Idiopathic Subglottic Stenosis: a Case Report

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ABSTRACT

Idiopathic subglottic stenosis (iSGS) is a rare fibro-inflammatory disease of unclear aetiology of the subglottic area[1,2] presented by dyspnea and stridor due to upper airway narrowing. The diagnosis is arrived at after history taking, endoscopic examination, and radiological and laboratory investigations to exclude other causes of stenosis. Many approaches have been described for the treatment of iSGS, of which the most common is the endoscopic approach which is less invasive than any open surgery approach.[3]

Case Presentation

A 61-year-old Caucasian white female patient, with a history of breathlessness on exertion for nearly five years, which was assumed to be because of asthma, was referred to our ear, nose and throat (ENT) clinic due to worsening of noisy breathing mainly on exertion and at times during rest. The noisy breathing was subsequently diagnosed as asthma, and she had used corticosteroid and bronchodilator inhalers for several years, but with relatively little clinical improvement. In recent months the stridor and shortness of breath had worsened before she was referred to ENT. Her co-morbidities included diabetes mellitus type 2 and hypertension and she had been admitted to the ICU for post covid pneumonia one year ago, where she was treated with oxygen therapy without intubation. She had no history of neck trauma, recurrent upper respiratory tract infections, hemoptysis, reflux oesophagitis or any chronic granulomatous upper airway disease.

At clinic she presented with biphasic stridor. Examination by flexible nasolaryngoscopy revealed subglottic stenosis. A subsequent CT scan (Figure 1) confirmed a short thin 7x8 mm subglottic stenosis approximately 2cm below the vocal cords with the thickest anterior base measuring 6mm craniocaudally, the central aperture measured 7x8 mm. This corresponds to Myer's-Cotton grade II stenosis corresponding to a 51-70% reduction in the cross-sectional area of the airway[4] and McCaffrey stage 1. Antinuclear cytoplasmic antibody (ANCA) was negative and ACE (angiotensin-converting enzyme) level was within normal range, ruling out granulomatosis with **polyangiitis** (GPA) and sarcoidosis, which are known aetiological factors for iSGS.

As part of the assessment of the overall impact of this stenosis, lung function tests were carried out. The lung function tests showed that there was an impairment of most of the spirometry tests of lung function - PEF, FEV_1, FEV_1/PEF, forced expiratory flow [FEF]. However, on the flow volume loop studies (Figure 2), the most significant reduction of the flow

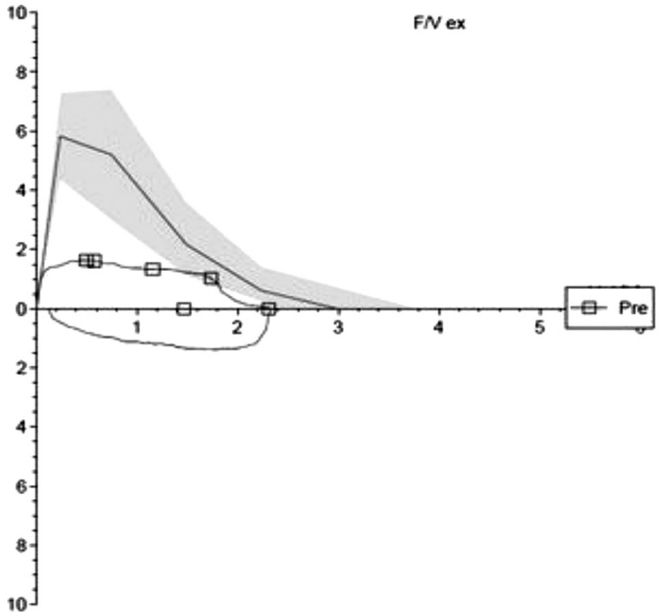

Figure 2: Flow-volume loop is demonstrating fixed upper airway obstruction [inner loop].

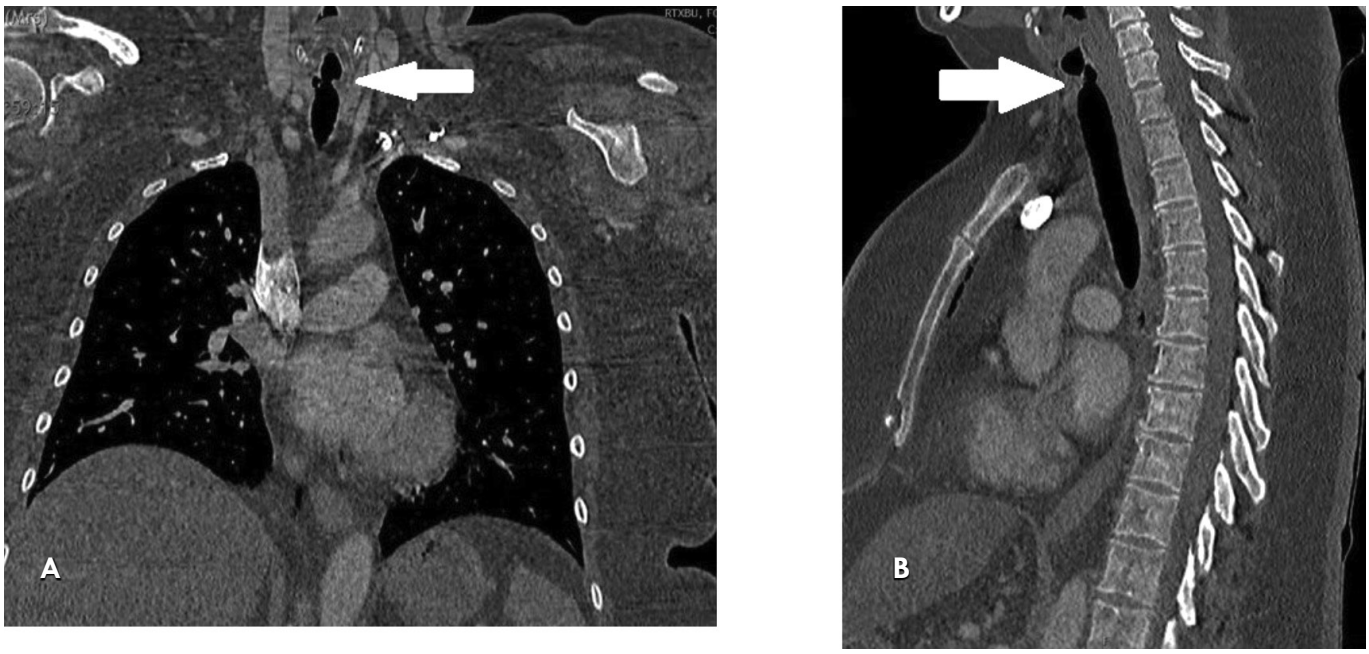

Figure 1: Coronal (A) and Sagittal (B) showing subglottic stenosis approximately 2 cm below vocal cord.

rates of both the inspiratory and expiratory curves was seen, with a typical flattening of the loop characteristic of fixed upper airway obstruction pattern.

A full evaluation was done by the otolaryngology, respiratory and anaesthesia teams. After discussing the possible lines of management, risks and complication with the patient, she underwent examination under anaesthesia and endoscopic balloon dilatation. Anaesthesia was delivered through the Jet ventilation technique. Dilatation was carried out using a Cook Hercules balloon 12-15mm then 15-18 mm (Figure 3); A and B show the significant improvement of the stenosis before and after the balloon dilatation without trauma to the vocal cords. Post-operatively the patient was transferred to surgical ITU for observation for 24 hours.

Two weeks after dilatation, the asthma inhalers were discontinued by the respiratory physicians as there was a significant improvement in her respiratory symptoms.

On follow up after three months, the patient reported continued improvement of her symptoms and appreciated her ability to do the usual daily activity with no shortness of breath and no noisy breathing anymore. She will continue to be followed up periodically, keeping in mind there is a chance of restenosis to occur in the future.

Case Discussion

Subglottic stenosis is a condition that is characterised by narrowing in the region bounded superiorly by a plane below the glottis and inferiorly by the first two tracheal rings.[5,6] It typically affects Caucasian women.[2] In one of the largest airway databases, the North American Airway Collaborative (NoAAC), 1056 patients were reported with iSGS and the demography of this condition showed it typically affects otherwise healthy, perimenopausal (mean age 50.4 years), Caucasian (95%) and female (98%) patients.[5]

Many etiological factors can lead to subglottic stenosis, the most common cause being mechanical trauma secondary to prolonged endotracheal intubation. Less frequently causes are external trauma, heat trauma, external beam radiotherapy, surgery, upper respiratory tract infections, granulomatosis with polyangiitis (former Wegener's granulomatosis), amyloidosis, or collagen vascular diseases.[7]

ISGS defined as subglottic stenosis of which the underlying cause cannot be determined[8] was first described by Brandenburg in 1972.[9] It is a rare disease with an estimated incidence of 1:400,000. It affects mainly Caucasian women aged 30–50 years and results in circumferential ring-like scar tissue in the subglottic area and upper trachea[10] that leads to significant morbidities, such as shortness of breath, respiratory distress, intolerance for physical activity and stridor.[11] Diagnosis of iSGS is based on the exclusion of other causes of SGS[12,13] and represents about 15% of cases of SGS.[2] Diagnosis is based on endoscopic examination of the larynx and trachea. Radiological investigations are crucial in determining the length and extent of the stenotic segment. Lung function tests show fixed upper airway obstruction pattern in the late stages of the disease while in early stages more subtle abnormalities could be seen. This explains why one-third of the patients misdiagnose with asthma or COPD at early stages.[1]

Apart from respiratory symptoms, some patients also report persistent coughing, as well as instances of voice changes. In several instances, the disease can present with recurrent stridor with the onset of upper respiratory infections and should be borne in mind as a potential cause of the symptoms.

Many hypotheses have been put forward to explain the pathophysiology of ISGS. Some authors believe that oestrogen may play a role in the development of stenosis, based on the fact that iSGS affects mainly women.[5,10] Other theories put forward to explain this condition include minor trauma from repeated cough,[13] a subtle manifestation of collagen vascular disease, an anatomical predisposition of the shorter female subglottis and gastro-oesophageal reflux disease (GORD).[14]

Treatment of iSGS is mainly surgical however use of anti-reflux treatment and corticosteroids can be used to decrease the recurrence. In 2020, Gelbard and his colleagues compared the most common surgical treatment approaches to iSGS (endoscopic dilation, endoscopic resection with adjuvant medical therapy, or cricotracheal resection. Endoscopic dilatation was the most popular approach.[5] Being a minimally invasive procedure, it was associated with lower risks and complications and does not affect the quality of voice. However, the rate of restenosis and the need for revision surgery may as high as 28%.[5]

Mitomycin-C has been used after dilatation or resection of scar to reduce re-stenosis but conclusive clinical data has been lacking about its efficacy in preventing re-stenosis.[15] Similarly, inhaled corticosteroids have also been used, but their efficacy is unknown for managing this condition from recurring in the long run after initial surgery.[16]

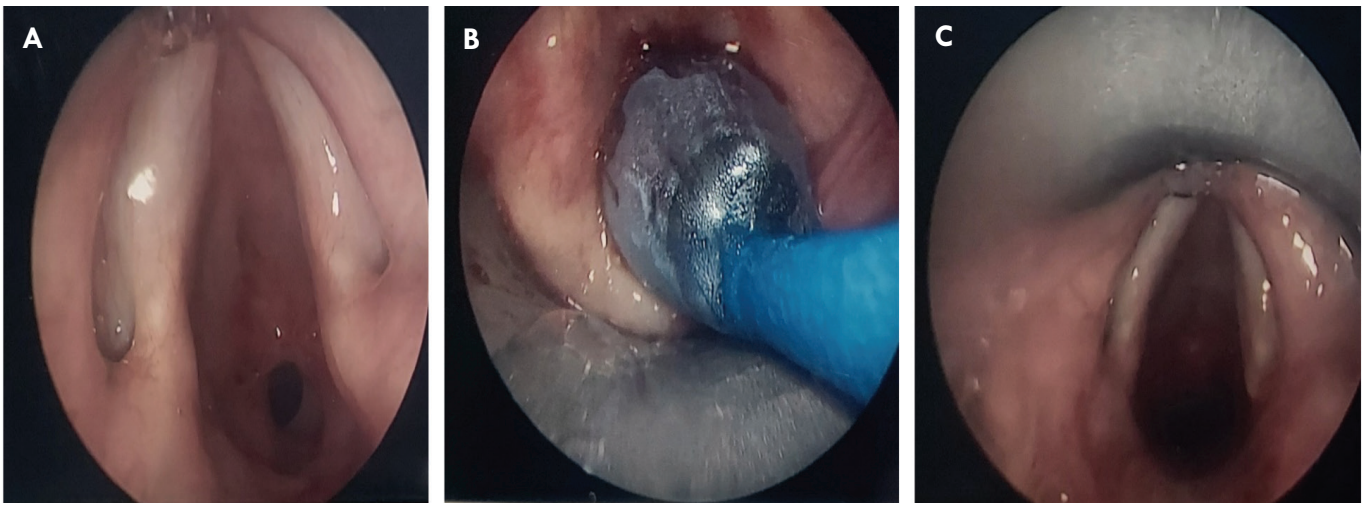

Figure 3: (A) Preoperative picture of iSGS, (B) Balloon dilatation using Cook Hercules balloo and (C) Post dilatation picture.

This case illustrates that being a relatively uncommon disorder, the diagnosis of iSGS may be delayed as a cause of long-standing shortness of breath. It implies that wherever shortness of breath and noisy breathing does not improve with medication, airway endoscopy should be a part of the next line of investigations.

Endoscopic balloon dilatation was the chosen surgical procedure in this case not only as it is less invasive for the patient, but also it ensures the best quality of voice in comparison with open surgery. However, long-term follow up is required due to the high rate of recurrence and the need for revision surgery.

Other surgical approaches can be divided into two main categories: (a) endoscopic resection of stenosis with prolonged medical treatment, afterwards with anti-reflux, antibacterial and inhaled corticosteroid medications. The resection has often been described with a C02 laser; and: (b) open surgery, either cricotracheal resection or laryngotracheoplasty, where the stenosed segment is resected and an end to end anastomosis of the upper trachea is carried out (cricotracheal resection) or cricoid cartilage is split and expanded with cartilage, usually with rib cartilage grafts. Although open procedures have good results, the main concern in this kind of surgery is the close proximity of the scar to the vocal cords and the persistent poor quality of voice. In our case, the stenosis was quite close to the vocal folds and was a major concern, which is why an endoscopic procedure was considered as the method of choice.

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