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Subglottic Stenosis due to Sjögren's Syndrome

Sjögren Sendromuna Bağlı Subglottik Darlık

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Abstract

A 36-year-old female patient diagnosed with asthma at an external center was referred to us after her complaints did not improve, at which point, hoarseness was added to the complaints. Stridor was identified in a respiratory system examination. Tracheal stenosis was seen in a postero-anterior chest X-ray and a fixed airway obstruction in a respiratory function test, upon which, the patient was referred to the ear, nose and throat department. Subglottic stenosis was detected on a neck computerized tomography and a bronchoscopy evaluation. The etiology of subglottic stenosis was evaluated, collagen tissue markers were positive, and the patient was asked to undergo a rheumatology consultation. The patient was subsequently diagnosed with Sjögren syndrome based on a salivary gland biopsy result. This rare case is presented to underline the need to keep Sjögren syndrome in mind as an etiology of subglottic stenosis.

Keywords: *Sjögren's syndrome, subglottic stenosis, chest disease.*

Öz

Otuz altı yaşında kadın hasta dış merkezde astım tanısı almış olup şikayetlerinin düzelmemesi üzerine tarafımıza başvurmuştur. Bu süreçte şikayetlerine ses kısıklığı da eklendi. Hastanın solunum sistemi muayenesinde stridor mevcuttu. Postero-anterior akciğer grafisinde trakeal darlık görüldü ve solunum fonksiyon testinde sabit hava yolu obstrüksiyonu mevcuttu. Bu neden ile hasta kulak burun boğaz konsülte edildi. Boyun bilgisayarlı tomografisi ve bronkoskopi değerlendirme sonucunda subglottik stenoz tespit edildi. Subglottik stenozun etiyolojisi açısından değerlendirildi ve kollajen doku belirteçlerinin pozitifti ve hastaya romatoloji konsültasyonu istendi. Tükürük bezi biyopsi sonucu ile hastaya Sjögren sendromu tanısı konuldu. Bu olgu, çok nadir de olsa, subglottik stenozun etiyolojisinden Sjögren sendromu akılda tutulması açısından sunulmuştur.

Anahtar Kelimeler: *Sjögren sendromu, subglottik stenoz, göğüs hastalıkları.*

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Connective tissue diseases are a heterogeneous spectrum of diseases with multiple organ involvement. Laryngeal involvement can be expected in connective tissue diseases, with the most commonly affected area being the cricoarytenoid joint. Failure to recognize laryngeal involvement due to connective tissue may be life-threatening due to the disruption of the airway (1).

Sjögren's syndrome is a chronic inflammatory, autoimmune disease characterized by lymphocytic infiltration of exocrine glands, affecting especially the salivary and lacrimal glands (1). The symptomatic findings of Sjögren's syndrome are diverse, as in addition to glandular findings there are also systemic clinical and extra-glandular findings (2). Dry eyes and mouth are the most common symptoms, caused by lymphocyte infiltration and dysfunction of the exocrine glands (3).

Although lung involvement is common in Sjögren's syndrome, the clinical symptoms are rarely important. The most common form of involvement is diffuse interstitial lung disease. Findings regarding pulmonary involvement in Sjögren's syndrome; It can manifest as interstitial lung disease, obstructive airway disease, pulmonary lymphoma and tracheobronchial dryness (4). The incidence of bronchitis, bronchiectasis, pneumonia and bronchiolitis increases due to the disruption of the secretion clearing mechanism and the drying of secretions (5). Wheezing and dry cough may develop due to the absence of secretions in the larynx, trachea and central bronchial branch points in cases with Sjögren's syndrome (6).

Subglottic stenosis was detected in the examination of the patient in the present study who presented to our clinic with complaints of hoarseness, dry cough and wheezing after being followed-up with a diagnosis of asthma and treatment at an external center. Examinations and consultations to clarify the etiology of the condition led the patient to be diagnosed with Sjögren's syndrome. We present this case of Sjögren's syndrome, a connective tissue disease, to highlight its ability to cause tracheal stenosis and particularly asthma, and due to the rareness of reports covering this condition in literature.

CASE

A 36-year-old female patient, whose consent was obtained for her detailing in this case report. presented to the chest diseases clinic of an external center with a 1-year history of shortness of breath and was subsequently diagnosed with asthma. The patient's complaints did not subside with medical treatment and hoarseness was added to the previously identified shortness of breath upon being hospitalized in our center for further diagnostic tests.

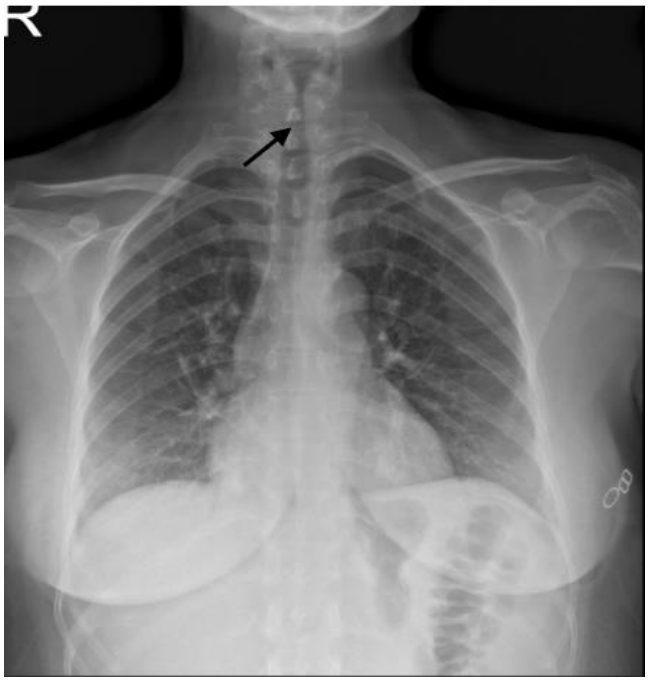

Figure 1: Narrowed lumen opening in the proximal trachea (Black arrow)

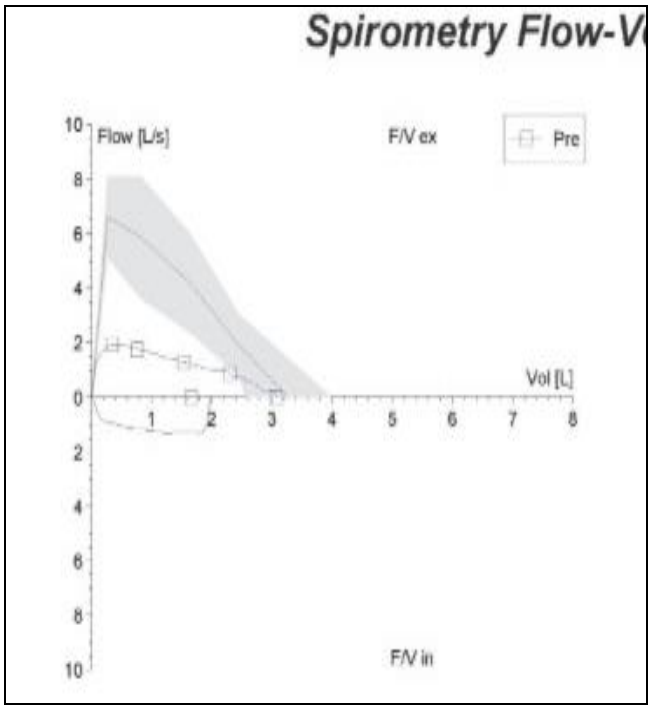

Figure 2: Plateau appearance in inspiration and expiration on spirogram

The patient had a history of hypothyroidism and was using levothyroxine sodium. She had no history of smoking and no history of recent surgery but had given birth to a cesarean delivery under spinal anesthesia 1 year earlier.

Here vital signs on admission were fever 36°C; Ta: 120/85 mmHg; pulse: 75/min; and respiratory rate: 19/min. Inspiratory stridor was identified during an examination of her respiratory system. Other system examination findings were normal, and her routine hemogram and biochemistry results were within normal limits.

A posteroanterior chest radiography was normal other than a suspicious calibration decrease identified in the upper sections of the trachea (Figure 1). In a respiratory function test, although the patient was unable to comply fully with the protocol, a flattening-plateau appearance was noted in both the inspiration and expiration loops in the flow-volume curve (compatible with fixed airway obstruction) (Figure 2).

The patient was asked to undergo an ear, nose and throat (ENT) consultation to identify the etiology of the stridor and hoarseness. A laryngoscopy performed by the ENT revealed stenosis in the subglottic area, and a neck CT was performed on the patient and neck CT interpretation: It was reported that the calibration of the tracheal diameter at the infraglottic level in the larynx decreased and was measured as 7 mm (stenosis?).

Based on the preliminary diagnosis of subglottic stenosis, we conducted a fiberoptic bronchoscopy (FOB) was performed ahead of surgical planning, revealing lesions in the form of mucosal swellings, starting at the vocal cord level, with 2–3 cm involvement proximal to the trachea, narrowing the lumen by 50%, and could not be progressed with bronchoscopy (Figure 3).

Lavage and biopsy were taken from the patient, who was subjected to rigid bronchoscopy and bougie dilation procedures by the thoracic surgeon, and the extracted samples were sent for microbiological and pathological examination, revealing intense mixed cellular inflammation, neovascularization and focal vasculitis-like leukocytoclastic activity in the sub-epithelial connective tissue.

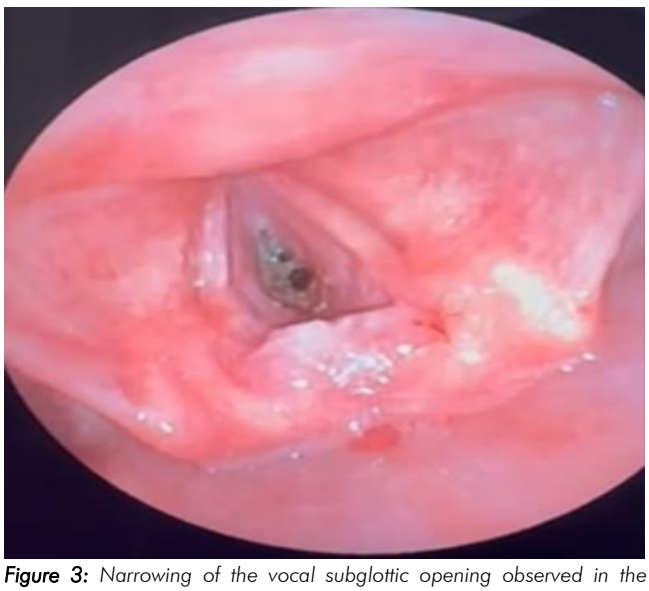

Figure 3: Narrowing of the vocal subglottic opening observed in the FOB

The patient's collagen tissue markers were studied revealing a positive ANA profile, and the SS-A/Ro 52 also came back positive, while the SS-B/La value and other markers came back negative. The patient had no family history of tuberculosis, and the ARB result from the sputum sample was negative with no bacterial growth identified in the sputum culture. Genetic testing for familial Mediterranean fever (FMF) and Behçet's disease were conducted with negative results, a Rose-Bengal test for brucellosis was also negative, and there was no recurrent polychondritis in the patient. When the patient's complaints were questioned in detail, she also reported a dry mouth, whereupon a salivary gland biopsy was performed by the ENT revealing chronic sialadenitis. The patient was diagnosed with Sjögren's disease based on her clinical, laboratory and pathological values by rheumatology, and was started on hydroxychloroquine and pilocarpine. A follow-up neck CT scan was performed at the end of the 4th month after the patient had started treatment, and the stenosis observed on the previous neck CT could not be detected (Figure 4). A control bronchoscopy performed on the patient 3 months later revealed the tracheal passage to be open with no evidence of stenosis (Figure 5). The patient's symptoms regressed with the disappearance of the tracheal stenosis.

DISCUSSION

Connective tissue diseases are known to affect the respiratory system at various levels to varying degrees. Airway involvement, especially in the trachea, is less common than lung parenchyma, and can be more difficult to diagnose. Sjögren's syndrome is a chronic autoimmune disease characterized by the lymphocytic infiltration of the exocrine glands, and especially the salivary and lacrimal glands (1). The effects on the mucus-secreting glands in the upper and lower respiratory tract lead to dryness in the trachea, nose and pharynx, and an increase in the frequency of pneumonia, bronchitis and bronchiectasis due to the disruption of the secretion-clearing mechanism and the drying of secretions (6,7).

In SLE, the most common lesions in the laryngeal region occur in the cricoarytenoid joint and glottis region, although other laryngeal regions may be affected. The symptoms of laryngeal involvement in SLE include hoarseness, dysphonia and sore throat (8). The diagnosis of SLE based on laryngeal findings can be quite difficult, and so micro-laryngoscopy and naso-endoscopy procedures may be required in SLE with upper respiratory tract symptoms (9).

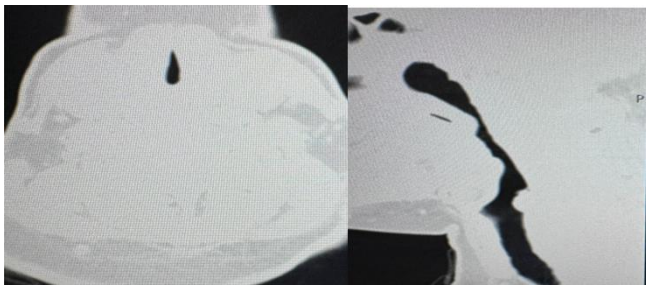

Figure 4: Post-treatment images of the patient's axial and sagittal CT sections

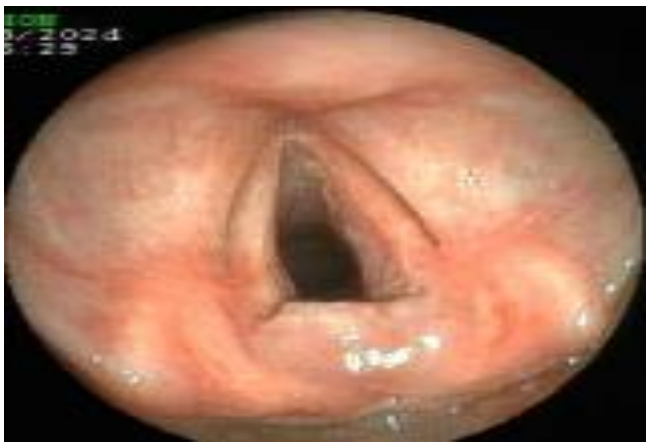

Figure 5: Subglottic opening achieved as observed in the control FOB

The subglottic region is the area extending from the vocal cords to the first tracheal ring. Subglottic stenosis that is unassociated with malignancy can be eliminated with simple surgical procedures (10). Subglottic stenosis is one of the main causes of chronic airway obstruction. Subglottic stenoses are congenital or acquired, intrinsic or extrinsic, and long segment or short segment (11).

The symptoms of subglottic stenosis vary depending on the size of the stenosis, with the most common symptom being aphonia. Stenosis may be confused with asthma and treated as such for years due to the presence of wheezing and cough, but can be correctly diagnosed when exertional dyspnea develops (12). The most common cause of subglottic space stenosis is abnormal wound healing resulting from damage caused by an endotracheal tube or tracheostomy. Other causes of subglottic stenosis include external factors such as malignancy, toxic inhalation, tuberculosis, vasculitis (especially granulomatous polyangiitis), diphtheria, polychondritis, sarcoidosis and goiters (13). Subglottic stenosis may occur alongside systemic inflammatory diseases, but may also be idiopathic. The condition occurs in 10–23% of patients with granulomatosis and polyangiitis (GPA) (14). While ear and nose involvement in GPA patients has a good prognosis, subglottic stenosis is a rare life-threatening finding (15). Subglottic stenosis can also occur alongside amyloidosis, sarcoidosis, cicatricial pemphigoid and inflammatory bowel disease (16), and the management of such forms of stenosis remains challenging, although systemic therapy and interventional endoscopy may be combined for a diagnostic biopsy along with specific procedures to improve morbidity and mortality (14). MALT lymphoma can be counted among the extranodal marginal zone B-cell lymphomas. Extragastric MALT lymphoma is thought to be predominant in such autoimmune diseases as rheumatoid arthritis, systemic lupus erythematosus, Sjögren's syndrome and Hashimoto's thyroiditis. Hematopoietic neoplasms are rarely found in the larynx, particularly in the subglottic region, and. can lead to subglottic stenosis (17). The primary imaging methods requested are lateral neck radiography and anteroposterior lung radiography. Chest X-ray can reveal any narrowing of the air passage in the laryngeal region. After tracheal stenosis has been detected, magnetic resonance and computed tomography imaging can be used to confirm the diagnosis (18).

Dryness and related infections have been found to develop due to the tracheobronchial involvements of Sjögren's syndrome, and may also lead to subglottic stenosis, as in the presented case. Similarly, our case leads us to believe that rheumatological diseases should also be kept in mind when investigating the etiological factors associated with tracheal stenosis. Another inference from this case is that making asthma in patients presenting with complaints such as shortness of breath, cough, wheezing can lead to delays in the correct diagnosis and treatment of the patient. As a final conclusion drawn from the presented case, while asthma may be the initial indication in patients presenting with such complaints as shortness of breath, cough and wheezing, tracheal stenosis should also be kept in mind in the differential diagnosis, and rheumatological diseases such as Sjögren's syndrome should be kept in mind in the differential diagnosis when investigating the etiology of subglottic stenosis.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - G.B.K., S.B., Ş. K.; Planning and Design - G.B.K., S.B., Ş. K.; Supervision - G.B.K., S.B., Ş. K.; Funding - G.B.K., S.B., Ş. K.; Materials - G.B.K., S.B., Ş. K.; Data Collection and/or Processing - G.B.K., S.B., Ş. K.; Analysis and/or Interpretation - G.B.K., S.B., Ş. K.; Literature Review - G.B.K., S.B., Ş. K.; Writing - G.B.K., S.B., Ş. K.; Critical Review - G.B.K., S.B., Ş. K.

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