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Idiopathic subglottic-tracheal stenosis about a case

Summary

Idiopathic subglottic stenosis is a rare disease that occurs almost exclusively in women between 20 and 60 years of age and whose cause is unknown. It is a narrowing of the laryngeal lumen at the level of the cricoid cartilage in the upper airway. Idiopathic subglottic stenosis is a rare entity of unknown etiology, the incidence of which has not been established to date. The clinical case of a 75-year-old woman with progressive inspiratory dyspnea is presented. After nasofibrolaryngoscopy, a decrease in the subglottic-tracheal lumen is observed, an entity that has been highlighted for its low frequency in the otorhinolaryngology and head and neck areas.

Keywords: stenosis, idiopathic, subglottis, trachea, dyspnea

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Introduction

Idiopathic subglottic stenosis is a rare disease. It is characterized by a nonspecific inflammation of the tracheal submucosa in its upper third or in the subglottic area, which can progress towards scarring and circumferential stenosis. It occurs almost exclusively in women between 20 and 60 years of age and whose cause is unknown.[1] Stenotic pathology is classified as congenital or acquired, in the latter case it responds to multiple etiologies and may be primary/idiopathic or secondary to post-intubation tracheal stenosis, tracheal trauma (external and internal; inhalation burns, irradiation), specific and non-specific tracheal infections (bacterial tracheitis, tuberculosis, histoplasmosis and diphtheria), collagen diseases and vasculitis (Wegener's granulomatosis, relapsing polychondritis, polyarteritis, scleroderma and sarcoidosis), amyloidosis and, finally, congenital causes.[2] Some authors, such as Walner et al[3] and Koufman[4], have The presence of gastroesophageal reflux is related to the appearance of laryngeal lesions.[5] In the absence of an identifiable cause, it is known as idiopathic subglottic stenosis.[6]

The subglottis is more predisposed to presenting stenosis due to its narrower diameter, its lining tissue being more fragile and having a bed of poor vascularization.[7] Its incidence and pathophysiology are unknown; Some authors have suggested hormonal involvement, although estrogen receptors have not been demonstrated.[8,9] Patients usually experience mild dyspnea, lasting months to years and progressing to dyspnea at rest, which correlates with the degree of stenosis of the lumen, as well as wheezing or stridor, persistent cough, barking cough, hoarseness, and increased phlegm.[5] This fact leads to many patients being misdiagnosed and treated for asthma.[1] To diagnose this disease, the patient must not have any history that is related to the appearance of stenosis of the upper airway.[10] The duration of the symptoms from its onset until it is diagnosed varies from 4 months to 15 years, with a mean of 1 to 3 years in most cases.

The clinic of laryngotracheal stenosis is similar in all the causal pathologies, therefore, the diagnosis must be based on the biopsy, the general clinical syndrome of each patient and the laboratory with serology and antibodies; in case no causal pathology is found, the entity will be called idiopathic subglottic stenosis.[5] The definitive diagnosis is made by fiberoptic bronchoscopy, complemented by a high-resolution computed tomography and three-dimensional reconstruction to assess whether the stenosis is segmental or patchy, and the arrangement of the adjacent tissues.[5] In most of the patients described so far, the stenosis is located in the laryngeal subglottic portion and with different degrees of severity.[10] The maximum degree of stenosis is usually located at the cricoid level and at the highest level of the trachea. The lesions are most often of the circumferential type and the function of the vocal cords is usually intact. From the histopathological point of view, the biopsy samples show a fibrosis dense keloid type, with thickening of the adventitia and lamina propria, and with small areas of "spindle" cell proliferation similar to that seen in fibrosing mediastinitis, Riedel's thyroiditis or retroperitoneal fibrosis. Unlike postintubation strictures, the cartilage is intact and there is no ischemic necrosis at any level.[11] Regarding treatment, it remains challenging and controversial. It must be individualized according to each patient and type of stenosis. In each case, the main objective will be to restore a sufficient caliber in the airway to allow breathing and prevent dyspnea.[5] In order to approach an adequate therapy, it is necessary to rule out the rest of the etiologies. The treatment of subglottic stenosis if it is idiopathic is localized dilation by laser, tracheal reconstruction or balloon dilation; these treatments being conservative.[5] However, some authors defend these treatments due to the possibility of recurrence of the lesion, the morbidity and mortality associated with surgery and the high percentage of need for palliative techniques after surgical resection.[12] The purpose of this article is to present an infrequent clinical case of this pathology of slow and progressive evolution with an unknown cause.

Clinical case

A 75-year-old female patient with no known pathological history, no surgical history. He came to our consultation referred by the pulmonology service due to progressive inspiratory dyspnea, laryngeal stridor with effort and rest, without dysphonia of 2 years of evolution, previously ruling out pathologies by pulmonology and cardiology. On physical examination, a nasofibrolaryngoscopy was performed, visualizing decreased subglottic lumen with good mobility of the vocal cords, normal mucosa and no laryngeal lesion.

A stroboscopy is performed, visualizing subglottic stenosis of more than 60% of the glottic lumen (Figure 1), confirming with Computed Axial Tomography (Figure 2), reporting an infraglottic membrane that conditions slight reduction of the airway with the presence of a false lumen of 10 x 3 mm on the left side and 3 x 2 mm on the right side of the airway. In addition, complementary analytical studies to rule out inflammatory diseases as a possible cause, these being negative. He was scheduled to perform tracheostomy plus laryngeal microsurgery with CO_2 laser resection and topical Mitomycin C. Pathological anatomy sample is sent reporting Fibro-leukocyte material, Subglottic Lesion. Currently exhaustive control by outpatient, patient is without dyspnea, without stridor and decannulated with more than 1 year post-surgery.

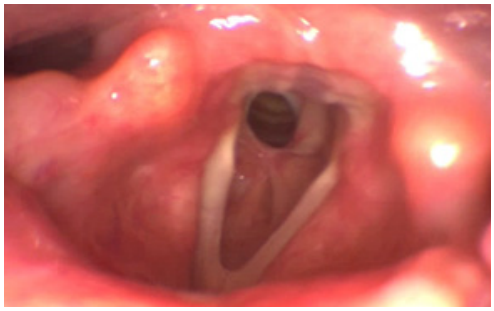

Figure 1 Glottic lumen.

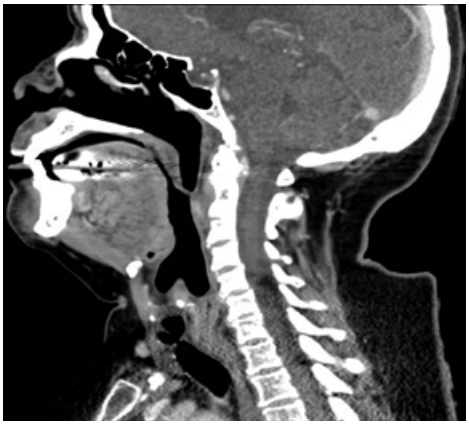

Figure 2 Computed axial tomography.

Discussion

Subglottic stenosis is the obstruction of the central airway in the region bounded superiorly by a plane below the glottis and inferiorly by the first two tracheal rings. When there is no obvious cause after a thorough evaluation, this disease process is known as idiopathic subglottic stenosis. Its estimated incidence is 1: 400,000. Being the first cases described by Brandenburg in 1972.[1]

In 2014, a retrospective cohort study gathered mostly female patients, with a diagnosis of idiopathic subglottic stenosis in 110 cases. The pathophysiology proposed by the authors speaks of lesions that occur in the laryngotracheal mucosa with abnormal healing and excessive fibroproliferative response. In this study, the objective was to evaluate the efficacy of endoscopic treatment with laser vaporization without dilation, and with infiltration of corticosteroids and mitomycin C at the lesion level. Recurrence at 5 years was 60%.[13]

In the retrospective study published in 2014 in the journal Laryngoscope, 92 patients with subglottic stenosis due to multiple causes were included, and the response to treatment was evaluated, always using endoscopy with different techniques; 45% required only one dilation procedure, while the cases that required a new procedure were approximately 14 months after the first intervention. There were no significant differences between the different endoscopic dilation techniques, and the use of local mitomycin C did not delay the need for a new intervention.[14,15] Between January 1, 1996 and January 1, 2005, 8 patients with idiopathic tracheal stenosis were treated in Valencia, in one case by surgery, so they were excluded from the study. All the patients were women and 7 of them were treated with periodic pneumatic tracheal dilations. Only 4 patients have required a dilation, who until now remain asymptomatic. Two have required a second dilation. The median symptom-free interval was 25.5 months. There was no associated mortality or morbidity.[10]

Grillo et al,[16] who in 1993 described the surgical treatment that is currently of choice in this disease: laryngotracheal resection in a single stage. In a series of 35 patients, excellent or good results were achieved in 91%. Mehta et al,[17] published a review and update of the series by Grillo et al,[16] and the results were consistent: of a total of 73 patients, the results were excellent or good in 91%. However, Dedo and Catten[12] believe that idiopathic tracheal stenosis is a progressive disease and that the best therapeutic option is periodic palliative treatment. In a recent series, in which Nd-YAG laser was used in 18 patients with concentric tracheal stenosis of different causes, only in two cases the etiology was unknown, in one of them the treatment was resolute and in the other it could not be performed. a follow-up of the patient. These two cases, unlike ours, were located in the distal third of the trachea.[18]

Axtel et al.,[18] reported their experience with single-stage laryngotracheal resection and reconstruction as definitive treatment for idiopathic subglottic stenosis. They reported a recurrence in 23 (8.7%) patients, determined by symptoms and bronchoscopic findings.[19] Black pudding et al. conducted a retrospective trial that included 64 patients who were treated by single-stage laryngotracheal resection, with a success rate of 97%.[20] These data suggest that, with respect to recurrence of stricture, open surgical techniques they have superior results compared to endoscopic techniques, but the minimally invasive nature and low complication rate make endoscopic treatment a good first-line treatment for idiopathic subglottic stenosis. However, given the enormous variation in the combination of endoscopic techniques used as well as all the confounding factors and the low quality of the studies, it is impossible to make any harsh statements about the "ideal endoscopic technique".

A study in 2022 by Alexander Gelbard et al.[20] comparing treatment outcomes for idiopathic subglottic stenosis with a total of 810 patients, 798 (98.5%) were female and 787 (97.2%) were white, with a median age of 50 years in the range of 43-58 years. The surgical index of the procedures were endoscopic dilation (n = 603, 74.4%), endoscopic recession with adjuvant medical therapy (n = 121, 14.9%), and cricotracheal recession (n = 86, 10.6%). Overall, 185 patients (22.8%) had a recurrent surgical procedure during the 3-year study, but recurrence differed by modality (cricotracheal recession, 1 patient [1.2%]; endoscopic recession with adjuvant medical therapy, 15 [12.4%], and Endoscopic dilatation, 169 [28.0%]).[20]

Conclusion

The clinic of subglottic-tracheal stenosis is similar for all the causal pathologies, with a slow and progressive evolution. Its diagnosis must be established with the clinic, examination, imaging studies, serology and biopsy to rule out the etiology, if the cause is not found we would be facing an idiopathic subglottic stenosis.

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Conflicts of interest

The authors state that there is no conflict of interest.

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