Unusual presentation of celiac disease in a child

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SUMMARY
Celiac disease is a genetic, immunologically mediated small bowel enteropathy that causes malabsorption. The immune inflammatory response to gluten frequently causes damage to many other tissues of the body. We report the association of celiac disease and alopecia areata in a girl. The alopecia developed after 11 years nonadherence to a gluten-free diet; the patient had HLA phenotype DR3/DQW2. Our patient had IgA class endomysial antibodies, IgA and IgG antigliadin antibodies and subtotal villous atrophy on jejunal biopsy. Administration of a gluten-free diet resulted complete hair growth.

Key words: celiac disease, alopecia areata

INTRODUCTION
Celiac disease (CD) is a gluten enteropathy occurring in both children and adults. The condition is characterized by a permanent sensitivity to gluten that results in inflammation and atrophy of the mucosa of the small intestine. Clinical manifestations include malabsorption with symptoms of diarrhea, steatorrhea and nutritional and vitamin deficiencies. CD has been reported in association with many other conditions, particularly those of autoimmune origin. Secondary immunologic illnesses may be the primary presentation¹².

Alopecia areata (AA) is a common, unpredictable, non-scarring form of hair loss. This disorder affects all age groups, with a higher prevalence in children and adolescents. The etiology of AA is as of yet unclear, but is presumed to be due to an autoimmune reaction. Consistent evidence of autoantibodies directed against anagen stage hair follicle structures are found both in affected humans and in mouse models³⁴.

We describe one patient with AA and CD:

CASE REPORT
A 13-year-old girl was referred to the Pediatric Department because of failure to thrive and anemia. Physical examination showed malnutrition (her weight was < -3 SD), marked abdominal distention, dry and pale skin and four vast areas of patchy AA on her scalp that appeared in 1997. Her personal and family history was negative for gastrointestinal disease and she never complained of diarrhea. Her parents had consulted various specialists in addition to dermatologists for their daughter’s hair loss. Previous treatments included systemic steroids and psoralen plus UVA, without achieving hair regrowth. Laboratory studies revealed serum hemoglobin: 6.5 mmol/L (normal: 8-9.9 mmol/L), hypochromia and microcytosis, iron: 7.3 μmol/L (normal: 9-26.9μmol/L), albumin: 30 g/L (normal: 35-50 g/L). Values of thyroid stimulating hormone (TSH) and thyroid hormone (T3, T4) were normal and autoantibodies for thyroid disease (antithyroid and antimicrosomal) were absent. Her HLA

Abbreviations:
CD: Celiac Disease
AA: Alopecia Areata
EMAs: Endomysial Antibodies
AGAs: Antigliadin Antibodies
HLA: Human Leucocyte Antigen
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phenotype was DR3/DQW2 and class IgA endomysial antibodies (EMAs) and antigliadin antibodies (AGAs). IgG and IgA were positive. Multiple endoscopic biopsy specimens were therefore taken at the distal duodenum, showing subtotal villous atrophy. After a year of gluten-free diet, a second duodenal biopsy was performed and findings were normalized. There was complete regrowth of hair; she had gained 5 kg in weight and the hematoclastic parameters returned to normal values. The patient continues to be under observation, while still on a gluten-free diet, and the situation remains unchanged.

DISCUSSION

An association between AA and CD has recently been reported5. Both CD and AA are thought to have an immunologic basis and, more specifically, a T-cell-immune dysregulation6. A frequent association of both conditions with many other autoimmune disorders has been described7,8; however, the association of these two conditions in the same patient is extremely rare.

In a prospective screening, 254 consecutive outpatients with alopecia areata were tested, using antigliadin and antiendomysial antibodies. Results were positive in three patients and, despite a lack of gastrointestinal symptoms, these patients underwent intestinal biopsy. All three were found to have celiac disease, and treatment with a gluten-free diet was initiated. Thus the observed frequency of association between these 2 entities is much greater than can be expected by chance5.

Celiac disease is a genetic, immunologically mediated, small intestine enteropathy in which mucosal villi are destroyed by cellular and humoral-mediated immunologic reactions to gliadin protein9. The diagnosis of celiac disease does not require further confirmation if the initial diagnosis is based firstly on the appearance of flat small intestinal mucosa with the histological features of hyperplastic villous atrophy while the patient is eating adequate amounts of gluten, and secondly on unequivocal and full clinical remission after withdrawing gluten from the diet. The finding of circulating antibodies (IgA gliadin, antireticulin, and antiendomysium) at time of diagnosis and their disappearance when the patient is on a gluten-free diet add weight to the diagnosis10. EMA screening was superior to that for either IgA AGA or IgG AGA in identifying untreated cases of celiac disease, and it had specificity comparable with that of IgA AGA11.

It is not clear why the association with alopecia is so rare in CD compared with other autoimmune disease. Alopecia may precede the clinical manifestations of intestinal disease and the gliadin antigens are the primary event leading to the alopecia. This may occur as a general immune response to the antigens or to a specific, shared antigenic mimicry between the gliadin and elements in the hair bulb23.

New data have determined that CD is more common than was previously thought. The diverse secondary clinical manifestations make it easy to miss. A high index of suspicion is required to diagnose CD in a patient with a confounding presentation. Studies with more patients may disclose the factors involved in these rare cases of CD associated with AA.

REFERENCES

4. Tobin DJ, Sundberg JP, King LE Jr, et al. Autoantibod-