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Case Report

A Case of Severe Duodenitis in an Adult Patient with Hyper IgESyndrome

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1. Abstract

We present the first case of severe duodenitis in the adult Japanese patient with hyper IgE syndrome (HIES). A 41 years male patient, who has a treatment in the dermatology of our hospital for severe dermatitis with hyper IgE syndrome, had a right frank pain with diarrhea for one week. Because he vomited with nausea two days ago, we were asked for his gastrointestinal examination. A gastroduodenal endoscopy showed severe erosive duodenitis with multiple erosions, submucosal hemorrhage and longitudinal ulceration. Although we doubted a vasculitis complicated with HIES, the findings of a vasculitis was not seen by histological examination in biopsy samples taken from duodenum. However, the diagnosis using skin biopsy samples was leukocytoclastic vasculitis. The patient received intravenous methylprednisolone (MP), which provided a complete resolution of abdominal pain within 48 hours. Thereafter, as a dose of MP was tapered, the gastroduodenal endoscopy showed almost normal findings with scattered atrophic villi at 28 hospital days. We suspect that this case was severe duodenitis induced by vasculitis complicated with HIES by endoscopic findings and response for MP treatment.

2. Keywords: Duodenitis; Vasculitis; Hyper IgE syndrome

3. Introduction

Hyper IgE syndrome (HIES) is a multisystem disorder characterized by eczema, recurrent skin and pulmonary infections and marked lyincreased levels of serum IgE [1]. HIES is a very rare primary immunodeficiency disease and is known to induce the multiple organ injuries [2]. However, there are few report about the gastrointestinal disorders in HIES [3, 4]. Here, we present the first case of severe duodenitis in the adult Japanese patient with HIES.

4. Case report

A 41 years male patient, who has a treatment in the dermatology of our hospital for severe dermatitis with hyper IgE syndrome (IgE value = over 20,000mg/dL) during over 13 years (Figure 1), had a right frank pain with diarrhea for one week. Because he vomited with nausea two days ago, we were asked for his gastrointestinal examination. Laboratory analyses showed the following: C-reactive protein (CRP) 6.19 mg/dL, D-D dimer 9.08 μg/mL, IgE 22,325 mg/dL. However, serum immunoglobulin levels (IgG, A and M) were within a normal range, P-ANCA (myeroperoxidase- anti- neutrophil cytoplasmic antibody) and C-ANCA (proteinase3- anti-neutrophil cytoplasmic antibody) were negative, and compliment activities (CH50) were very high. Urinalysis showed macroscopic hematuria (over 100 erythrocytes/HPF) and protein urea (100 mg/dL). A gastroduodenal endoscopy showed severe erosive duodenitis with multi pleerosions, submucosal hemorrhage and longitudinal ulceration (Figure 2). Although we doubted a vasculitis complicated with HIES from these findings, the findings of a vasculitis was not seen by histological examination in biopsy samples taken from duodenum. However, the diagnosis using skin biopsy samples was leukocytoclastic vasculitis (LCV), but IgA deposition was

not found (Figure 3). The involvement of the other gastrointestinal tracts was not seen by colonoscopy and capsule endoscopy. From these findings, we diagnosed this duodenitis as an involvement of LCV secondary to chronic infections complicated with HIES. The patient received intravenous methylprednisolone (MP) with Piperacillin-Tazobactum (co-administered for one week), which provided a complete resolution of abdominal pain within 48 hours. Thereafter, as a dose of MP was tapered, normal finding of urinalysis was seen at the 7 hospital days. At the 28 hospital days, the values of D-D dimer and CRP became negative and the gastroduodenal endoscopy showed almost normal findings with scattered atrophic villi (Figure 4).

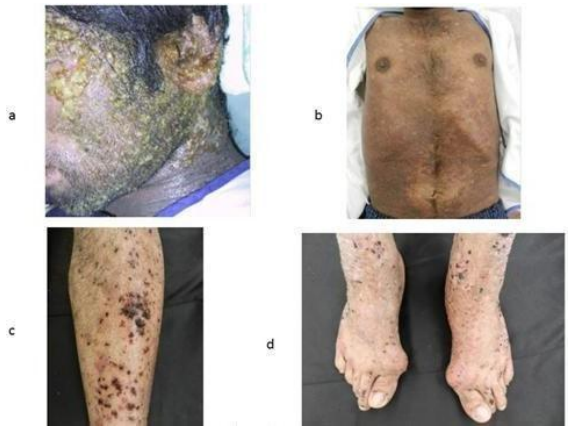

Figure 1: A patient with hyper IgE syndrome having a treatment in dermatology
a): Photograph when he had a medical examination in dermatology for an severe skin infection in 2003
b), c) and d): Photographs of body,dorsum of footand lower leg when he consulted gastroenterology in 2016.

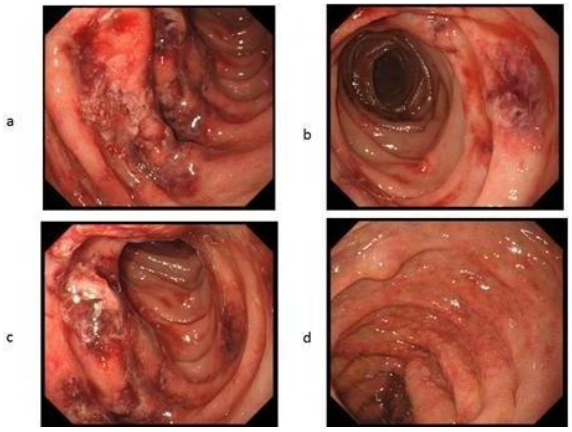

Figure 2: The first gastroduodenal endoscopic findings showed severe erosive duodenitis with multiple erosions, submucosal hemorrhage and longitudinal ulceration.

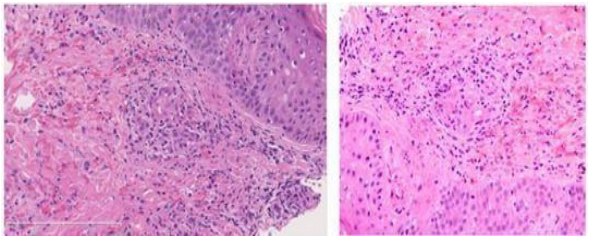

Figure 3: The pathological findings from skin biopsy specimen sowed leukocytoclastic vasculitis.

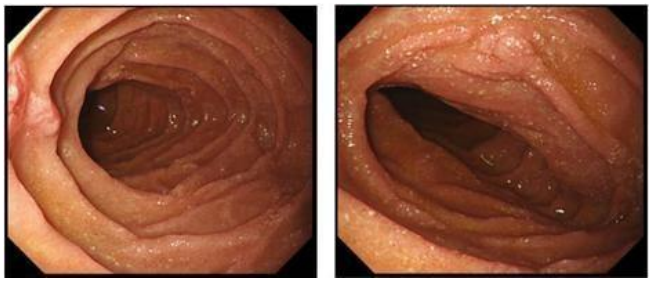

Figure 4: The gastroduodenal endoscopic findings after methylprednisolone treatment showed almost normal findings with scattered atrophic villi.

5. Discussion

Almost patients with HIES have recurrent various bacterial infections, including Staphylococcus aureus, and fungal infections [2]. Regarding as infection sites, the skin and lungs are predominated, but the ears, eyes, oral mucosa, sinuses, joints, blood, and even viscera were also involved. There were few reports about the complications of gastrointestinal tracts with HIES. Alberti-Flor et al. reported a case of Ileocecal histoplasmosis mimicking Crohn's disease in a patient with HIES [3]. Steiner SJ et al. also reported a similar case [4]. However, the other gastrointestinal tract injuries with HIES are not reported.

This is the first case report about severe duodenitis with HIES in the world. Although we suspected the duodenitis seen in this case was caused by vasculitis from endoscopic findings, histological findings of vasculitis were not seen in duodenal biopsy samples. However, LCV was histologically diagnosed using skin biopsy samples. In the laboratory analyses on the admission, serum IgA levels were within a normal range, P-ANCA and C-ANCA were negative, and CH50was not in low level. In addition, IgA deposition was not seen in biopsy specimen. Therefore, we diagnosed duodenitis as an involvement ofLCV, neither IgA vasculitis nor ANCA associatedvasculitis.

LCV may be secondary to medications, underlying infection, collagen-vascular disorders, or malignancy, although approximately half of cases are idiopathic [5]. LCV has been described primarily in the skin. In addition, many patients have associated systemic manifestations because of other organ involvement such as kidneys and gastrointestinal system [6].We suspected the duodenitis in our case is involved by LCV resulted from a recurrent Staphylococcus aureus infection in the skin. In fact, a few cases of vasculitis with HIES [7] and several cases of LCV complicated HIV infection are recently reported [8, 9]. Our case was characterized by a peculiarity such as the other gastrointestinal involvements were not seen except duodenum. This reason is unclear at present. In cases of the IgA vasculitis with gastrointestinal involvements, the most important finding is severe erosive duodenitis (in the second part of the duodenum), characteristic, but not pathognomonic for IgA vasculitis on the gastrointestinal endoscopy [10].The gastrointestinal involvement of LCV may be similar to that of IgAvasculitis.

Clinicians should keep an attention to vasculitis secondary to infections for the patients with immunodeficiency diseases, when they have an abdominal discomfort.

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