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Case report ■

Re-expansion of an Atelectatic Lung through Flexible Bronchoscopy in a Child with Dermatomyositis and Celiac Disease

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SUMMARY

This paper describes the utility of flexible bronchoscopy in a sick child diagnosed with dermatomyositis, celiac disease. Mucus plug is a common medical cause of lung atelectasis. Due to deteriorated respiratory condition, the child was highly febrile, cyanotic, liver 6-7 cm, palpable under the right rib arch. We described a child with dermatomyositis and lung atelectasis. Atelectasis causes difficulty breathing and decreased oxygen saturation, so the child was intubated and put on a complete mechanical ventilation.

Before intubation, the number of respirations exceeded 40/min, O2 saturation on the pulse oximeter fell under 73%. Pulse rate was 173/min, blood pressure 94/37 mmHg. Before intubation, the gas analysis of blood showed: Ph below 7.30 and pCO2 9 kPa, pO2 in the blood below 4.9 kPa. After flexible bronchoscopy was performed, therapeutic and diagnostic, lung re-expansion was enabled. After performed bronchoalveolar lavage with 0.9% NaCL 1 ml per kg TT, twice repeated, corticosteroids were introduced at the site of the changed mucus membrane. Mechanical ventilation parameters: Fio2, number of respirations and inspiratory pressure decreased. Values of gas analysis: ph improvement above 7.30 Pco2 3.6kPa, pO2 in the blood 11. O2 saturation 95%, pulse rate 120/min. The five-year-old child patient was extubated five days after bronchoscopy and was transferred to the standard Pulmonology Ward. Blood derivatives were obtained on several occasions. The condition improved, methotrexat therapy was introduced, with corticosteroids once a week, 3x40 mg i.v., on other week days Pronison 5 mg 4x1.

Key words: **dermatomyositis, atelectasis, flexible bronchoscopy with bronchoalveolar lavage**

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INTRODUCTION

Dermatomyositis is a connective tissue disease characterized by inflammatory skin and muscle processes. This is a rare disease with female children affected in a larger percentage (1). Usually, proximal myopathy is used to determine the level of muscle weakness. Hypercapnia in respiratory failure is likely to occur, tests of pulmonary function and respiratory muscle strength can suggest muscle weakness. Respiratory muscle strength was determined based on maximal static efforts; in half of the patients, both inspiratory and expiratory muscle strengths were less than 50% of the normal. In most of the patients without lung disease, respiratory muscle weakness was accompanied by significant decreases in vital capacity, total lung capacity, and maximum voluntary ventilation, by significant increases in the residual volume and arterial carbon dioxide tension (PaCO2) and a greater likelihood of dependence on ventilations, atelectasis and pneumonia. Hypercapnia was particularly likely whenever respiratory muscle strength was less than 30% of its normal value, in uncomplicated myopathy and when vital capacity was less than 55% of the predicted volume in any patient (2, 3). Pathogenesis: this is most likely a cellular automechanism, given that lymphocytes from the infected focal points of patients in vitro destroy the striated muscles. Skin contains perivasculitis, connective tissue swelling and edema, while the muscle system has inflammations with edema. Clinically: face with purple color with symmetric skin modifications on the eye-lids, mouth, cheeks, extensory parts of hands. Subskin and skin edemas result in a swollen face. Efflorescences are painless, no itches. They could be peeled off so that they reminded of psoriasis.

Myositis is often an accompanying phenomenon, often of a proximal structure. Laboratory-increased values of enzymes in the serum (creatine - kynasis, transaminasis, aldosis) indicate a degree of muscle degeneration.

Muscle biopsy shows inflammation infiltrates and muscle fibers necroses. Other signs of inflammation can also appear: accelerated erythrocyte sedimentation, increased gammaglobuline fraction.

After setting a safety diagnosis, the medicine of choice is the corticosteroids - large doses of prednisone (60 mg/m^2 or 1-2 mg/kg daily). A success of the treatment is visible after a few days only, through the withdrawal of exanthema and edema, and the increase in muscle strength. In case of dermatomyositis, it is recommended to apply corticosteroids for 2-3 months without any fear of the Cushing syndrome side-effects (osteopetrosis, vertebra compression, muscle system atrophy).

If there is no improvement within 6 to 8 weeks, one should introduce Azathioprin or Cyclosporin A, reducing the corticosteroid doses.

AIM

The paper aims to describe the significance of bronchoscopy with bronchoalveolar lavage in resolving atelectasis of a sick child diagnosed with dermatomyositis, celiac disease.

METHOD

This case report addresses the issue of dermatomyositis, which, due to the basic disease, grew into respiratory insufficiency. The child's atalectic lung was treated with flexible bronchoscopy, therapeutically.

Children-patients - the child was admitted with symptoms of heavy breathing, cyanosis, high febrility, swelling. Due to deteriorated respiratory condition, the child was transferred to the Intensive Care Ward. General condition deteriorated with highly febrile, cyanotic liver 6-7 cm in size, palpable under the right rib arch. Increased sedimentation rate 44/97, RBC 3.52, Hgb 95, Hct 0.28, EMNG (electromyelography): neurophysiological parameters leading towards myoneuropathy. Muscle fascia biopsy: muscle fibers are atrophic, focal, with signs of degeneration and regeneration, while between them there are inflammation elements of the lymphocytes and cell plasma type. Deterioration in terms of difficulty breathing, decreased saturation, and an auscultatory lung finding. The lack of muscle control and strength leads to an inability to clear secretions from the tracheobronchial tree. This was the cause of an acute respiratory failure. Highly febrile, cyanotic liver augmented by 6-7 cm, with periorbital edema and shin edema. Due to deterioration in terms of respiratory insufficiency, the child was intubated and put on IPPV mechanical ventilation (Figure 1).

Before intubation, the number of respirations exceeded 40/min, O2 saturation on the pulse oximeter fell under 73%. Pulse 173 /min TA 94/37 mmHg.

Before intubation, blood gas analysis Ph was below 7.30 and pCO2 9 kPa, pO2 in the blood below 4.9 kPa.

After the flexible bronchoscopy was performed, therapeutic and diagnostic, lung re-expansion was enabled. After the performed bronchoalveolar lavage with 0.9% NaCL 1 ml per kg TT, twice repeated, corticosteroids were installed at the site of the changed mucus membrane. Bronchoscopy was performed and a large mucus plug was found obscuring the right upper lobe.

RESULTS

A follow-up chest x ray after bronchoscopy demonstrated a resolution of atelectasis (Figure 2). Mechanical ventilation parameters: FiO2, number of respirations, the frequency and inspiratory pressure were all decreasing. Values of gas analysis pH improvement above 7.30 Pco2 3.6kPa, pO_2 in the blood 11. O_2 saturation

95%, pulse 120/min. The child was extubated five days after flexible bronchoscopy and transferred to the standard Pulmonology Ward. Blood derivatives were taken on several occasions. The condition improved, so that methotrexate therapy was introduced, with corticosteroids once a week, 3x40 mg i.v., on other week days Pronison 5 mg 4x1, with Brufen, gastal and cellianic diet.

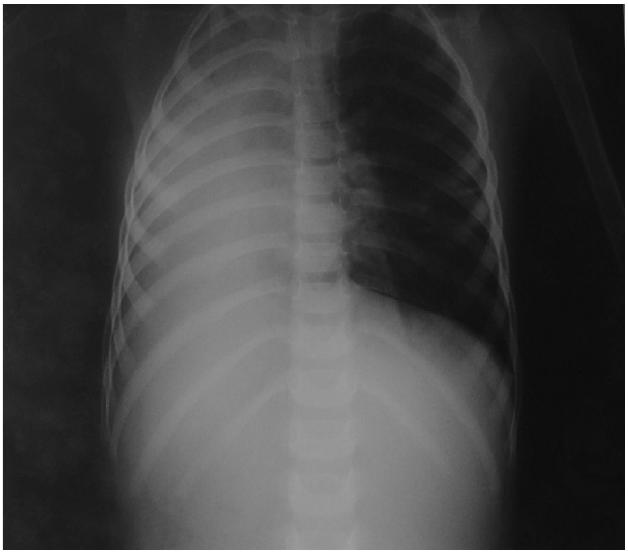

Figure 1. *Chest X ray prior to bronchoscopy (A. A.)*

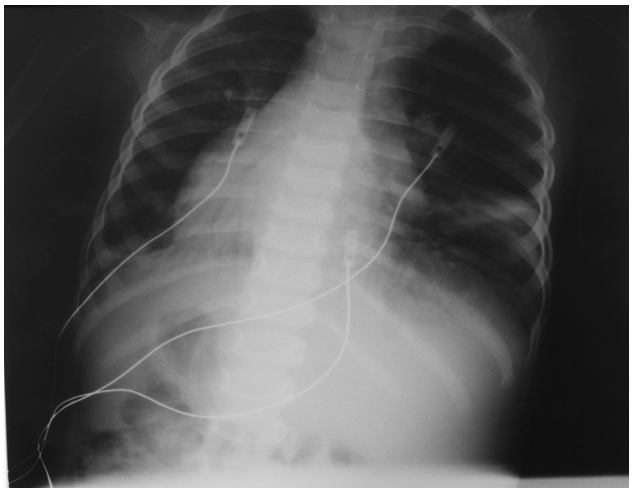

Figure 2. *Lungs radiogram after bronchoscopy*

DISCUSSION

Inflammatory myopathies fall into a group of acquired diseases characterized by proximal myopathy caused by an inflammatory infiltrate of the skeletal muscle. The three major diseases are dermatomyositis, polymyositis and inclusion body myositis. Dermatomyositis is a systematic autoimmune connective tissue disease, which most frequently affects the skin and muscles, but may also affect the joints, esophagus, lungs, and heart (1, 2). The course of disease is difficult to predict, usually, it has a long course with remission and exacerbations (3). The estimated incidence of dermatomyositis is about 10 cases per one million, twice as common in women as in men. Dermatomyositis can occur in people of any age. In children, the peak age is 5-10 years (4). Dermatomyositis may cause death because of muscle weakness or cardiopulmonary involvement (5).

Pulmonary manifestation of dermatomyosistis has often been reported and can occur in three patterns: (a) acute onset of fever, dyspnea, and lung infiltrates; (b) insidious onset of dyspnea and diffuse interstitial fibrosis; and (c) abnormal chest radiographs without respiratory symptoms (5, 6). Some patients may have severe, progressive, and ultimately fatal lung involvement despite the use of corticosteroids and immunosuppressive agents (7, 8). Children with neuromuscular disease such as dermatomyositis which manifests as a proximal symmetrical muscle weakness can develop acute respiratory failure extremely quickly. The lack of muscle control and strength can lead to an inability to clear secretions from the tracheobronchial tree (9, 10).

Bronchoscopy and bronchoalveolar lavage (BAL) are particularly important in excluding infection as a cause of lung infiltrates (6-8). The most common indication for bronchoscopy is the presence of retained secretions and atelectasis (11, 12).

Celiac disease is usually associated with autoimmune disorders and has occasionally been reported in patients with inflammatory myopathies. The association between dermatomyositis and celiac disease in children has been well documented. In the adult population, however, the association has not been clearly established (13-18).

An adequate treatment should be initiated as soon as possible. The drug of choice is the corticosteroids (prednisone 60 mg /m^2 or 1-2 mg /kg daily) for 2-3 months. If there is no improvement in 6 to 8 weeks, we should introduce Azathioprin or Cyclosporin A (19, 20). It is suggested that patients with newly diagnosed dermatomyositis be investigated for concomitant celiac disease even in the absence of gastrointestinal symptoms.

CONCLUSION

Bronchoscopy and bronchoalveolar lavage are particularly important in excluding infection as a cause of lung infiltrates (6-8). The most common indication for bronchoscopy is the presence of retained secretions and atelectasis (11, 12). The child's atalectic lung is treated with flexible bronchoscopy, therapeutically.

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RE-EKSPANZIJA PLUĆA ZAHVAĆENIH ATELEKTAZOM PUTEM FLEKSIBILNE BRONHOSKOPIJE KOD DETETA SA DERMATOMIOZITISOM I CELIJAKIJOM

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Sažetak

Ovaj rad ukazuje na značaj fleksibilne bronhoskopije kod obolelog deteta sa dijagnozom dermatomyositis, coeliakia. Sluzni čep u bronhu je čest medicinski uzrok atelektaze kod ovih bolesnika. Usled pogoršanog respiratornog stanja, obolela deca mogu imati visoku temperaturu, cijanozu, uvećanu jetru do 6-7 cm. Prikazujemo dete sa dermatomiozitisom, koje je imalo atelektazu pluća. Atelektaza pluća uzrokuje otežano disanje, smanjenje saturacije, zbog čega je obolelo dete intubirano i priključeno na totalnu mehaničku ventilaciju. Broj respiracija pre intubacije prelazio je 40 min, saturacija oksigena saturacija na oksimetru pulsa pada ispod 73%. Puls 173/min, krvni pritisak 94/37 mmHg. Pre intubacije vrednosti plinova u krvi - pH obično su ispod 7,30 i pCO2 9 kPa, pO2 u krvi ispod 4,9 kPa. Posle izvedene fleksibilne bronhoskopije, dijagnostičke i terapijske sa bronhoalveolarnom lavažom, nastupa reekspanzija pluća. Posle izvedene bronhoalveolarne lavaže sa 0,9% NaCL 1 ml/kg TT, dva puta ponovljene, uvedeni su kortikosteroidi na mestu promene na sluznici obolelog bronha. Parametri mehaničke ventilacije: FiO2, smanjen broj respiracija i vrednosti inspiratornog pritiska. Vrednosti pH arterijske krvi se popravljaju iznad 7,30 Pco2 3,6 kPa, pO2 u krvi 11. Saturacija kiseonikom 95%, puls 120/min. Petogodišnja devojčica je pet dana kasnije ekstubirana i premeštena na standardno odeljenje pulmologije. Krvni derivati su uključeni u nekoliko navrata. Stanje se poboljšalo, tako da je methotreksat uveden u terapiju, sa kortikosteroidima jednom sedmično, 3x40 mg i.v., a drugim danima u sedmici Pronison 5 mg 4x1.

Ključne reči: **dermatomiozitis, atalektaza, fleksibilna bronhoskopija sa BAL-om**