

# Tracheal Hamartoma Causing Unique Stridor and a Review of the Literature

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A 47-year-old male, who had been treated under the diagnosis of bronchial asthma, was admitted to this department with the complaints of progressive dyspnea of 3 years duration and upper air way obstruction causing unique stridor. Through roentgenography and bronchoscopy, lower tracheal hamartoma was confirmed, but a pulmonary function test precipitated acute hypoxia with mental somnolence. The tumor was removed as quickly as possible by multiple punch biopsy, through a rigid bronchoscope, for life saving air way maintenance.

The term hamartoma was first described by Albrecht in 1904 (Butler and Kleinerman 1969).

Hamartoma of the lung and bronchus is rarely seen in the literature.

Especially in the case of hamartoma of the trachea, we were able to find only three previous reports from the English literature. (Hurst and Nelson 1977, Engelking 1959, Kaneko *et al* 1978).

This case is of interest not only because of its rarity but also because it was a unique cause of stridor with over 90 percent of the tracheal lumen obstructed, and also the patient became able to breath comfortably after life saving rigid bronchoscopic procedures.

## CASE REPORT

A 47-year-old Korean male teacher was

admitted to Severance Hospital because of respiratory distress on Sep. 10, 1979.

He was in good health until 3 years before when he began to suffer from mild but sudden onset of dyspnea on exertion. He took some medicine including herbs under the diagnosis of bronchial asthma without any benefit. He continued to have cough with some mucoid phlegm and during those years before entry the hospital he experienced progressive respiratory difficulty accompanied by a loss of 20Kg in weight.

Five months before admission wheezing respirations began to occur.

Two weeks before entry he entered another hospital where upper air way obstruction was suspected and he was referred to this hospital for further evaluation and treatment.

There was no history of smoking and he denied previous tuberculosis, hemoptysis or

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Fig. 1 Tomogram of the trachea shows a definite mass almost completely occluding the trachea.

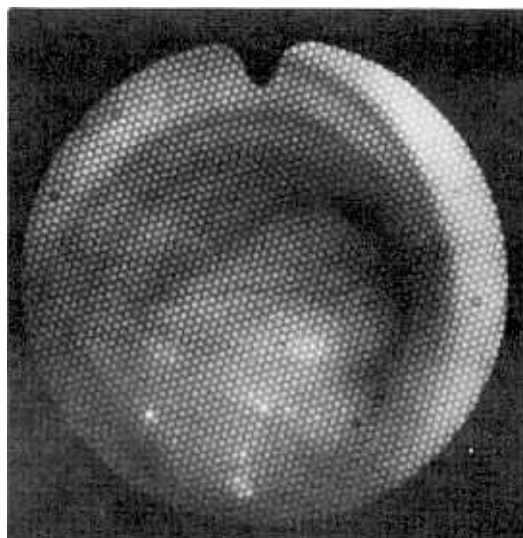


Fig. 2. Bronchofiberscopic picture by Machida 6 TL; round mass obstructing the tracheal lumen with sparing crescentic slit on right lateral side of the trachea.

allergic episodes.

On admission to the hospital the patient was in severe respiratory distress.

He had dyspnea with inspiratory and expiratory stridor, dilated neck veins and prominent movement of the accessory muscles in his neck. The skin was not cyanotic.

The sputum cytology showed no malignant or atypical cells. A sharply circumscribed mass density could be seen more clearly at the level of 2 cm above the carina, on the lateral view of the chest roentgenogram rather than the postero-anterior view. Tomogram of the trachea confirmed that the mass was in the trachea (Fig. 1) and showed some scattered calcified densities on the margin of the tumor. Bronchofiberoptic examination revealed a lobulated and pinkish hard tumor attached to the left lateral wall of the trachea obliterating almost 90 percent of the tracheal lumen. There was only a small crescentic slit of airway near the right lateral wall (Fig. 2).

The mass was too hard to get the biopsy specimen through bronchofiberscope. No further attempt was made to get the specimen. After trying the pulmonary function test the patient began to suffer from more severe respiratory difficulty with mental somnolence. Arterial blood gases while breathing oxygen at 2 liters per minute through a nasal prong showed an arterial  $P_{O_2}$  of 58 mmHg,  $PCO_2$  of 75 mmHg, and arterial pH of 7.18 with a standard bicarbonate concentration of 22.1 mEq per liter and actual bicarbonate concentration of 26.5 mEq. Pulmonary function tests revealed a forced vital capacity (FVC) of 1.6L (50 percent of predicted), forced expired volume (FEV1) in one second of 20 percent, and FEV1/FVC was 16 percent. Maximal expiratory flow-volume loops could not be obtained because of severe respiratory distress. Early on the morning of the 4th hospital day the patient became drowsy and the respiration was weak with distant breathing sound over both lungs. Emergency rigid

bronchoscopy was performed for life saving maintenance of the air way and the mass in the distal trachea was removed by multiple punch biopsies.

A portex endotracheal tube was inserted to maintain the air way. The patient was examined by bronchofiberscope again on the 9th hospital day and this disclosed ulceration of the carina and the left lateral wall of the trachea where the tumor had been located. Concomittantly the endotracheal tube was removed without difficulty.

Histologic sections of the mass, protruding into the lumen of the trachea, was roughly composed of two major types of tissue, the adult fat and

the hyaline cartilage, and was covered by squamous epithelium.

The squamous covering was irregularly acanthotic and thought to be metaplastic compared to the tracheobronchial ciliated epithelium.

Dominate was the adult fat tissue, where islands of mature hyaline cartilage and a third minor component, the groups of glands, that simulated the tracheobronchial mucous gland, were intervened (Fig. 3). Focal linear calcification was also noted along the margins of the cartilage lobules (Fig. 4).

In addition, this patient has been under close follow up by bronchofiberscopy without any evidence of regrowth of the tumor for the last  $2\frac{1}{2}$  years.

## DISCUSSION

The first tracheal tumor was recorded in 1761, it was a tracheal fibroma found at post-mortem examination (Kaneko et al, 1978; Weber and Grillo, 1978). Tumors of the trachea remain relatively rare.

Compared with laryngeal lesions, tumors of the trachea occur with less frequency. Gilbert and colleagues compiled the reported primary tracheal tumors (Gilbert et al, 1953). His review revealed that the most common tracheal tumor was osteochondroma, followed by papilloma and fibroma in the adult.

Weber and Grillo analysed 84 cases over a 17 year period. He reported only 8 cases of benign tumors among the 84 cases of tracheal tumors.

There was no case of hamartoma in that series. Hamartomas occur commonly in the lung and less frequently in the bronchi, and are exceedingly rare in the trachea.

Table 1 illustrated 3 cases reported previously. They were all males.

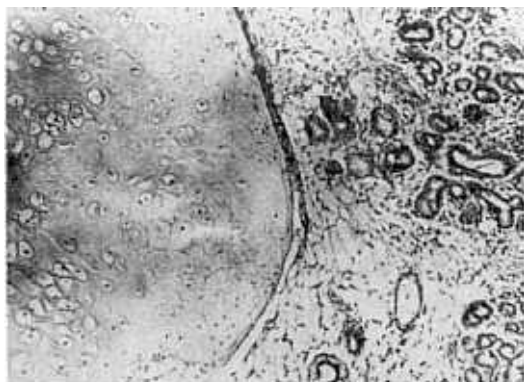


Fig. 3. The tumor is composed of mature hyaline cartilage(left) and adult fat tissue where mucous secreting glands were grouped (H-E stain, x 430).



Fig. 4. Linear calcification is seen along the margin of a cartilage lobule (H-E stain, x 430).

Table 1. Reported Cases of Hamartoma of the Trachea

Reference (reported year)	Sex, Age and Race	Diagnosis on admission	Duration till diagnosis	Location and gross appearance	Treatment	Follow up without recurrence
Engelking 1952	M 51 White	Bronchial Asthma	20 years	Right mid trachea post-lateral, lobulated, hard and pinkish	Resection	4 years
Hurst et al. 1977	M 75 White	COPD	10 years	Left lateral, round, pinkish-yellow	Segmental resection (3 ring)	At least 5 months
Kaneko 1978	M 34 Japanese	Bronchitis	1 year	Anterior, 3cm above carina round, lobulated, bright red	Electric cauterization thru BF*	9 months
Present Case 1981	M 45 Korean	Bronchial asthma	3 years	Left, 2cm above carina post-lateral, round, lobulated hard, pinkish	Multiple punch biopsy thru rigid BF	2 $\frac{1}{2}$ years

The origin of the hamartoma is controversial. However, a common concept is that it is tumor-like and non-neoplastic, but arises from embryonal connective tissue cells. Tracheal hamartoma may often resemble chondroma in its clinical and roentgenological picture.

Sometimes, however, hamartoma may likewise demonstrate calcification.

Definite differentiation is usually made on a histologic basis. The hamartoma contains cartilage but it also contains other fatty tissue, lymphoid tissue and epithelial components or leave out.<sup>(1)</sup>

Tracheal tumors are usually recognized late in the course and should be differentiated from obstructive air way disease.

Weber and Grillo suggested that the tracheal lumen is often more than 75 percent compromised before any localizing signs and symptoms appear.

In malignant lesions of the trachea the average duration of symptoms from the onset to the final

diagnosis is about 6 to 12 months. In benign lesions, however, patients did not notice symptoms for months or years. Usually they noted only mild exertional dyspnea for a long period and were treated under the diagnosis of obstructive air way disease such as asthma or allergic bronchitis (Gilbert et al 1953). All of the tracheal hamartomas had the same clinical pictures with a duration of one to 20 years until a final diagnosis (Table 1). In our case the patient had been treated under the diagnosis of asthma for 3 years. In the last 3 months he began to suffer from stridor. Tracheal stridor due to tracheal tumor brings about this usual suffering. Weber and Grillo reported 12 cases that suffered from tracheal stridor among 84 cases of tracheal tumors. Fleetham and colleagues reported one case of tracheal leiomyosarcoma with stridor (Fleetham et al 1977), and the clinical course and roentgenographic picture was very similar to our case except for hemoptysis.

Another similar case was reviewed in a "case

record" of Massachusetts General Hospital (Scully et al 1975), and recently Beller and colleagues reported a case of coccidioidal granuloma with wheezing and stridor secondary to obstruction of the right main stem bronchus and lower trachea (Beller et al, 1979).

Routine posteroanterior chest roentgenogram may not reveal a mass shadow in the trachea. Hurst and Nelson reported one case of tracheal hamartoma with a normal chest roentgenogram (Hurst and Nelson 1977).

Daniels and colleagues also reported a similar case of primary chondrosarcoma (Daniels et al, 1967).

In reviewing the cases bronchoscopy seemed to be important in the diagnosis of tracheal tumor and the gross findings revealed that the hamartoma was commonly round, lobulated, hard and bright red in color (Table 1).

However, special procedures should be performed very carefully because the patient with tracheal obstruction may become dyspneic and began to wheeze after bronchoscopic examination, position change (Scully et al, 1975) or even talking (Fleetham et al, 1977; Scully et al, 1975).

In this patient, bronchoscopic examination was done without difficulty under topical anesthesia but unfortunately acute hypoxia was precipitated during the performance of a pulmonary function test on the 3rd hospital day.

Treatment might vary according to the character and the location of the tumors. For example, if the tumor is benign in type and located in the lower trachea in location, resection through the rigid bronchoscope would be considered. In our patient the tumor was removed through the rigid bronchoscope without any complication. There were some similar trials with good results in other types of tumor (Daniels et al 1967; Beller et al 1979) and in tracheal hamartoma (Kaneko et al 1978).

In conclusion, we would like to suggest the

following points for early detection of the tracheal tumor; 1) high index of suspicion for tracheal tumor in a patient with dyspnea and wheezing 2) progressive asthmatic wheezing unresponsive to antiasthmatic therapy 3) patient with stridor 4) roentgenographic studies of posteroanterior view with lateral projection 5) bronchofiberscopy, especially in case with recent onset of respiratory distress.

In addition it is recommended that care be taken in performing a pulmonary function test, which can be a dangerous risk in known severe tracheal obstruction.

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