

Surgical Treatment of Invasive IgG4-related Sclerosing Esophagitis Inflammatory Pseudotumor: A Case Report and Literature Review

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Abstract

IgG4-related sclerosing disease (IgG4-RSD) is an uncommon autoimmune entity, manifesting as tumefactive lesions and fibrosis in a spectrum of organs, including the pancreas, biliary tract, and salivary glands. Involvement of the esophagus is exceedingly rare. We present herein a case of an invasive IgG4-related sclerosing esophagitis pseudotumor, confirmed through surgical intervention and pathological assessment. A 26-year-old female exhibited symptoms of dysphagia and significant weight loss over a six-month period. Esophagoscopy disclosed a mass-like lesion localized to the upper esophagus. Preoperative biopsies, performed on two occasions, identified squamous epithelial cells, pointing towards a mesenchymal tumor in the differential diagnosis. Following a comprehensive multidisciplinary consultation, surgical resection was elected owing to the lesion's extensive nature and the patient's pronounced symptoms. Histopathological examination substantiated the presence of an esophageal tumor with the hallmarks of invasive IgG4-related sclerosing esophagitis. Postoperative recovery was uneventful, and the patient has remained asymptomatic throughout the follow-up period. This case illustrates the necessity of including IgG4-related disease in the differential diagnosis of esophageal tumors and accentuates the significance of surgical management in symptomatic and invasive instances of IgG4-related sclerosing esophagitis pseudotumors.

Keywords

IgG4-related disease; sclerosing esophagitis pseudotumor; surgical treatment; case report; literature review

1. Introduction

IgG4-related disease (IgG4-RD) is a fibroinflammatory condition characterized by tumefactive lesions, lymphoplasmacytic infiltration rich in IgG4-positive plasma cells, and tissue fibrosis in affected organs [1]. Initially identified in autoimmune pancreatitis, IgG4-RD can affect virtually any organ system, including the biliary tree, salivary glands, retroperitoneum, and lungs [2]. Esophageal involvement in IgG4-RD is exceedingly rare, with only a few cases reported in the literature [3]. Here, we present a case of invasive IgG4-related sclerosing esophagitis pseudotumor in a young female, treated and diagnosed surgically, along with a review of related literature.

2. Case Report

A 26-year-old female presented with a six-month history of progressive dysphagia and a significant weight loss of

10 kg. She reported no prior episodes of gastroesophageal reflux disease, autoimmune conditions, or recent infections. Upon physical examination, no notable abnormalities were detected. Comprehensive laboratory investigations, inclusive of serum IgG4 levels, were unremarkable, remaining within the normal parameters. An upper gastrointestinal endoscopic evaluation exposed a circumferential, elevated mass within the upper esophagus, characterized by a smooth mucosal surface and causing substantial eccentric luminal narrowing, which impeded the advancement of an ultra-thin endoscope (Figure 1). The lesion underwent two separate biopsy procedures, the findings of which were indicative of squamous epithelial cells, pointing towards a mesenchymal tumor in the differential diagnosis. No malignant characteristics were identified. Chest and abdominal computed tomography (CT) scans delineated extra-luminal extension and pronounced involvement of adjacent structures (Figure 2). Considering the symptomatic and invasive attributes of the esophageal tumor, coupled with the inadequacy of conservative management, a multidisciplinary panel comprising thoracic surgeons, radiologists, and pathologists engaged in a thorough discussion regarding the therapeutic strategy. Ultimately, due to the tumor's invasive nature and the lack of response to non-surgical interventions, a surgical resection was deemed the most appropriate course of action.

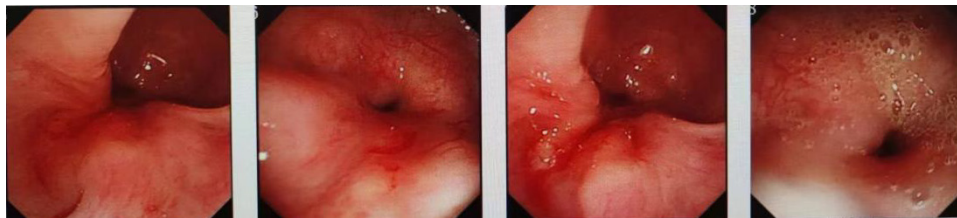


Figure 1. Endoscopic findings of esophageal lesion.

Upper gastrointestinal endoscopy revealed a circumferential, elevated mass within the upper esophagus, characterized by a smooth mucosal surface and causing substantial eccentric luminal narrowing, which impeded the advancement of an ultra-thin endoscope.



Figure 2. Computed tomography (CT) imaging of the esophageal lesion.

Chest and abdominal CT scans delineated extra-luminal extension of the esophageal mass and pronounced involvement of adjacent structures. The images demonstrate the circumferential thickening of the esophageal wall and the extent of tumor invasion.

The patient underwent thoracoscopic-assisted McKeown esophagectomy. Intraoperatively, the lesion was found to involve the muscularis propria but had not extended to adjacent structures. Gross examination of the resected specimen revealed a 4 cm diameter, firm, white circumferential lesion (Figure 3).



Figure 3. Gross pathological examination of the resected esophageal specimen.

Intraoperative and gross pathological images of the resected esophageal specimen following thoracoscopic-assisted McKeown esophagectomy. The lesion measured approximately 4 cm in diameter, presenting as a firm, white, circumferential mass involving the muscularis propria without extension to adjacent structures.

Histopathological examination showed chronic inflammation of the entire esophageal wall with fibrosis, dense lymphoplasmacytic infiltration, focal osseous and bone marrow metaplasia, and partial involvement of the circumferential resection margin (Figure 4). Both resection margins were unremarkable. Immunohistochemistry revealed IgG4-positive plasma cells with an IgG4/IgG ratio of approximately 5%. The diagnosis was confirmed as invasive IgG4-related sclerosing esophagitis pseudotumor.

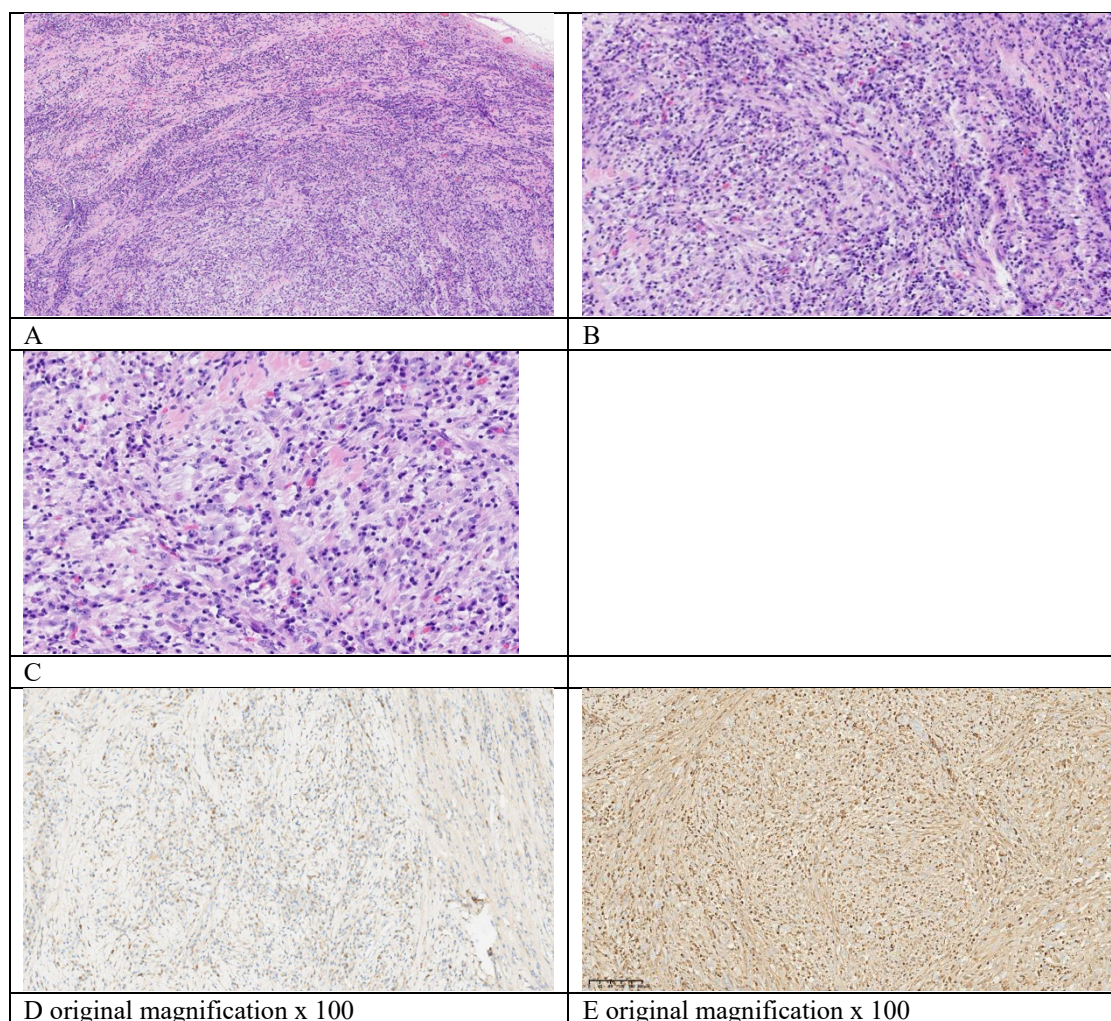


Figure 4. Histopathological examination of the resected esophageal lesion.

(A) Low-power view (original magnification $\times 4$) showing chronic inflammation of the entire esophageal wall with fibrosis and dense lymphoplasmacytic infiltration. (B) Medium-power view (original magnification $\times 10$) demonstrating dense lymphoplasmacytic infiltration and focal osseous and bone marrow metaplasia. (C) Higher magnification (original magnification $\times 20$) highlighting the inflammatory infiltrate and fibrotic changes. (D) IgG4 immunohistochemical staining (original magnification $\times 100$) revealing IgG4-positive plasma cells. (E) Additional IgG4 immunostaining (original magnification $\times 100$) showing the IgG4/IgG ratio of approximately 5%, confirming the diagnosis of invasive IgG4-related sclerosing esophagitis pseudotumor.

Postoperatively, the patient had an uneventful recovery and was discharged on the seventh postoperative day. During follow-up, the patient remained asymptomatic, with resolution of dysphagia and weight recovery. Serial imaging studies showed no evidence of disease recurrence.

3. Discussion

IgG4-related disease (IgG4-RD) is a systemic fibroinflammatory condition [3]. In 2003, Kamisawa et al. described the close relationship between autoimmune pancreatitis and various inflammatory disorders, thereby characterizing IgG4-RD as a systemic disease [4]. Stone and colleagues subsequently published a comprehensive review on IgG4-RD in 2012 [1]. IgG4-RD presents with a wide range of clinical manifestations, potentially affecting nearly any organ system. Common presentations include severe swelling of the salivary and lacrimal glands, orbital disease,

autoimmune pancreatitis, retroperitoneal fibrosis, and tubulointerstitial nephritis. Elevated serum IgG4 concentrations are diagnostically significant but not mandatory, as approximately 30-50% of patients exhibit normal serum IgG4 levels [5, 6]. Currently, IgG4-RD is diagnosed histopathologically, with key pathological features including dense lymphoplasmacytic infiltrate, storiform fibrosis, and obliterative phlebitis [7].

IgG4-related esophageal disease is rare, with most literature comprising case reports. Obiorah et al. evaluated chronic esophagitis specimens over six years and found evidence of IgG4-RD in eight patients, all of whom exhibited gastroesophageal reflux symptoms. Three of these patients had esophageal strictures. Glucocorticoids are the mainstay of treatment but are prone to relapse [8]. Immunosuppressants may be used for persistent disease activity [9].

Diagnosing IgG4-related esophageal disease is challenging, with literature reporting diagnostic timelines ranging from three months to 20 years, with an average exceeding six years [10]. Dysphagia is a common early symptom. Early diagnosis and treatment with glucocorticoids can promptly alleviate symptoms. However, due to the lack of reliable diagnostic markers, some patients may progress to IgG4-related sclerosing esophagitis pseudotumor, as in the case we reported. At this advanced stage, surgical resection of the affected esophagus becomes the only effective treatment [11].

IgG4-related sclerosing esophagitis pseudotumor is a rare entity that should be considered in the differential diagnosis of esophageal tumors, particularly when characteristic histopathological features and elevated IgG4 levels are present. While corticosteroids are the primary treatment for IgG4-RD, surgical resection may be necessary for symptomatic or locally invasive lesions. The role of surgery in treating IgG4-related esophageal disease requires further elucidation through larger studies and long-term follow-up data.

4. Conclusion

This case of rare invasive IgG4-related sclerosing esophagitis pseudotumor was successfully treated with surgery. This case emphasizes the importance of considering IgG4-related disease in the differential diagnosis of esophageal tumors and the potential role of surgery in treating symptomatic and invasive lesions. Further research is needed to better understand the optimal treatment strategies for IgG4-related esophageal disease.

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Conflicts of Interest

The authors declare no conflicts of interest.

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