# GASTRIC ANTRAL STRICTURE IN A PATIENT WITH CHRONIC GRANULOMATOUS DISEASE*

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**SUMMARY: Metin A, Sanal Ö, Tezcan İ, Ersoy F, Berkel Aİ. (Immunology Unit, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey). Gastric antral stricture in a patient with chronic granulomatous disease. Turk J Pediatr 1999; 41: 369-373.**

**Chronic granulomatous disease (CGD) is a rare disorder of phagocytic cell oxidative metabolism. Patients have recurrent infections with catalase-positive organisms and granulomatous lesions throughout the body. Gastric antrum can be an occult site of involvement. We describe a four-year old boy with chronic granulomatous disease who was admitted with the complaints of persistent vomiting and weight loss. Gastric antral narrowing was diagnosed according to radiological findings. Treatment with steroid and antibiotics yielded a good clinical response in 15 days with a relief of the obstruction. This case report emphasizes the beneficial effect of this form of therapy in preventing life-threatening obstruction of vital organs in CGD.** *Key words: chronic granulomatous disease, complications.*

Chronic granulomatous disease (CGD) is a rare disease affecting about 1 in 500,000 individuals. It is characterized by recurrent, life-threatening infections with catalase-positive microorganisms and excessive inflammatory reactions that lead to granuloma formation. Neutrophils do not respond to various stimuli with respiratory burst, therefore failing to reduce molecular oxygen to superoxide which is required for the generation of other toxic metabolites such as hydrogen peroxide and hydroxyl radicals. Several different mutations of NADPH-oxidase are associated with this disorder[1]. In addition to recurrent and severe pulmonary and skin infections, CGD patients have inflammatory disease of the gastrointestinal and urinary systems which leads to luminal narowing[2-8]. The general management of CGD includes prophylactic antimicrobial agents in order to prevent infection and immunomodulatory therapy with interferon-gamma[9]. Here we present a CGD patient with granulomatous narrowing of the gastric antrum who was treated successfully with corticosteroid and antibiotic.

## Case Report

A four-year-old boy diagnosed as having CGD at the age of two months was admitted to Hacettepe Children's Hospital with persistent vomiting and weight loss. There was a medical history of a recent cranial trauma, the complaints of

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which had been evaluated at a local hospital. His cranial computed tomography was (CT) normal, and he was treated for the diagnosis of peptic ulcerative disease because of an antral abnormality shown by roentgenograms. His first admission to our hospital was at two months of age. He was the fourth child of parents who are first-degree relatives. The first and second children were healthy boys, but the older sister of the patient had also been admitted to our hospital for generalized lymphadenopathy two months after BCG vaccination. Culture of the left axillary discharge grew M. bovis (BCG strain). She was diagnosed with CGD by slide NBT test and died at eight months of age after developing pneumonia at home.

On the first admittance to our hospital at two months of age, our patient's physical examination revealed generalized pustular rash; a liver and spleen palpable 3 and 2 cm below the costal margins, respectively, and a suppurative lymphadenitis on the left inguinal region. Cultures were positive for coagulase-positive staphylococci, and the patient was diagnosed as his sister with CGD. Medical management from infancy included administration of trimethoprim-sulfamethoxazole (TMP-SMX) and itraconazole. Over a three-year period he was admitted regularly for controls and treatment of occasional upper and lower respiratory tract infections, and he was hospitalized for a deep neck infection at three years of age. He achieved age-appropriate developmental milestones between the $3^{rd}$-$10^{th}$ percentile. Hepatosplenomegaly disappeared. He was initially maintained on prophylactic TMP-SMX and itraconazole but for the previous 13 months, his parents were unsuccessful in administering the medication due to the child's resistance.

On this admission at the age of four years with persistent emesis of one-month duration, findings remained unchanged. He had several small, mobile, non-tender cervical and inguinal lymph nodes and scars which were fully healed. There was no hepatosplenomegaly or pulmonary symptoms. He was midly dehydrated. His complete blood count showed a hemoglobin level of 12 g/dl, a leukocyte count of 10,000/µl with 55 percent neutrophils, 3 percent banded neutrophils, 36 percent lymphocytes, 6 percent monocytes and a platelet count of 385,000/µl.

Esophagus, stomach, and duodenum radiograms with barium swallow taken to investigate a gastric outlet obstruction showed antral narrowing (Fig. 1). Treatment with oral methylprednisolone (2 mg/kg/day, bid) and clindamycin (25 mg/kg/day, qid) decreased the frequency of emesis in a week and the patient began tolerating oral feeding. After 14 days, the methylprednisolone dose was slowly tapered over two months and stopped. He has been doing well four about eight months without recurrence of symptoms, and control radiographic studies are normal (Fig. 2). Daily prophylactic TMP-SMX and itraconazole therapies were started again and have been continued to date.

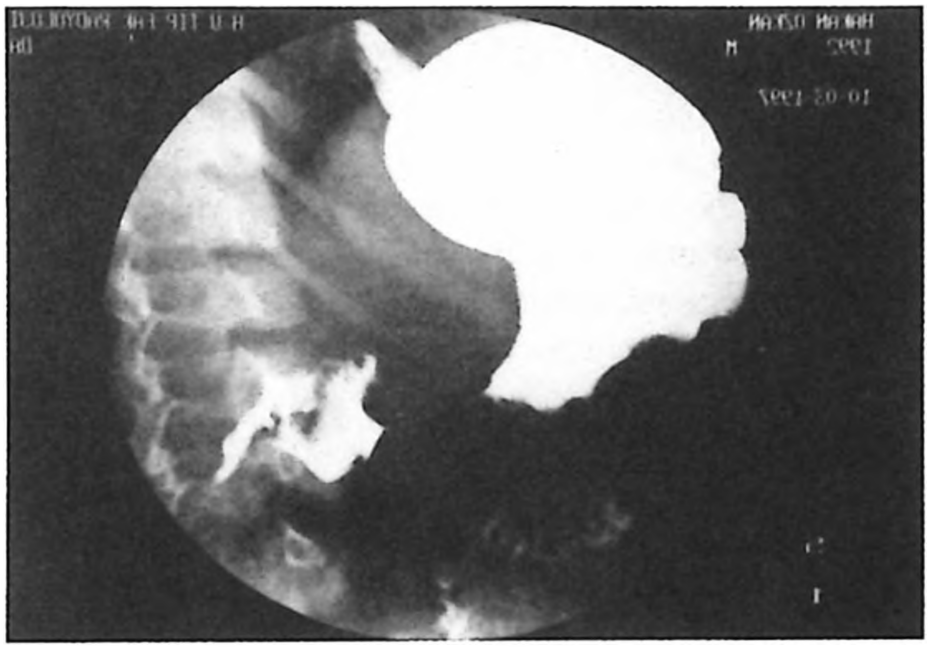

Fig. 1: Upper gastrointestinal radiography shows nearly complete gastric outlet obstruction.

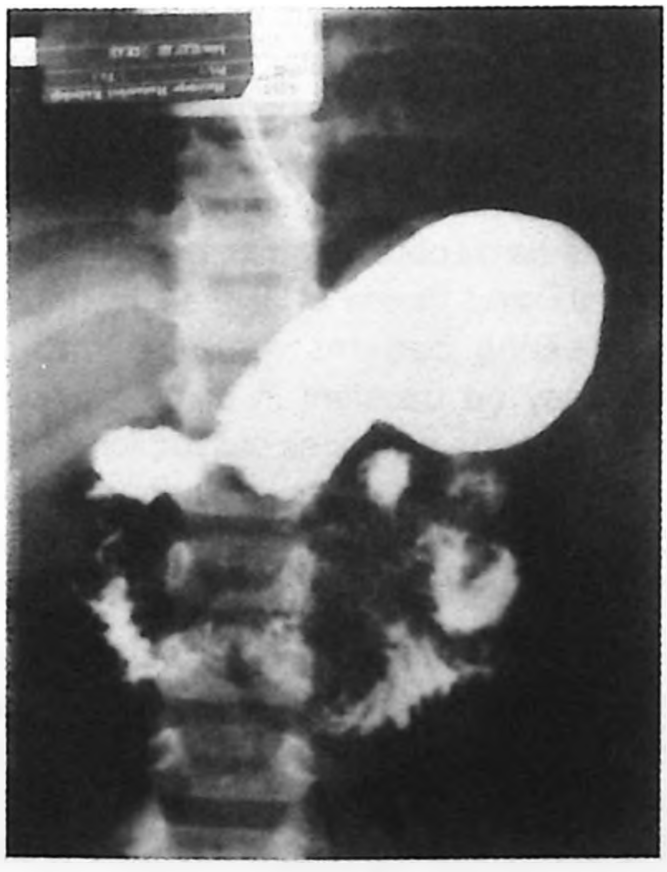

Fig. 2: Appearance of antrum after steroid therapy.

## Discussion

Gastrointestinal manifestations of CGD include persistent diarrhea, vitamin $B_{12}$ malabsorption, steatorrhea, and diffuse or localized granulomatous and obstructive lesions in the esophagus, gastric outlet or entire intestine due to incomplete resolution of the inflammatory response[10]. Involvement of the terminal ileum and colon results in enteritis and colitis similar to that of Crohn's disease. Gastric antral narrowing or obstruction may lead to vomiting, delayed gastric emptying and malnutrition. Genitourinary tract may also be an occult site of involvement. A retrospective chart review revealed that seven of 60 CGD patients (10%) had ureteral strictures[6]. Histopathological changes leading to obstruction in luminous organs are the same as in other parts of the body, characterized by sterile non-caseating granulomas with an accumulation of phagocytes and giant histiocytes and focal necrosis in the muscle layers.

Griscom et al.[2] first reported gastrointestinal obstruction as a major complication in CGD in 1974. Since this initial report, 20 patients with CGD and gastric outlet obstruction have been reported.

Surgery must be considered in the treatment of certain obstructions in CGD patients. However, because of the possibility of postoperative complications due to excessive granuloma formation in the site of operation, defective wound healing and fistulization, corticosteroid and antibiotic treatment is preferred. Indeed, this protocol has been effectively used in many patients to reverse the excessive inflammatory process in CGD[2,3,5,6]. In our patient, symptomatic relief occurred within two weeks and allowed outpatient management, as in the patient described by Chin et al.[3] and we thus avoided surgical intervention. However, responses of different patients or of the same patient at different episodes may vary. Patients described by Bowen et al.[7] and Varma et al.[8] received therapy for five to six months before normal eating patterns returned. Improvement with this combination of therapy may be transient in some patients for variable time periods. Some patients showed recurrences of symptoms[2,3]. On the other hand, most of the patients described to date showed urinary system involvement as well (granulomatous cystitis, bladder outlet obstruction or ureteral obstruction[2,3,6]. Therefore, therapy for these complications has been difficult.

Differential diagnosis of gastic antral narrowing in a child should include Crohn's disease, peptic ulcerative disease and eosinophilic gastritis, but these conditions seldom produce the abrupt annular narrowing of the antrum usually seen in CGD patients. Annular narrowing may occasionally be the presenting symptom of CGD[8]. Therefore, when an antral stenosis is discovered, an NBT test should be done to exclude CGD.

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