Case Report

Cukurova Anestezi ve Cerrahi Bilimler Dergisi

AA Amyloidosis with Gastrointestinal, Mesenteric, and Omental Involvement in a Patient with Crohn's Disease and Familial Mediterranean Fever: A Rare Case Report

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Abstract

Purpose: Amyloidosis comprises a heterogeneous group of disorders characterized by extracellular deposition of misfolded proteins, which may result in progressive organ dysfunction. Gastrointestinal involvement is uncommon and often presents with non-specific symptoms, making diagnosis challenging. Involvement of the mesentery and omentum is particularly rare. **Case Presentation:** A 29-year-old male with a known history of Crohn's disease and Familial Mediterranean Fever (FMF) presented with nausea, vomiting, and inability to pass gas or stool. Radiological evaluation revealed intestinal obstruction accompanied by nodular lesions in the mesentery and omentum. Due to persistent clinical findings, surgical intervention was performed, including adhesiolysis and omental biopsy. Histopathological examination confirmed amyloid deposition. The patient had an uneventful postoperative course and was discharged on the fifth day. **Conclusion:** This case highlights a rare presentation of AA amyloidosis involving the gastrointestinal tract, mesentery, and omentum in the setting of chronic inflammatory disease. Amyloidosis should be considered in the differential diagnosis of patients with unexplained gastrointestinal symptoms, particularly in those with underlying inflammatory conditions. Early diagnosis and histopathological confirmation are crucial for appropriate management and improved prognosis.

Keywords: Amyloidosis; gastrointestinal involvement; omentum; Crohn's disease; Familial Mediterranean Fever

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1. Introduction

Amyloidosis represents a heterogeneous group of disorders characterized by the extracellular deposition of misfolded protein fibrils, which can lead to progressive organ dysfunction over time and may affect multiple organ systems[1]. The clinical presentation varies widely depending on the extent of disease and the organs involved, often resulting in delayed diagnosis and recognition at advanced stages[1,2]. Although gastrointestinal involvement is relatively uncommon, it may be easily overlooked in clinical practice due to its non-specific manifestations. Furthermore, involvement of the mesentery and omentum

is exceedingly rare and has been reported only in a limited number of cases in the literature[3,4]. Depending on the underlying pathophysiological mechanism, amyloidosis may present as a systemic disease or, less frequently, as a localized process confined to a specific organ or tissue[2]. In this study, we aimed to present a rare case of AA amyloidosis involving the gastrointestinal tract, mesentery, and omentum in a patient with a history of chronic inflammatory disease.

2. Case Presentation

A 29-year-old male with a long-standing history of Crohn's disease and Familial Mediterranean Fever (FMF) presented to the emergency department with complaints of nausea, recurrent episodes of vomiting, and inability to pass gas or stool for the past 3–4 days. His medical history was notable for prior cholecystectomy and appendectomy. Approximately six months earlier, a gastric biopsy obtained during endoscopy had been reported as histopathologically consistent with amyloidosis.

On physical examination, marked abdominal distension and diffuse tenderness were observed, along with decreased bowel sounds. An upright abdominal radiograph demonstrated multiple air–fluid levels, raising suspicion for intestinal obstruction, and the patient was subsequently admitted to the general surgery department for further evaluation and management. Nasogastric decompression was initiated, oral intake was discontinued, and conservative treatment was commenced. Contrast-enhanced computed tomography revealed adhesions likely related to previous surgical interventions, as well as multiple nodular lesions in the mesentery and omentum, raising concern for an infiltrative process or lymphadenopathy[4,5]. Due to the persistence of clinical findings and radiological suspicion, surgical intervention was undertaken. During the operation, adhesiolysis was performed to relieve bowel obstruction, and an omental biopsy was obtained for diagnostic purposes (Figure 1). The patient's postoperative course was uneventful, with gradual clinical improvement, resumption of bowel function, and tolerance of oral intake. He was discharged in good condition on postoperative day 5. Histopathological examination of the omental tissue confirmed amyloid deposition, establishing the diagnosis (Figure 2, Figure 3).

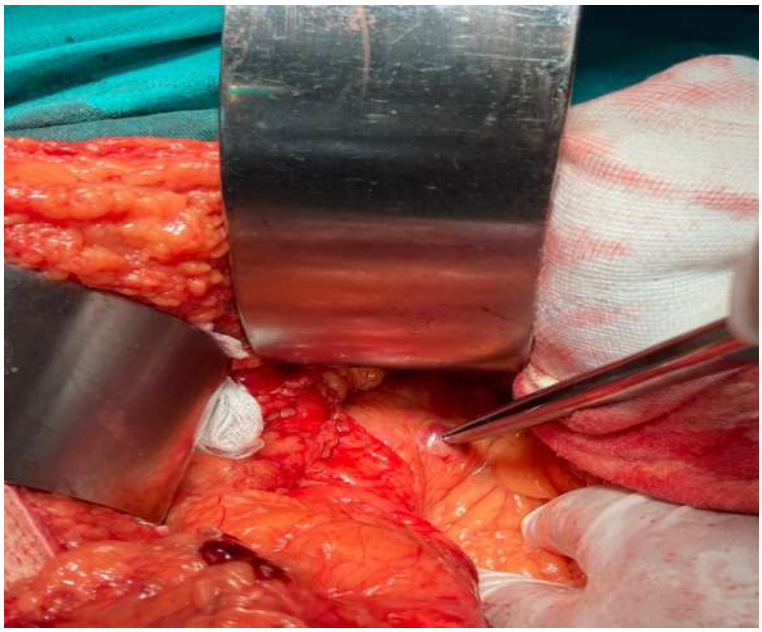

Figure 1. Intraoperative image showing nodular involvement of the omentum and mesentery.

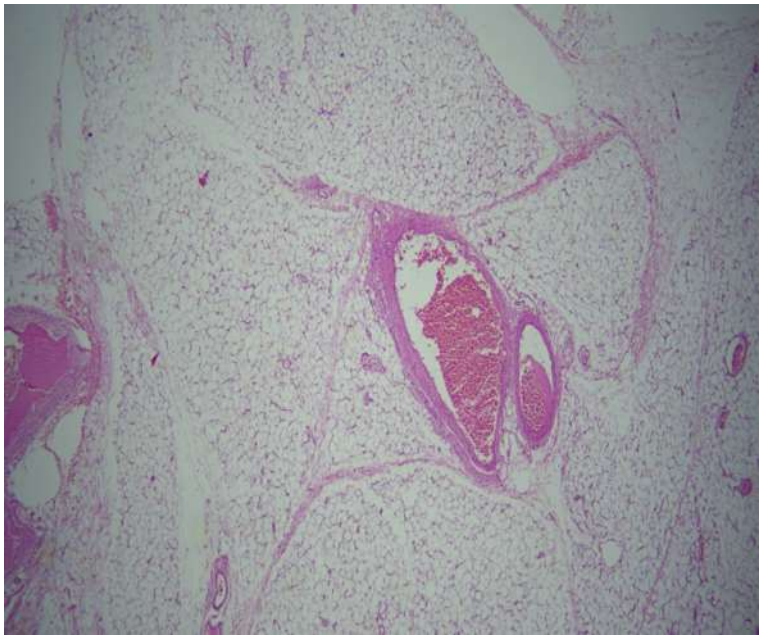

Figure 2. Histopathological examination showing amorphous eosinophilic deposits in omental tissue (Hematoxylin-Eosin staining).

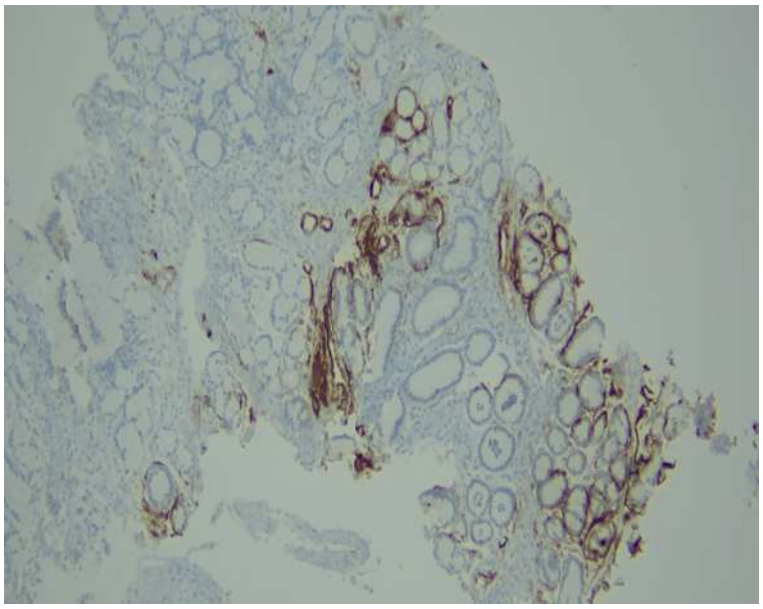

Figure 3. Congo red staining demonstrating amyloid deposition with characteristic apple-green birefringence under polarized light.

3. Discussion

Amyloidosis represents a complex and heterogeneous group of disorders characterized by the abnormal misfolding of various precursor proteins into insoluble fibrillar structures, which subsequently accumulate within the extracellular matrix of tissues, ultimately leading to progressive organ dysfunction and involvement of multiple organ systems[1,2]. The gold standard for diagnosis remains Congo red staining, through which amyloid deposits exhibit the characteristic apple-green birefringence under polarized light, providing a definitive histopathological confirmation[6]. Among the various subtypes, AL and AA amyloidosis are the most frequently encountered in clinical practice, with AA amyloidosis being particularly associated with chronic inflammatory conditions such as Crohn's disease and Familial Mediterranean Fever (FMF); this association is largely

attributed to persistently elevated levels of serum amyloid A protein secondary to ongoing inflammation[7,8]. Although gastrointestinal involvement is relatively uncommon, it poses significant diagnostic challenges due to its tendency to present with non-specific symptoms, including abdominal pain, diarrhea, malabsorption, and weight loss, all of which may overlap with a wide range of other gastrointestinal disorders[9,10]. Consequently, this nonspecific clinical presentation often leads to delayed diagnosis or incidental detection during evaluation for other conditions. Indeed, gastrointestinal amyloidosis remains a rare entity in clinical practice, and its subtle and variable symptomatology frequently contributes to under-recognition or misdiagnosis[11]. Furthermore, systemic amyloidosis is characterized by a highly variable clinical course owing to its ability to affect multiple organs to differing extents, thereby necessitating a multidisciplinary approach for accurate diagnosis, comprehensive evaluation, and optimal management[12]. Involvement of the mesentery and omentum is exceedingly rare and has been reported only in a limited number of cases in the literature. Importantly, such involvement may radiologically mimic malignant or lymphoproliferative processes, thereby complicating the differential diagnosis and often prompting further invasive diagnostic procedures[4,5]. For this reason, definitive diagnosis in such cases typically relies on histopathological examination. In the present case, it is considered that the coexistence of Crohn's disease and FMF contributed to the development of AA amyloidosis through chronic inflammatory mechanisms, with disease progression leading to simultaneous involvement of the gastrointestinal tract, mesentery, and omentum. Moreover, the intraoperatively observed omental nodularity supports the macroscopic infiltrative nature of amyloid deposition, further corroborating the pathological findings.

4. Conclusion

This case illustrates a rare presentation of AA amyloidosis involving the gastrointestinal tract, mesentery, and omentum, and highlights the variable and often insidious clinical course of the disease. Amyloidosis should be considered in the differential diagnosis of patients presenting with unexplained gastrointestinal symptoms, particularly in those with a history of chronic inflammatory conditions. Early recognition, together with appropriate histopathological evaluation, is essential for improving patient outcomes and ensuring optimal management of the disease process.

genAI: No artificial intelligence-based tools or generative AI technologies were used in this study. The entire content of the manuscript was originally prepared, reviewed, and approved by all authors.

Statement of Ethics: This study was conducted in accordance with the principles of the Declaration of Helsinki. Written informed consent was obtained from the patient for the publication of this case report and the accompanying images. In accordance with institutional policies, ethics committee approval was not required.

Data Availability: The datasets used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

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