Case Report

Primary Tracheal Schwannoma Masquerading as Bronchial Asthma: A Case Report and Review of Literature

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Abstract

We report an unusual case of primary tracheal schwannoma and related treatment challenges in a 30-year-old male who was symptomatic with breathlessness on exertion for the last one year. On computed tomography (CT) an intra-tracheal mass lesion was detected. Fibreoptic bronchoscopy revealed an intra-tracheal polypoid mass obstructing more than 80% of the lumen and histopathology of bronchoscopic biopsy diagnosed it as benign nerve sheath tumour. It was managed effectively with electrocautery through fibreoptic bronchoscopy. **[Indian J Chest Dis Allied Sci 2019;61;43-45]**

Key words: Tracheal schwannoma, Fixed airway obstruction, Electrocautery.

Introduction

Primary tracheal malignancies are very rare tumours and their incidence is approximately 0.1 in every 100,000 persons per year.[1] The differential diagnosis of a tracheal mass comprises of certain malignant (squamous cell carcinoma, adenoid cystic carcinoma, and carcinoid) as well as benign tumours, such as papilloma, haemangioma, hamartoma, and neurilemmomas and neurofibromas.[2] Squamous cell carcinoma is the most common cause of intra-tracheal mass followed by adenoid cystic carcinoma and together these account for about two-thirds of adult primary tracheal tumours.[3] Benign tracheal tumours are less common than the malignant ones and among these squamous papilloma, leiomyoma and haemangiomas are most frequently seen. We describe a case of primary tracheal schwannoma which is extremely rare tumour in this report. *To the best of our knowledge,* only a single case has been reported till date in the Pubmed indexed Indian journals.

Case Report

A 32-year-old male, non-smoker and without any significant past medical history initially reported to a peripheral health centre with complaints of cough, breathlessness on exertion associated with wheezing for the last eight months. There were no aggravating or relieving factors. There was no history of fever, haemoptysis, joint pains, skin rashes, nasal symptoms and weight loss. He was initially diagnosed to have bronchial asthma in a peripheral health centre and was treated with inhaled corticosteroids and bronchodilators without any relief.

On examination, he was dyspnoeic at rest which aggravates in supine position and had noisy breathing suggestive of stridor, indicating a central airway obstruction. Auscultation of the chest revealed bilateral fixed inspiratory and expiratory wheeze. Other systemic examination was unremarkable. Haematological and biochemical investigations were within normal limits. Chest radiograph showed an opacity involving the lower part of the trachea. Spirometry demonstrated severe airway obstruction, there was flattening of both inspiratory and expiratory flows on flow–volume loop graph, indicative of fixed upper airway obstruction with a forced expiratory volume in one second (FEV_1) of 0.97 litres (26% of predicted), forced vital capacity (FVC) 2.67 litres (58% of predicted) and FEV_1/FVC ratio was 36.3%. Arterial blood gas values on room air were normal. Contrast enhanced computed tomography of the chest showed intra-tracheal rounded mass (12.8 mm × 20 mm) 9 cm below the cords with no extra-tracheal extension (Figure 1). Flexible bronchoscopy showed a large, rounded pedunculated mass (Figure 2), arising from the left tracheal wall in the lower trachea, occluding more than 80% the lumen.

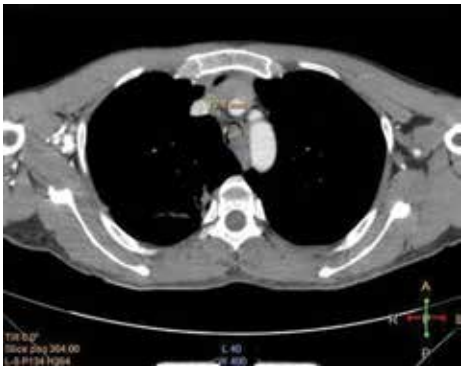

Figure 1. Contrast enhanced computed tomography of the chest showing an intra-tracheal rounded mass (12.8mm×20mm in diameter), 9cm below the vocal cords with no extra-tracheal extension.

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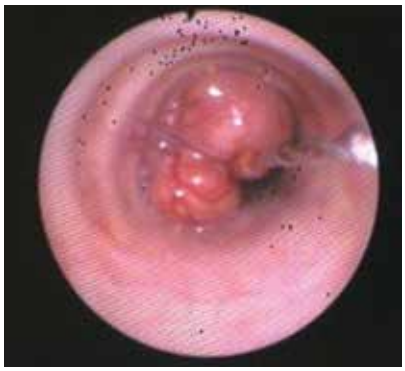

Figure 2. Fibreoptic bronchoscopy revealed a large rounded pedunculated mass arising from the left tracheal wall, in the lower trachea, occluding more than 80% of the lumen.

Bronchoscope was negotiated distally around the mass and distal to this endotracheal mass tracheobronchial tree was found to be normal. Endotracheal brushing, multiple biopsies were taken from the endotracheal mass and bleeding was controlled with the application of argon plasma coagulation. Histopathology of endotracheal biopsy specimens revealed a spindle cell tumour with interspersed inflammatory cell infiltrate without any mitosis. This was thought initially to represent a myofibroblastic tumour. However, on subsequent immunohistochemical testing, tumour stained strongly positive with antibodies to S100 protein and vimentin, and was negative for SMA, cytokeratin indicative of a nerve-sheath tumour or schwannoma. Ki-67 labelling index (ki-67 Li), a marker of proliferation was less than 1% which confirmed the benign nature of the tumour. As there was no extra-tracheal extension of the tumour and due to its proximity to the carina (2cm from the carina) patient was taken up for endotracheal resection of the tumour. Anesthetic considerations were discussed with cardiothoracic anesthetist, administering muscle relaxants were considered to be risky in this patient because it may cause muscle laxity and has potential to aggravate obstruction in already compromised airway leading to total closure of the airways. Moreover, endotracheal intubation or tracheostomy could not be of any use in this patient due to the distal location of the tumour.

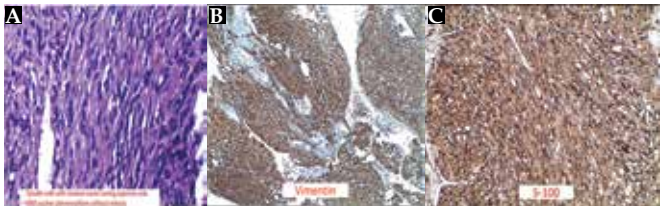

Figure 3. Photomicrographs showing (A) spindle cell tumour (Haematoxylin and Eosinx200) (B) on immunohistochemistry it stained strongly positive with antibodies to vimentin (IHCx100) and (C) S100 protein (IHCx200)

Considering the risks associated with administration of muscle relaxants, fibreoptic bronchoscopy was planned instead of rigid bronchoscopy. However, vascular cannulation was also established for venovenous extra-corporeal membranous oxygenation (ECMO) to tide over any emergency arising due to total airway occlusion during the procedure. Under conscious sedation electrocautery electrode was introduced through flexible bronchoscope and the patient underwent spray coagulation of the tumour using electrocautery electrodes (Figure 4 A). Whole of the tumour was coagulated leaving fully patent trachea (Figure 4 B) and remaining bits of tissue was removed using biopsy forceps. During bronchoscopic procedure small amount of bleeding occurred which was controlled effectively with the application of argon plasma laser. Patient had significant symptomatic improvement post coagulation.

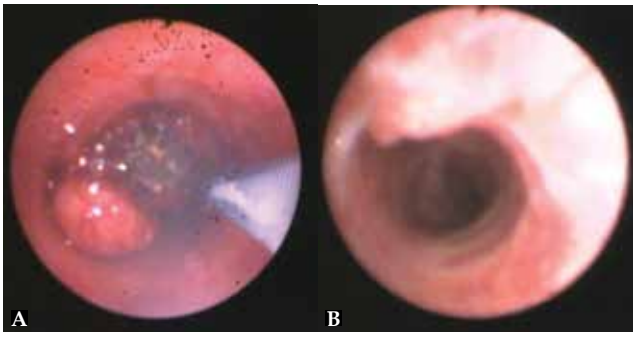

Figure 4. Fibreoptic bronchoscopy guided electrocoagulation of the tumour and (B) post-procedure, patent tracheal lumen

Discussion

Primary tracheal tumours are quite rare. A high level of clinical acumen is required to diagnose tracheal tumours. Diagnosis of tracheal tumours is to be considered in patients presenting with breathlessness unresponsive to bronchodilators and showing characteristic flattening of flow-volume loops on spirometry, as was in our case. Thorough knowledge of flow-volume loops on spirometry at the primary care level may help in avoiding delay in diagnosis and misinterpretation of tracheal tumours as asthma. As per World Health Organization (WHO) classification tracheal tumours are classified accordingly to their cell origins: malignant epithelial tumours, benign epithelial tumours, neuroendocrine tumours, malignant soft tissue tumours and benign soft tissue tumours comprising of lipoma, leiomyoma, haemangioma, nerve sheath tumours: schwannoma and neurofibroma.[4] Primary neurogenic tumours of the trachea (schwannomas or neurilemmomas) are extremely rare. Straus and Guckien were the first ones to describe tracheal neurilemmomas in 1951.[5] The presenting symptoms are usually chronic dry cough, and progressive exertional dyspnoea that are not relieved with medication. Wheezing is usually generalised, and the presence of stridor may help in the diagnosis. Clinically it is often mis-diagnosed as asthma, and the diagnosis is usually delayed for an average of 10 to 15 months.[6] In our patient also initial diagnosis of asthma was made and was prescribed bronchodilators which failed to show any relief. Schwannoma occurs most frequently in the distal of the trachea, followed by decreasing order in the proximal and

middle thirds. Tissue for the confirmation of diagnosis is obtained by the bronchoscopy. Amongst the neurogenic tumours of the trachea: neurofibromas are usually multiple, unencapsulated lesions showing proliferation of all the elements of the nerve including the Schwann cells, and nerve axons.[7] However, neurilemmomas (schwannomas) are usually single, encapsulated lesions composed of schwann cells attached to a nerve but containing no axons. Microscopically, schwannomas look as spindle cells tumour in which cells are arranged in compact fashion as well as in loose myxoid fashion without any capsular invasion or mitoses. On immunohistochemical stains these tumours stain positively for S-100 protein and vimentin differentiating them from the smooth muscle tumours.[8] These were the findings in our case also. Treatment modalities of schwannoma comprises of endoscopic excision, sleeve resection and tracheal resection.[9] There is no consensus as to the optimal management of tracheal schwannoma because of its rarity. The choice of treatment is influenced by the clinical presentation and whether the tumour is pedunculated or sessile, the risks involved in the tracheal resection, and the presence or absence of an extra-tracheal component. Endoscopic resection is the preferred treatment if tumour is pedunculated and there is no extra-tracheal component.[10] Endoscopic resection can be accomplished with Nd-YAG laser, argon plasma coagulation, cryotherapy and electrocautery. Our case met the criteria for endoscopic resection of the tumour and was resected completely using electrocautery.

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