CASE REPORT

Paediatric feather duvet hypersensitivity pneumonitis

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SUMMARY

A previously well 12-year-old boy was admitted with a second insidious episode of dyspnoea, dry cough, anorexia, weight loss and chest pain. At admission, he had an oxygen requirement, significantly impaired lung function and reduced exercise tolerance. Initial forced expiratory volume in 1 s was 26%; a 3 min exercise test stopped at 1 min 50 when saturations dropped to 85%. CT scan showed ground-glass nodularity with lymphadenopathy. Bronchoalveolar lavage (BAL) for *Pneumocystis carinii* pneumonia and viruses were negative, and microbiology results for the BAL were reported in the absence of histology. This is because at the time the BAL samples were collected, a lung biopsy was performed. The biopsy was consistent with hypersensitivity pneumonitis. Echo was normal and CT pulmonary angiography negative. After taking a thorough history, exposure to feather duvets prior to each episode was elicited. IgG of avian precipitants was raised at 10.6 mgA/L (normal <10 mgA/L). Clinical improvement began with avoidance of exposure, while the boy was an inpatient. Antigen avoidance continued on discharge. He continues to improve since discharge. The condition was diagnosed as hypersensitivity pneumonitis secondary to exposure to antigens from feather duvets.

BACKGROUND

Childhood interstitial lung disease is a rare group of disorders that mostly affect the alveolar and peri-alveolar lung tissue, which can lead to a restrictive pathology with fibrosis. Interstitial lung disease (ILD) in children is different to that seen in the adult population and therefore needs a separate approach. Some cases in children have an identifiable cause after taking a thorough history and excluding differential diagnoses.

Hypersensitivity pneumonitis is a form of immune-mediated ILD. It is a complex syndrome with a wide variety of organic, high-molecular-weight inciting antigens or inorganic low-molecular-weight antigens. Both varieties have a number of possible presentations. There is a high risk of missed diagnosis. The majority of reported cases has been in the adult population; for example, Bird Fancier's Lung, Farmer's Lung and Cheese Worker's Lung. Those that have been reported in children have been related to exposure to domestic birds rather than feather bedding.¹

It is possible that members of the paediatric population are being misdiagnosed with more common, similarly presenting conditions such as asthma, episodic wheeze or atypical infection.

We think that it is important to consider this diagnosis with relevantly presenting cases because

if it is not thought about, it will not be investigated for and therefore left undiagnosed. Left untreated it can result in irreversible disease. The use of feather duvets in the home and the presence of domestic birds is not unusual, and is easy to elicit while taking a thorough history.

CASE PRESENTATION

Our patient first presented in 2010 to his general practitioner and later to his local hospital, with a history of diurnal dyspnoea, reduced exercise tolerance and weight loss. He did not benefit from inhalers. He gradually returned to his premorbid state over a period of months. There was no history of significant medical problems including respiratory illnesses, reflux or obstructive sleep apnoea. He had not recently travelled abroad, but his father had travelled with the military prior to both episodes (Kenya and Afghanistan). He lived at home with his parents and a cat. He had no known allergies.

In 2013, he re-presented with a second episode. His symptoms included dyspnoea, dry cough and anorexia, with weight loss and chest pain. His mother recognised the symptoms from 2010, and he presented to his local hospital acutely. On admission, he had an oxygen requirement, significantly impaired lung function with reduced exercise tolerance. He improved while in hospital, which retrospectively was considered to be secondary to avoidance of avian antigen.

The house was not damp, no smokers lived in it and the rest of the household were well. The boy's mother had run a part-time beauty therapy service from home and therefore there had been various chemicals that our patient could have come into contact with.

Alongside these chemicals were a number of plug-in air fresheners and recent interior wall painting. However, these are non-organic substances and less likely to cause this presentation. The boy's mother had noted that both in 2010 (single duvet) and 2013 (double duvet), prior to the episodes of illness, new feather duvets had been purchased. There have been reports of hypersensitivity reactions to feather duvets from doves and geese.^{2,3} The boy's initial investigations were suggestive of ILD.

INVESTIGATIONS

Chest X-ray (CXR) (figure 1): ground-glass appearance, most prominent in lower zones.

Initial forced expiratory volume in 1 s (FEV1) 26%; forced vital capacity 27%; forced expiratory flow 25–75 56% (restrictive small airways disease).

Diffusing capacity for CO: 35% predicted (disease at alveolar level).

Echo: normal.

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Figure 1 The patient's chest X-ray: ground-glass appearance, most prominent in lower zones.

Exercise testing 3 min step: stopped at 1 min 50 with saturations dropping to 85%.

Haemoglobin 159, white cell count 8.75, platelets 388, C reactive protein <4, erythrocyte sedimentation rate 2.

Immunoglobulins: normal.

Negative antinuclear antigen screen/antiglomerular basement membrane abs/AAFB/virology/HIV/*Mycoplasma*/*Aspergillus*.

CT scan (figure 2): diffuse ground-glass nodularity with some lymphadenopathy (CT pulmonary angiography negative).

Lung biopsy: interstitial chronic inflammatory cell infiltrate (lymphoplasmic with occasional eosinophils) with greater intensity around bronchovascular bundles with several small but discrete non-necrotising epithelioid and giant cell granulomas (consistent with hypersensitivity pneumonitis).

The case was discussed with Professor Bush, Royal Brompton Hospital, who felt that clinical history and image pathology was classical for hypersensitivity pneumonitis in association with feather duvets. It was advised to test for IgG avian precipitants.

IgG avian precipitants 10.6 mgA/L (normal <10 mgA/L). ImmunoCAP testing to pigeon serum antigen is used in our laboratory as it has similarities to other antigens.⁴ Any results over 10 mgA/L IgG antibody are said to be highly indicative of exposure to avian antigens.⁵

Figure 2 The patient's chest CT scan: diffuse ground-glass nodularity with some lymphadenopathy.

DIFFERENTIAL DIAGNOSIS

- ▶ Asthma
- ▶ Atypical infection
- ▶ Malignancy
- ▶ Chemical hypersensitivity pneumonitis
- ▶ Other causes of interstitial lung disease, for example, Wegener's, Sarcoidosis, Goodpasture syndrome.

TREATMENT

The family was advised to remove the feather bedding from the house and the patient was recommended lifelong avian antigen avoidance. It was also advised, if the house contained a chimney, to check for birds nesting within it.

The patient was already taking lansoprazole due to symptoms of chest pain, before transfer from the local hospital, which he continued until the steroid treatment was completed.

Two month steroid treatment course:

Four weeks high-dose prednisolone (60 mg);

One week 40 mg prednisolone;

One week 30 mg prednisolone;

One week 20 mg prednisolone;

One week 10 mg prednisolone.

OUTCOME AND FOLLOW-UP

- ▶ Four-week hospital stay (10 days oxygen requirement)
- ▶ First week home: walking stairs (treated for *Haemophilus*-positive sputum sample)
- ▶ Third week home: return to school
- ▶ The boy had gradual improvement in formal lung function tests, which began with avoidance of exposure
- ▶ He had regular clinical follow-up and monitoring of lung function, CXR and overall progress
- ▶ Initial FEV1 26%, FEV1 86% 3 months after admission.

DISCUSSION

Hypersensitivity pneumonitis in children is under-recognised, but clinically significant, usually resulting from repeated exposure to avian antigens. It is an immune-mediated disease process (type III and IV hypersensitivity) that relies on an exposure to an inciting antigen, and a genetically susceptible and suitably sensitised host. IgG-mediated sensitisation will not necessarily lead to overt disease, but for some, repeated exposure can lead to irreversible disease with lung damage in the form of interstitial and intra-alveolar fibrosis.

1. In 2010, another retrospective review of medical records of patients with Feather Duvet Lung was published. In that review, the patient population was exclusively woman with a median age of 53 years. The authors proposed that the patients had become sensitised to bird antigens in their childhood/adolescence, either from bird (domestic) or feather exposure. All were positive for specific IgG antibodies and were shown to have a restrictive impairment of lung function. High resolution CT (HRCT) revealed ground-glass appearances and fibrosis. Lymphocytic alveolitis was present on bronchoalveolar lavage (BAL) and biopsy. The authors highlighted the importance of a thorough history considering this diagnosis in those presenting with clinical findings suggestive of extrinsic allergic alveolitis.
2. In an earlier review from 2002, the median age for reported cases of hypersensitivity pneumonitis in children was found to be 10 years. The majority of cases were caused by exposure to domestic birds. In the acute stages, the patients experienced a flu-like illness with dyspnoea, dry cough and

lethargy. If exposure ceased, there was a gradual reduction in symptoms. With chronic exposure, there was progressive illness with increasing symptoms such as weight loss, exercise intolerance and signs of longstanding respiratory pathology in the form of clubbing, hypoxaemia and pulmonary hypertension. Restrictive lung defects were more common, although the presence of an additional obstructive element could be seen. In the majority of patients, the CXR was abnormal, HRCT showed a ground-glass appearance and positive precipitins were found. Lymphocytosis was demonstrated on BAL and biopsy. In such cases, antigen avoidance is the mainstay of treatment. The use of steroids may be indicated, and should lead to resolution of symptoms and reversal of radiographic/lung function abnormalities.

Learning points

- ▶ Hypersensitivity pneumonitis is rare, but important to consider as it can mimic other more common disorders.
- ▶ It is important to take a thorough history of the onset, symptoms and possible causes.
- ▶ Environmental (inciting antigen) and host factors determine occurrence and severity.
- ▶ Symptoms develop over a prolonged period and can be intermittent/chronic.
- ▶ The diagnosis requires an index of suspicion—if we do not consider it, we will not diagnose it.
- ▶ It is IgG antibody mediated, so IgE Rast has no diagnostic value and wheeze is not a symptom.
- ▶ Treatment and long-term management begins with antigen avoidance.
- ▶ Untreated cases can result in irreversible damage.

In the paediatric population, prognosis is good with a prompt diagnosis, antigen avoidance and steroid treatment.

3. In 2006, an adult study was published that had focused on the clinical features of Bird Fancier's Lung secondary to feather duvets. It was a retrospective review of patients from 2000 to 2003 under the care of Tokyo Medical and Dental University, in Japan. All of the acute patients had positive specific antibodies against avian antigens. Whether acute or chronic, lymphocyte proliferation in blood or BAL was found. The authors warned physicians to consider the diagnosis as a possibility in relevant cases as the use of feather duvets was getting increasingly popular.

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