Unusual cause of respiratory distress misdiagnosed as refractory asthma

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Abstract:

We report a young lady, who was labeled as a case of refractory asthma for a few years, based on history of shortness of breath on minimal exertion, noisy breathing and normal chest radiograph. Repeated upper airway exam by an otolaryngologist and computerized tomography scan, were normal. On presentation to our hospital, she was diagnosed to have fixed upper airway obstruction, based on classical flow-volume loop findings. Fibroptic bronchoscopy revealed a web-shaped subglottic stenosis. The histopathology of a biopsy taken from that area, showed non-specific inflammation. No cause for this stenosis could be identified. The patient was managed with rigid bronchoscopy dilatation, without recurrence. We report this case as idiopathic subglottic stenosis, that was misdiagnosed as refractory bronchial asthma, stressing the importance of performing spirometry in the clinic.

Key words:

Asthma, subglottic stenosis, flow volume loop.

Subglottic stenosis is a serious and sometimes life- threatening condition. It results from narrowing of the subglottic airway, which is housed in the cricoid cartilage. It is the narrowest area of the airway, since it is a complete, non expandable and a non pliable ring. Stenosis in this area can be congenital, which is usually diagnosed during childhood, or acquired. Typically, acquired subglottic stenosis has an insidious onset, where early manifestations are usually mistaken for other respiratory disorders, like bronchial asthma and bronchitis.

We report a patient with acquired subglottic stenosis of unknown etiology. She was misdiagnosed as a case of refractory bronchial asthma for few years, until her condition was finally diagnosed with the help of spirometry and fibroptic bronchoscopy. Successful dilatation was performed with rigid bronchoscope, without recurrence.

Case Report

A 26-year-old lady, not known to have any medical problems, was referred to the pulmonary clinic, as a case of a refractory asthma, not responding to bronchodilators and multiple courses of systemic steroids. Her main complaint was shortness of breath that started 4 years ago and became progressively worse over the past year, until she became dyspneic on minimal exertion. She noticed that her dyspnea increased on exposure to perfumes and was associated with noisy breathing. There was no associated cough, chest pain, orthopnea, or heart burn. There was no history of recurrent sinusitis, nasal discharge, eye complaint, recurrent pneumonitis, or ingestion of corrosives. Her past medical and surgical history was insignificant. There was no prior intubation or upper airway trauma.

Physical examination revealed a young lady in obvious respiratory distress, on minimal exertion with noisy breathing. Respiratory rate was 22/min. Chest examination revealed inspiratory stridor, without obvious wheeze. Neck examination showed no obvious goiter or masses. Other systems were normal. Chest radiograph was normal. Blood works including complete blood count, renal function, liver function, arterial blood gases (at room air and rest) and thyroid function tests, were normal. Direct laryngoscopy in the clinic revealed no abnormality. Spirometry revealed a box-like flow-volume loop compatible with fixed upper airway obstruction [Figure 1a]. Detailed ENT examination was normal. CT scan neck, sinuses and chest were reported initially, as normal. However, reconstruction CT scan was requested and showed an area of narrowing, just below vocal cords [Figure 2a and b]. Bronchoscopy revealed subglottic stenosis and a thin web, resulting in significant narrowing [Figure 3]. Bronchoalveolar lavage and brush were negative for gram stain and acid fast bacilli stain and culture and malignant cells. Serology for ANA and ANCA was negative. Dilatation under

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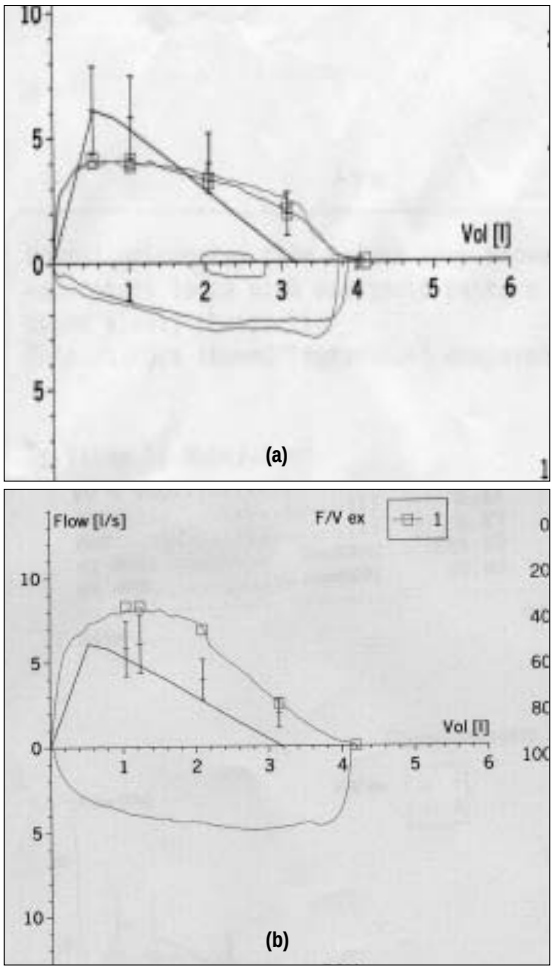

Figure 1: Flow-volume loop of the patient before treatment (a) and two days after dilatation (b).

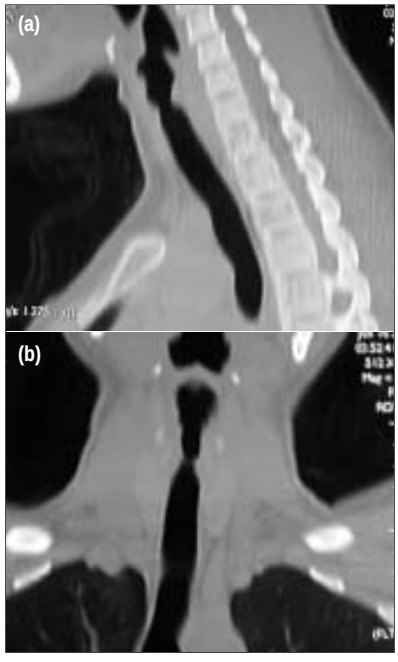

Figure 2, a and b: Reconstruction of CT scan of the upper airway reveals the subglottic narrowing.

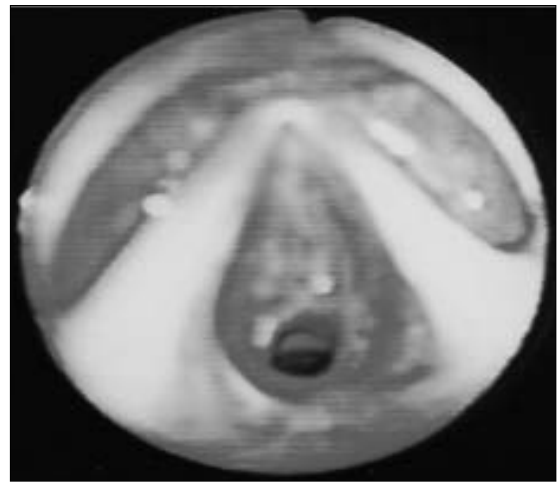

Figure 3: Bronchoscopic view of the web like narrowing of the subglottic area.

general anesthesia was performed, using rigid bronchoscope size 6, then 7.4, and lastly size 8. Two days after dilation, flow volume loop was normal [Figure 1b]. Histopathology report from the web revealed non-specific inflammation.

For the past two years, the patient has no recurrence of the above problem and no changes in the flow volume loop.

Discussion

Acquired subglottic stenosis is secondary to endotracheal intubation, in 90% of cases. The reported rate of stenosis following intubation, ranges from 0.9-8.3%.[1] Other causes include external trauma, tracheostomy, radiation necrosis,[2,3] chronic infection e.g., syphilis, tuberculosis, typhoid and diphtheria[4] and other chronic inflammatory conditions such as Wegner's granulomatosis,[5] relapsing polychondritis, sarcoidosis, scleroderma[6] and systemic lupus.[7] Neoplasms have been also reported to cause subglottic stenosis, especially Chondroma.[8] Our patient has no history of endotracheal intubation, or upper airway trauma. Work up for the above mentioned chronic infections and inflammatory conditions and neoplasms, was negative. Idiopathic subglottic stenosis was reported in 52 cases over 34 years, by Dedo, *et al*.[9] All but one were females, suggesting hormonal cause. Moreover, Bruce described 15 females with idiopathic subglottic stenosis, between 1980-1994.[10] In a similar report of 16 patients with idiopathic subglottic stenosis, Valdez, *et al* reported two males only. Although the presence of sex hormone receptors has been demonstrated in the normal larynx by other investigators, these authors were unable to find any estrogen receptors in the 8 patients who were tested by immune histochemical analysis in Dedo's series nor in Valdez.[9,11]

A possible role for laryngopharyngeal reflux was proposed, but not confirmed. Maronian *et al*, focused on the role of laryngopharyngeal reflux as a possible causative factor in idiopathic subglottic stenosis.[12] Between 1991-1999, the authors reported nine cases of idiopathic subglottic stenosis and ten cases of idiopathic subglottic stenosis, associated with a concommitant disease state, including sarcoidosis, Wegeners, laryngeal trauma and history of intubation. In the

above series, 14 patients underwent a 24 hour ambulatory pH probe testing, with 3 or 4 port probes. The proximal port was placed directly behind the larynx. In 12 patients (86%), the pH was < 4.0. Five of the seven patients (71%) with idiopathic subglottic stenosis and seven of seven (100%) of idiopathic subglottic stenosis and concomitant disease, tested positive for reflux, despite therapy with proton pump inhibitiors, at the time of testing. Therefore, the author suggested a strong association between laryngopharyngeal reflux and idiopathic subglottic stenosis and a possible causation effect.

Similarly Jindal *et al*, reported seven women with idiopathic subglottic stenosis, with an age range of 39-66 years.[13] Six patients responded to surgical treatment only, after treating gatroesophageal reflux. This suggests that medical management of reflux is vital to successful treatment of idiopathic subglottic stenosis. The patient we described, had no symptoms suggestive of gastroesophageal reflux and upper gastroscopy was normal. The patient refused twenty four-hour pH testing.

Establishing the diagnosis of subglottic stenosis, should start with a high index of clinical suspicion, especially in cases labeled as refractory, bronchial asthma and patients with co-morbidities known to cause subglottic stenosis. This report stresses the importance of performing office spirometry and flow-volume loop, for patients with chronic respiratory complaints. Spirometry becomes an essential component of the clinical practice for the pulmonologist. Normal CT neck and chest does not rule out the diagnosis. Two reports described the use of helical CT imaging, to evaluate laryngotracheal stenosis, in patients who had previously undergone laryngotracheal resection.[14,15] The use of three dimensional skeletanization and helical CT scan, improves the detection rate.[14,15]

The basic principle for repair of subglottic stenosis, is providing substantial circular architecture, over which respiratory epithelium may regenerate.[16] Some of the endoscopic treatment methods used are, endoscopic dilatation, intralesional steroid injection,[17] carbon dioxide (CO_2) laser excision[9,18] or mucosal flaps. Each of these endoscopic methods has been successful in some cases, where the integrity of cricoid cartilage is not affected, as in web like stenosis of limited thickness,[8] in which simple dilation or CO_2 laser excision has a high success rate. However, in thick stenosis that compromises the cricoid cartilage either by collapse or scarring, simple endoscopic excision is not effective and the use of CO_2 laser is associated with high recurrence, which, could be more severe than the initial problem.[8] As the patient we described had a thin subglottic web, simple dilatation with rigid bronchoscope, was sufficient to restore the patency of the upper airways, without recurrence.

In Summary, upper airway obstruction, secondary to subglottic stenosis, may mimic bronchial asthma, which in turn may result in wrong diagnosis and treatment and a delay in reaching the correct diagnosis, that may jeopardize the patient's life. We stress the importance of performing office spirometry and assessment of the flow-volume loop, in patients with respiratory distress.

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