Case Report

A rare aspect of Crohn's disease: Pulmonary involvement in a child

Ebru Atike Ongun, Reha Artan[1], Aysen Bingol[2], Oguz Dursun

Abstract

Crohn's disease (CD), known as the disease of gastrointestinal system, is a granulamatous systemic disorder with extraintestinal manifestations including the respiratory system. The resemblance in the embriological origins and the immunities of both organ systems' mucosae, also the circulating immune complexes and the autoantibodies are accepted as contributing factors. The shift of inflammation may become prominent when the colon is removed after colectomy and independent of the bowel disease activity; pulmonary involvement may be exarbecated. In the pediatric population, CD associated pulmonary involvement is very rare, mainly in the form of subclinical alterations and the data are limited mostly to case reports. Therefore, it is possibly overlooked since the diagnosis relies on suspicion. We represent a 5-year-old CD patient with previous bronchiolitis episodes that might have resulted from CD-associated pulmonary involvement; whom later developed severe pneumonia resulting in acute respiratory distress syndrome and bronchiectasia following a colectomy operation.

Keywords: Acute respiratory distress syndrome, colectomy, Crohn, infliximab, pulmonary

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Introduction

Pulmonary involvement is a rare manifestation of Crohn's disease (CD). The clinical spectrum is wide from subclinical alterations to life-threatening respiratory problems.[1] In the pediatric population, the data are limited mostly to case reports;[2] subclinical alterations in the form of latent pulmonary abnormalities are observed.[3] Unlike the observations in adults, symptomatic lung disease appears uncommon in children.[4] Following a colectomy, which is accepted as a triggering factor, the shift of inflammation results in prominent pulmonary disease.[5]

We represent a 5-year-old CD patient with previously unknown CD-related lung disease, developed severe pneumonia, acute respiratory distress syndrome (ARDS), and bronchiectasis following a colectomy operation.

Case Report

The case is a 5-year-old CD patient, diagnosed at the age of three, with an older brother suffering from the same disease. He had a history of bronchiolitis episodes in the past and was given anti-tumor necrosis factor drug (infliximab) thrice in the past 4 months due to protein-losing enteropathy and recurrent intestinal pseudo-obstruction. He was admitted to hospital with abdominal pain and bloody stool; 40 cm of gangrenous bowel of the ascending colon was removed. In the postoperative period, he developed

From:
Department of Pediatrics, Division of Pediatric Critical Care, School of Medicine, Akdeniz University, [1]Department of Pediatrics, Division of Pediatric Gastroenterology, School of Medicine, Akdeniz University, [2]Department of Pediatrics, Division of Pediatric Pulmonology, School of Medicine, Akdeniz University, Antalya, Turkey

Correspondence:
Dr. Ebru Atike Ongun, Department of Pediatrics, Division of Pediatric Critical Care, School of Medicine, Akdeniz University, Dumlupınar Bulvarı 07058 Kampus, Antalya, Turkey.
E-mail: ebruongun@akdeniz.edu.tr

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pneumonia leading to ARDS (PaO_2/FiO_2 <100) and ventilated according to lung protective ventilation strategy in Pediatric Intensive Care Unit (PICU). He had a fever, hypotension, tachycardia, tachypnea, and bilateral crackles in auscultation. The laboratory results were as followed - arterial blood pH: 7.10, pCO_2: 55, pO_2: 95, HCO_3: 17, BE: −7, O_2 saturation: 98%, lactate: 6.8; hemoglobin: 9.9 g/dL, white blood cell count: 19,300/mm^3, platelets: 61,000/mm^3, peripheral blood smear: Increased neutrophil band count, C-reactive protein (CRP): 10 mg/dL (0–0.5 mg/dL), procalsitonin: 100 ng/mL (0–0.5 ng/mL). Chest radiography showed diffuse infiltration and consolidation. Antibiotics and vasopressors were initiated to treat pneumonia and septic shock. Pseudomonas aeruginosa was detected in tracheal and blood cultures. The patient developed oliguric renal failure and received continuous venovenous hemodialysis for 10 days. Later on the course, the fever had subsided down; laboratory findings were back to normal except CRP. Cultures taken in the following days were sterile. Despite a general well-being in the clinic, weaning attempt had failed at day 14 and re-intubated. The ventilator settings showed a restrictive pattern with low compliance (pressure/volume slope).

The need for ventilatory support and the previous history of the patient before progression of ARDS had led us to evaluate possible inflammatory problems caused by CD. High-resolution computerized tomography (HRCT) showed interlobular septal thickening, air bronchograms, consolidated areas with bronchiectasia, and ground glass opacities. Histopathological study could not be done due to parental disapproval for bronchoscopy. Instead, we studied cytological evaluation of the deep tracheal aspirates, demonstrating neutrophilic infiltration. The aspirates were also evaluated for mycobacterium tuberculosis and viral respiratory organisms to rule out infliximab-related infections (all were negative). The immunological profile (immunoglobulins) was normal. Tracheostomy was performed at day 23. We initiated prednisolone 1 mg/kg/day to resolve the restrictive respiratory pattern and mediate anti-inflammatory effect caused by a possible lung involvement as revised in the literature.[5] By the 96th h of initiation of steroid, the patient's clinical condition and pulmonary functions improved dramatically; CRP returned to normal. After 32 days of intubation, without mechanical ventilator support, he was discharged from PICU with a tracheostomy.

Discussion

CD-related pulmonary involvement in pediatrics is often disregarded since the literature is limited mostly to the case reports.[2] The clinical spectrum is wide from subclinical alterations, airway, and parenchymal disease to pleural disease or drug-induced disease.[1,6] Asymptomatic pulmonary involvement has been widely reported.[7] Pulmonary function tests (PFTs) are recommended;[5] decreased diffusing capacity and restrictive pattern in pulmonary functions along with bronchial hyperresponsiveness (BHR) even in the absence of clinical evidence have been documented at all ages.[3,8] Relevant with the previous data, our patient's compliance (pressure/volume) and resistance (flow/volume) slopes on the ventilator showed predominantly restrictive pattern of a respiratory problem.

Symptomatic lung disease appears uncommon and occurs mainly in adults.[4] Bronchiolitis is the main form of small airway disease, occurs at younger ages, earlier in the disease course, and pulmonary disease precedes intestinal disease in one-third of cases.[4] Our patient had a background of bronchiolitis episodes, but no PFT was applied to determine BHR and subclinical alterations in the past. In terms of airway manifestation, the most common form is the bronchial inflammation with or without bronchiectasis[1] and may become apparent after bowel surgery.[9] Our patient's HRCT result was consistent with the literature; since bronchial wall thickening, bronchiectasia, cysts, emphysema, ground-glass, and reticulonodular opacities can be observed in HRCT.[6] The bronchoalveolar lavage (BAL) studies point out neutrophilic or lymphocytic inflammation in the bronchi and alveoli of the patients.[6] We could not examine BAL due to parental disapproval for broncoscopy, nevertheless, the cytological evaluation of the deep tracheal aspirations obtained from our patient, which demonstrated neutrophilia consisted with inflammation in the lungs. Since CRP is a well-known marker for disease activity in inflammatory bowel disease,[10] a rapid ameliorate in CRP after initiation of prednisolone, which is accepted as a treatment option in CD,[1] has led us to think the inflammation might be the result of a CD-associated pulmonary disease.

Not only the disease itself, but the drugs used in CD might also cause lung damage. Infliximab is associated with opportunistic infections, ARDS, diffuse alveolar hemorrhage and interstitial lung disease.[11] The complications are reported due to delayed immunological lung damage in the form of the interstitial disease has been reported.[11,12] Due to parental disapproval for bronchoscopy, we could not differentiate the cause of the inflammatory lung disease by histopathological examination. Nevertheless, the patient's history of bronchiolitis episodes in the past suspecting of a

subclinical pulmonary involvement, the aggravation of respiratory symptoms after colonic surgery which is accepted as a triggering factor,[5] a sudden decline in CRP and dramatic clinical improvement after the initiation of prednisolone appointed us the higher possibility of CD-related pulmonary involvement.

Finally, the studies on CD related pulmonary involvement in children are very rare and possibly overlooked. Suspicion is the key point to diagnose and treat the patient accordingly in case of refractory respiratory problems when the histopathological examination is unavailable. Performing comprehensive analyses of pulmonary functions prior to progression of symptomatic respiratory problems are recommended during routine follow-up of a CD patient.

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Conflicts of interest

There are no conflicts of interest.

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