Primary Light-chain Amyloidosis Featuring Worm-like Small Bowel Polyposis

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A 66-year-old man with a history of asthma presented with abdominal distension of one month in duration. Computed tomography revealed jejunal wall thickening and distended loops of small bowel (Picture 1). Double-balloon enteroscopy revealed numerous polypoid, worm-like, 1-5 cm masses, while the mucosa in the duodenum and ileum had a normal appearance (Picture 2, 3). One of the masses was removed with endoscopic mucosal resection. A histological

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examination revealed the presence of Congo red-positive material in the submucosal layer (Picture 4). An immunohistochemical examination revealed that the material was positive for amyloid light-chain lambda protein. The endoscopic findings of the esophagus, stomach and colon were normal. The patient had no chronic disorders that might predispose him to secondary amyloidosis. Thus, primary amyloid light-chain amyloidosis of the small bowel was diagnosed. The polyps were considered to be amyloidomas. The patient’s symptoms resolved with conservative therapy, and he has not required surgery during the two-year follow-up period. Thickening of the valvular conniventes and polypoid protrusions are endoscopic features of AL amyloidosis of the small intestine (1). This is the first report of light-chain amyloidosis featuring small bowel polyposis causing intestinal obstruction.

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Reference


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