



CrossMark

To cite: Devaraja K, Sagar P, Chirom AS. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-220567

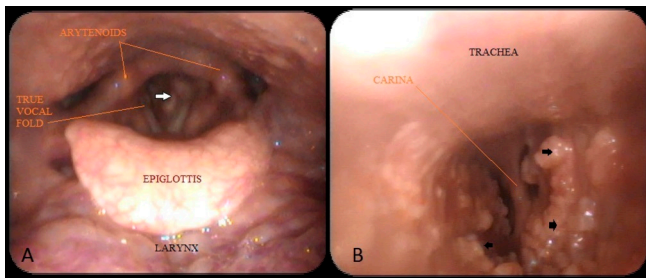


Figure 1 (A) Laryngoscopy showing subglottic narrowing (white arrow). (B) Fibre-optic bronchoscopy image with irregular nodular thickening (black arrows) along the entire length, sparing the posterior wall.

the membranous part. These lesions are ‘rock-garden’ like thickenings representing osseous or metaplastic cartilaginous nodules of 1–3 mm underneath normal respiratory mucosal layer.⁶ Amyloidosis, sarcoidosis and papillomatosis could also have a similar endoscopic appearance, but none of these would selectively spare posterior membranous wall.

CT shows irregular calcification along the inner lining of the tracheobronchial tree, typically, sparing the posterior membranous trachea.^{14 16} This nodular lesions on the inner surface of tracheal cartilage are usually sessile and diffuse, but could be focal and may not show calcification at times.^{14 17} The other conditions with possible similar radiological picture include relapsing poly-chondritis and old age. However, in contrast to TO, the cartilage gets thickened and deformed without any intraluminal nodules in relapsing poly-chondritis. The calcification due to ageing is usually uniform and not extensive like that of TO.¹⁶

Symptomology of TO may overlap with that of laryngeal amyloidosis, sarcoidosis, respiratory papillomatosis, dystrophic calcification of trachea, relapsing poly-chondritis and other connective tissue disorders. As the prevalence of TO is rare, these differential diagnoses should be considered while making the diagnosis of TO. However, most of these lesions have clinical appearances and radiological features that are distinct from characteristic clinicoradiological features of TO. Nevertheless, the presence of concomitant systemic features should not be overlooked and, whenever in doubt, further investigations should be done accordingly, which may include the biopsy from the lesion(s).

The histopathological feature of distinct cartilaginous, as well as mineralised osseous elements underneath intact mucosa, confirms the diagnosis of TO, but a biopsy is not necessary in all cases.^{1 2 18} TO has been found to be associated with certain

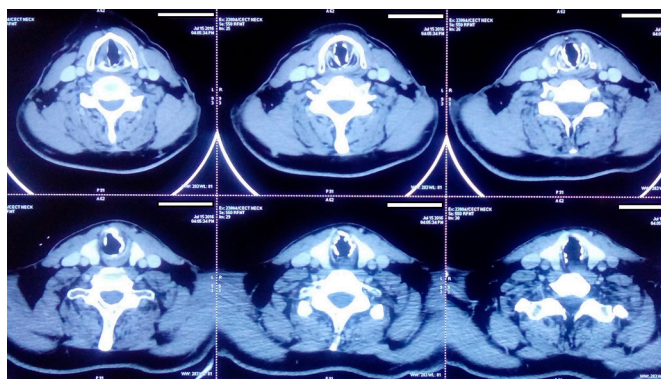


Figure 2 CT images showing irregular submucosal nodular calcification.

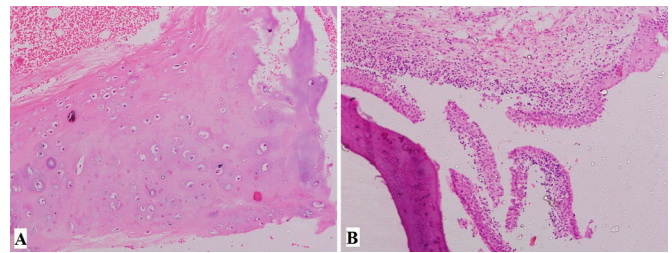


Figure 3 Histopathological pictures. (A) Typical submucosal cartilaginous and osseous deposits underneath the respiratory epithelium (40×). (B) Squamous metaplasia of the ciliated columnar epithelium of respiratory mucosa representing chronic inflammatory reaction (40×).

malignancies like skin cancer and multiple myeloma.^{3 9} However, this association is probably coincidental as there is no pathophysiological correlation between TO and malignancy.⁸ Various other diseases like polyarteritis nodosa and IgA deficiency have also been found concomitantly with TO.^{5 11} In the index case, there were no signs and symptoms of malignancy, bronchiectasis, polyarteritis nodosa or any other previously described associations.

TO is a benign disease with the stable course over years. It doesn't progress in half of the cases on repeated bronchoscopic examinations and most of these patients are managed conservatively without any active treatment.^{1 4 8} Affected individuals have had a non-progressive course as long as 20 years after diagnosis.¹⁹ However, it is important to explain the patients about the condition along with implications and consequences. These patients may have exertional distress and should have adequate treatment available to relieve distress at that time. They should be warned about the risk of respiratory failure during an episode of superadded airway infection, which is due to further narrowing of the lumen.²⁰ Affected individuals are also at a risk of difficult intubation during any elective surgery.¹⁰ If such a need arises, patients should be able to discuss the diagnosis with concerned anaesthesiologists to avoid unanticipated complicated airway. Topical steroids and chest physiotherapy could be beneficial in controlling cough and dyspnoea.^{2 15} Surgical intervention is indicated for lesions causing symptomatic narrowing.^{11 12 18} Endobronchial Nd:YAG laser, coring with a rigid bronchoscope and

Patient's perspective

The patient is very happy to get the right diagnosis and management after many years of onset of symptoms. The patient himself along with family members has voluntarily insisted on this publication so that it generates interests among medical professionals and helps further patients of the condition.

Learning points

- ▶ The possibility of tracheobronchopathia osteochondroplastica must be exercised while dealing with tracheobronchial disease, especially in adults with a long-standing history.
- ▶ The typical clinicoradiological features are often diagnostic, but the treating doctor should be aware of the condition.
- ▶ This may help in avoiding the morbidity of unnecessary treatment.

endoluminal stents are the various surgical modalities used to provide adequate airway in these cases.²¹

Contributors All the authors: involved in the manuscript preparation. KD, PS and AS: involved in concepts and design; planning; definition of intellectual content; literature search; clinical work; data acquisition; data analysis; manuscript preparation; manuscript editing; and manuscript review.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2017. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- Luo S, Wu L, Zhou J, et al. Tracheobronchopathia osteochondroplastica: two cases and a review of the literature. *Int J Clin Exp Pathol* 2015;8:9681–6.
- Wang N, Long F, Jiang S. Tracheobronchopathia Osteochondroplastica: two cases reports and review of Literature. *Medicine* 2016;95:e3396.
- Agarwal A, Klair J, Joiner A, et al. Nodular trachea: tracheobronchopathia osteochondroplastica. *BMJ Case Rep* 2015;2015:bcr2015209860.
- Sant'Anna CC, Pires-de-Mello P, Morgado MF, et al. Tracheobronchopathia osteochondroplastica in a 5 year-old girl. *Indian Pediatr* 2012;49:985–6.
- Dincer HE, Dunitz JM. Tracheobronchopathia osteochondroplastica and selective IgA deficiency. *J Bronchology Interv Pulmonol* 2012;19:54–6.
- Fois AG, Arcadu A, Santoru L, et al. Tracheobronchopathia Osteochondroplastica: a rare case report of a non-smoker and non-atopic patient, with a long history of wheezing since childhood. *Multidiscip Respir Med* 2016;11:16.
- Willms H, Wiechmann V, Sack U, et al. Tracheobronchopathia osteochondroplastica: a rare cause of chronic cough with haemoptysis. *Cough* 2008;4:4.
- Leske V, Lazor R, Coetmeur D, et al. Tracheobronchopathia osteochondroplastica: a study of 41 patients. *Medicine* 2001;80:378–90.
- Laine M, Elfihri S, Kettani F, et al. Tracheobronchopathia osteochondroplastica associated with skin Cancer: a case report and review of the literature. *BMC Res Notes* 2014;7:637.
- Warner MA, Chestnut DH, Thompson G, et al. Tracheobronchopathia osteochondroplastica and difficult intubation: case report and perioperative recommendations for anesthesiologists. *J Clin Anesth* 2013;25:659–61.
- Uchimura K, Yamasaki K, Yatera K, et al. Multiple Tracheobronchial Polyposis caused by Tracheobronchopathia Osteochondroplastica. *Intern Med* 2016;55:3165–7.
- Nielsen SW, Stevens JR, Dion GR, et al. Dyspnea, Dysphonia, and Cough: varied presentations of Tracheobronchopathia Osteochondroplastica. *Ann Otol Rhinol Laryngol* 2015;124:829–33.
- Simmons C, Vinh D, Donovan DT, et al. Tracheobronchopathia osteochondroplastica. *Laryngoscope* 2016;126:2006–9.
- Ribeiro GM, Natal MR, Silva EF, et al. Tracheobronchopathia osteochondroplastica: computed tomography, bronchoscopy and histopathological findings. *Radiol Bras* 2016;49:56–7.
- Hayes D. Tracheopathia osteoplastica misdiagnosed as asthma. *J Asthma* 2007;44:253–5.
- Jindal S, Nath A, Neyaz Z, et al. Tracheobronchopathia osteochondroplastica--a rare or an overlooked entity? *J Radiol Case Rep* 2013;7:16–25.
- Raess PW, Cowan SW, Haas AR, et al. Tracheobronchopathia osteochondroplastica presenting as a single dominant tracheal mass. *Ann Diagn Pathol* 2011;15:431–5.
- Chroneou A, Zias N, Gonzalez AV, et al. Tracheobronchopathia osteochondroplastica. An underrecognized entity? *Monaldi Arch Chest Dis* 2008;69:65–9.
- Brandén E. A 20-year follow-up of a case with tracheobronchopathia osteochondroplastica. *J Bronchology Interv Pulmonol* 2013;20:84–6.
- Danckers M, Raad RA, Zamuco R, et al. A complication of tracheobronchopathia osteochondroplastica presenting as acute hypercapnic respiratory failure. *Am J Case Rep* 2015;16:45–9.
- Jabbarjarjani HR, Radpey B, Kharabian S, et al. Tracheobronchopathia osteochondroplastica: presentation of ten cases and review of the literature. *Lung* 2008;186:293–7.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow