A Three Years Old Indian Girl with Abdominal Distension and Bizarre Eating Habit

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Abstract

Abdominal distension is a common but non-specific sign of a variety of diseases and conditions that always warrant meticulous and thorough physical examination as well as investigations. An Indian girl who presented with a grossly distended abdomen was admitted for suspected child neglect and malnutrition. History of pica was noted and a big gastric bezoar was removed surgically. Investigations came up with the diagnosis of coeliac sprue. Low level of awareness would overlook this disease entity easily, particularly in a region with exceedingly low prevalence like our locality. Indeed, screenings of at-risk groups in Caucasians have resulted in identification of considerable numbers of atypical and asymptomatic cases.

Key words   Bezoars; Coeliac disease; Jejunal biopsy; Malabsorption; Pica

Case Report

The Indian girl, aged 34 months, was born here in Hong Kong. Birth history was unremarkable. She returned to India since eight months old and was fed mainly on cow's milk, rice and bread. She was noticed to have pica since early childhood, eating own hair, dirt and soil from floor, cotton threads and fragments from clothes and bed linen. Abdominal distension was present for a long time but she was brought to medical attention when vomiting and abdominal pain was developed in the recent six months. Bowel habit was normal. Investigations done in India suggested foreign body in the gut causing partial intestinal obstruction. No intervention was done however. She returned to Hong Kong in April 2002 and was admitted to hospital for gross abdominal distension. Iron deficiency anaemia was found but parents refused further investigations. The provisional diagnosis was malnourishment. Social worker brought her to our hospital two weeks later for suspected child neglect.

Physical examination revealed a small child with wasted limbs (Figure 1). Body weight (10 kg) fell on the 3rd centile while her body height (80.6 cm) was 4.5 cm below the 3rd centile (Figure 2). She was pale, but neither jaundiced nor dehydrated. Her abdomen was soft and non-tender but grossly distended. Abdominal girth was 62.5 cm at umbilical level. There was no palpable mass in the abdomen. Bowel opened once to three times daily with non-greasy, yellowish formed stool. Pica was observed, she ate her own hair and cotton threads from clothes, bed linen and even torn fragments of diaper. Her appetite was good and no vomiting was noted after admission.

Iron deficiency anaemia with haemoglobin level of 8.2 g/dL was noted, serum iron level was 4 µmol/L (normal ranged 10.7-31.3) and total iron binding capacity (TIBC) was 75 µmol/L (normal ranged 45-72). Hypoalbuminaemia (30 g/L) was documented. Serum zinc level was low (6.3 µmol/L; normal ranged 9-29). Stool specimens showed no ova, cysts or enteric pathogens. Barium meal and follow-through showed intraluminal filling defects in stomach, duodenum and jejunum compatible with bezoars. Irregularly thickened folds were noted in duodenum and jejunum but transit time was normal. IgA anti-gliadin antibodies (AGA) and IgA anti-endomysial antibodies

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Figure 1 Physique of patient before starting gluten-free diet (left); and 8 weeks after gluten-free diet (right). Note the obvious gain in weight and muscle bulk as well as the marked improvement of the abdominal distension.
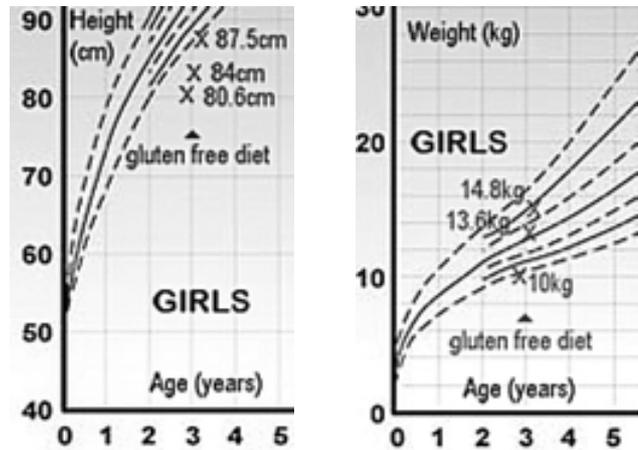

Figure 2 Growth parameters of the patient. (Growth charts adopted from Hong Kong Growth Survey 1993 by the Faculty of Medicine, the Chinese University of Hong Kong in collaboration with the Department of Health and the Hospital Authority).

(EMA) were positive, titre of the former above 200 RU/ml (normally less than 20 RU/ml). Impaired D-xylose absorption (plasma xylose 0.12 mmol/L at 2 hours after 5 gram dose, normal level >1.7) and elevated faecal fat (65 mmol/24 hours; normal ranged 11-18) were demonstrated. The IgA AGA titre level of her mother was elevated to 69.6 RU/ml and her IgA EMA was positive. Her father was IgA AGA-negative but weakly positive for IgA EMA, while her elder sister got negative results for both. They were all asymptomatic.

A big, hard trichobezoar entangled with cotton wool fragments was removed surgically (Figure 3); it occupied most of the stomach and extended down to the duodenum. Small bowel wall was mildly thickened and multiple mesenteric lymph nodes were noted. A jejunal biopsy taken at laparotomy for removal of the gastric bezoar revealed that the jejunum was remarkable for nearly total villous atrophy with hyperplastic crypt region and an increase in intraepithelial lymphocytes (Figure 4). Mesenteric lymph node showed reactive lymphoid hyperplasia.

She was put on iron and multi-vitamin supplements and a gluten-free diet (Table 1). Eight weeks later (37 months

Figure 3 The bezoar. It was moulded to the shape of the stomach with long extension to the duodenum. It is composed of hair, i.e. trichobezoar, entangled with cotton wool threads and fragments.
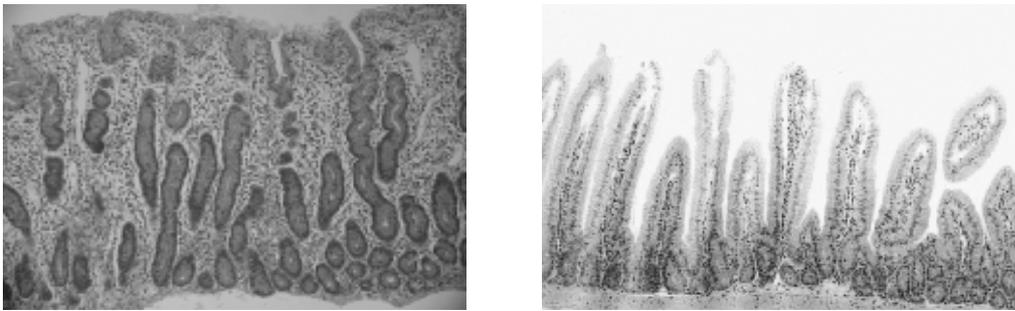
Figure 4   Jejunal biopsy of the patient before starting gluten-free diet, showing nearly total villous atrophy, hyperplastic crypts and an increase in intraepithelial lymphocyst (left). A normal slide from other subject for comparison (right).

Table 1   Gluten free diet (suggested by dietitians in Princess Margaret Hospital)

	Food allowed	Foods avoided
Cereals and cereal products	Rice, corn, millet, buckwheat, potato, corn starch, sago ( ), rice vermicelli ( ), some common Chinese rice cereal products and gluten free wheat flour (, , , , , , , , , ,)	Wheat, rye, oat, barley, wheat flour, breads, breaded food, bread crumbs, pastas, macaroni ( ), spaghetti, muffins, meat and fruit pies, biscuits, pizzas
Fruits	All fresh fruits and fruit juices, canned fruits	Commercial fruit pie fillings
Vegetables and beans	All fresh vegetables and beans	Vegetables with dressings, canned ketchup bean
Meat and eggs	All fresh fish, meat and poultry (**prepared without wheat, rye, oats, barley, bread crumbs, fried powder**); eggs	Sausages, beef and pork patties ( ), fried fish fillet, luncheon meat, canned meat (**contained gluten stabilisers or fillers**)
Milk and dairy products	Milk, evaporated milk ( ), condensed milk ( ), skimmed milk, cheese	Milkshakes (**additives contained gluten**)
Others	Salt, sugar, honey, pepper, vanilla and other herbs ( ), vinegar, condiments and seasonings (including ,), nuts, tea, coffee, candies, jelly	**Any commercially prepared products that may contained wheat flour or gluten:** Chinese vegetarian food ( ), canned creamy soup, packed soup, ketchup, seafood sauce, chocolate beans, chocolate powder, ice-cream, ovaltine, horlicks, malted milk ( ) or flavourings, curry powder, chewing gums, salad dressings, etc.

old), her height was 84 cm (3 cm below 3rd centile) and bodyweight 13.6 kg (50th centile) (Figure 2). The abdominal girth was reduced to 56 cm (Figure 1); IgA AGA fell to 70 RU/ml and EMA was weakly positive. Iron deficiency anaemia was corrected and supplements were taken off. Pica was no longer observed. Dietary compliance was satisfactory. Her parents were referred to medical unit for further management. Linkage study was not performed.

Discussion

With the growing identification of atypical or asymptomatic cases by sensitive serological markers, it has become increasingly evident that coeliac disease is more common than previously thought. Studies showed that coeliac disease is relatively common in Europe and North America, affecting up to 1 of every 200 to 300 persons.[1,2] Indian studies even give a prevalence of 5 to 6.8 percent.[3,4]

It is, however, still rare among Chinese, Japanese or people of African-Caribbean background.

The hereditary basis of coeliac disease is supported by the finding that it is diagnosed in around 10 percent of first-degree relatives of an individual with coeliac disease.[1] In addition, coeliac disease is concordant in up to 70 percent of identical twins.[2] Linkage disequilibrium with HLA-DQ2 is the main concern when genetic predisposition is considered. Over 90 percent of European patients with coeliac disease express the HLA-DQ heterodimer DQα1*0501/DQβ1*0201.[5] Association with another major histocompatibility complex (MHC) Class II molecule, HLA-DR, is also present. Individuals with HLA-DR3 status will carry both the α and β DQ molecules on the same chromosome and express them on the antigen-presenting cells. The same condition occurs in those with HLA-DR5 or HLA-DR7, but they carry one specific HLA-DQ molecule on each chromosome.[5] The antigen-presenting cells present the gluten-derived gliadin peptide in conjunction with the HLA-DQ2 dimer and induce an abnormal T-cell response. Tissue transglutaminase (the principal component of the endomysium autoantigen), IgA AGA and IgA EMA are produced by activated plasma cells. The former then modify the gliadin peptide and elicit an even stronger immune response causing cytokines release from mucosal lymphocytes, which finally damage the enterocytes.[1]

A wide range of gastrointestinal or extrainstestinal manifestations may occur singularly or in combination. In children, symptoms do not occur until gluten is introduced into an infant's diet, so late onset is also possible. Our patient presented with pallor, impaired growth and abdominal distension in early childhood, somewhat resembling the classical description in young children.[2] Vomiting and chronic diarrhoea, two common symptoms in coeliac disease, are however absent. Intussusception, which may be recurrent, is not rare.[6,7] The malabsorption of carbohydrate results in bacterial fermentation producing gas and adding a foul smell to faeces while malabsorbed fat leads to steatorrhoea and diarrhoea. Impaired xylose absorption and elevated faecal fat were documented in our case. Abdominal bloating, general wasting, lactose intolerance, and signs of multiple vitamin and mineral deficiencies are also common findings. Our patient did suffer from hypoalbuminaemia, iron deficiency anaemia and her serum zinc level was low. Behavioural disturbances, such as depression and irritability, are recognised in children with coeliac disease. Korman reported pica as a presenting symptom in three children with coeliac disease.[8] In these cases pica evidently resulted from iron deficiency secondary to malabsorption. A dramatic growth spurt and complete resolution of pica were observed after a gluten-free diet.[8] The progress of our patient follows the same pattern so far.

Availability of reliable serological markers drove the European Society of Paediatric Gastroenterology and Nutrition (ESPGAN) to revise the diagnostic criteria for coeliac disease in 1990. The diagnosis of coeliac disease does not require further confirmation if the initial diagnosis is based firstly on positive serologic and histologic findings while the patient is taking adequate amounts of gluten, and secondly on unequivocal and full clinical remission after gluten withdrawal.[9] A re-biopsy is necessary only for unsatisfactory or equivocal clinical response to a strict gluten-free diet; while gluten challenge is reserved for patients with doubtful initial diagnosis who do not have characteristic serologic or histologic findings.[1] IgA EMA is species specific,[10] with a reported sensitivity and specificity of 85 to 98 percent and 97 to 100 percent respectively in different reports.[1] It is believed to be the most reliable among all the available serological markers (including tissue transglutaminase and anti-reticulin antibodies) with the highest positive and negative predictive values.[11] IgA AGA is far less specific but still with moderate sensitivity (75 to 90 percent), and is more useful for monitoring dietary adherence because it is easier to quantify.[1] It is generally recommended that any patient with a positive test for EMA should undergo small bowel biopsy to establish the diagnosis of coeliac disease.

It is becoming more evident that silent or atypical subclinical disease is common in the general population. Some authors also recommend treating silent cases with standard gluten-free diet because of unpredictable potential clinical and histologic changes.[10] Those with short stature, insulin-dependent diabetes mellitus (IDDM) or being first-degree relatives of patients with coeliac disease are usually being screened. Rossi et al (1996) found that the positive rate was 8% in family members, 4% in IDDM and 1.7% in short stature, using IgA EMA as screening tool. Similar results were reported in European studies.[11] Using AGA, several large-scale screening studies in Europe have shown a prevalence rate of silent cases of 1 in 200.[10,12] AGA can also be positive in many clinical conditions such as atopic eczema, pemiphigus, dermatitis herpetiformis, Sjögren's syndrome and rheumatoid arthritis.[12] EMA measurement is an unreliable screening test in patients with selective IgA deficiency.[1,13]

Symptomatic improvement occurred within two weeks in around 70% of patients after starting gluten-free diet.[1]

Weight gain should be noted within three weeks. Normal weight is expected within six months followed by catching up of height around two months later.[14] In children, mood is no longer disturbed after days with appetite returning. Stools return to normal within few days or weeks. Abdominal distension takes months to resolve. IgA AGA levels usually normalise within 3 months whereas normal small gut histology may not be restored until 12 months.[14] Serial measurements of the IgA AGA level serve as a mean of assessing clinical response and monitoring dietary compliance. Inadequate adherence of diet should be considered if clinical response to gluten exclusion is not satisfactory.

The concept of lifetime commitment to gluten avoidance is being challenged recently. There is growing evidence supporting a long-term trend that favours greater gluten tolerance.[14] Schmitz (1996) reported children with coeliac disease who were left on normal diet could attain greater-than-predicted final height and reach normal puberty. Restoration of normal mucosal architecture was observed in some of them.[14] However, taking into account that enteropathy-associated T-cell lymphoma as well as carcinoma of oesophagus and small gut occur more often in complicated refractory and prolonged undiagnosed patients, it seems more prudent to recommend a strict dietary adherence and follow original directions. AGA levels show no correlation with malignancy complicating coeliac disease.[12] Surveillance imaging procedures, including radiograph, barium studies and computed tomography are usually unnecessary to screen for malignancy initially, but should be performed in refractory cases or those with unexpected relapse of signs and symptoms.[1] Long-term follow-up is mandatory for all patients with coeliac disease.

References

1. Farrell RJ, Kelly CP. Celiac sprue. N Engl J Med 2002;346: 180-8.
2. Feighery C. Fortnightly review: coeliac disease. BMJ 1999;319: 236-9.
3. Mohindra S, Yachha SK, Srivastava A, et al. Coeliac disease in Indian children: assessment of clinical, nutritional and pathologic characteristics. J Health Popul Nutr 2001;19:204-8.
4. Khoshoo V, Bhan MK, Jain R, et al. Coeliac disease as cause of protracted diarrhoea in Indian children. Lancet 1988;1:126-7.
5. Murch S, Walker-Smith J. Etiology and Pathogenesis of Childhood Coeliac Disease. International Seminars in Paediatric Gastroenterology and Nutrition 1996;5:11-15.
6. Mushtaq N, Marven S, Walker J, Puntis JW, Rudolf M, Stringer MD. Small bowel intussusception in celiac disease. J Paediatr Surg 1999;34:1833-5.
7. Martinez G, Israel NR, White JJ. Celiac disease presenting as entero-enteral intussusception. Pediatr Surg Int 2001;17: 68-70.
8. Korman SH. Pica as a presenting symptom in childhood celiac disease, Am J Clin Nutr 1990;51:139-41.
9. Walker-Smith JA, Guandalini S, Schmitz J, Shmerling DH, Visakorpi JK. Report of Working Group of European Society of Paediatric Gastroenterology and Nutrition. Revised criteria for diagnosis of celiac disease. Arch Dis Child 1990; 65:909-11.
10. Catassi C, Fabiani E, Ratsch IM, Coppa GV, Giorgi PL. Celiac Disease in the General Population: Should We Treat Asymptomatic Cases? J Pediatr Gastroenterol Nutr 1997;24:S10-S13.
11. Rossi TM, Tjota A. Serologic Indicators of Celiac Disease. J Pediatr Gastroenterol Nutr 1998;26:205-10.
12. Troncone R, Ferguson A. Anti-gliadin Antibodies [review]. J Pediatr Gastroenterol Nutr 1991;12:150-8.
13. Bousvaros A. Coeliac Disease: Introduction. Int Sem Paediatr Gastroenterol Nutr 1996;5:1-2.
14. Schmitz J. Epidemiology, Diagnosis and Monitoring of Childhood Coeliac Disease. Int Sem Paediatr Gastroenterol Nutr 1996;5:2-7.