【 CASE REPORT 】

Megaesophagus and Megaduodenum Found Incidentally on a Routine Chest Radiograph During a Health Examination

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Abstract:

Chronic idiopathic intestinal pseudo-obstruction (CIIP) caused by impaired intestinal peristalsis leads to intestinal obstructive symptoms. A 20-year-old man had marked esophageal dilatation that was found incidentally on chest radiography during a health examination. Chest/abdominal contrast-enhanced computed tomography and endoscopy showed marked esophageal and duodenal dilatation without mechanical obstruction. Upper gastrointestinal series and high-resolution esophageal manometry revealed absent peristalsis in the dilated part. CIIP was suspected in the patient's father, suggesting familial CIIP. The patient likely had signs of pre-onset CIIP. This is the first case of suspected CIIP in which detailed gastrointestinal tract examinations were performed before symptoms appeared.

Key words: chronic idiopathic intestinal pseudo-obstruction, gastrointestinal motility disorder, intestinal dilatation, gastrointestinal series, esophageal manometry, chest radiograph

(Intern Med 60: 2039-2046, 2021)
(DOI: 10.2169/internalmedicine.6324-20)

Introduction

When gastrointestinal dilatation is detected during an imaging examination, it is necessary to first exclude any mechanical obstruction before diagnosing intestinal dilatation due to gastrointestinal motility disorder associated with decreased intestinal peristalsis. The causes of intestinal dysmotility are classified into primary (idiopathic) and secondary. The main cause is known as chronic idiopathic intestinal pseudo-obstruction (CIIP), a pathological condition of unknown etiology that causes intestinal dysmotility and dilatation (1-3). Secondary intestinal dysmotility is caused by collagen and endocrine diseases and drug-induced conditions; the administration of drugs, such as antidepressants, anticholinergics, and opioids, can lead to secondary chronic intestinal pseudo-obstruction.

The term chronic intestinal pseudo-obstruction encompasses both CIIP and secondary chronic intestinal pseudo-obstruction. CIIP is a refractory disease characterized by chronic, irreversible progression of intestinal dysmotility, leading to malnutrition due to dilatation-associated intestinal obstructive symptoms and dysfunction of the small intestine. Many cases are sporadic, but familial occurrence has been reported as well, and the disease can affect any part of the gastrointestinal tract (4-12).

We herein report a case of a young man with megaesophagus and megaduodenum that was detected incidentally on a routine chest radiograph obtained during a health examination. CIIP was suspected in the patient's father; therefore, the patient was diagnosed with intestinal dilatation before the onset of CIIP, although he was asymptomatic. To our knowledge, this is the first reported case of suspected CIIP in which detailed examinations of the gastro-

Table 1.   Blood Test Findings.

Blood cell counts		Blood biochemistry findings			
WBC (/μL)	7,600	AST (IU/L)	22	Glucose (mg/dL)	93
Granulocytes	63.8%	ALT (IU/L)	24	HbA1c (%)	5.5
Lymphocytes	27.5%	LDH (IU/L)	141	TSH (μIU/mL)	2.15
Monocytes	6.1%	T-Bil (mg/dL)	0.7	FT3 (pg/mL)	3.65
Eosinophils	2.2%	ALP (IU/L)	251	FT4 (ng/dL)	1.47
Basophils	0.0%	γ-GTP (IU/L)	14	PTH-intact (pg/mL)	33
RBC (/μL)	$544×10^4$	TP (g/dL)	7.7	ANA	1:40
Hb (g/dL)	16.6	Alb (g/dL)	4.5	Anti-SM	Negative
MCV (fL)	87.7	CK (IU/L)	69	Anti-Scl-70	Negative
Ht	47.7%	BUN (mg/dL)	11	Anti-Jo-1	Negative
Plt (/μL)	$21.7×10^4$	Cr (mg/dL)	0.73	ACA	Negative
		Na (mEq/L)	142	$1,25\text{-}(OH)_2$ Vitamin D (pg/mL)	84.0
		K (mEq/L)	3.8		
		Cl (mEq/L)	103		
		Ca (mg/dL)	9.8		
		CRP (mg/dL)	0.12		

ACA: anticentromere antibody, Alb: albumin, ALP: alkaline phosphatase, ALT: alanine aminotransferase, ANA: antinuclear antibody, AST: aspartate aminotransferase, BUN: blood urea nitrogen, CK: creatine kinase, Cl: chloride, Cr: creatinine, CRP: C-reactive protein, Hb: hemoglobin, Ht: hematocrit, K: potassium, LDH: lactate dehydrogenase, MCV: mean corpuscular volume, Na: sodium, Plt: platelet, PTH: parathyroid hormone, RBC: red blood cell count, T-Bil: total bilirubin, TP: total protein, TSH: thyroid-stimulating hormone, WBC: white blood cell count, γ-GTP: gamma-glutamyl transpeptidase
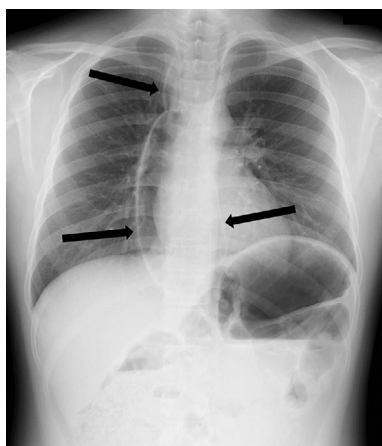

Figure 1.   Chest radiography findings. Marked dilatation of the esophagus (arrows). A prominent gastric bubble and niveau in the stomach and duodenum are visible.

Figure 2.   Abdominal radiography findings. Gas in the large intestine is not prominently seen.

intestinal tract were performed before the appearance of abdominal symptoms.

Case Report

The patient was a 20-year-old man who had been diagnosed with epilepsy at 5 years old. He was taking oral sodium valproate 600 mg/day. He visited our hospital due to marked dilatation of the esophagus detected on a chest radiograph during a health examination.

He was 162 cm tall and weighed 63 kg, and his vital signs were normal. He had no history of smoking or drinking and had no known food or drug allergies. He had no

subjective symptoms, including abdominal symptoms such as bloating, nausea/vomiting, abdominal pain, or abnormal bowel movements. No abnormal findings were noted on a physical examination. Blood test results were within the normal range and revealed no abnormalities in electrolytes or anemia (Table 1).

A chest radiograph showed marked dilatation of the esophagus with a prominent gastric bubble and niveau in the stomach and duodenum (Fig. 1). Gas in the large intestine was not prominent in an abdominal radiograph (Fig. 2). Contrast-enhanced chest/abdominal computed tomography

Figure 3.   Chest and abdominal contrast-enhanced computed tomography findings. (a) Axial plane image showing marked dilatation from the upper to lower esophagus. (b) Axial plane image showing dilatation of the stomach. (c) Axial plane image showing marked dilatation of the duodenum. (d) Coronal plane image showing marked dilatation from the bulb to transverse part of the duodenum. No obstruction is evident.

(CT) showed marked dilatation from the upper to lower esophagus and mild dilatation of the stomach. In addition, marked dilatation was observed from the bulb to the transverse part of the duodenum. No obstruction was noted in the digestive tract, and no dilatation was seen in the jejunum, ileum, or large intestine (Fig. 3). Peroral balloon-assisted endoscopy showed marked dilatation and fluid retention from the upper to lower esophagus. The esophagogastric junction showed no organic disease that could cause stenosis, and passage of the fiberscope was easy. Marked dilatation was seen from the bulb to the transverse part of the duodenum. In addition, no organic disease that could cause obstruction was noted at the transition point between the dilated and non-dilated parts of the duodenum, and passage of the fiberscope was easy. Endoscopy revealed no organic disease up to 100 cm distal to the ligament of Treitz (Fig. 4). Upper gastrointestinal series revealed marked dilatation from the upper to lower esophagus; however, no apparent stenosis was present at the esophagogastric junction. Although esophageal peristalsis was absent, delayed passage of the contrast medium to the stomach was observed. The stomach was slightly dilated, and although the peristalsis decreased,

outflow of contrast medium to the duodenum was adequate. Marked dilatation was observed from the bulb to the transverse part of the duodenum. Although peristalsis of the dilated duodenum was absent, delayed outflow of contrast medium to the anal side of the small intestine was noted (Fig. 5). High-resolution manometry showed an integrated relaxation pressure value of 15.1 mmHg, which was within the normal range, indicating that relaxation of the lower esophageal sphincter was not impaired. The rate of failed contractions was 100%, indicating the absence of esophageal peristalsis. Achalasia was ruled out according to the Chicago Classification of Motility Disorders v3.0 (13), and absent contractility was diagnosed (Fig. 6).

The present case met the following four of seven diagnostic criteria for CIIP (Table 2): 1) presence of marked dilatation of the esophagus and duodenum, 2) no organic disease causing digestive tract obstruction, 3) absence of peristalsis of the dilated digestive tract, and 4) no underlying disease. The patient's father was being treated for intestinal Behçet's disease in our hospital and was suspected of having CIIP because of dilatation of the digestive tract without organic obstruction. He had started to experience abdominal symp-
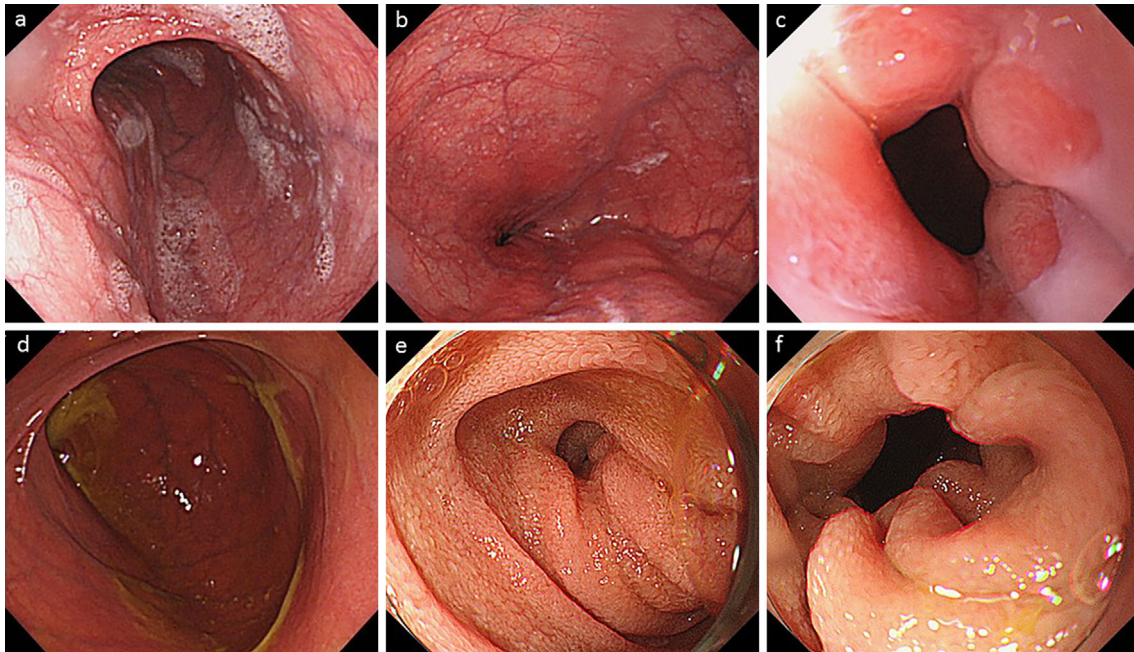
Figure 4. Peroral balloon-assisted endoscopic findings. (a) Esophagus. Marked dilatation and fluid retention is noted from the upper to lower esophagus. (b) Esophagogastric junction. (c) Enlarged image from (b). The presence of organic diseases causing stenosis is not noted, and the fiberscope could pass easily. (d) Descending part of the duodenum. Marked dilatation from the bulb to the transverse part of the duodenum. (e) Transition point between the dilated and non-dilated duodenum. (f) Enlarged image from (e). The presence of organic diseases causing stenosis is not noted, and the fiberscope could pass easily.

Figure 5. Upper gastrointestinal series. (a) Marked dilatation from the upper to lower esophagus. Apparent stenosis at the esophagogastric junction is not observed. Although esophageal peristalsis is absent, delayed passage of the contrast medium to the stomach is observed. (b) Slight dilation in the stomach. Peristalsis is decreased, but outflow of the contrast medium to the duodenum is adequate. (c) Marked dilatation from the bulb to the transverse part of the duodenum. Although peristalsis of the dilated duodenum is not observed, delayed outflow of contrast medium to the anal side of the small intestine is observed.

toms in his 20s, and bowel obstruction due to superior mesenteric artery syndrome was suspected. Therefore, duodenal resection had been performed when the father was 34 years old at another hospital. After the surgery, he visited our hospital. He sometimes had symptoms of intestinal obstruction that required decompression with an ileus tube. We performed contrast-enhanced chest/abdominal CT, upper gastrointestinal endoscopy, colonoscopy, and small bowel series. The father's intestinal Behçet's disease was well controlled with mesalazine, and no inflammation or intestinal stenosis
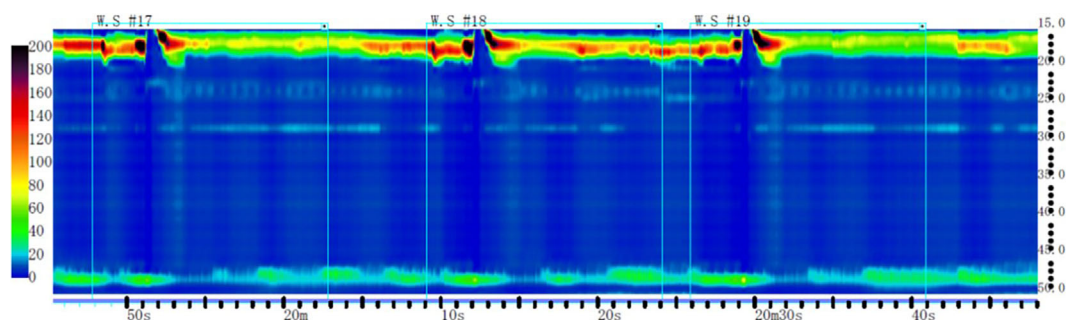
Figure 6. High-resolution manometry findings. The integrated relaxation pressure value is within the normal range, indicating that relaxation of the lower esophageal sphincter is not impaired. The rate of failed contractions was 100%, indicating the absence of esophageal peristalsis. Based on the Chicago Classification of Motility Disorders v3.0 (13), absence of contractility is diagnosed.

Table 2.   Diagnostic Criteria for Chronic Idiopathic Intestinal Pseudo-obstruction.

The following 7 criteria must be met:

1. Severe symptoms of intestinal obstruction that require hospitalization, such as abdominal distention, nausea/vomiting, and abdominal pain, persist or recur over a long period.

2. The duration of symptoms is ≥ 2 months for neonatal-onset cases and ≥6 months for cases with post-infancy onset.

3. Dilated gastrointestinal tract and niveau are observed on imaging examinations (1).

4. Organic lesions blocking the gastrointestinal tract are not observed.

5. Hematoxylin-eosin (HE) staining of a full-thickness gastrointestinal wall biopsy specimen shows no morphological abnormalities in the neural plexus (2).

6. In pediatric patients, megacystis-microcolon-intestinal hypoperistalsis syndrome (MMIHS) and segmental dilatation of intestine should be ruled out.

7. Secondary chronic intestinal pseudo-obstruction (CIPO) should be ruled out.

1) In neonates, the observation of niveau in upright plain radiographs of the abdomen is not necessary for diagnosis.

2) If full-thickness gastrointestinal wall biopsy is difficult, evidence of apparent dysmotility centering on the small intestine is shown on cine magnetic resonance imaging or gastrointestinal manometry.

Adapted from the Japan Intractable Diseases Information Center.
https://www.nanbyou.or.jp/entry/3961 (September 2020)(34).

had been observed. Contrast-enhanced chest/abdominal CT showed marked dilatation of the stomach and small and large intestines without intestinal stricture and no findings of intestinal adhesion after duodenal resection (Fig. 7). No dilatation was seen in the esophagus. Small bowel series using the ileus tube revealed marked dilatation of small intestine but no apparent stenosis (Fig. 8). Although peristalsis of the dilated small intestine was not observed, delayed outflow of contrast medium to the anal side of the digestive tract was noted. Based on the examination findings, we suspected that the father had CIIP. He is currently 60 years old and is receiving diet and bowel control therapy with the administration of magnesium oxide and acotiamide hydrochloride for abdominal symptoms, including bloating, nausea, and abnormal bowel movements.

Based on the above findings, the intestinal dilatation in the present case can be regarded as a pre-symptomatic condition of CIIP. No abdominal symptoms have occurred as of 16 months since the first visit to our hospital; however, because the patient meets most of the diagnostic criteria for CIIP, careful follow-up is planned in order to evaluate whether or not abdominal symptoms persisting for more than six months will appear.

Discussion

CIIP, named by Maldonado et al. in 1970 (1), is a chronic and irreversible disease that causes pathological dilatation due to impaired peristalsis of the intestine in the absence of organic disease, resulting in persistent intestinal obstructive symptoms, such as bloating, abdominal pain, and vomiting (1-3, 14). The small intestine is mainly affected; however, impairment of peristalsis can occur anywhere in the gastrointestinal tract, from the esophagus to the rectum, synchronously or asynchronously. Some reports have used terms such as megacolon, megaduodenum, and megaesophagus to refer to the major lesion of the dilated digestive tract.

CIIP is classified into the following four pathological categories: 1) visceral myopathy, 2) visceral neuropathy, 3) abnormality of interstitial cells of Cajal, and 4) mixed type (15-18). Peristalsis occurs due to the interaction between the intestinal smooth muscles and the enteric nervous

Figure 7. Abdominal contrast-enhanced computed tomography scans of the patient's father. Axial plane image showing marked dilatation of the stomach and small and large intestines. No obstruction is seen in the digestive tract.
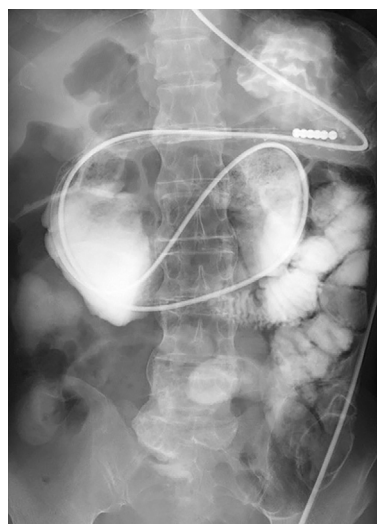

Figure 8. Small bowel series of the patient's father. Small bowel series using the ileus tube revealed marked dilatation of small intestine. No stenosis is apparent. Although peristalsis of the dilated small intestine is not observed, delayed outflow of contrast medium to the anal side of the digestive tract is noted.

system (ENS). The ENS consists of Auerbach's and Meissner's plexus, which are present throughout the entire gastrointestinal tract and regulate sensory signals and intestinal motility. It is thought that congenital and acquired anomalies in the ENS can cause CIIP, the occurrence of which can be either sporadic or familial, and the severity of gastrointestinal motility disorders, affected locations, and histological findings are not always consistent, indicating that it is a heterogeneous syndrome rather than a single genetic abnormality (19, 20). The prevalence in Japan is 1.0 per 100,000 men and 0.80 per 100,000 women. with no significant sex difference (21). The onset of CIIP can occur at any age over a wide range, from youth to old age; however, it is reported to be particularly frequent in young people in their teens and 20s (22).

All seven criteria shown in Table 2 must be met for the diagnosis of CIIP to be made. In the early stage of CIIP, abdominal symptoms are absent or very mild if present at all, due to the body's compensatory mechanisms (compensatory phase). In this phase, nutrient absorption by the intestinal tract is maintained, and patients can live a normal daily life with only a proper diet and bowel movement control. However, once the compensatory mechanisms fail to function and the transit of food slows down, the retained contents of the intestine lead to dilatation of the digestive tract, which manifests as abdominal symptoms, such as bloating, nausea/vomiting, and abdominal pain (non-compensatory phase). In this phase, digestion and absorption are impaired due to small bowel dysfunction, causing malnutrition. The disruption of the intestinal barrier function due to the excessive elevation of intraluminal pressure causes bacterial translocation, which often results in sepsis. In the non-compensatory phase, adequate nutritional management and decompression therapy are required. If oral intake is possible, small, frequent meals and elemental diet should be provided, and if oral intake becomes impossible, intravenous hyperalimentation (IVH) is necessary.

CIIP is a chronic intestinal obstruction that requires continuous decompression. Therefore, treatment using ileus tubes is limited. Ohkubo et al. reported the efficacy of decompression therapy by percutaneous endoscopic gastrojejunostomy (PEG-J), in which a jejunal tube is placed through an endoscopically created gastrostomy to percutaneously drain the contents of the small intestine (23). According to their report, this method enabled continuous decompression without inserting a trans-nasal tube, which helped improve subjective symptoms and the nutritional status and dramatically shortened the hospital stay. In principle, resection of the affected intestine should not be performed without careful consideration because of the risk of dilatation in the residual intestine, often resulting in the exacerbation of symptoms (24). Surgical therapy is limited to cases where perforation, torsion, or strangulation of the intestine occurs. Although drug therapy is not very effective, bowel movement control is important, and stimulant laxatives, prokinetics, mosapride, and metoclopramide are used in combination. The reported 10-year survival rate of patients in the non-compensatory phase who require home IVH therapy is 75% (25). The causes of death include malnutrition, IVH-associated catheter infection, bacterial translocation-related sepsis, and IVH-associated liver failure/pulmonary embolism (25).

Most CIIP cases are sporadic; however, some show familial occurrence, and the question of their genetic basis has attracted attention (4-12). Many reports have described an autosomal dominant inheritance pattern (5, 6, 15); however, one study reported a case of an autosomal recessive pattern (4). A mutation in the ACTG2 gene has also been identified in CIIP patients (26-28).

Sodium valproate elicits antiepileptic activity by increasing the concentration of gamma-aminobutyric acid (GABA), the principal inhibitory brain neurotransmitter (29). GABAergic neurons play an important role in controlling

peristalsis and GABA can decrease gastrointestinal motility (30, 31). Mo et al. reported a case of paralytic ileus with abdominal symptoms, including severe abdominal distension and vomiting, within four days after the administration of sodium valproate (32). It has been reported that sodium valproate increases the lower esophageal sphincter (LES) resting pressure without affecting postdeglutition relaxation of the LES and peristaltic activity of the esophagus (33). In our case, the patient has been taking sodium valproate for 15 years, since 5 years of age, and high-resolution manometry showed a normal LES pressure and the absence of esophageal peristalsis. Therefore, whether or not sodium valproate caused intestinal dilation in this case is unclear.

In the present case, the patient had no abdominal symptoms and did not meet all of the CIIP diagnostic criteria. When the intestinal dilatation started or whether or not it was progressing was unclear, so it may not have been pathogenic. However, based on the examination findings, the patient may have presented with esophageal and duodenal dilatation due to pre-onset CIIP. The patient is currently asymptomatic, but if symptoms of intestinal obstruction appear in the future, we plan to treat him through dietary modifications (small, frequent meals and low-residue and elemental diet) and bowel movement control. If the patient is ultimately diagnosed with CIIP, we may be able to conclude the following: 1) CIIP is a chronic, progressive condition; 2) nutrition therapy with IVH and decompression therapy, such as using an ileus tube or PEG-J, is required in the long term; and 3) CIIP may be inherited. Since there is no radical treatment for this disease, the main principles of treatment are to avoid small intestine dysfunction and to improve the patient's quality of life.

The authors state that they have no Conflict of Interest (COI).

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