Medical Journal of the
Islamic Republic of Iran

Volume 15
Number 2
Summer 1380
August 2001

Case Reports

CHRONIC DIARRHEA DUE TO GANGLIONEUROMA IN A CHILD

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ABSTRACT

Ganglioneuroma is a very rare cause of chronic diarrhea in children. This benign tumor usually presents with diarrhea and failure to thrive. It is necessary for physicians to be familiar with this rare cause of chronic diarrhea. Here we present and discuss such a case.

MJIRI, Vol. 15, No. 2, 115-116, 2001.

Keywords: Ganglioneuroma, children, chronic diarrhea, celiac disease.

INTRODUCTION

Chronic diarrhea is a challenger for the physician because of variability in the signs of disease and difficulty in evaluating its severity. Moreover, the extensive differential diagnosis and the different available diagnostic tests complicate the matter.

Secretory chronic diarrhea may be a clinical manifestation of neural crest tumors in children which persists even when the patients is ordered to remain NPO. Diarrhea is massive and results in fluid and electrolyte imbalance and failure to thrive. Diagnosis is based on the presence of secretory watery diarrhea, extra-intestinal manifestations, results of hormone analysis and imaging techniques. Surgical resection of the tumor is the treatment of choice.

CASE REPORT

A 4-year-old girl was admitted to the emergency department with chronic diarrhea, polyuria and polydipsia. She had been otherwise healthy till the age of 8 months when, after a common cold, she developed foul smelling and bulky diarrhea with a frequency of 7-8 times a day. Routine laboratory tests were negative and she was treated for viral gastroenteritis. Three months later, when she was 11 months old, her urine culture was positive and she was admitted to hospital for a 2-week course of parenteral antibiotics.

Gradually, she failed to thrive and developed intermittent polyuria and polydipsia. At the age of 3, she was hospitalized for head injury due to a fall and developed seizures and vomiting. Brain CT scan and lumbar puncture were normal, so she was discharged after one week. Small bowel biopsy revealed villous atrophy compatible with celiac disease. On admission the girl appeared ill and her mucous membranes were markedly dry. Her abdomen was midly distended but organomegaly or a palpable mass were not detected. Vital signs were as follows: blood pressure, 100/50 mmHg; axillary temperature, 36.2°C (97.2°F); pulse rate, 62 beats/min; respiratory rate, 14 breaths/min and weight, 11kg. Laboratory findings were as follows: serum potassium, 2.8 mEq/L; serum chloride, 100mEq/L; pH, 7.38; P co_2, 11.4%; base excess, 12.4 mmol/L; erythrocyte sedimentation rate, 47 mm/h. Urine analysis revealed a specific gravity of 1.010 and a pH of 5. Abdominal ultrasonography revealed an enlarged right kidney with hydronephrosis. Intravenous

pyelography showed duplication of the right renal pelvis and ureter. Dimercaptosuccinic acid scans showed a filling defect in the right kidney. Final diagnosis was revealed after surgical intervention for the kidney anomaly.

DISCUSSION

Chronic diarrhea in children is defined as a daily stool output greater than 10g/kg/24hr lasting for at least 2 weeks. The two major factors resulting in diarrhea are intraluminal and mucosal factors. Evaluation of patients with chronic diarrhea includes clinical history, physical examination, stool examination, hematological studies, sweat test, endoscopy, biopsy, barium studies, and hormonal studies.

FTT usually refers to growth below the 3rd percentile or a change in growth that has crossed two major growth percentiles. The cause of organic FTT are numerous and any organ may be involved. After taking a history, performing a physical examination, and observing the parent-child interaction, it may be helpful to approach the diagnosis based on signs and symptoms.[2]

In this patient, primary investigation was normal but small bowel biopsy revealed villous atrophy compatible with celiac disease; a restricted gluten-free diet was not successful. It should be noted that villous atrophy can occur in other conditions as well.

Although it is more favorable to consider all the patient's signs and symptoms as a single disease, the history of head injury is probably a separate incident. Episodic weakness known as periodic paralysis is associated with transient alterations of serum potassium levels, usually hypoalbuminemia, which may be due to many factors. Urethral duplication is the most common urethral anomaly, which may be incidental. The patient underwent surgery for vesicourethral reflux resulting from urethral duplication. During the operation, a well-defined mass was found near the spine and pathologic examination revealed a ganglioneuroma. Ganglioneuroma is the fully differentiated and benign counterpart of neuroblastoma[3] and is principally composed of mature ganglion cells, neutrophils and Schwann cells.[4] This tumor is one of the uncommon causes of chronic diarrhea in childhood. Ganglioneuroma has been found most commonly in the abdomen. The immediate cessation of diarrhea after removal of the tumor suggests that a neuro-hormonal substance, which affects intestinal function, is secreted by ganglioneuromas. Vasoactive intestinal peptide (VIP) is probably the most common agent knwon to induce watery diarrhea. An abdominal CT scan, MRI, bone scan and bone marrow biopsy were performed after surgery. Results were negative and vanilylmandelic acid was 2.8 mg/g creatinine. Two years after the management. The girl did not mention repeated diarrhea.

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