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Bird fancier's lung disease in children: a single center experience

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Summary

Bird fancier's lung disease is an immune-mediated lung disease caused by organic antigens of birds. It is a rare hypersensitivity pneumonitis in children. Here, we presented four children who presented with recurrent respiratory system complaints and were diagnosed with Bird fancier's lung disease. To the best of our knowledge, our first patient is the youngest patient in English literature. Bird fancier's lung disease should be considered in children who present with symtoms of chronic respiratory disease and history of bird feeding should be persistently questioned. *(Turk Arch Ped 2013; 48: 327-331)*

Key words: Bird fancier's lung disease, child, hypersensitivity pneumonitis

Introduction

Hypersensitivity pneumonitis (HP) also called "extrinsic allergic alveolitis" is a lung disease characterized with sensitivity to various organic aerosols and low-moleclar-weight chemical antigens and involvement of interstitium, bronchioles and alveoles as a result of recurrent inhalation of these substances. It is mostly observed in farmers and bird fanciers. Bird fancier's disease (BFD) is a HP which develops against the antigens included in the feathers and stools of birds. It is the most common subtype of hypersensitivity pneumonitis. HP which is observed rarely in the childhood is mostly in the form of bird fancier's disease (BFD) (1,2,3,4). In this article, four pediatric patients who presented with recurrent respiratory system complaints and were diagnosed with BFD were presented.

Case 1

A 22-month male patient presented with complaints including recurrent cough, wheezing, fever and exercise-induced cough. The patient had had recurrent complaints since the age of 6 months. He had been hospitalized with the same complaints and treated in the last 3-4 months. There was consanguinity between his mother and father and his father was breeding pigeons at home. His familial history was negative. On pyhsical examination, he had a fever of 36.7 °C and an oxygen saturation of 97%. Systemic examination findings were found to be normal. Laboratory findings were as follows: WBCt:18 380/mL (52% neutrophils, 42% lymphocytes, 4% monocytes, 2% eosinophils), hemoglobin: 10.8 g/dL, hematocrit: 34.1% platelets: 312 000 mm^3, erythrocyte sedimantation rate (ESR): 14 mm/h (N:0-10 mm/h), C-reactive protein (CRP): 4.43 mg/L (N:0-8 mg/L). Immunoglobulin (Ig) G, A, M, E levels, tuberculin skin test, postero-anterior lung graphy, echocardiography and sweat test were found to be normal. No sensitivity was found in skin prick tests performed with general aeroallergens (house dust, hayfield, weed pollens, animal dander and cockroach) and foods. Thoracal high-resolution computarized tomography (HRCT) revealed

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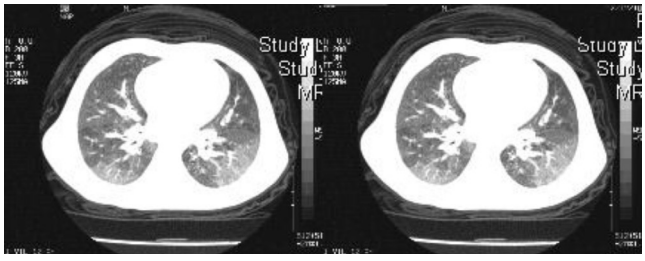
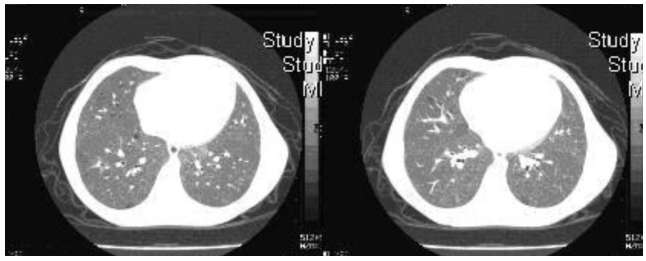
ground-glass areas predominantly in the superior parts of the lower lobes in both lungs (more extensive in the left side) and in the form of pathces in the posterior basal segments of the lower lobes and lymphadenopathies in the paratracheal area at the mediastinal level with the largest one having a dimension smaller than 1 cm (Picture 1). Precipitating antibody was found to be positive in the blood which was tested with a prediagnosis of bird fancier's disease and the patient was diagnosed with BFD. Pigeon contact was stopped and high dose inhaled fluticazone propionate treatment was started. The patient's complaints regressed following treatment and discontinuance of pigeon contact.

Case 2

An eight-year-old male patient presented with complaints including cough, wheezing, sputum production, dispnea and exercise-induced cough. The patient was hospitalized because of pneumonia at the ages of 4, 5 and 7. He had complaints for the last two weeks. His mother had a diagnosis of asthma. Pigeons had been bred at home for a long time. Physical examination revelaed no pathology except for bilateral crepitant rales. Hemogram, tuberculin skin test and Ig A, E, M were found to be normal. Ig G was found to be 3530 mg/dL (N:608–1572 mg/dL) and ESR was found to be 58 mm/h. No sensitivity was found in skin tests. Pulmonary function test (PFT) results were as follows: forced vital capacity (FVC):56, forced expiratory volume in the first second (FEV1):63, FEV1/FVC:112, peak expiratory flow rate (PEF):54, medium expiratory flow rate (MEF25-75):102. Lung graphy revealed right paracardiac-perihilar involvement. Thoracal HRCT revealed ground-glass appearance in all lobes and segments in both lungs and extensive centrilobular millimetric nodularities, regionally increased aeration especially in the lower lobes and fibrotic changes in the anterobasal segment of the lower lobe in the right lung (Picture 2). Precipitating antibody was found to be positive in the blood which was tested with a prediagnosis of bird fancier's disease and the patient was diagnosed with BFD. Pigeon contact was stopped and high dose inhaled fluticazone propionate treatment was started. His complaints regressed in the follow-up.



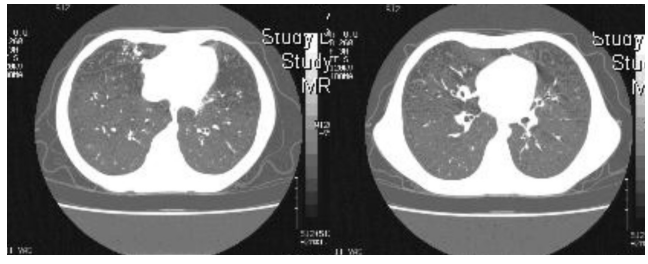
Picture 1. Appearance of Thoracal high resonance computarized tomography findings of the first patient



Picture 2. Appearance of Thoracal high resonance computarized tomography findings of the second patient

Case 3

An eleven-year-old female patient presented with complaints including recurrent cough, wheezing, sputum production, malaise and exercise-induced cough. She had had bronchiolitis at the age of 6 months and measles at the age of one year. She had recurrent complaints since the age of 3 years. Her mother and father had consanguinity and her older brother was being followed up in our division with a diagnosis of bronchiectasis. Physical examination revealed no pathology except for crepitant rales in the right side. Hemogram, tuberculin skin test, lung graphy, immunoglobulins, sweat test, saccarin test and ESR were found to be normal. No sensitivity was found in skin prick tests. Pulmonary function test results were as follows: FVC:67, FEV1:65, FEV1/FVC:97, PEF:54, FEF25-75: 51. Reversibility was positive (23% change in FEV1). Inhaled corticosteroid treatment was started. Thoracal HRCT which was performed in terms of bronchiectasis revealed tubular bronchiectasis which was central in the superior segments in the lower lobes of both lungs and peripheral in the posteror basal segment of the lower lobe in the left lung, millimetric nodular centrilobular involvement in the medial segment of the middle lobe in the right lung and in the posteror basal segments of the lower lobe in the left lung and pleural retractions in the lateral basal segment of the lower lobe in the right lung and in the inferior lingular segment of the



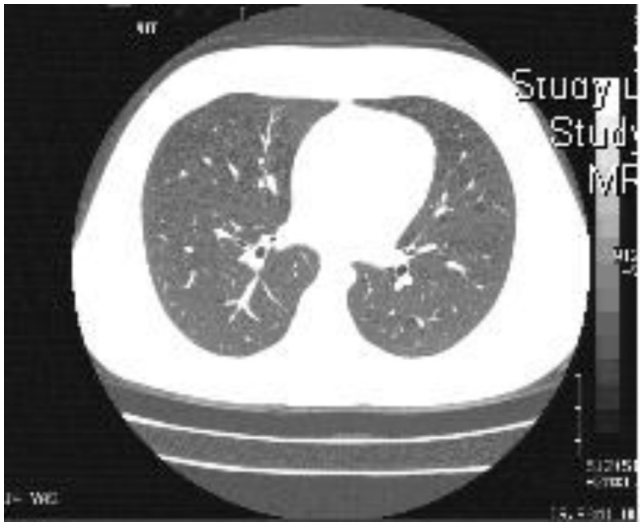
Picture 3. Appearance of Thoracal high resonance computarized tomography findings of the third patient

left lung (Picture 3). The patient's complaints continued despite inhaled corticosteroid treatment. When her history was questioned again, it was learned that they had been breeding pigeons at home for 7 years. Precipitating antibody was found to be positive in the blood which was tested with a prediagnosis of bird fancier's disease and the patient was diagnosed with BFD. Her complaints regressed following discontinuance of pigeon contact and high dose inhaled corticosteroid treatment.

Case 4

A 13-year-old female patient presented with complaints including recurrent cough, wheezing and exercise-induced cough. She had complaints for the last one month and was in contact with pigeons for the last two months. Pigeons were present in their garden and her complaints occured especially when she was outside home. Her familial history was negative. Her physical examination was normal. Complete blood count revealed mild leukocytosis (11 010/mm^3). Tuberculin skin test, immunoglobulins and skin prick test were found to be normal. Pulmonary function test results were as follows: FVC:103, FEV1:86, FEV1/FVC: 82, PEF: 78 MEF25-75: 56. Lung graphy revealed right paracardiac-perihilar involvement. Precipitating antibody was found to be positive in the blood of the patient who was thought to have bird fancier's disease. Thoracal HRCT was found to be normal (Picture 4). Contact with pigeons was discontinued and high dose inhaled fluticazone propionate treatment was started with a diagnosis of BFD. Her complaints regressed in the follow-up.

Demographic and clinical properties of our patients are shown in Table 1 and laboratory and radiological findings are shown in Table 2.



Picture 4. Appearance of Thoracal high resonance computarized tomography findings of the fourth patient

Discussion

Hypersensitivity pneumonitis is an immune-mediated lung disease. Bird fancier's disease is the most common subtype of hypersensitivity pneumonitis and mostly defined in breeders of pigeon and lovebird (4,5). In a recent study performed in our country, it was found that the most common cause in patients with HP was BFD. The most common bird leading to disease was found to be pigeon (6). The most common cause of HP in children is also BFD. As far as we know, our first patient is the youngest patient in the English literature. The extension of hypersensitivity pneumonitis is affected by environmental factors including antigen intensity, exposure period and frequency, solubility of antigen, particle size and use of respiratory protective device. Additionally, individual sensitivity is the most important factor in transformation from exposure to disease (2,7). It is predicted that the frequency is between 1-100 and 1/1000 among bird breeders (8). Publications related with children in our country are moslty case reports (9,10,11,12).

Hypersensitivity penumonies are classically clasified as acute, subacute and chronic according to exposure period and intensity (13).

In the acute form, symptoms which may be confused with influenza including shiver, fever, malaise, sweating, muscle pain, headache and nausea occur 2-9 hours after contact with antigen. The signs make a peak at the 6-24^{th} hour. Cough and dyspnea are observed frequently. Physical examination may reveal fever, tachypnea, tachycardia and rales. The signs disappear spontaneously in a few days.

The subacute form occurs in a few days-weeks. Cough and dyspnea are predominant. Severe dyspnea and cyanosis requiring hospitalization may occur.

The chronic form is a picture with an insidious course which develops in months characterized with increased cough and effort dyspnea. Sputum production, weight loss, dyspnea and malaise are present. Physical examination frequently reveals rales. Physical examination may be normal in approximately 20% of the patients (4,5). The complaints of our patients were compatible with the literature, while two of our patients had normal physical examination findings at presentation. This may be related with the fact that the history of contact with birds was shorter in these patients compared to the other patients and they were not examined during the period of acute disease. Since clinical findings may be frequently confused, it is difficult to differentiate these forms from each other definetely. The chronic and subacute forms are observed more frequetly in children compared to the acute form (14). Our first three patients are being followed up with a diagnosis of chronic BFD and our last patient is being followed up with a diagnosis of subacute BFD.

A different classification made according to progression of disease classifies the disease into four classes including acute intermittent, acute progressive, chronic intermittent and chronic progressive (15). According to this classification our first three patients were thought to have chronic progressive BFD and our fourth patient was thought to have acute intermittent BFD.

Laboratory findings may reveal mildly increased SR and CRP. Immunoglobulin G, A and M may also be increased. Sometimes increased Ig may be prominent. Allergen skin tests are not useful for the diagnosis (16). Increased ESR was found in two of our patients and increased IgG was found in one of our patients. No sensitivity was found in any of our patients in skin prick tests.

Lung graphy is used in excluding other causes rather than making a diagnosis of BFD. Ground-glass appearance, increased nodular or streaky pathcy intensities may be observed in acute BFD. These involvements are generally distributed extensively. They are frequently localized in the lower lobes in the subacute form. None of these findings are specific for BFD. Lung graphy may be normal in 20% of the patients (17). Involvement was found in lung graphy in two of our patients.

The classical thoracal HRCT findings in hypersensitivity pneumonitis include mosaic perfusion, ground-glass appearance and centrilobular micronodules. Ground-glass appearance may be the predominant finding or the only finding. HRCT findings may be normal, when the disease is diagnosed early (18). Fibrosis, increased reticular intensity, loss of lobar volume and honeycomb appearance are obserbed on thoracal HRCT of the patients with chronic hypersensitivity pneumonitis. The prominent finding may be mosaic perfusion. Mild reactive mediastinal lymphadenopathy is a frequntly observed finding. Lung cysts may be observed in subacute HP (19). Thoracal HRCT findings of our patients were as follows: ground-glass appearance in two patients, centrilobular micronodules in two patients, mediastinal LAP in one patient, fibrotic changes in one patient and normal findings in one patient (our 4th patient). This may be related with the fact that our fourth patient was diagnosed in the early period (two-month exposure). Lung cyst which may be observed in the subacute period was found in our second patient previously. In the follow-up, the cyst disappeared and other chronic period findings developed.

In acute BFD, restrictive dysfunction is found in PFTs classically. PFT may be normal in the subacute form. In the chronic and subacute forms, PFTs may reveal obstructive, restrictive or mixed dysfunction. In two of our patients restrictive dysfunction was found in PFT. One of our patients had decreased MEF25-75. In the study performed by Hasani et al. (20), decreased MEF25-75 was found, while FEV1 and FVC were in the normal limits in precipitating antibody positive BFD patients and it was reported that this might reflect the early period of chronic bronchitis.

Presence of precipitating antibody in the serum shows that the host has had sufficient exposure to develop immune response. It is a helpful test in the diagnosis of bird fancier's disease. Its positivity does not make a definite diagnosis and its negativity does not exclude the diagnosis of BFD. In all our patients, serum precipitating antibody was positive.

Specific bronchial stimulation test, bronchoalveolar lavage (BAL) and lung biopsy which are used in the diagnosis of bird fancier's disease were not performed, since the history and clinical, radiological and serological findings of our patients were compatible with BFD.

Various diagnostic criteria have been defined in the diagnosis of bird fancier's disease (13,21,22). However, the accuracy of these criteria have not been proven. Most criteria include presence of exposure, effort dyspnea, rales and lymphocytic alveolitis (if Bal has been performed). Supportive criteria include recurrent fever, increased intensities on lung graphy, decreased carbondioxide diffusion capacity, presence of precipitating antibody, granuloma on lung bioopsy (generally not necessary) and improvement with avoidance of contact (2). A history explaining the relation between exposure to the agent and occurence of symptoms is very important.

The prognosis is usually good in acute BFD. The findings regress in days after exposure to the agent is discontinued. Mild PFT abnormalities regress in a few months. Sometimes, BFD may be progressive even if contact is discontinued after a single attack. Subacute and chronic BFD have a more insidious course. The severity of the disease increases with exposure. It progresses to pulmonary fibrosis, if not treated. The prognosis is worse in patients with fibrotic chronic BFD compared to patients with non-fibrotic BFD (23).

The main treatment in bird fancier's disease is discontinuance of contact with the agent. In cases where it is impossible to eliminate the antigen or the cause can not be found, the patient is moved away from the setting which contains antigen. Inhaled and systemic steroids are used in medical treatment of BFD. There no randomized controlled studies related with the effect of systemic steroids in acute BFD. Systemic steroid is started in severe attacks. It is continued until the findings and function improve. If improvement occurs, the lowest dose is started and continued. If no improvement occurs, the dose is tapered and discontinued. Similar results have been obtained with high dose inhaled steroid and systemic steroids (24).

Steroids have no beneficial effect on the long-term prognosis. Supportive treatment should be administered in all forms (24). In our patients, we started treatment with high dose inhaled corticosteroid. We decreased the dose

after clinical improvement and/or improvement in PFT was obtained.

Conclusively, BFD is the most common subtype of HP in children. The prognosis is poor in cases of delayed diagnosis and treatment. BFD which is observed rarely in the childhood should be considered in children presenting with chronic respiratory system complaints and history of bird breeding should be questioned persistently.

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