Laryngo-tracheo-bronchial stenosis in a patient with primary pulmonary amyloidosis: a case report and brief review

[Une sténose laryngo-trachéo-bronchique chez un patient atteint d'amylose pulmonaire primaire : une étude de cas et une brève revue]

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Purpose: To report a case of lower respiratory tract obstruction occurring in a patient with primary pulmonary amyloidosis and discuss anesthetic management.

Clinical features: A 53-yr-old man was referred to our institution for microlaryngoscopy and laser treatment of the larynx. He presented with a five-year history of primary laryngo-tracheo-bronchial amyloidosis and symptoms consistent with narrowing of the conducting airways. General anesthesia was induced with iv propofol 150 mg and remifentanyl 50 µg. Mivacurium 20 mg provided muscle relaxation for endotracheal intubation. Following endotracheal intubation, the airway became obstructed and patient ventilation impossible. The endotracheal tube was removed and a Dedo laryngoscope inserted. Gas exchange was maintained using supra-glottic jet ventilation via a distal port of the laryngoscope. Rigid bronchoscopy showed tissue partially obstructing the lumen of the lower trachea. This was removed and the airway appeared patent. At the end of the case, a further episode of lower airway obstruction occurred requiring reinsertion of the laryngoscope and resumption of jet ventilation. Extensive debridement through the bronchoscope was required before adequate ventilation could be restored. Some days later when the patient's condition deteriorated again and he required further debridement of the trachea and insertion of a tracheostomy, guide wires were positioned in the femoral vessels in the event that cardiopulmonary bypass was required for gas exchange.

Conclusions: Primary laryngo-tracheo-bronchial amyloidosis is a recurrent disease, requiring repetitive surgical procedures. Airway compromise can be a persistent problem. Awareness of this uncommon disease process and its presentation may serve to caution the anesthesiologist presented with this type of case.

Objectif : Présenter un cas d'obstruction des voies respiratoires inférieures chez un patient atteint d'amylose pulmonaire primaire et discuter de la démarche anesthésique.

Éléments cliniques : Un homme de 53 ans a été dirigé vers notre institution pour une microlaryngoscopie et un traitement au laser du larynx. Il souffrait depuis cinq ans d'amylose laryngo-trachéo-bronchique primaire et de symptômes caractéristiques d'un rétrécissement des voies aériennes inférieures. L'anesthésie générale a été induite avec 150 mg de propofol iv et 50 µg de rémifentanyl. Une dose de 20 mg de mivacurium a permis le relâchement musculaire favorable à l'intubation endotrachéale. Après l'intubation, les voies aériennes se sont obstruées et la ventilation est devenue impossible. Nous avons retiré le tube endotrachéal et inséré un laryngoscope Dedo. Les échanges gazeux ont été maintenus avec la ventilation en jet supraglottique par une ouverture distale du laryngoscope. La bronchoscopie rigide a montré une obstruction tissulaire partielle de la lumière trachéale basse. Le tissu a été enlevé et la trachée est apparue dégagée. Après quoi, un autre épisode d'obstruction est survenue et a nécessité la réinsertion du laryngoscope et la reprise de la ventilation en jet. Un débridement étendu, fait en passant par le bronchoscope, a été nécessaire pour restaurer une ventilation adéquate. Quelques jours plus tard, l'état du patient s'est de nouveau détériorée et a requis un autre débridement de la trachée et l'insertion d'une trachéostomie ; un guide métallique a alors été mis en place dans les vaisseaux fémoraux au cas où la circulation extracorporelle serait exigée pour les échanges gazeux.

Conclusion : L'amylose laryngo-trachéo-bronchique primaire est une affection récurrente qui exige des interventions chirurgicales répétées. Une obstruction des voies aériennes peut persister. La connaissance de ce processus morbide rare et de sa manifestation peut servir de mise en garde à l'anesthésiologiste confronté à ce genre de situation.

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P RIMARY laryngo-tracheo-bronchial amyloidosis is an uncommon presentation of the heterogeneous group of disorders known as amyloidosis. Unlike systemic amyloidosis where there is widespread interstitial pulmonary amyloid deposition, primary pulmonary amyloidosis is characterized by the deposition of amyloid predominantly along the laryngo-tracheo-bronchial tree and uncommonly in the interstitial pulmonary tissue.^{1,2}

We describe a case of lower respiratory tract obstruction occurring in a patient with laryngo-tracheo-bronchial amyloidosis who presented for microlaryngoscopy and laser excision of laryngeal amyloid.

Case report

A 53-yr-old man was referred to our institution for microlaryngoscopy and laser treatment of the larynx. He had a five-year history of primary pulmonary amyloidosis with involvement of the larynx, trachea and bronchi. He had been assessed and followed up at regular intervals over the preceding five years. He complained of associated exertional dyspnea resulting in a limitation of some physical activities. He had also complained of hoarseness for a number of years. His medical and surgical history was otherwise unremarkable. Computerized tomography (CT) of the thorax prior to admission demonstrated extensive thickening and calcification of the trachea and proximal bronchial walls with associated narrowing of the lumen. Precise measurements were not reported. The patient was scheduled for laryngoscopy and laser treatment of amyloid lesions in the larynx in an effort to improve phonation.

Routine monitoring during the procedure consisted of electrocardiography, pulse oximetry, PetCO₂ and blood pressure monitoring. General anesthesia was induced with *iv* propofol 150 mg and remifentanyl 50 µg. Mivacurium 20 mg provided muscle relaxation for endotracheal intubation.

A 5.5-mm laser shielded endotracheal tube was inserted initially into the trachea but following insertion of the endotracheal tube, the airway became obstructed and ventilation impossible. The endotracheal tube was removed and a Dedo laryngoscope was inserted: this is an anterior commissure suspension laryngoscope, it has a straight enclosed barrel, a recessed light and a distal port which allows for supraglottic jet ventilation while also allowing direct visualization of the larynx.

Although chest wall movement was limited, gas exchange was sufficient so as to maintain oxygen saturation. Anesthesia was maintained using *iv* infusions of propofol (0.2 mg·kg⁻¹·min⁻¹) and remifentanyl

FIGURE 1A View at endoscopy of the laryngeal inlet. Arrows indicate amyloid infiltration of the vocal cords and supraglottic region. The normal anatomy of the glottis and supraglottic region has been distorted by amyloid infiltration.

FIGURE 1B Trans-axial computed tomography view of the thorax at the level of the thoracic inlet showing a thickened tracheal lumen with associated narrowing of the lumen. Arrows indicate circumferential submucosal amyloid infiltration.

(0.25 µg·kg⁻¹·min⁻¹) and muscle relaxation using mivacurium in increments of 0.1 mg·kg⁻¹. Direct inspection with a rigid bronchoscope showed the lower trachea to be reduced to an opening of 4 to 5 mm in diameter. The obstruction was due to flaps of mucosal tissue crossing the trachea and occluding it both anteriorly and posteriorly. Using a cup forceps, some of this material was removed and although this initially created further flaps, after removing more tis-

sue, the airway appeared patent. The Dedo laryngoscope was removed and a laryngeal mask airway (LMA) was inserted. It was not possible, however, to ventilate the lungs using the LMA, therefore the laryngoscope was reinserted and jet ventilation resumed. Maintaining adequate ventilation however was difficult. In an effort to improve ventilation the trachea was intubated using a 5-mm plain, cuffed endotracheal tube. With the endotracheal tube in place however, ventilation was impossible and this resulted in a brief period of hypoxia with the patient's oxygen saturation dropping to around 50% while the Dedo laryngoscope was reinserted and jet ventilation recommenced.

With the assistance of a thoracic surgeon, more tissue was removed using a biopsy forceps to debulk the trachea until an 8-mm endotracheal tube could be accommodated. The patient was then transferred to the main postoperative recovery area where he was ventilated mechanically overnight and extubated the following morning.

During the following week, the patient became increasingly dyspneic and was unable to lie flat. This was most likely due to a combination of aspiration of blood and obstruction of the trachea with amyloid tissue. He was started on a helium-oxygen mixture (80:20) at 6 L·min⁻¹ via a face mask in order to decrease the work of breathing and returned to the operating room for repeat bronchoscopy and elective fashioning of a surgical tracheostomy to improve his symptoms. Prior to induction of anesthesia, invasive arterial monitoring was instituted and, as a precaution, guide wires were positioned in the femoral vessels to expedite access in the event that lung ventilation became impossible and cardiopulmonary bypass was required for gas exchange. Anesthesia was induced with fentanyl 50 µg and propofol (1000 µg·kg⁻¹·min⁻¹) and remifentanyl (1 µg·kg⁻¹·min⁻¹) infusions. Anesthesia was maintained with propofol (80 µg·kg⁻¹·min⁻¹) and remifentanyl (0.25 µg·kg⁻¹·min⁻¹) infusions. Muscle relaxants were not administered. Ventilation was maintained using jet ventilation and endotracheal intubation was avoided. Using the bronchoscope, tracheal tissue was removed from the subglottic area down to the carina. A tracheostomy was then fashioned and a tracheostomy tube inserted at the level of the fourth tracheal ring. Using a flexible bronchoscope, the distal end of the tube was confirmed to be in a satisfactory position immediately above the carina. The remainder of the procedure was uneventful and the patient was transferred to the postanesthetic care unit for observation.

Discussion

Primary pulmonary amyloidosis may be isolated to the larynx, the tracheobronchial tree or the laryngotrachea. More unusually, they may be extensive involvement of the entire laryngo-tracheo-bronchial tree.³ Laryngeal amyloidosis represents 0.5 to 1% of benign laryngeal disease. It occurs predominantly in the supraglottic area and presents with hoarseness, stridor, or less commonly, choking or dyspnea on exertion.⁴ Tracheobronchial amyloidosis typically presents after the fifth decade with deposits producing diffuse or multifocal plaques, or less commonly, solitary masses mimicking endobronchial tumours. Diffuse involvement of the tracheobronchial tree usually results in symptoms of cough, stridor and hemoptysis whereas localized masses result in signs and symptoms related to airway obstruction. Involvement of the tracheobronchial tree may be radiographically evident as atelectatic changes distal to the site of obstruction.⁵

Treatment of symptomatic laryngeal and tracheobronchial amyloidosis usually involves either stenting of the narrowed airway segments, piecemeal resection of tumour masses, open thoracotomy or more recently laser ablation therapy.⁶

As primary pulmonary amyloidosis is a rare occurrence, there is little information available concerning perioperative anesthetic management. Pulmonary function tests (PFTs) allow an objective assessment of the degree of airway obstruction and serial PFTs allow monitoring of disease progression. Changes in PFTs will usually necessitate radiological clarification with CT scanning to show the precise distribution of lesions.⁷ Tracheobronchial narrowing and wall thickening can be demonstrated and measurements of the length and internal diameter of the flow limiting segments involved can be taken.⁸

The cause of the airway obstruction during the initial operative procedure was most likely due to disturbance of the tumour during intubation of the trachea creating a flap obstruction. Due to the friable 'waxy' nature of amyloid deposits and their occurrence often at multiple sites, there is an increased risk of airway obstruction or bleeding either on intubation or on instrumentation of the airway.

Given the extent of tumour infiltration and associated airway narrowing in this case, avoidance of muscle relaxants and maintenance of spontaneous ventilation may have been a more appropriate technique.

The decision to cannulate the femoral vessels for the second procedure was undertaken in expectation of a similar intraoperative event occurring again and cardiopulmonary bypass being necessary for gas exchange. This procedure was previously reported as

part of the anesthetic management of an anterior mediastinal mass, where the possibility of precipitating airway obstruction existed.⁹

In summary, we have reported a case of intraoperative lower respiratory tract obstruction in a patient with primary laryngo-tracheo-bronchial amyloidosis. This is a recurrent disease and despite repeated treatments with bronchoscopic procedures, airway compromise can be a persistent problem. Awareness of this uncommon disease process and its presentation may serve to caution the anesthesiologist presented with a similar case.

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