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한림병원 내과

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Hypereosinophilic Syndrome Presenting as Intussusception in Adults

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Hypereosinophilic syndrome (HES) is characterized by persistent eosinophilia associated with damage to multiple organs. Although the diagnostic criteria for HES include sustained hypereosinophilia for at least 6 months, early initiation of therapy may be recommended in patients exhibiting HES symptoms. Eosinophilic enteritis has been reported as a cause of intussusception in several cases. However, HES as a cause of intussusception has not yet been reported. In the present report, we describe a case of HES that manifested as jejunojejunal intussusception. Although long-standing hypereosinophilia was not confirmed, the patient required eosinophil-lowering therapy for an intestinal obstruction. The patient was treated with systemic corticosteroids, after which the symptoms and multiple organ involvement, including intussusception, improved dramatically, as noted on the radiological investigation. Moreover, surgery was not necessary. (Korean J Med 2014;87:585-588)

Keywords: Hypereosinophilic syndrome; Intussusception

INTRODUCTION

Hypereosinophilic syndrome (HES) is characterized by persistent eosinophilia associated with damage to multiple organs. Any organ system may be involved in a patient with HES. The heart, central nervous system, skin, and respiratory tract are most commonly involved, whereas HES with predominantly gastrointestinal symptoms is very rare [1]. Thus far, only a few cases of HES with gastrointestinal involvement in the form of

eosinophilic colitis have been reported [2,3]. HES presenting as intussusception in adults has not been reported. We describe such a case of HES presenting as intussusception that was treated with steroids.

CASE REPORT

A 19-year-old male was admitted to our hospital with abdominal pain, nausea, and vomiting for 3 days. He had no his-

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tory of medication use, atopy, allergy, or parasitic infestation. His blood pressure was 120/70 mmHg, pulse rate was 67/min, respiratory rate was 20/min, and body temperature was 37.7°C on admission. A physical examination showed mild tenderness

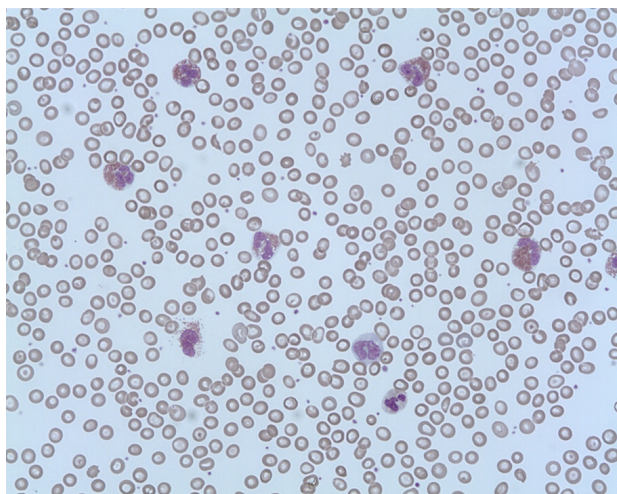


Figure 1. Peripheral blood smear revealed an increase in the number of eosinophils without blasts (Wright stain, $\times 400$).

in the left lower abdomen with normal bowel sounds. Cardiovascular, respiratory, and neurological examinations yielded normal results. Laboratory investigations revealed a white blood cell count of $11,540/\text{mm}^3$ with 31.7% eosinophils, eosinophil count of $3,658/\text{mm}^3$, hemoglobin level of 17.3 g/dL, and platelet count of $395,000/\text{mm}^3$. A peripheral blood smear showed marked eosinophilia without blasts (Fig. 1). Other serum values were within normal ranges. Negative test results were obtained for viral markers (hepatitis A, B, C, and human immunodeficiency virus), serum antibodies to parasites, autoimmune antibodies (anti-nuclear and anti-neutrophil cytoplasmic antibodies and rheumatoid factor), tumor markers (alpha-fetoprotein, carbohydrate antigen 19-9, and carcinoembryonic antigen), a stool examination (parasites, protozoa, and egg count), and a skin prick test. The findings of plain chest and abdominal radiographs were normal.

Abdominal computed tomography (CT) showed a jejunojejunal intussusception (Fig. 2A), distal duodenal and jejunal wall edema (Fig. 2B), a small volume of fluid collected in the pelvis (Fig. 2C), multiple irregular hypoattenuated lesions in both liver lobes, suggesting multiple eosinophilic liver abscesses (Fig. 2D), and a few small nodules surrounded by ground-glass opacities in both lungs, suggesting eosinophilic pneumonitis (Fig. 2E).

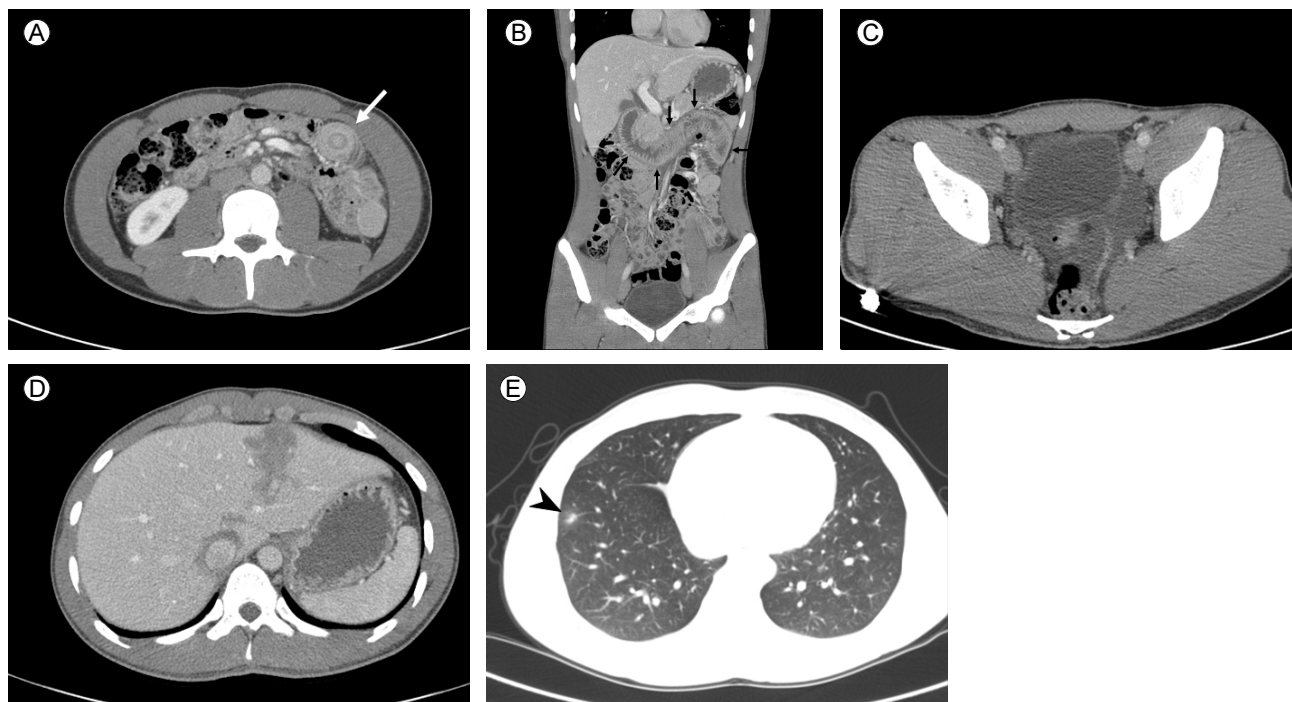


Figure 2. High-resolution abdominal and chest computed tomography. (A) The target sign is seen in the jejunum, indicating a jejunojejunal intussusception (white arrow). (B) Diffuse wall edema is noted in the distal duodenal and jejunum (black arrow). (C) A small volume of fluid collected in the pelvis. (D) Multiple irregular hypoattenuated lesions are noted in both liver lobes, suggesting multiple eosinophilic liver abscesses. (E) A few small nodules surrounded by ground-glass opacities are seen in both lungs, suggesting eosinophilic pneumonitis (arrowhead).

(Fig. 2C), and multiple irregular hypoattenuated lesions in both lobes of the liver (Fig. 2D), suggesting multiple eosinophilic liver abscesses. High-resolution chest CT showed a few small nodules surrounded by ground-glass opacities in both lungs, suggesting eosinophilic pneumonitis (Fig. 2E). Other investigations were performed to assess the presence of eosinophil infiltration in other organs. However, the echocardiography findings were unremarkable. Thereafter, gas passage was confirmed during 2 days of fasting, and we performed esophagogastroduodenoscopy and colonoscopy to investigate the esophagus, stomach, duodenal bulb, second portion of the terminal ileum, colon, and rectum. Although endoscopy showed normal mucosa, we performed multiple endoscopic biopsies. A histological examination revealed few mucosal and intraepithelial eosinophils ($< 20/\text{high-power field}$). These findings were not sufficient to diagnose eosinophilic enteritis.

Although we were unable to confirm long-standing eosinophilia, the patient was finally diagnosed with HES based on the presence of idiopathic hypereosinophilia and multiple organ

involvement. Intravenous corticosteroid therapy (prednisolone, 60 mg/day) was initiated for the HES with an intestinal obstruction due to intussusception. A marked reduction in abdominal pain was noted after 2 days of treatment. A follow-up abdominal CT after 7 days of intravenous corticosteroid therapy revealed a significant reduction in distal duodenal and jejunal wall edema, fluid collection in the pelvis, multiple ground glass opacities in the lungs, and liver abscesses as well as disappearance of the jejunojejunal intussusception (Fig. 3). The eosinophil count was $434/\text{mm}^3$, which was within the normal range. The patient was discharged, and the steroid dose was tapered slowly.

DISCUSSION

Intussusception in adults is a rare condition, accounting for 1-5% cases of bowel obstruction. It is usually primary and benign in children. However, almost 90% of intussusception cases in adults are secondary to a pathological condition [4]. Approx-

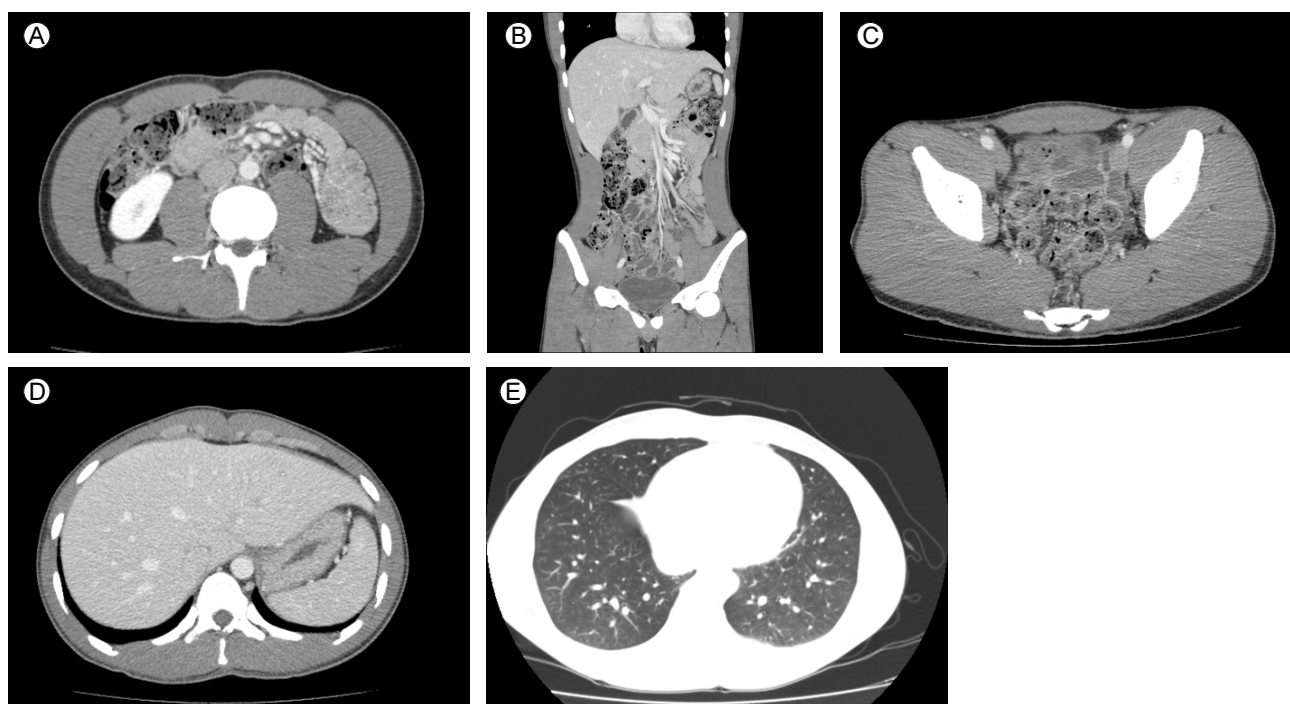


Figure 3. Follow-up computed tomography after 7 days of systemic steroid therapy. (A) Previously noted jejunojejunal intussusception is absent. (B) Distal duodenal and jejunal wall edema improved. (C) No fluid collection in the pelvis. (D) The eosinophilic liver abscesses are reduced markedly. (E) Complete resolution of multiple nodules surrounded by ground-glass opacities in both lungs.

imately 70-90% of intussusception cases in adults require treatment. Surgical resection is the most prevalent treatment [5]. Eosinophilic enteritis has been described as a rare cause of intussusception in two case reports [6,7]; however, HES presenting as intussusception has not been reported.

Eosinophilic disease of the gastrointestinal tract is poorly understood, and there are many probable differential diagnoses. The possibility of HES should be considered because it can involve multiple organs and has a progressive and potentially fatal course [8]. HES is defined by the following: (1) persistently elevated eosinophil count ($> 1,500/\text{mm}^3$) for at least 6 months; (2) eosinophilia with no recognizable cause such as parasitic infestation, cancer, or allergic condition; and (3) symptoms of eosinophilia-mediated organ dysfunction. However, a shorter period of hypereosinophilia with symptoms requiring eosinophil-lowering therapy is also sufficient for diagnosis [9]. In the present case, early initiation of systemic corticosteroid therapy resulted in a marked response in a patient with HES presenting as intussusception, and thus obviated the need for surgery.

The clinical manifestations of eosinophilic disease of the gastrointestinal tract vary according to the site and depth of eosinophilic infiltration on the bowel wall. Klein et al. [10] classified this condition into three forms based on the bowel wall layer that is predominantly involved. Symptoms such as diarrhea, hematochezia, and cramping abdominal pain usually occur in the mucosal type. In the muscular type, thickening of the muscular layer narrows the intestinal lumen and can lead to intestinal obstruction or perforation. Ascites may develop if the subserosal layer is involved. In the present case, endoscopic findings and random biopsies did not suggest eosinophilic infiltration. However, we suspected outer layer (muscular or subserosal) involvement due to the presence of intussusception and fluid collection in the pelvic cavity, as noted on abdominal CT.

The present case is the first known case of HES presenting as intussusception in a 19-year-old male. The patient was successfully treated with steroids, and surgery was not necessary.

중심 단어: 과호산구 증후군; 장중첩증

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