Case report

A case of idiopathic tracheal stenosis

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ABSTRACT

Acquired tracheal stenosis is a condition that often results from trauma, neoplasm, infection, vasculitis, inflammatory or infiltrative processes. Idiopathic tracheal stenosis is a rare entity and represents a diagnosis of exclusion. We report a case of severe localized extrathoracic tracheal stenosis in a 35 year old female who was initially suspected to have asthma as the cause of several months of chronic dyspnea.

1. Case report

A 35 year old female was referred to the outpatient pulmonary clinic for evaluation of abnormal pulmonary function tests done as a work up for chronic dyspnea and suspected asthma. Five months earlier she was hospitalized with bilateral pneumonia treated with a third generation cephalosporin and a macrolide with no specific pathogen identified. Following the hospitalization, the patient noticed persistent and progressive shortness of breath with exertion in addition to recurrent cough with occasional production of mucus slightly tinged with blood. The patient was involved in a motor vehicle accident two years prior to this presentation. Cervical MRI at that time showed degenerative disc disease at C6–C7 and osteophytes at C2–C3 without cord compression. No history of asthma, rhino-sinusitis, vasculitis, connective tissue disease or airway instrumentation.

Pulmonary function testing (Fig. 1) showed significant flattening of the inspiratory and expiratory flow volume loop. The pattern was suggestive of a fixed large airway stenosis. There was no significant bronchodilator response on spirometry.

Neck soft tissues enhanced CT with 3D reconstruction (Figs. 2A, B and 3) showed an area of concentric tracheal narrowing just above the level of the thoracic inlet with a diameter of 6.5 mm at its narrowest point. On a chest CT scan done 5 months earlier, the trachea appeared normal at that level.

On rigid bronchoscopy the lesion was measured 1.5–2 cm and began about 2 cm below the cricoid ring. Tracheal dilatation and...
Fig. 2. (A, B). CT images of the soft tissue neck with contrast shows area of narrowing in the trachea above the level of the thoracic inlet (arrow). The lesion is localized in the extrathoracic trachea, adjacent to the level of the thyroid gland, about 3.5 cm below the vocal cords.

Fig. 3. (A, B). 3D reconstruction of the CT scan shows localized area of severe tracheal narrowing (arrow). The lesion is localized in the extrathoracic trachea, adjacent to the level of the thyroid gland, about 3.5 cm below the vocal cords.
several biopsies were performed. About 4 weeks later the patient underwent tracheal resection with primary anastomosis. The pathology report showed acute ulcerative tracheitis with squamous metaplasia associated with dense and widespread submucosal fibrosis and scarring with smooth muscle metaplasia and intense lymphoplasmacytic infiltration. No evidence of granulomatous disease or malignancy was present.

Vasculitis and connective tissue disease serologies were negative.

The patient had complete clinical recovery. Repeat flow-volume loop about 2.5 years later revealed significant improvement with only mild truncation of the inspiratory loop (Fig. 4). The expiratory loop was unremarkable.

2. Discussion

The diagnosis of idiopathic tracheal stenosis (ITS) is one of exclusion and is made based on history, clinical presentation, physical examination, and pathological findings. The condition is more common in middle aged females [1,2]. Some authors have postulated that the incidence of ITS is higher in post-menopausal females secondary to the absence of estrogen receptors at the site of stenosis. This leads to the increase of fibroblast growth factor at that site, which in turn results in the formation of a stenotic lesion [3]. Patients usually present with progressive shortness of breath, hoarseness, stridor, and wheezing [2]. This entity is often initially misdiagnosed as asthma [1].

Causes of tracheal stenosis include iatrogenic (endotracheal intubation, tracheostomy, radiotherapy, surgery), congenital, external injury, tumors, autoimmune conditions (sarcoidosis, amyloidosis, polychondritis, Wegener’s granulomatosis), tracheopathia osteoplastica, bacterial infection (rhinolaryngosclerosis, TB, bacterial tracheitis, tuberculosis, diphtheria, histoplasmosis). For that reason history of neck trauma, inhalational burns, irradiation, infection, neoplasm, intubation, connective tissue diseases need to be ruled out prior to establishing the diagnosis.

A PubMed literature search using the key words “pneumonia” and “tracheal stenosis” yielded no case reports describing the link between pneumonia and tracheal stenosis.

Spirometry, CT scan and bronchoscopy are important modalities that aid in the diagnosis. Even though the spirometry numerical data might not show definite obstruction, inspection of the flow volume loop in those patients will reveal flattening of both the inspiratory and expiratory loop. At that point examination of the patency of the large airway either by bronchoscopy or by imaging studies is indicated.

Tracheal stenosis can be focal or diffuse. Systemic conditions such as tuberculosis, systemic lupus erythematosis, histoplasmosis, sarcoidosis usually cause diffuse tracheal stenosis [2]. Neck trauma and intubation usually result in focal tracheal stenosis [2]. In a review of the radiologic studies of 15 patients with idiopathic tracheal stenosis Bhalla et al. concluded that ITS leads to focal concentric or eccentric tracheal stenosis [2]. Its dimensions are between 2 and 4 cm in length and no more than 5 mm in diameter [2]. Thus, in patients with focal tracheal stenosis and no prior history of trauma or intubation ITS should be considered high in the differential diagnosis.

On pathological examination the lesions in ITS reveal keloidal fibrosis of the adventitia and lamina propria [2]. The mucosa, muscularis propria and cartilage appear intact [2]. Mark at al. [4] performed a clinicopathologic review of 63 tracheal resections in patients diagnosed with idiopathic tracheal stenosis and compared them to 34 cases of tracheal stenosis secondary to chondromalacia (CM) after mechanical injury. The authors noted that all 63 cases of ITS occurred in females. When compared to CM, on pathologic examination ITS showed normal cartilage with smooth inner and normal perichondrium, keloid fibrosis and dilated mucous glands. In contrast, CM showed cartilage degeneration with irregular inner perichondrium. The authors concluded that in the majority of cases, ITS can be pathologically distinguished from CM. Wegener’s granulomatosis can be excluded on the basis of negative ANCA and microscopically by the absence of granulomatous features and absence of microabscesses beneath the epithelium [4]. Polychondritis, rheumatoid arthritis and systemic lupus erythematosus can be excluded on the lack of systemic signs and symptoms. Pathological examination in this case showed acute ulcerative tracheitis with squamous metaplasia associated with dense and widespread submucosal fibrosis and scarring. There was no evidence of granulomatous disease or malignancy. The above findings were compatible with idiopathic tracheal stenosis as a probable diagnosis.

There are several options for the management of ITS [5–8]. Balloon dilatation is a conservative treatment approach that can be employed in some cases [3]. Other modalities such as airway stenting, laser treatment, surgical reconstruction or resection are available [3]. In cases where ITS involves only the upper trachea segmental resection with end-to-end anastomosis can be performed [5].

3. Conclusion

ITS is a rare serious entity that should be considered in the differential diagnosis in patients with shortness of breath and cough that are unresponsive to treatment. The diagnosis is based on the combination of findings on history, physical exam and diagnostic testing. Pulmonary function testing shows flattening of both

**Fig. 4.** Pulmonary flow volume tracing shows mild truncation in the inspiratory loop. The expiratory loop is unremarkable.
the inspiratory and expiratory loops and plays an important role in the diagnostic work-up. Definitive diagnosis is based on histopathologic findings. The choice of surgical versus endoscopic treatment is made on an individual basis based on the patient’s characteristics.

Conflict of interest

The authors declare that there is no conflict of interest.

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