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Review Article

Saber Sheath Trachea- Functional and Clinical Correlations

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Abstract

Saber sheath trachea refers to diffuse coronal narrowing of the intra-thoracic portion of the trachea with concomitant widening of the sagittal diameter. It is considered to be widely associated with Chronic Obstructive Pulmonary Disease (COPD). The diagnosis is based on the calculation of the tracheal index which is the ratio of coronal to sagittal length in the axial plane measured 1cm above the upper margin of the aortic arch. Saber sheath trachea is considered to be present when the tracheal index is less than 0.67. The tracheal index has been associated with severity of obstruction and most notably with hyperinflation and extent of emphysema in COPD patients. Thus, it can have clinical importance. The presence of saber sheath trachea and its clinical implications should be further explored in COPD as well as in other diseases in which emphysema represents a major component, as Combined Pulmonary Fibrosis Emphysema (CPFE).

Tracheal Anatomy

The trachea is a cartilaginous and fibro muscular tubular structure. It extends from the inferior aspect of the cricoid cartilage (at the level of the sixth cervical vertebra) to the carina (at the level of the fifth thoracic vertebra). Total length usually varies from 8 to 13 cm. The trachea is divided into an extrathoracic (2-4 cm in length) and intrathoracic portion (6-9 cm in length) at the level it passes posterior to the manubrium. The intrathoracic portion is subject to the pleural pressure [1].

The trachea is comprised of 16 to 22 C-shaped cartilages, which form the anterior and lateral walls. The posterior wall does not have any cartilage elements and is supported by the trachealis muscle. The trachealis muscle is composed by transverse smooth muscle fibers. Their contraction results in narrowing of the tracheal lumen [1]. The trachea cartilages are linked longitudinally by annular ligaments of fibrous and connective tissue [1,2]. The above mentioned cartilaginous structure plays a supportive role, maintaining the patency of the trachea and on the same time it allows the trachea to follow the various movements of the neck without the risk of getting folded.

CT Appearance of the Normal Trachea

On axial CT images, the normal tracheal lumen usually demonstrates an oval, round, or horseshoe shape [3]. The tracheal wall is usually visible as a soft tissue stripe, with a thickness of 1-2 mm. It is delineated by air in the lumen and is outlined by mediastinal fat externally. The posterior tracheal membrane usually appears thinner than the anterior and lateral tracheal walls and can vary in shape due to the absence of cartilaginous support.

The posterior membranous wall of the trachea bows forward during expiration in normal subjects [4]. This can be used as a useful sign to evaluate whether the CT scan was captured during inspiration or expiration.

The coronal diameter of the normal trachea ranges from 13 to 25 mm in men and from 10 to 21 mm in women, and the sagittal diameter of the normal trachea ranges from 13 to 27 mm in men and from 10 to 23 mm in women [1].

Saber Sheath Trachea

The term “saber sheath trachea” was first introduced by Greene and Lechner [5].

The recognition of the saber sheath trachea is based on the calculation of the tracheal index.

Tracheal index is defined as the ratio of coronal to sagittal length in the axial plane. It is usually measured at a plane 1cm above the upper margin of the aortic arch. Hence, a tracheal index of 1 corresponds to a trachea with a conventional rounded cross section. When the coronal length is less than the two thirds of the sagittal length (i.e. tracheal index<0.67), then saber sheath trachea is present (Figure 1). Saber sheath trachea has been widely associated with COPD. In saber sheath trachea the inner wall of the trachea is smooth with no nodularity and no thickening. These findings are helpful in the differentiation from other diseases with tracheal involvement (tracheobronchomalacia, tracheopathia osteochondroplastica, relapsing polychondritis, amyloidosis, sarcoidosis, granulomatosis with polyangiitis) [6] (Table 1).

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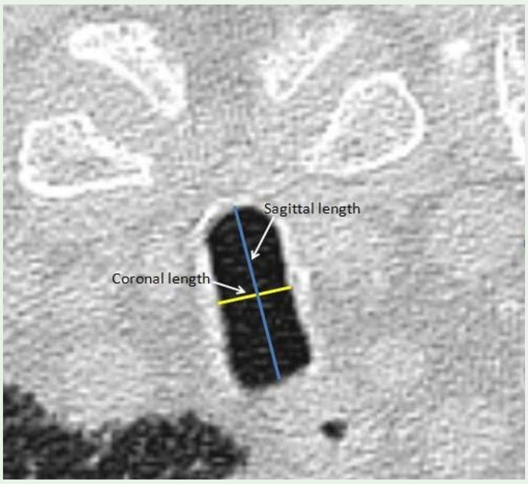

Figure 1: CT scan of saber sheath trachea. Sagittal length = 10.1 mm, Coronal length =27mm. Tracheal index is 37.4%.

Etiology

Regarding etiology, the causative mechanisms responsible for the formation of the "saber sheath" shape are not fully understood. Several mechanisms have been proposed:

1) Chronic cough, often a symptom of COPD patients' leads to recurrent injuries in the tracheal cartilages and the subsequent remodeling leads to the "saber sheath" shape [7,8].

2) In COPD patients, especially those with a more severe degree of obstruction, trachea acquires an elongated shape. This leads to the exertion of vertical forces upon the cartilaginous rings.

3) Finally, lung hyperinflation can lead to "saber sheath" deformity in two ways:

Firstly, hyperinflation of the upper lobes directly results in an increased pressure on the lateral walls of the trachea.

Secondly, hyperinflation is the result of the narrowing of the small airways. This leads to an increased pleural pressure during expiration. This increased pleural pressure is exerted on the intrathoracic trachea which finally acquires the characteristic "saber sheath" shape. Tracheal index is measured in the intrathoracic part of the trachea, i.e. 1-2 cm above the upper margin of the aortic arch.

The latter theory is supported by papers that demonstrate a correlation between the tracheal index and parameters indicative of hyperinflation, such as Functional Residual Capacity (FRC) [9]. Also, in another study [10], saber sheath trachea was identified in 10 out of 43 emphysematous COPD patients' candidates for Lung Volume Reduction Surgery (LVRS). The percentage of residual volume divided by total lung capacity (RV/TLC%, a strong indicator of hyperinflation) was significantly different between the COPD patients with and without saber sheath trachea.

Furthermore, in COPD patients with saber sheath trachea, the extrathoracic trachea has a normal shape [11]. Also, as mentioned before, although the tracheal index is measured at a plane 1-2 cm above the aortic arch, the saber sheath shape is recognized consistently across the intrathoracic trachea [3,5].

Hence, it seems that lung hyperinflation plays a significant role in the pathogenesis of saber sheath trachea. Intrinsic factors such as the degree of rigidity of the tracheal cartilages, must determine in which cases the forces that are exerted upon them will result in the "saber sheath" deformity.

Aging does not seem to be implicated in the etiology of saber sheath trachea. In a study in 83 healthy adults (age range: 21-83 years), tracheal index was not correlated with age [12].

Papers investigating the incidence of saber sheath trachea in COPD and its correlation with various radiological and functional parameters of the disease are rather limited. A summary of the available studies is shown in table 2.

Trigaux, et al. [9] studied 20 patients with saber sheath trachea (Tracheal index<67%). This study also included as control group 20 respiratory patients without saber sheath trachea (Tracheal index>70%). In the first group, there was a significant correlation of the tracheal index with parameters indicative of hyperinflation such as FRC and sternum-spine distance. These two parameters also exhibited a significant difference between the two groups.

Gupta, et al. [13] studied 40 male patients with spirometrically confirmed COPD. Saber sheath trachea (Tracheal index<0.67) was found in 14 patients (35%). Tracheal index exhibited a significant negative correlation with age, duration of illness, pack years, dyspnea scale, and Airways Questionnaire 30 (AQ30) score. It also had a significant positive correlation with of FEV1, FEV1/FVC and PEFR.

Leader, et al. [10] examined the size and morphology of the trachea before and 3 months after Lung Volume Reduction Surgery

Table 1: Differential Diagnosis of Diffuse Tracheal Diseases.

Saber Sheath Trachea	Tracheal index<0.67 No tracheal wall thickening Inner wall smooth with no nodularity
Tracheobronchomalacia	Airway narrowing accentuated by expiration (lunate appearance) Decrease of cross sectional area by at least 50% at end expiration is considered diagnostic
Tracheopathia Osteochondroplastica	Multifocal calcified mural nodules in the anterior and lateral tracheal walls typically sparing the posterior tracheal membrane
Relapsing Polychondritis	Thickening of the tracheal wall typically sparing the posterior tracheal membrane
Amyloidosis	Nodular thickening of tracheal wall. Nodules may be calcified. Posterior tracheal membrane may be involved
Sarcoidosis	Due to extrinsic pressure from enlarged lymph nodes or formation of non caseating granulomas in tracheal walls
Granulomatosis with Polyangiitis	Subglottic (typically) or diffuse tracheal involvement Thickening of tracheal wall (smooth or nodular) No sparing of the posterior tracheal membrane

Table 2: Studies on Saber Sheath Trachea.

Studies	Study Population	Results	Conclusions
Trigeaux, et al. 1994 [9]	20 patients with SST, and 20 respiratory patients without SST as control group	Significant correlation of Ti with FRC and sternum-spine distance	SST related to hyperinflation
Muro, et al. 2000 [18]	35 patients with spirometrically confirmed COPD and 24 healthy individuals as control group	No correlation of Ti neither to pulmonary function nor to CT parameters of hyperinflation	SST not related neither to hyperinflation nor functional obstruction
Leader, et al. 2004 [10]	24 male and 19 female patients with emphysema fulfilling the criteria for LVRS	Significant correlation of Ti with the RV/TLC ratio of before surgery	SST related to hyperinflation
Gupta, et al. 2008 [13]	40 male patients with spirometrically confirmed COPD	Significant correlation of Ti with of FEV1, FEV1/ FVC and PEFR.	SST related to functional obstruction
Lee, et al. 2011 [16]	115 male patients with spirometrically confirmed COPD and 92	Significant correlation of Ti with FEV1, FEV1/ FVC in the COPD group. Significant correlation of Ti with CT quantification parameters indicative of hyperinflation	SST related to hyperinflation and functional obstruction
Ciccarese, et al. [17] [2014]	71 patients with spirometrically confirmed COPD	Greater prevalence of SST in patientswith lower Tiffenau Index	SST related to functional obstruction

Abbreviations: SST: Saber Sheath Trachea; TI: Tracheal index

(LVRS). This study included 24 male and 19 female patients. We should emphasize that this is a very specific group of emphysematous COPD patients that fulfill strictly defined selection criteria [14,15]. In this specific group, saber sheath trachea was identified in 10 out of 43 patients (23%). In the "saber sheath" group, there was a positive correlation between the tracheal index and the ratio of Residual Volume to the Total Lung Capacity (RV/TLC %) before surgery, supporting the role of hyperinflation in the pathogenesis of saber sheath trachea. However, there was no correlation of the tracheal index with pulmonary function test parameters.

The largest study included 115 male patients with spirometrically confirmed COPD and 92 healthy individuals as control group who underwent volumetric inspiration/expiration CT scanning [16]. All participants also underwent pulmonary function test evaluation on the same day. An important advantage of this study is that it examined tracheal morphology not only in inspiration, but also in end-expiratory phase. Another advantage was that it used volumetric CT in sub millimeter resolution, thus increasing the accuracy of the measurements.

Several parameters regarding tracheal morphology were calculated on CT as tracheal index, Mean Lung Density (MLD), Lung Volume (LV) and the volume fraction of the lung below -950 Hounsfield Units (EI). MLD and EI are very important indexes as they positively correlate to the extent of emphysema. To assess air trapping, the ratio of the MLD on expiration and inspiration (CT air trapping index, CT–ATI) was used.

The only significant morphologic change in COPD patients compared to the healthy group was the tracheal index. In the COPD group, the tracheal index showed a significant correlation with FEV1 as well as the ratio of FEV1/FVC. There was a significant correlation of the tracheal index with the EI on inspiration CT. This correlation was further enhanced on expiration CT indicating the importance of air-trapping. Indeed, the tracheal index showed a significant correlation with CT-ATI. Finally, the tracheal index exhibited a significant correlation with MLD and LV only on expiration CT scans.

Ciccarese, et al. studied a cohort of 71 COPD patients [17]. Saber sheath trachea was found in 18 (25%) and was linked to the functional severity of airway obstruction as there was a greater prevalence in patients with lower Tiffenau Index (p=0,02).

Finally, in a single study by Muro, et al. [18], there was no correlation of tracheal index neither to pulmonary function nor to CT parameters of hyperinflation.

Summarizing, there are studies correlating the presence of saber sheath trachea with airway obstruction, although this finding is not consistent across all studies. What is rather consistent is the correlation between saber sheath trachea and indexes of lung hyperinflation (FRC, RV/TLC %), as well as the presence and extent of emphysema. This supports the theory that lung hyperinflation may be the more important etiologic factor leading to the saber sheath deformity of the trachea.

The clinician should bear in mind that in COPD patients with saber sheath trachea who require intubation, air leaking around the cuff of the endotracheal tube could be the cause of problematic ventilation [19,20].

In some patients emphysema coexists with pulmonary fibrosis (Combined Pulmonary Fibrosis Emphysema-CPFE) [21]. These patients are smokers and emphysema is usually encountered in the upper lobes preceding fibrosis of the lower lobes. Usually, the pulmonary fibrosis follows a definitive or possible Usual Interstitial Pneumonia (UIP) radiology pattern on HRCT [22]. A characteristic of patients with CPFE is that they usually present with relatively preserved lung volumes in relation to the extent of fibrosis. That is because in these patients the decreased lung compliance that is caused by fibrosis is counterbalanced by the increased lung compliance that is usually seen in patients with emphysema.

For the above mentioned physiological reasons, it would be very interesting to study not only the prevalence of saber sheath trachea in CPFE patients, but also its probable correlation with clinical, physiological and radiological parameters.

Conclusion

Saber sheath trachea is a characteristic tracheal deformity that is widely associated with COPD. The diagnosis is based on the calculation of the tracheal index which is the ratio of coronal to sagittal length in the axial plane measured 1cm above the upper margin of the aortic arch. Saber sheath trachea is considered to be presented when the tracheal index is less than 0.67. Several hypotheses have been proposed regarding its etiology. Lung hyperinflation seems to play the most important role. The calculation of the tracheal index could have clinical significance as it is correlated with the level of obstruction, lung hyperinflation and the extent of emphysema. Its potential clinical role should be further explored not just in COPD but also in diseases in which emphysema is a major component, as CPFE.

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