Original article

RARE TRACHEAL TUMOR MASQUERADING AS ASTHMA

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ABSTRACT

BACKGROUND: Rare Tracheal Tumor is often misdiagnosed as asthma and is treated with inhaled steroids and bronchodilators without resolution.

DESIGN: Retrospective descriptive observational study.

PLACE AND DURATION: Thoracic Surgery Unit, Post Graduate Medical Institute Lady Reading Hospital, Peshawar from Jan 2002 to Dec 2009.

MATERIAL AND METHODS: Computerized clinical data of 9 cases of tracheal tumor was retrospectively analyzed over a period of 7 years. Patients with all ages, sex, diagnosed case of operable polypoidal tracheal tumor and with the clinical presentation of asthma were included in the study. Inoperable tracheal tumor, medically unfit patients and non polypoidal tracheal tumor was excluded from the study. In all cases tumor was removed bronchscopically. Post operative follow-up was done for six months with the help of bronchoscopy and computed tomography at 3 month interval.

RESULTS: Out of 9 cases, 6 were female and 3 were male, age group was 20 to 40 years with a mean age of 28 years. Biopsy showed spindle cell tumor in 4 cases and benign polyp in 5 cases. During 6 months follow-up, recurrence was observed in 2 cases of spindle cell tumor of the trachea. There was no mortality and morbidity was 11.1% including postoperative pneumonia in one case.

CONCLUSION: It is critical to bear in mind that not all wheezes are asthma and if a patient does not respond to appropriate anti-asthma therapy, localized obstructions should be ruled out before establishing the diagnosis of asthma.

KEY WORDS: Tracheal tumor, Polyp, Spindle cell tumor, Asthma.

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INTRODUCTION:

Tracheal neoplasms occur infrequently, accounting for less than 1% of all malignancies.[1,2,3]. The inclusion of tracheal tumors in the differential diagnosis of adult-onset dyspnoea and wheeze is even more uncommon than the tumours themselves. Most authors reporting this condition have remarked on the delay in accurate diagnosis. Tracheal tumours frequently present with symptoms suggestive of asthma or chronic bronchitis and the patients are usually initially managed accordingly. The symptoms may be insidious in onset and the tumour may occlude three-quarters of the lumen before symptoms are reported. The single most common symptom is wheeze, but cough, dyspnoea and haemoptysis are frequent. Pain, weight loss and malaise are unusual[4]. When upper airways obstruction is present, a large reduction of expiratory flow occurs at high lung volumes only, so that the FEV1 is almost normal. There is thus a disparity between PEFR and FEV1, values, and the ratio FEV1 /PEFR is therefore often increased. The inappropriate PEFR value for the FEV1, value may serve to alert the physician to the possibility of upper airways obstruction. The routine PA chest radiograph is classically normal. If the 'asthmatic' patient's progress is less than satisfactory, the physician should consider obstructing lesions of the major airways. This should involve more probing investigations including high-penetration chest radiography, tracheal tomography and examination of flow-volume loops. Bronchoscopy is definitive.Adenoid cystic carcinoma probably accounts for up to one-third of malignant tracheal tumours [5]. It tends to occur more commonly in females and has not demonstrated association with cigarette smoking. It has a striking tendency to local spread, probably because of poor encapsulation. Distant metastases are a late event. Where possible, the treatment of choice is surgical but recurrence is frequent. Chemotherapy alone has not been useful [6], but good response after cobalt-60 therapy has been described[7] . Delay in diagnosis remains the principal problem. Only nine cases of operable polypoidal tracheal growth were presented to us over a period of seven years with initial diagnosis of asthma. This shows the rarity of tracheal tumor. The purpose of our study is to share our experience of rare tracheal tumor mimicking as asthma.

MATERIAL AND METHODS:

Computerized clinical data of 9 cases of tracheal tumor was retrospectively analyzed over a period of 7 years from January 2002 to December 2009 at Thoracic Surgery Unit , Postgraduate Medical Institute, Lady Reading Hospital Peshawar.Patients with all ages, sex, diagnosed case of operable polypoidal tracheal tumor and with the clinical presentation of asthma was included in the study. Inoperable tracheal tumor, medically unfit patients and non polypoidal tracheal tumor was excluded from the study. In all cases preoperative evaluation included pulmonary function tests, computed tomography and bronchoscopy. Tumor was removed bronchscopically in all cases by local improvisation, the technique is to insulate the biopsy forcep and resect the tracheal tumor by cautry via rigid bronchoscope, the resected tumor removed by using forcep and suction. Post operative follow-up was done for six months with the help of bronchoscopy and computed tomography at 3 month interval.

RESULTS:

Out of 9 cases, 6 were female and 3 were male; ages ranged between 20 to 40 years with a mean age of 28 years. Eight cases were labeled as bronchial asthma/COPD while one case was treated as brittle asthma. Average duration of symptoms was 3.3 months. The treatment included bronchodilators and steroids in all cases before referral to our unit. Computer tomography showed operable polypoidal tracheal growth and removed via rigid bronchoscope in all cases. Biopsy showed spindle cell tumor in 4 cases and benign polyp in 5 cases. During 6 months follow-up, recurrence was observed in 2/9 (23%) cases of spindle cell tumor of the trachea. There was no mortality and morbidity was 11.1% including postoperative pneumonia in one case.

DISCUSSION:

Tracheal tumours are rare and comprise only 1-2% of all lung tumours. Of the tracheal tumours, approximately 85% are carcinoid tumours, 10% adenoid cystic carcinomas and 5% mucoepidermoid tumours. Mixed tumours are rare. Of all primary tumors of the trachea, 80% are malignant; adenoid cystic carcinoma (ACC) and squamous cell carcinoma (SCC) are the most common, comprising two thirds of all tracheal tumors. The remainder is widely varied and includes both malignant and benign histotypes. Despite their rarity, their usually insidious onset often leads to a delay in diagnosis, making these potentially treatable lesions difficult to treat and often fatal. Benign tumors can arise from any of the tissues present in the trachea. Malignant tumors probably follow a carcinogenesis similar to that of lung cancers. Most of these tumors occur sporadically. Apart from squamous papillomas, which have been associated with viral infection, no consistent etiology has been found. Smoking is a known risk factor[2].In our 5 cases were diagnosed as squamous papilloma on histopathology.

Tracheal tumours are difficult to diagnose as they simulate other illnesses like bronchial asthma and chronic bronchitis[8, 9].The common symptoms include wheezing, cough, dyspnoea and haemoptysis[10, 11]. They, thus, mimic asthma closely. To compound the problem they may respond temporarily to bronchodilators and steroids, further delaying the real diagnosis[12]. Delay in diagnosis occurs because the pulmonary fields remain normal on a chest radiograph[2].In our study 8 cases were diagnosed as a case of bronchial asthma and one pt was treated as a case of brittle asthma before conclusion of polypoidal tracheal growth by computed tomography .The average duration of symptoms was 3.3 months and all the patients were on bronchodilator and steroids. One patient in our study diagnosed as a case of brittle asthma was also put on ventilator as well for some time because of respiratory failure secondary to occlusion of tracheobronchial tree by the tracheal tumor before referring our unit. After differentiating stridor from wheeze the computed tomography was done in all cases followed by bronchoscopy. The main reason for misdiagnosis of tracheal tumor from bronchial asthma in our setup was the failure to differentiate stridor from wheeze in the peripheral health unit, which further necessitates evaluation by computer tomography followed by bronchoscopy for final diagnosis and treatment.

One quarter of tracheal tumors are neither ACC nor SCC. This heterogeneous group of tumors has varying degrees of malignancy and includes both epithelial and mesenchymal histotypes. Unusual tumor types included carcinoid, mucoepidermoid, nonsquamous bronchogenic carcinoma, lymphoma, melanoma spindle cell sarcoma[13].In our study 4 cases were reported as spindle cell tumor on histopathology

Perineural and intraneural infiltration may explain high local tumour recurrence after resection[14]. In our study 2 of 4 cases of spindle cell tumor had local recurrence. Metastases occur late in the disease, with bone and lung being common sites. Tracheal resection with reconstruction carried out with pericardial or synthetic graft remains the treatment modality of choice for resectable tracheal tumours. Radiation may produce good palliative therapy. Use of radiotherapy in conjunction with chemotherapy has also been advocated. Chemotherapy alone has not been used. Endotracheal radioactive iridium insertion has shown dramatic palliative results in recurrence of tumour[15]. In our study 4 of 9 cases was reported as spindle cell tumor which were referred to oncologist for adjuvant therapy.

A high index of suspicion is needed to diagnose tracheal tumours. This entity needs to be kept in mind when one is confronted with asthma poorly responsive to therapy. In such cases, overpenetrated chest x-ray or fluoroscopy of trachea may help in clinching the diagnosis. Appropriate timely therapy has yielded remission for many years. Symptoms associated with tracheal tumors often mimic those of asthma. Flow-volume curves, however, distinguish them from asthma by a flattening of the expiratory or inspiratory limb.

Spindle cell tumor of the trachea is the rarest neoplasm of the lower respiratory tract [16-18]. In our study four cases were reported as a case of spindle cell tumor. A screening chest radiograph is often normal, but it can also identify lung metastases or synchronous primary tumors of the lung. Computed tomography is the most effective method to determine the extent of mediastinal spread of the tumor. Barium swallow is useful in the evaluation of posterior tracheal masses, but a “positive” barium esophagram is nonspecific and unable to distinguish esophageal invasion from external esophageal compression. The most important staging test is rigid bronchoscopy performed by the surgeon planning a resection. Regardless of the results of previous diagnostic bronchoscopies, it is absolutely essential that the operating surgeon perform a bronchoscopy to assess the extent of the lesion and the amount of uninvolved trachea available for reconstruction. The rigid bronchoscope is preferred because it allows more accurate measurements than the flexible bronchoscope. In addition, it allows more secure control of the obstructed airway and control of any bleeding that may occur following biopsy. The important measurements include the length of the trachea from the vocal cords to the carina, the distance from the cords to the top of the tumor, the length of the tumor, and the distance from the bottom of the tumor to the carina. Rigid bronchoscopy allows “coring out”of an obstructing tumor to allow clearing of a postobstructive pneumonia or to allow weaning from corticosteroids prior to definitive resection and reconstruction. In our study we have done coring out of the tracheal tumor via rigid bronchoscopy in all cases by local improvisation. The technique is to insulate the biopsy forcep and resect the tracheal tumor by cautry via rigid bronchoscope; the resected tumor is removed by using forcep and suction. Esophagoscopy should also be performed under the same anesthesia as the bronchoscopy if, based on the size and location of the tracheal

tumor, esophageal involvement is suspected.Mediastinoscopy is a valuable part of the assessment of some tracheal neoplasms[19].

CONCLUSION:

It is critical to bear in mind that not all wheezes are asthma and if a patient does not respond to appropriate anti-asthma therapy, localized obstructions should be ruled out before establishing the diagnosis of asthma.

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