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Case Report

Solitary intratracheal neurofibroma: a rare tracheal tumor

Misauq Mazcuri*, Ambreen Abid, Tanveer Ahmad

Department of Thoracic surgery, Jinnah Postgraduate Medical Centre, Karachi, Sindh, Pakistan

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***Correspondence:**
Dr. Misauq Mazcuri,
E-mail: misauq229@gmail.com

ABSTRACT

Intratracheal neurofibromas are rare benign tumors with a tendency to transform into malignant form. Intratracheal neurofibroma remain asymptomatic and are incidentally found during routine radiography or may present with wheeze, cough and lower respiratory tract infection. We recently operated a 38 year old gentleman, who presented with progressive shortness of breath and chronic cough for two years. Examination was normal. CT identified a supracarinal, intratracheal mass. Rigid bronchoscopy revealed a hyper vascular mass, attached to membranous trachea. We performed resection anastomosis via right posterolateral thoracotomy. Histopathology revealed neurofibroma. Patient recovered well and was discharged after eight days.

Keywords: Intratracheal neurofibroma, Resection anastomosis, Membranous trachea, Supracarinal

INTRODUCTION

Primary neurogenic tracheal tumors are extremely uncommon making up to 0.5% of all primary tracheal tumors.[1,2] Intratracheal neurofibromas are rare entity and tend to develop from proliferation of schwann cells or fibroblast cells.[2,3] They can occur at any age with more predilections in males and have tendency to transform into malignant variant.[1,2] Literature reports only 28 cases of tracheobronchial neurofibroma.[2] Here we report a case of intratracheal neurofibroma treated with resection anastomosis via right posteriolateral thoracotomy.

CASE REPORT

A 38 year old male, ex-smoker, presented with complains of shortness of breath, dry cough and fever for the last two years. Symptoms were gradual in onset and progressively worsened over the last six months. He was treated for asthma with bronchodilators and corticosteroids but developed high grade fever and productive cough with shortness of breath, repeatedly. Work up for pulmonary tuberculosis and heart failure was negative. CT was done showing a well-defined hypodense lesion 1.8×1.6×1.9 cm in the lumen of distal trachea, arising from right posterolateral wall of trachea and extending into carina causing marked luminal narrowing and old changes representing recurrent bilateral pneumonia (Figure 1). His general physical and cardiovascular examinations were normal except for rhonchi. Pulmonary functions revealed pattern of fixed upper airway obstruction. Pre-operative laboratory values were normal. We planned to perform rigid bronchoscopy followed by surgery in the same stage.

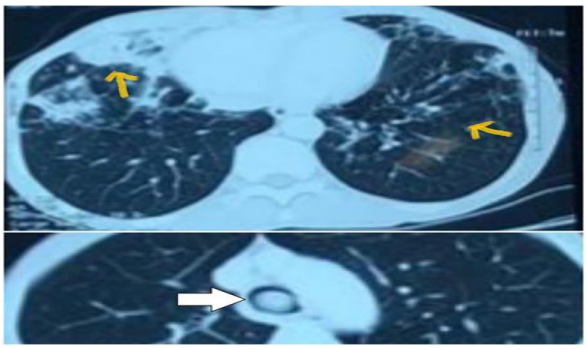

Figure 1: CT scan showing intratracheal tumor with bilateral pneumonic changes and fibrosis.

Procedure

Rigid bronchoscopy was done to visualize the tumor and confirm patency of the left bronchus. Patient was intubated with a size six single lumen endotracheal tube to bypass the tumor into the left bronchus. Right posterolateral thoracotomy was done and chest entered through the bed of fifth rib. Carina was identified. Tumor was localized using palpation and confirming with air aspiration. Trachea was incised just below the tumor attachment and the distal end of trachea was secured with silk stays. At the same time the endotracheal tube was replaced by a cross field armor tube size seven in the distal trachea for ventilation. The proximal extent of the tumor was identified and the involved trachea about 3.5 cm was excised taking care not to injure the underlying esophagus. Lateral dissection was confined to the resected segment of the trachea. Posterior wall of trachea was repaired with interrupted 3/0 vicryl and the armor tube was replaced with a size seven endotracheal tube. Anterior layer was repaired with interrupted 3/0 vicryl. The anastomosis integrity was confirmed with ventilation under saline. Para tracheal lymph nodes were excised for histopathology. Thoracotomy was closed conventionally. Guard sutures were applied and patient was extubated on table. Biopsy showed solitary neurofibroma with clear resected tumor margins.

Grossly the tumour measured 2.6×1.9×1.6 cm and clear margins. Histopathology revealed a neoplastic lesion in a loose myxoid background, composed of spindle shaped cells with moderate cytoplasm and round to oval nuclei with no significant pleomorphism, necrosis or mitotis. Lymph nodes only showed reactive changes (Figure 2).

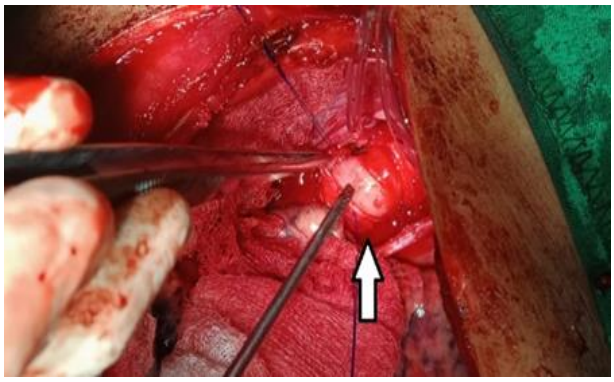

Figure 2: Arrow representing tumor.

DISCUSSION

Primary tracheal tumors are rare, accounting for up to 0.2% of all respiratory and 1% of overall malignancies, with 90% being malignant.[1,4] Common benign tumors of trachea and bronchus are papilloma, fibroma, hemangioma followed by neurogenic tumors.[3,5]

Benign neurogenic tumors include neurilemmoma, neurofibroma, and neuroma.[3,5] Neurofibroma develop from schwann or perineural cells They are usually multiple and occasionally associated with neurofibromatosis (NF) type 1.[3,6] In association with NF-1, it has around 10-12% chances of malignant transformation.[2,5,6] Neurofibroma may arise from any nerve in the mediastinum, lung or airway with trachea, being the most common site.[5,6] Neurilemmoma are more common intra tracheal neurogenic tumors than neurofibroma.[5] On basis of growth pattern neurofibroma are classified into solitary, plexiform or diffuse.[3]

Neurofibromas grow slowly and often remain asymptomatic for years or may present with symptoms of airway obstruction depending on size and location.[7] Symptoms include cough, dyspnea, wheezing with occasional hemoptysis, chest pain and history of recurrent chest infections.[2,4,5]

Chest X-ray is of less diagnostic yield and may lead to delayed diagnosis. CT scan should be carried out in all symptomatic patients un responsive to medical management.[1,2] It helps define the pathology and its relation to other structures.[5,8] Neurofibroma may be seen as a hypodense mass with enhancement on contrast study.[7] Bronchoscopy certainly remains gold standard for assessment, diagnosis and restorative purposes.[1,4]

Treatment for tracheal tumors depends on size, location and symptoms.[3,5] Various management methods include endoscopic resection techniques such as laser cauterization or snaring, radiotherapy and surgery.[2,3,4,9] Endobronchial methods were considered first therapy for neurofibromas, but failed to achieve complete clearance.[1,2] Surgery offers not only an early control of airway but also definitive cure for benign and low grade tumors and histopathological clearance.[2-4]

We performed rigid bronchoscopy to assess the patency of airway, anatomical attachment of tumour with trachea and its distance from carina. Our goals for surgical resection were to gain early control of airway and to prevent any untoward complications like hemorrhage leading to failure of ventilation and desaturation.[2,6] In another recent report by Angeliki et al surgical resection with R0 intent is favored to prevent any local recurrence or metastatic spread.[10] Resection with tracheal mobilization with end to end anastomosis remains the procedure of choice.[2,8]

CONCLUSION

Neurofibroma is a rare tracheal tumor with malignant potential. Recent onset of asthma like symptoms which fail to resolve with treatment should be investigated for an intratracheal tumor. Radiological and endoscopic investigations should be done with early referral to surgical department.

Bronchoscopic procedure can be performed in selective cases but surgical resection ensures better control of airway, clear margins and a long term cure.

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