Primary Rectal Amyloidosis in a Patient with Human Immunodeficiency Virus

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Amyloidosis can involve a single organ or multiple organs. The gastrointestinal tract is a common site of amyloid deposition. We report a rare case of solitary rectal amyloidosis in a patient with human immunodeficiency virus who had experienced recurrent oral candidiasis for 3 months and intermittent diarrhea for 6 months. The lesion was confirmed histologically and there were no detectable amyloid lesions at other gastrointestinal sites or systemic involvement. (*Gut and Liver* 2009;3:57-59)

Key Words: Rectum; Amyloidosis; Human immunodeficiency virus

INTRODUCTION

Amyloidosis is not a single disease but a term used to describe diseases that share a common feature: the extracellular deposition of pathological insoluble fibrillar proteins in organs and tissues.¹ The gastrointestinal (GI) tract is a common site of amyloid deposition. However, organ-specific, solitary amyloid deposition is very rare in the GI tract. We could not find a report of a case of primary rectal amyloidosis associated with human immunodeficiency virus (HIV) in the medical literature. Here we report on a HIV-infected patient who was diagnosed with rectal amyloidosis.

CASE REPORT

A 40-year-old male presented to our hospital on June 22, 2007, with the chief complaint of recurrent oral candidiasis for 3 months and intermittent diarrhea for 6 months. On physical examination, he had oral candidiasis.

Otherwise the physical findings were not remarkable. In routine laboratory finding, the leukocyte count was 7,540/ μ L, the serum total protein 8.3 g/dL, albumin 4.1 g/dL, BUN 11.5 mg/dL and creatinine 0.8 mg/dL. AST was 36 IU/L, ALT 37 IU/L, and serum alkaline phosphatase was 278 IU/L, LDH 424 IU/L, total bilirubin 0.67 mg/dL. ESR was 37 mm/hr, C-reactive protein 5.5 mg/dL. Urinalysis showed normal finding (no proteinuria). The patient had a CD4 count of 80 cells/ μ L and a HIV RNA viral load of 174,000 copies/mL. A gastro-duodenal endoscopy revealed esophageal candidiasis and atrophic gastritis. A colonoscopy showed two 0.5 cm sized polyps in the sigmoid colon and rectum (Fig. 1). There were no abnormalities in the other parts of the colon. The sigmoid colon polyp was a tubular adenoma

Fig. 1. Colonoscopy finding. Colonoscopic view of the rectum showing a polyp covered by erythematous mucosa.

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Fig. 2. Photomicrographs of the rectal polyp. (A) An eosinophilic and amorphous deposit was mainly evident in the submucosal layer (H&E stain, $\times 100$). (B) Amorphous material that stained positive with Congo red under polarized light was deposited at the submucosa (Congo-red stain, $\times 200$).

and the rectal polyp was diagnosed as amyloidosis. Serum protein electrophoresis revealed an elevated α_1 fraction (9.2%) and γ fraction (32.2%). Serum immunofixation showed a markedly elevated λ light chain band (3,950 mg/dL). After 2 weeks, random endoscopic biopsies were performed on the tongue, esophagus, stomach, duodenum, terminal ileum, colon, and rectum. The microscopic examination of the rectal biopsy showed focal deposition of amorphous material with congo-red staining positive under polarized light (Fig. 2). However, amyloid deposition was not identified in the random biopsy specimens from the tongue, esophagus, stomach, duodenum, terminal ileum, and colon. The liver, heart and kidneys were ruled out for multiorgan involvement. Radiographs of the chest and abdomen revealed no abnormalities. The abdominal CT scan revealed no abnormalities except for several renal cysts and a GB stone. The Doppler echocardiography showed a normal ventricle wall thickness and ejection fraction.

DISCUSSION

A case of rectal amyloidosis in an HIV-infected patient was presented. There are four main types of amyloidosis, categorized by their underlying etiology. Primary amyloidosis (AL) is a multisystem disease of unknown etiology characterized by deposition of amyloid fibrils composed of variable portions of the monoclonal light chain. The second subtype, AA amyloidosis, occurs most frequently as a complication of chronic infection or chronic inflammatory disease. The third type is hemodialysis-related and the final type is familial amyloidotic polyneuropathy.^{1,2} Amyloi-

dosis can involve single organ or multiple organs; the involvement may take the form of a focal, tumor like lesion or an infiltrative process. The GI tract is a common site of amyloid deposition, occurring in 70% of cases.^{2,4} Biopsies of affected organs are often performed to confirm the diagnosis. Radiological and angiographic diagnostic studies are generally insensitive and nonspecific for the diagnosis.¹ Endoscopic findings such as a fine granular pattern and polypoid protrusions are common and may reflect amyloid deposition in the mucosa or submucosa of the GI tract.⁵ The characteristic histological finding is an amorphous material with apple green birefringence under polarized light after Congo red staining.^{1,2} The deposition of AL type amyloid protein in the GI tract was marked in the muscular layer of the mucosa, and in the blood vessel walls of the submucosa and muscularis propria, whereas for the AA type the lamina propria is primarily involved.^{2,3} The most common GI manifestations include bleeding, malabsorption, obstruction or pseudo-obstruction, poor motility, infarction, ischemia, nodules or ulceration.^{1,5,6} Our patient had intermittent diarrhea. AA amyloidosis has been reported in intravenous or subcutaneous ("skin-popping") drug abusers, some of whom have been HIV-infected. Among drug abusers, it is postulated that prolonged inflammation of the skin is important in the pathogenesis of amyloidosis.⁷⁻⁹ Recently, case reports of HIV-associated amyloidosis involving the skin, liver, kidney, and lung have been increasing but solitary rectal amyloidosis in HIV-infected patients has not been previously reported. Our patient had no history of viral hepatitis, tuberculosis or hemodialysis. The pattern of amyloid deposition, the absence of

amyloid in the lamina propria, no history of infection, serum protein electrophoresis and serum immunofixation supported the diagnosis of primary amyloidosis rather than the AA type. This is the first case report of primary rectal amyloidosis in an HIV-infected patient.

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