

THERAPEUTIC FLEXIBLE BRONCHOSCOPY IN CHILD WITH CYSTIC FIBROSIS

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ABSTRACT

The report deals with the case of a 10-year-old girl with chronic cystic fibrosis. She has been repeatedly treated at the hospital. She has been hospitalized due to respiratory deterioration. Cystic fibrosis is a rare disease, inherited autosomally recessively, but is very complex in terms of diagnostic and treatment (2). The diagnosis is confirmed based on a clinical picture of the child, measure of Chloride in the sweat, chest X-ray, CT thorax, laboratory findings - genetic confirmation CFTR (cystic fibrosis transmembrane conductance regulator) genes (3), which result in the production of hyper-viscous mucus and chloride malabsorption in the sweat glands ducts (5,6). Bronchial thickening and plugging and ring shadows suggesting bronchiectasis, segmental or lobar atelectasis are often. Computer tomography of the chest can be used to detect and localize thickening of bronchial airways walls, mucus plugging, hyperinflation and early bronchiectasiae. Pulmonary therapy: the object is to clear secretions from airways and to control infection (7). The diagnosis is originally set when she was 4 years old. She is now admitted due to a deterioration of the main disease. Day before admission in the hospital had a higher bodily temperature, cough and difficult breathing. She already treated conservatively (Ceftazidim, Ceftriakson, Kloksacillin) Since the girl is a chronic patient with bronchiectasie chronic walls of bronchi changes full of the mucus, who is not responding to conservative treatment (antibiotics), therapeutic and diagnostic flexible bronchoscopy had to be performed, resulting in a gram-negative bacteria *pseudomonas aeruginosa* – a typical bacteria for chronically sick C. F. patient. A *pseudomonas* therapy was prescribed according to the sensitive antibiogram, during which bronchoscopy was given locally on changes mucous pulmozyme and garamycin. Flexible bronchoscopy was performed as therapeutic. Local bronchoscopy findings: by aspiration of tracheo-bronchial truncus it was found hyperemia and a lot of mucous sticky secretion inside of tracheobronchial tree, especially middle lobe right side, lingual and basals part of the lungs. It was performed broncho-alveolar lavage and given steroids on the place of changed inflamed mucous membranae of the bronchi. It was also given pulmozyme to destroid mucous and make better spontaneously expectorations. Control chest x ray was performed and it was better.

KEY WORDS: therapeutic flexible bronchoscopy, cystic fibrosis.

INTRODUCTION

Cystic fibrosis or mucoviscidosis is an autosomal recessive disease which is manifested as a multisystemic disease of mostly exocrine glands. The majority of C.F. patients also have a chronic lung disease, pancreas insufficiency and an increased level of chloride in the sweat (over 60 mmol/l). The gene responsible for cystic fibrosis is located at 7q 31,2. By its gene product, it is marked as a CFTR gene (cystic fibrosis transmembranous regulator). It is a protein that functions as a chloride channel regulated by a cAMP that controls the flow of chloride through the cell membrane. Radiologically, the changes we find are bronchiectasias. The therapy is life-long (inhalator corticosteroids ICS-Flixotide, inhalation with garamycin, steroids, tobramycin and 0,9 % NaCl) and antibiotic therapy (ceftazidim, gentamycin, ceftriakson, azitromicin). Physical therapy: massage vibratione to remove as much as possible of the mucous plug from the bronchial tree. In oil resolvable vitamins with pancreatin is a life-long therapy of the digestive tract.

THE CASE REPORT OBJECTIVE

This is a case report about a 10-year-old girl with cystic fibrosis, who has been treated with therapeutic flexible bronchoscopy. A ten-year-old girl K.A. was repeatedly hospitalized at the Pediatric Clinic of the Kosevo University Hospital in Sarajevo due to respiratory insufficiency. The diagnosis was originally made when she was 4 years of age. The early-development anamnestical data are normal. Family history: her younger sister also has C.F. Anamnestically: a respiratory deterioration occurred in february 2001. The patient had already been treated conservatively, but she was still very sick, so she was hospitalized. Therapeutic flexible bronchoscopy was performed with a broncho-alveolar lavage in general anaesthesia. On admission: a 10.5-year-old girl, BW 19.5 kg BL 135 cm, afebrile, tahidyspnoic with perioral cyanosis, pale. Auscultatory findings on the lungs: crepitations, bilaterally. Chest X-ray shows a diffuse-like spot shadows with bronchiectasie. Laboratory findings: ABS- respiratory acidosis, poor oxygen saturations 68 %.

DISCUSSION

The case report describes a 10-year-old girl with cystic fibrosis, who was hospitalized at our clinic. The girl had been under the Longacef conservative therapy. Therapeutic flexible bronchoscopy was performed in general



FIGURE 1. Chest X-ray before bronchoscopy (K. A.)

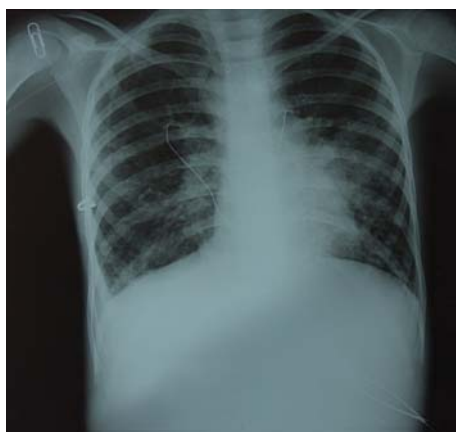


FIGURE 2. Chest X ray after bronchoscopy (K.A.)

anaesthesia. "Local bronchoscopy findings:by aspiration of tracheo-bronchal truncus it was found hyperemia and a lot of mucous sticky secretion inside of tracheobronchal tree, especially middle lobe right side , lingual and basals part of the lungs. It was performed broncho-alveolar lavage and given steroids on the place of changed inflamed mucous membranae of the bronchi. It was also given pulmozyme to destroid mucous and make better spontaneously expectorations. Control chest x ray was performed and it was better. The auscultatory findings on the lungs turned out to be better. The infection control has been enabled, as well as spontaneous expectorations.

CONCLUSION

The treatment of children with cystic fibrosis is very complex, and besides the conservative treatment it also requires therapeutic flexible bronchoscopy. C.F. children must undergo regular control in order to control the infection and avoid spreading bacterial infection. (3). Local findings of the tracheobronchial tree is very important and wall of the bronchi must be control because of huge amount of sticky mucus.

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