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CASE REPORT – OPEN ACCESS

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Tracheal Stenosis Related to Ulcerative Colitis

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ABSTRACT

Background: Ulcerative colitis (UC) is a chronic inflammatory disease of the intestines. UC-related respiratory disease is fairly rare. Early diagnosis is important and can prevent significant potential effects, but can be challenging, due to mild symptoms and inconclusive radiology findings. This report describes some elements that can assist with early diagnosis.

Case Report: A 41-year-old female with diagnosed UC and Takayasu arteritis presented with a dry cough ongoing for 3 weeks. The patient had experienced no recent symptoms related to UC and had discontinued use of medication. A pulmonary function test (PFT) indicated an obstruction during inhalation and exhalation in the flow-volume loop. Irregular diffuse tracheal wall thickening was observed on chest computed tomography images. Bronchoscopy revealed that the trachea/bronchus had a rough, hyperemic appearance. Lymphoplasmacytic inflammation observed in the bronchus biopsy sample was compatible with UC-related respiratory disease. No symptoms or obstruction were seen in the PFT after 2 months of treatment with azathioprine and methylprednisolone.

Conclusion: Early bronchoscopy in patients with unclear radiological findings is important for an early diagnosis and the prevention of potentially irreversible complications in UC-related respiratory disease.

Keywords: Bronchi, stenosis, trachea, ulcerative colitis, vasculitis

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INTRODUCTION

Ulcerative colitis (UC) is a chronic inflammatory disease of the gastrointestinal tract. Numerous extraintestinal manifestations are possible. Pulmonary involvement is fairly rare; however, it can present in the form of airway or parenchymal disease. The most common respiratory manifestation is bronchiectasis, which is a result of irreversible airway destruction secondary to inflammation (1, 2). In addition, larger airways, such as the trachea, may be affected in UC, but the diagnosis may be delayed and result in stenosis. The primary reasons for delay are mild respiratory symptoms in the beginning, unclear radiological findings, and no gastrointestinal activation symptoms. This report describes the case of a patient with UC-related tracheobronchitis diagnosed early after a detailed evaluation.

CASE REPORT

A 41-year-old woman presented with a severe, dry cough ongoing for 3 weeks. She had no other symptoms. She had been diagnosed with UC 7 years prior and recently diagnosed with Takayasu arteritis, which was identified after the evaluation of asymmetrical blood pressure between the right and left upper extremities. There were no symptoms related to Takayasu arteritis. From time to time, she had experienced abdominal pain or hematochezia in the past, but she had no recent symptoms of activation. She had been taking mesalazine, azathioprine, and methylprednisolone, but had discontinued use of her medication 10 days prior to presentation. She had no pet at home and no occupational exposure that might explain the cough. Her family history was unremarkable. A physical examination was normal, with the exception of a stridor that was appreciable during both forced inspiration and expiration. Biochemical test results and a blood cell count revealed anemia, an elevated C-reactive protein level, and a positive finding for perinuclear antineutrophil cytoplasmic antibodies (pANCA). Occult blood was present in the stool with no parasites observed in a direct examination. A pulmonary function test (PFT) revealed a mild decrease in both forced expiratory volume in 1 second (FEV1) and forced vital capacity (FVC), with a ratio of 0.74 and an obstruction during inhalation and exhalation in the flow-volume loop (Fig. 1a). A posteroanterior chest X-ray was normal. A chest computed tomography (CT) image revealed irregular diffuse tracheal wall thickening, starting 6 cm below the vocal cords and extending to the left main bronchus (Fig. 2). Tumor positron emission tomography-CT results were normal. Evaluation of the trachea for possible tracheal pathologies, such as amyloido-

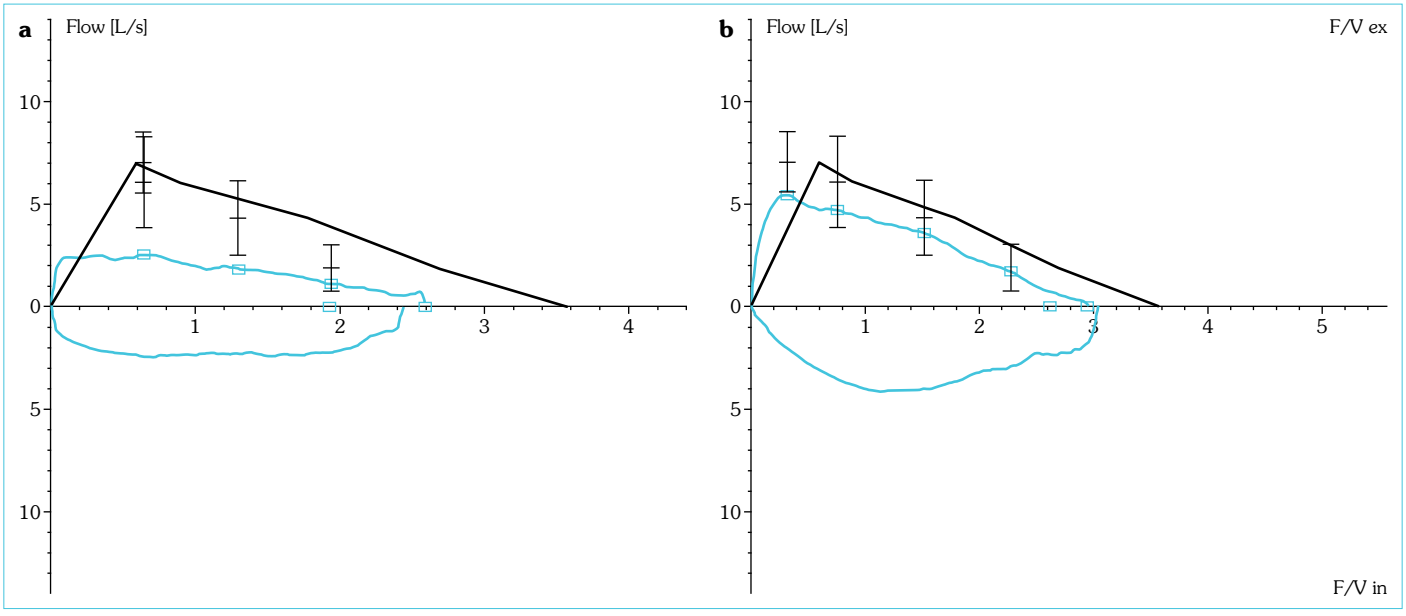


Figure 1. (a) Pre-treatment pulmonary function test with a plateau visible in the expiratory and inspiratory flow/volume loop; (b) Post-treatment pulmonary function test with a normal flow/volume loop

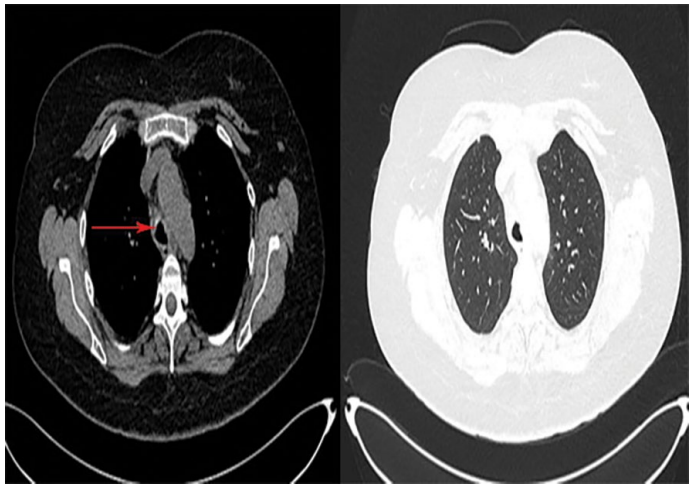


Figure 2. Chest computed tomography images with visible irregular diffuse tracheal wall thickening

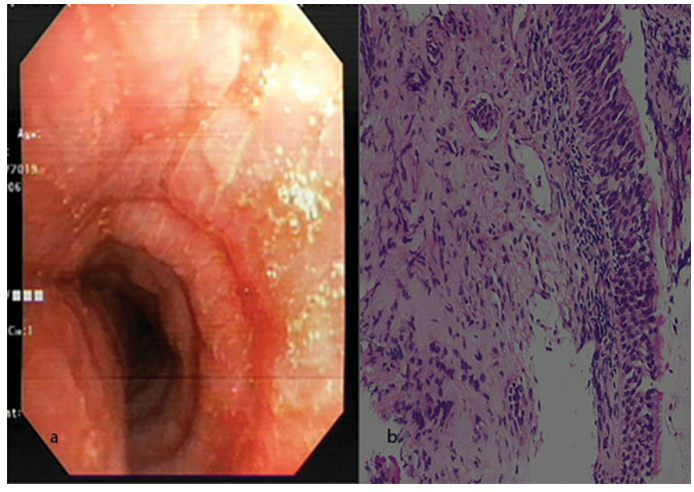


Figure 3. (a) Diffuse and irregular tracheal wall thickening with a cobblestone-like appearance seen with fiberoptic bronchoscopy; (b) Histology image of lymphocytic and plasmocytic inflammation and vascular proliferation under the epithelium of the bronchus

sis, Wegener's granulomatosis, papillomatosis, sarcoidosis, or UC-related tracheobronchitis was performed via flexible fiberoptic bronchoscopy. The vocal cords and the subglottic third of the trachea were normal. The remainder of the trachea and the left main bronchus walls were slightly hyperemic and rough in appearance, resembling cobblestones (Fig. 3a). A histopathological evaluation of the bronchus biopsy sample revealed vascular proliferation and lymphocytic and plasmacytic inflammation under the epithelium (Fig. 3b). Following consultation with a rheumatologist and a gastroenterologist, it was decided that she did not have ANCA-associated vasculitis, the Takayasu arteritis was stable, and that the UC was active. Treatment with azathioprine and methylprednisolone was reinitiated. Her cough had decreased on the fifth day of treatment. After 2 months, there was no cough or stridor. Pulmonary function tests showed a normal FEV1 and FVC, with a ratio of 0.86, and a normal flow-volume loop (Fig. 1b).

DISCUSSION

UC is an inflammatory bowel disease with an unknown cause. Sometimes, inflammation is not limited to the intestines. Extraintestinal signs and symptoms can vary according to the organ affected by inflammation related to UC. Although the pathogenesis of respiratory tract involvement in UC is unknown, it is believed that an undetermined systemic stimulating factor may be responsible for the common inflammatory response in both the intestinal and bronchial epithelium (3). Inflammatory changes in the airways are thought to be similar to inflammatory changes in the small intestine during active disease, based on the fact that both systems develop from the primitive intestine and have similar columnar cells in their mucous membrane. Airway mucosa surfaces have been shown to

have an appearance suggestive of cobblestones, similar to those in the affected intestine (4). Similarly, endobronchial biopsies often show chronic lymphocytic inflammation in the lamina propria and the epithelium without granuloma formation. In our case, there was evidence of vascular proliferation, lymphocytes, and plasma cells under the epithelium of the bronchi.

Respiratory system diseases associated with UC may vary from airway disease (tracheobronchitis, tracheal stenosis, bronchiectasis, granulomatous bronchiolitis) to parenchymal lung disease (diffuse or localized interstitial pneumonitis and fibrosis) (5). Airway involvement is more common than parenchymal involvement. Approximately 66% of airway involvement is in the form of bronchiectasis, while the majority of the rest is in the form of acute or chronic tracheobronchitis, and rarely, tracheal stenosis (1). Patients with tracheal involvement are often misdiagnosed with infectious tracheobronchitis and inappropriately prescribed antibiotics. The fact that the early signs and symptoms are typically mild and may appear independently of gastrointestinal activation makes diagnosis difficult. However, delayed diagnosis and treatment can result in tracheal inflammation becoming tracheal stenosis (6). A wide variety of drugs are now available for the treatment of UC. Steroids are highly effective for acute UC, but to treat patients with a more complicated disease course, other drugs, including azathioprine, biological agents, Janus kinase inhibitors, and calcineurin inhibitors, may be preferred (7). With appropriate and timely treatment, the airway mucosa may return to normal appearance. But delay may result in stenosis. Tracheal and bronchial stenosis can be treated with bronchoscopic interventions, such as balloon dilation and stent application. Stenosis can also be destroyed using an endoscopic yttrium aluminum garnet laser in order to achieve tracheobronchial patency (8).

Bronchiolitis or small airway involvement is a less common form of UC-related lower airway involvement. Tree-in-bud opacities, mosaic attenuation, and centrilobular ground-glass nodules are common CT findings of bronchiolitis. UC patients presenting with scattered tree-in-bud opacities (bronchiolitis) and diffuse tracheobronchial wall thickening (tracheobronchitis) can also respond well to steroid therapy with no need for any further immunosuppressive medication (9).

Although our patient had a cough for 3 weeks and did not have diarrhea at the time of presentation, the presence of hemoglobin in the stool and intermittent gastrointestinal complaints suggested UC activation. Furthermore, a chest CT, which was performed following the determination of airway obstruction with the PFT and the presence of stridor, revealed diffuse thickening of the trachea. However, it should be kept in mind that sometimes radiological findings may be uncertain and evaluated as normal. Therefore, even if the radiological finding is normal, patients with stridor due to the tracheal narrowing observed in the physical examination and airway obstruction in a PFT should be evaluated by bronchoscopy. Aside from the necessity of bronchoscopy for the diagnosis of UC-related airway disease, it is required to exclude other tracheal diseases, such as amyloidosis, tracheobroncopathia osteochondroplastica, Wegener's granulomatosis, tuberculosis, and tracheal papillomatosis. Our case was a rare example of UC-related endobronchial involvement and represents a valuable contribution to the literature.

In conclusion, large airway involvement may vary from mild chronic lymphocytic inflammation to severe stenosis. Airway diseases associated with UC can be missed in the early stages due to the nonspecific, mild signs and symptoms, especially in the absence of symptoms of gastrointestinal disease activation. Prevention of irreversible complications of endobronchial involvement requires an early diagnosis and treatment. Therefore, early bronchoscopic examination in patients with respiratory symptoms is important.

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