

A CASE OF ENDOBRONCHIAL TUMOUR – NEUROFIBROMA**Dr. Prabhat B. Nichkaode***

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ABSTRACT

Among benign tracheobronchial neoplasms, neurofibromas of neurogenic origin are exceedingly rare. In a search of world literature, only few cases of endotracheobronchial neurofibromas were reported. We report another case of a 17 years female who presented to our hospital with the symptoms of recurrent fever, intermittent nonproductive cough and short of breath on exertion. Chest X ray shows total opacity on right side, tracheal shift on right and well compensated Left lung. Bronchoscopy revealed a round tumor obstructing the lumen of the right main bronchus. Computed tomography demonstrated a homogenous tumor located within the right main bronchus with obstructive pneumonitis of the right lung. Right posterolateral thoracotomy done. Bronchotomy done for excision of endobronchial tumour followed by end to end bronchoplasty with live pericardial flap. Endobronchial neurofibroma was confirmed by pathologic examination and immunohistochemical examination.

KEYWORDS: Endobronchial neurofibroma; Thoracotomy; Lung; Neurofibromatosis; Bronchus.**INTRODUCTION**

Neurofibroma belongs to the benign tumors and it is categorized as neuroectodermal tumor. Neurofibromas are most frequently found in the posterior mediastinum, their endobronchial localization is rather rare. Tumors arising from tracheobronchial trees are not common. Most of these tumors are malignant in origin, such as squamous cell carcinoma, adenoid cystic carcinoma, or malignant tumors of thyroid or lung with secondary tracheal or bronchial involvement.^[1] Benign tumors of tracheobronchial trees include papilloma, leiomyoma, hemangiomas and tumors of neurogenic origin.^[2] Benign neurogenic tumors originate from the peripheral nervous system, are composed of Schwann cells, and appear as neurilemmas or neurofibromas. Neurilemmas (Schwannomas) are single, encapsulated lesions attached to a nerve but containing no neurites.^[3]

Neurofibromas are usually multiple, not encapsulated, occur within the nerve sheath, and contain neurites. These two tumors can be found in the mediastinum, lung parenchyma, or within the tracheobronchial trees.^[4,5] The incidence of neurofibromas involving the trachea or bronchus is lower than that of neurilemmas. In a search of the world literature, there were only 23 cases of tracheobronchial neurofibromas reported.^[5]

CASE REPORT

A 17 years female, presented with complaints of recurrent history of fever with chills, nonproductive cough, short of breath on exertion, no hemoptysis. No other comorbid conditions and familial disease. General

examination was normal. No evidence of neurofibromatosis. On respiratory examination, no breath sounds on right side of chest. Routine blood examination was normal. Pulmonary Function tests also well compensated, Mild restrictive. FEV 1-1. 89 lt. Chest X ray shows total opacity on right side, tracheal shift on right and well compensated Left lung (Fig. 1). Bronchoscopy revealed a round tumor growth with a wide base arising from the posterior wall of the right main bronchus, about 5 mm distal to the carina, obstructing nearly the whole lumen of the right main bronchus (Fig. 2). Bronchoscopic brush cytology s/o Bronchial adenoma. Computed tomography demonstrated a homogenous tumor located within the right main bronchus with obstructive pneumonitis of the right lung (Fig 3, 4, 5). The patient underwent right-sided posterolateral thoracotomy. A firm yellowish-brown, well-defined tumor was found at the posteromedial wall of the right main bronchus. The lumen of the right main bronchus was completely occluded. There was no invasion of adjacent structure. We exposed the right main bronchus and bronchotomy done for excision of endobronchial tumour (Fig. 6). Endotracheal Tube of 4 Fr. passed in to distal Right Bronchus and Jet Ventilation given to see the Inflation of Lung. End to end bronchoplasty done followed by reinforcement of bronchoplasty with live pericardial flap (Fig. 7). Histopathologically, the tumor consisted of interlacing, waving, spindle-shaped cells strongly positive for S-100 protein and negative for desmin, and actin immunohistochemically, which confirmed the diagnosis of Benign Endobronchial neurofibroma. Fully

expansion of lung seen in 3 weeks on chest X ray. (Fig.8).

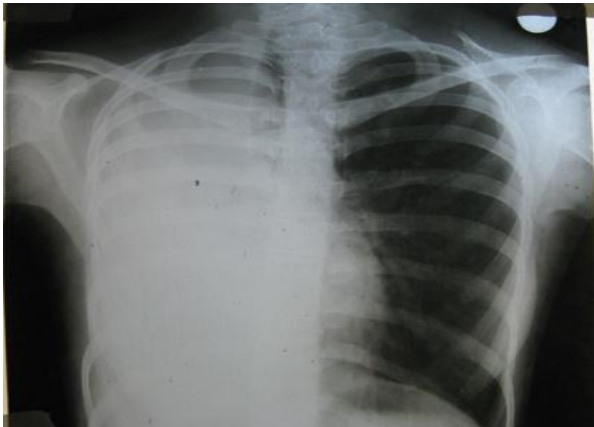


Fig. 1: X ray chest PA view – Complete Opacity Rt. Lung.

Trachea shifted to Right, Notching at right bronchus.



Fig. 2: Rt. Bronchial tumor Obstructing the main right bronchus.

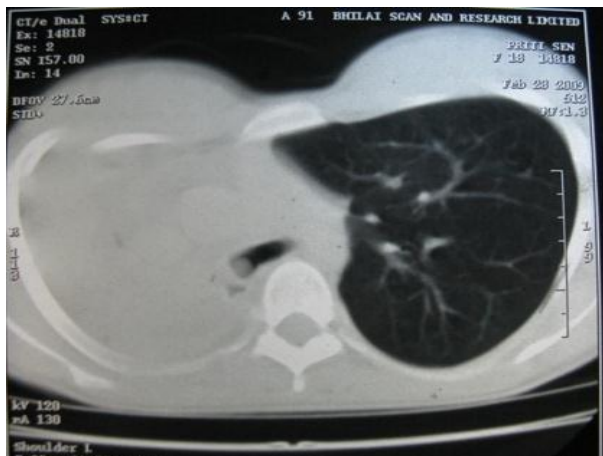


Fig 3: CT Scan– Showing Rt. Main Bronchus –mass.

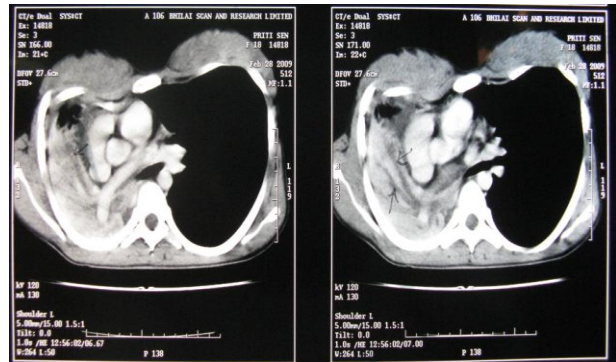


Figure 4: Rt. Bronchial Obstruction just a cm. away from Carina.

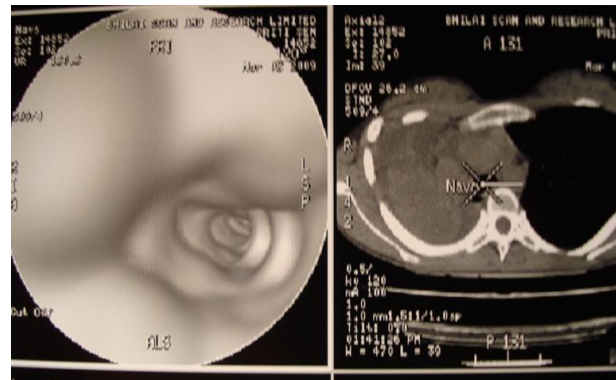


Fig. 5: Virtual Bronchoscopy – after CT Scan.



Fig. 6: Bronchotomy for excision of Intrabronchial tumour.

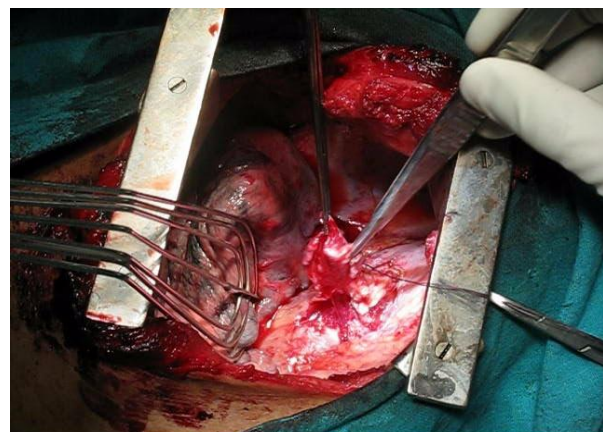


Fig.7: Live pericardial Flap- over Bronchoplasty sutures.

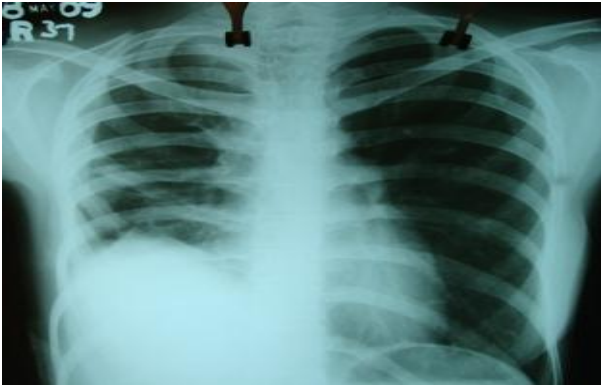


Fig. 8: Near complete expansion of right lung within 3 weeks.

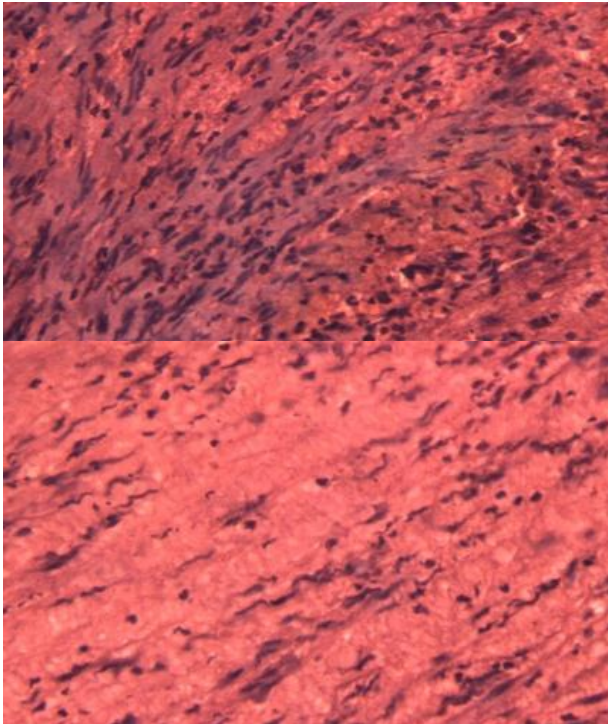


Fig. 9: Neurofibroma- Confirmed- Spindle shaped cells.+ve-for S-100 proteinNegative for Desmin. Prabhat.

Table 1. Cases of Endotracheobronchial Neurofibromas

Source (Y)	Age	Sex	Location	Symptoms or Signs	Operative Procedure
Dodge et al. (1977)	60	M	Distal trachea	Wheezing, dyspnea	Thoracotomy
Meredith et al. (1978)	60	M	Distal trachea	Wheezing, cough, exertional dyspnea	Thoracotomy
Thijs-van Nies et al. (1978)	36	M	Mid-trachea	Cough, hemoptysis	Bronchoscopic removal
Xu et al. (1982)	11	M	Distal trachea	NA	Lateral wall resection of trachea
Xu et al. (1982)	25	M	RMB	NA	Lateral wall resection of bronchus
Neal Rutledge et al. (1983)	18	F	Distal trachea	Cough, wheezing, SOB	Sternotomy
Hutton et al. (1983)	64	M	Right lower lobe bronchus, larynx	Cough, pneumonia	Bronchoscopic biopsy
Pearson et al. (1984)	NA		Trachea	NA	Surgery
Goode et al. (1986)	8	M	Distal trachea	Respiratory tract infection, cough, stridor	Bronchoscopic removal
Lossos et al. (1988)	45	M	Distal trachea	SOB	Bronchoscopic removal
Li et al. (1988)	32	F	RMB	Dyspnea, cough, fever	Surgery
Bagwell et al. (1989)	NA	NA	NA	NA	CO ₂ laser
Krawczynski et al. (1989)	10	F	NA	NA	NA
Marty-Ane et al. (1992)	30	M	Trachea	SOB	Laser, segmental resection of trachea
Kilani et al. (1992)	32	M	RMB	NA	Pneumnectomy
Takiguchi et al. (1993)	34	M	RMB	Persistent cough	Bronchoscopic removal, Nd:YAG laser
Davies et al. (1993)	63	F	Mid-trachea	Cough, SOB	Bronchoscopic removal
Khan et al. (1995)	23	M	RMB, RMB	Dyspnea	Nd:YAG laser
Stah et al. (1995)	23	F	Trachea	NA	Nd:YAG laser
Gjonaj et al. (1997)	10	M	Distal trachea	Pneumonia, wheezing, pneumothorax, chest pain	Bronchoscopic removal
Elmhorst et al. (1997)	42	F	Right upper lobe bronchus	Cough, hemoptysis	Right upper lobe resection
Present case (2000)	52	M	RMB	Productive cough, pneumonia	Sleeve pneumnectomy
	2009	17	f RMB		→ Proximal resection / bronchotomy

NA = not available; Nd:YAG = neodymiumyttrium-aluminum garnet; RMB = bronchus intermedius; RMB = right main bronchus; SOB = shortness of breath.

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DISCUSSION

Benign tumors originating from the tracheobronchial tree are not common. Papilloma, polyps, fibroma and hamartoma account for most of the benign tumors arising in the tracheobronchial trees, followed by leiomyoma and neurogenic tumors.^[1,2] Neurogenic tumors include neurilemoma (Schwannoma), neurofibroma and neuroma, which can also be seen in the mediastinum or lung parenchyma. Some can cause external compression of the superior vena cava or bronchus.^[3,4] They rarely occur within the endotracheobronchial trees as primary tumors.^[5] Neurilemmas are rarely associated with von Recklinghausen's neurofibromatosis, rarely undergo malignant change and often contain degenerative changes. Neurofibromas are frequently associated with von Recklinghausen's neurofibromatosis and 12% undergo malignant change. In our review of the literature, we found 14 male and 6 female patients with tracheobronchial neurofibroma, including our present case (Table 1). The ages ranged from 8 to 64 years (mean 33.9). Perelman and coworkers reported 2 tracheal neurofibroma cases in their series including 144 tracheal tumors.^[6] The trachea was the most frequent site involved in the cases of endotracheobronchial neurofibromas.^[6] One patient had neurofibromas in the right intermedium bronchus and the right main bronchus when presented, while the other patient had the neurofibromas in the right lower lobe bronchus and the larynx removed in different periods of time. It is surprising that no lesion from left-sided bronchial trees was reported.^[6] Four out of 18 patients had von

Recklinghausen's neurofibromatosis. The ages of these 4 patients were from 18 to 45 years, average 29 years. Three of 4 patients were male (75%). The clinical symptoms and signs of patients with tracheobronchial neurofibromas were mostly related to airway obstruction resembling patients with tracheal tumors. The initial symptoms presented clinically included cough, wheezing, and dyspnea, which are related to airway obstruction. Repeated respiratory tract infection, hemoptysis, and chest pain are occasionally seen. The procedures for tumor removal consisted of operation, laser vaporization, and bronchoscopic removal. The size of the tumor was recorded in only 5 surgical cases and varied from 1.8 to 6 cm in diameter. There has been no recurrence reported to date. Patients with tracheobronchial tumors may be asymptomatic for years. Chest roentgenography may reveal.

Infiltration and atelectasis, though the majority of the patients have normal chest films in the initial presentation. Delayed diagnosis is not unusual, as overlying soft.

Tissue obscures the trachea in posteroanterior radiography. Some reported that final diagnosis might be realized after a 10 to 15 month delay from the onset of symptoms.^[7] The other diagnostic tools include computed tomography and magnetic resonance imaging, which can demonstrate the size of the tumor and the relationship between the tumor and adjacent tissue. Bronchoscopy may disclose a round, protruded and sometimes hyper vascular tumor. Diagnosis can be obtained through biopsy, though carries some risk of hemorrhage. The treatment for the endotracheobronchial neurofibroma depends on the size and the location of the tumor and should be conservative. Some small tumors can be removed endoscopically with or without laser. However, if the tumors invade adjacent tissue or result in obstructive pneumonitis, surgery should be undertaken.

CONCLUSION

Endobronchial neurofibroma is a rare clinical entity. CT is helpful to find small lesions in airway or lesions in the lung; bronchoscopy is not only used to explore the airway, but also to biopsy in order to understand the etiology. After diagnosis of the neurofibromas, the treatment depends on the size and the location of the tumors. Some small tumors can be removed endoscopically with or without laser. However, if the tumors invade adjacent tissue, surgery should be carried out.

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