Unusual association of diseases/symptoms

Coeliac disease presenting as chronic cough in an 8-year-old child

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Patients with the 'classical' gastrointestinal symptoms of coeliac disease (CD) are usually readily diagnosed and treated with a gluten exclusion diet. However, the advent of sensitive serological investigations has revealed that over half of childhood CD remains asymptomatic or presents with more subtle non-gastrointestinal symptoms, with a significant risk to the health of undiagnosed children. In view of its changing presentation there is an increasing need for clinicians to have a low threshold to investigate patients for this disease. Rare cases of CD associated with respiratory symptoms have been reported. The authors report an interesting case presenting as chronic cough in an otherwise asymptomatic 8-year-old girl. As a result of having a low threshold of suspicion for underlying CD, a diagnosis has been made and the long-term health risks of undiagnosed disease have been improved in both the patient and her immediate family.

BACKGROUND

There is increasing evidence that coeliac disease (CD) is more prevalent than previously thought, and serological studies in asymptomatic children without a family history of auto-immune conditions have suggested the prevalence may be as high as 1%. A significant burden of this disease is undiagnosed in childhood, potentially with a considerable impact on morbidity.¹

CASE PRESENTATION

An 8-year-old girl presented to the outpatient department with an 18 month history of persistent non-productive cough. Her father reported that the cough began in the summer months and had progressively worsened. There was no association with wheeze, chest tightness, nor shortness of breath on exertion. Similarly, there was no association with posture and the cough was not heard by the parents at night.

She was born at term following a healthy pregnancy to non-smoking parents. She breast fed up to 4 months after which she was successfully weaned by 6 months. There were no concerns with her growth and development. Her height and weight continued to lie on the 9th centile. Over the years she had received steroid treatment for frequent bouts of viral croup, the last being 12 months previously. Mild asthma suffered by her father was the only significant family history.

INVESTIGATIONS

Prior to her attendance at paediatric clinic she had a flexible bronchoscopy in the ear, nose and throat department which was reported as normal. Physical examination was unremarkable. Normal investigations included full blood count, renal and liver function tests, bone profile, erythrocyte sedimentation rate and thyroid function tests. Immunological testing of radioallergosorbent test against

house dust mite, cat, grass and tree pollen was negative. A chest radiograph and sweat test were normal.

However, an antitissue transglutaminase (tTG) IgA antibody titre was positive at 21 U/ml (0–10). On direct questioning she had no gastrointestinal symptoms, nor personal or family history of auto-immune conditions. However, in view of the elevated tTG level, endoscopic duodenal biopsies were obtained, a photomicrograph from which is shown below (figure 1).

OUTCOME AND FOLLOW-UP

A diagnosis of CD was made. A gluten exclusion diet was commenced and on review 6 weeks later her symptoms had entirely resolved. Subsequent serological testing of the immediate family led to confirmation of asymptomatic CD in the mother of the patient.

DISCUSSION

The well-recognised triad in diagnosing CD is positive serology, positive intestinal biopsy and resolution of symptoms on gluten free diet. Patients with the 'classical' gastrointestinal symptoms of CD are usually readily diagnosed and treated with a gluten exclusion diet. With the advent of serological testing there is increasing evidence that the majority of cases of CD remain undetected or presents with more subtle manifestations of the disease, usually with symptoms associated with mineral or vitamin deficiencies.¹ A case series of 50 children presenting with CD between 1999 and 2004 at the University Hospital of Wales, Cardiff, UK, revealed 42% of patients presented with gastrointestinal symptoms, typically diarrhoea, weight loss, abdominal pain and constipation. Thirty two per cent presented with non-gastrointestinal manifestations, including mouth ulcers, short stature and iron deficiency and 26% presenting after serological screening of at risk groups.¹

Figure 1 Duodenal biopsy at 200x magnification with (H&E) stain showing subtotal villous atrophy with crypt regenerative hyperplasia and focal intraepithelial lymphocytosis consistent with coeliac disease, Marsh stage 3b.

Rarely CD has been associated with respiratory conditions in adults, including pulmonary haemosiderosis² and cavitating lung disease.³ Reports of CD associated with respiratory symptoms are even more uncommon in the paediatric population, although case reports of idiopathic pulmonary haemosiderosis associated with CD have been published.²

We report on a case of an 8-year-old girl presenting with chronic cough as the only presenting feature of underlying CD. The cough rapidly resolved after the commencement of a strict gluten exclusion diet. There is no obvious explanation for the chronic cough in our patient and its resolution with a gluten exclusion diet, especially in the absence of abnormal bronchoscopy findings. There is a case report of a 65-year-old adult presenting with cough and lymphocytic infiltration as the presenting feature of CD.⁴ Another possible hint at the underlying mechanism may be related to the discovery of increased pulmonary vascular permeability in patients with CD⁵ although given the rarity of this presentation no significant work in this area has been published.

It is possible that the resolution of the cough coincided with the institution of the gluten free diet. However, the temporal relationship is intriguing. As it is no longer advised that the response to a gluten challenge is necessary to confirm the diagnosis, we do not plan to recommend that she eats gluten again to see if her symptoms recur.

CONCLUSION

The increasing use of serological tests indicates that a significant number of people with CD remains clinically undetected and can present in a manner that is more subtle than previously appreciated. Our case highlights the changing

nature of CD, and the need to consider this diagnosis in cases of chronic unexplained respiratory symptoms. As a result of having a low threshold of suspicion for underlying CD a diagnosis has been made and the long-term health risks of undiagnosed disease have been improved in both the patient and her immediate family.

Learning points

- ▶ The prevalence of CD is 1%, much higher than previously thought.
- ▶ Approximately 60% of patients do not have classical gastroenterological features.
- ▶ Chronic respiratory symptoms may be a presenting feature of occult CD.
- ▶ Occult CD in family members should be considered.

Competing interests None.

Patient consent Obtained.

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