RESOLUTION OF BRONCHOMALACIA PRESENTING AS SEVERE ASTHMA BY ENDOSCOPIC INTERVENTION

Difficult to control asthma should make us think of other concomitant airway diseases, mainly if the treatment for the initial presumed diagnosis is failing. A 42-year-old female smoker was referred to our allergy department because of symptoms of urticaria and facial and eyelid edema in the month before her visit. A detailed allergy history also revealed intermittent cough and dyspnea with no apparent trigger or seasonal changes. The patient had no known allergies or atopic diseases. A physical examination revealed diffuse wheezing in both lungs. The results of a test for dermographism were negative. Results of skin prick tests with a panel of food and aeroallergens were negative. Spirometry revealed a clear improvement (FVC, 4.07 L; FEV1, 2.04 L; FEV1/FVC ratio, 65%), and a bronchodilator test result was negative. Results of plethysmography and a diffusion test, the results of both of which were normal. Induced sputum analysis showed Curschmann spirals and leukocytes (99% neutrophils, 1% eosinophils).

A chest computed tomogram with contrast showed mild emphysema in the upper lobes of the lungs. Respiratory infections persisted for the following 2 months, and some exacerbation required care from the emergency department. Spirometries showed no improvement. Because of a lack of improvement, a bronchoscopy was performed.

Bronchoscopy revealed a bronchial collapsibility, with significant loss of airway caliber in the left main bronchus, resulting in severe bronchomalacia. Days later, the bronchoscopist placed a Dumon prosthesis in the left main bronchus using an Efer-Dumon rigid bronchoscope (Fig 1).

In subsequent monthly visits, the patient reported good tolerance to the prosthesis and demonstrated clinical improvement. Spirometry also revealed a clear improvement (FVC, 4.07 L; FEV1, 2.04 L; FEV1/FVC ratio, 65%), and a bronchodilator test result was negative. Tracheobronchomalacia is a central airway disease characterized by an increased susceptibility of the tracheal and bronchial lumen to collapse during expiration. Occurring rarely, broncho-

Figure 1. Placement of the prosthesis in the left bronchus.
malacia is often mistaken for other more common obstructive airway disorders. This disorder is encountered in both pediatric and adult medicine. Its prevalence in adults is unknown because the available data are derived from studies conducted in selected populations and not in the general population, but it can be a more common finding in patients with chronic obstructive pulmonary disease and asthma.

In our case, flow-volume loops did not help to establish the diagnosis because typical findings, such as a rapid reduction in maximum expiratory flow after the initial peak, notching of the expiratory descent, and oscillations in the curve, were shown, likely because the trachea was not affected enough to induce a collapse that can be detected in flow-volume curves. The computed tomogram was not performed in inspiration and expiration; therefore, it was not possible to evaluate the reduction in the cross-sectional area from the bronchus. The bronchoscopy is the “gold standard” technique that allows a definitive diagnosis. In conclusion, we describe a patient with severe nonallergic asthma whose outcome was poor despite the prescribed treatment. An endoscopic examination revealed severe bronchomalacia. Placing a Duman prosthesis on the more affected bronchus resulted in clear clinical and spirometric improvement. Bronchial endoscopy should be considered in cases of non-controlled severe asthma to rule out endobronchial diseases.

EOSINOPHILIC FASCIITIS MIMICKING ANGIOEDEMA AND TREATMENT RESPONSE TO INFILXIMAB IN A PEDIATRIC PATIENT

Eosinophilic fasciitis (EF) is a rare scleroderma-like condition that was first described by Shulman in 1974 and characterized by eosinophilic inflammatory fascial infiltrate. The origin of EF remains unclear, and there have been only approximately 200 cases reported to date. Although the onset of symptoms has been linked with severe exercise, use of medication, trauma, and insect bites, no singular inciting event or trigger has been identified. Symptoms characterize-ically include progressive and symmetric erythema, edema, stiffness, painful extremities, and skin induration. The face, hands, and feet are usually spared. EF is associated with peripheral blood eosinophilia (eosinophil count >500/μL) in 65% to 80% of patients. Hypergammaglobulinemia and elevated acute-phase reactants are also commonly seen. Because the mean age of onset is 40 to 50 years, EF is extremely uncommon in the pediatric population.

Full-thickness (skin, fascia, and muscle) biopsy is the “gold standard” test to diagnose EF and shows inflammation and thickening of collagen bundles in muscle fascia with infiltration of lymphocytes, plasma cells, and eosinophils. Magnetic resonance imaging (MRI) is also a useful tool to diagnose and follow up EF. It detects fascia thickening and shows high signal intensity on T2-weighted sequence of muscle fascia while muscle pattern is normal. Most patients respond to moderate- or high-dose corticosteroids. Methotrexate, azathioprine, cyclophosphamide, and infliximab have been used in corticosteroid-resistant patients.

A 5-year-old, previously healthy boy developed acute, diffuse edema and erythema of the face, abdomen, and extremities, including the hands and feet. The swelling and erythema were painless. There was no precipitating trauma or history of exposure to new foods or medications. His parents reported that he was playing outdoors just before the episode began. On the basis of his symptoms and physical examination results, he was initially diagnosed as having idiopathic angioedema and treated with antihistamines and oral steroids. When the symptoms recurred 2 weeks later, further evaluation was conducted. Family history was significant for a first cousin with EF. The patient’s white blood cell count was 12,600/μL, with 51% eosinophils (absolute eosinophil count of 6426/μL). The patient’s white blood cell count was 12,600/μL, with 51% eosinophils (absolute eosinophil count of 6426/μL). The eosinophilia and persistence of symptoms led to an inpatient admission. Infections, malignant neoplasm, immunodeficiency, and drug reaction were ruled out. Tests for parasitic infections were unrevealing. Lyme titers were negative. Bone marrow biopsy results were normal. T-cell and B-cell subsets were normal in number, morphology, and function. C3, C4, C1q, C1 esterase inhibitor, and immunoglobulin levels were normal. To rule out myeloproliferative hypereosinophilic syndrome, genetic testing was performed, and the results were negative for gene deletion associated with FIP1L1/ PDGFRα fusion. Lower-extremity MRI showed thickened and increased signal in the subcutaneous soft tissue fascia surrounding the muscles. A full-thickness skin biopsy specimen showed fascia and adjacent adipose tissue with perivascular eosinophilic infiltrates and eosinophils in a deep dermal perivascular pattern consistent with EF (Fig 1).

On the basis of the biopsy and MRI results, the patient was diagnosed as having EF. He was initially treated with oral steroids (2 mg/kg) and methotrexate (20 mg) but continued to have progressive subcutaneous induration and developed lower-extremity peau d’orange changes. The therapy was escalated to weekly intravenous methylprednisolone pulses (30 mg/kg), oral steroids (2 mg/kg), subcutaneous methotrexate (25 mg), and infliximab (initially 10 mg/kg and then 15 mg/kg). With this regimen, his edema, erythema, and induration markedly improved and his eosinophil count decreased during several months to 2.5% or 153/μL. During this time,