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【 CASE REPORT 】

IgG4-related Diaphragmatic Inflammatory Pseudotumor

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Abstract:
A 71-year-old man underwent surgery for a pancreatic neuroendocrine tumor. Follow-up imaging showed swelling of the remnant pancreas, and he was histologically diagnosed with autoimmune pancreatitis based on endoscopic ultrasonography-guided fine-needle aspiration specimens. After two years, a tumor appeared on the liver surface. Although we planned to perform laparoscopic partial hepatectomy, the intraoperative findings showed that the tumor was located in the diaphragm. Partial resection of the diaphragm was performed, and the final diagnosis was an immunoglobulin G4-related inflammatory pseudotumor in the diaphragm. To our knowledge, this is the first reported case of an immunoglobulin G4-related diaphragmatic inflammatory pseudotumor.

Key words: IgG4-related disease, IgG4, autoimmune pancreatitis, pancreatic neuroendocrine tumor, inflammatory pseudotumor

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Introduction

Autoimmune pancreatitis (AIP) is a rare type of pancreatitis with autoimmune mechanisms that was first described by Yoshida et al. in 1995 (1). AIP is currently considered to be a pancreatic manifestation of immunoglobulin (Ig)G4-related diseases (IgG4-RDs) (2). IgG4-RDs are characterized by the histological infiltration of lymphocytes and IgG4-positive plasma cells, storiform fibrosis, and obliterative phlebitis with elevated serum IgG4 levels (3). IgG4-RDs commonly involve the pancreas, bile ducts, salivary glands, lacrimal glands, kidneys, retroperitoneum, and lungs (4).

Inflammatory pseudotumors (IPTs), which occur at various sites in the body, are tumor-like mass lesions associated with both acute and chronic inflammation. Histologically, IPTs show variable amounts of fibrosis with polymorphous inflammatory infiltrates, including lymphocytes, plasma cells, and myofibroblastic spindle cells (5). Recently, some IPTs have been considered manifestations of IgG4-RDs and categorized as IgG4-related IPTs (6).

We herein report a case of an IgG4-related IPT that developed in the diaphragm during follow-up of AIP.

Case Report

A 71-year-old man underwent subtotal stomach-preserving pancreaticoduodenectomy for a pancreatic neuroendocrine tumor (PanNET) in the pancreatic head in 2012. The tumor grade was classified as G2 according to the World Health Organization classification 2010 (7). In 2015, follow-up computed tomography (CT) revealed localized swelling in the remnant pancreatic tail with a capsule-like rim (Fig. 1A). The patient had no symptoms, and a physical examination found no abnormalities. Laboratory data revealed an increased serum IgG4 level (353 mg/dL; normal range, 5-117 mg/dL). The levels of pancreatic enzymes and tumor markers, including carcinoembryonic antigen and carbohydrate antigen 19-9, were not elevated. ^{18}F-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography (PET)/CT showed an abnormal accumulation in the swollen remnant pancreatic tail [maximum standardized uptake value

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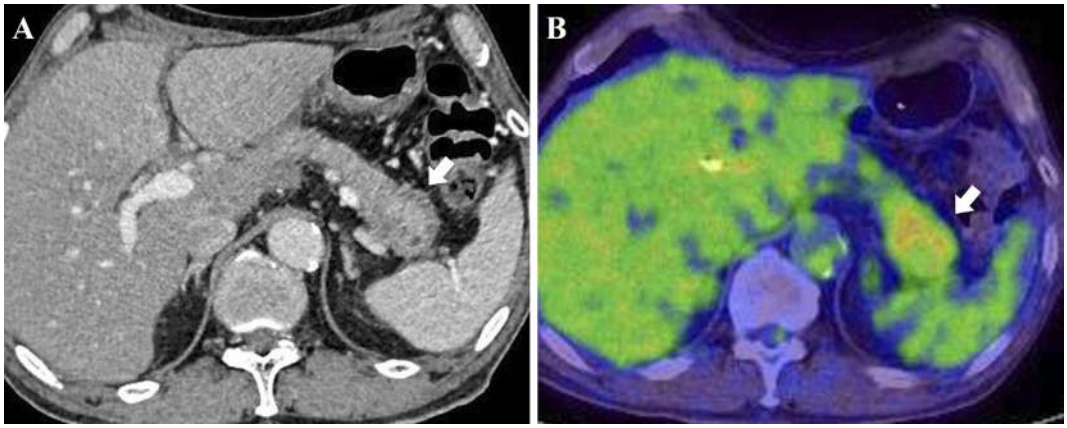

Figure 1. **CT and ^{18}F-FDG PET/CT findings of the remnant pancreatic tail in 2015. (A) CT showed swelling in the remnant pancreatic tail with a capsule-like rim (white arrow). (B) ^{18}F-FDG PET/CT showed an abnormal accumulation in the swollen remnant pancreatic tail (SUV_{max} 3.9) (white arrow). ^{18}F-FDG: ^{18}F-fluorodeoxyglucose, CT: computed tomography, PET: positron emission tomography, SUV_{max}: maximum standardized uptake value**

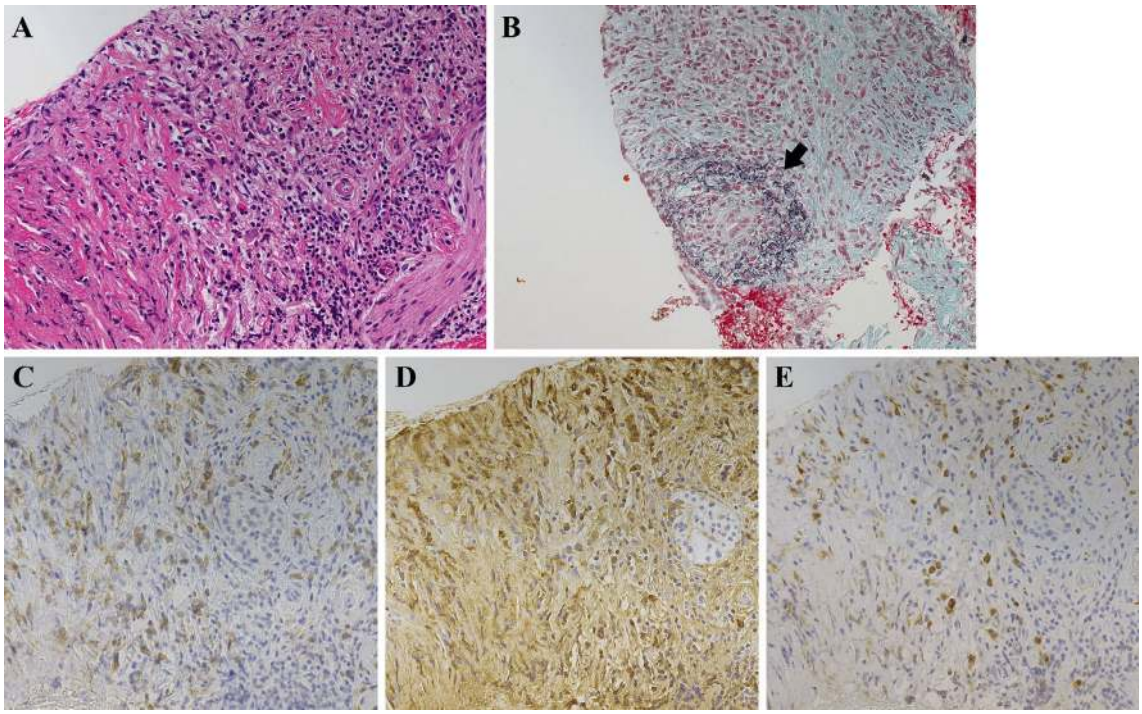

Figure 2. **Histological findings of the remnant pancreatic tail based on EUS-FNA specimens. (A) The histological findings revealed marked lymphoplasmacytic infiltration and storiform fibrosis (Hematoxylin and Eosin staining). (B) Obliterative phlebitis was observed (arrow) (Elastica–Masson's staining). An immunohistochemistry assessment demonstrated (C) CD38-, (D) IgG-, and (E) IgG4-positive plasma cells. More than 10 IgG4-positive plasma cells per HPF were observed. All figures are shown at a magnification of ×200. CD38: cluster of differentiation 38, EUS-FNA: endoscopic ultrasonography-guided fine-needle aspiration, HPF: high-power field, Ig: immunoglobulin**

(SUV_{max}) 3.9] (Fig. 1B). Endoscopic retrograde cholangiopancreatography was not performed because of the surgically altered anatomy. We performed endoscopic ultrasonography-guided fine-needle aspiration (EUS-FNA) using a 19-gauge needle (Expect™; Boston Scientific Japan, Tokyo, Japan). A histological examination showed marked lymphoplasmacytic infiltration, storiform fibrosis (Fig. 2A), and obliterative phlebitis (Fig. 2B). Immunohistochemistry showed the infiltration of cluster of differentiation 38 (CD 38)- and IgG-positive plasma cells (Fig. 2C, D) and more than 10 IgG4-positive plasma cells per high-power field (HPF) (Fig. 2E). These findings met the level 1 histological criteria for type 1 AIP according to the International Consensus Diagnostic Criteria for AIP (8). The patient was diag-

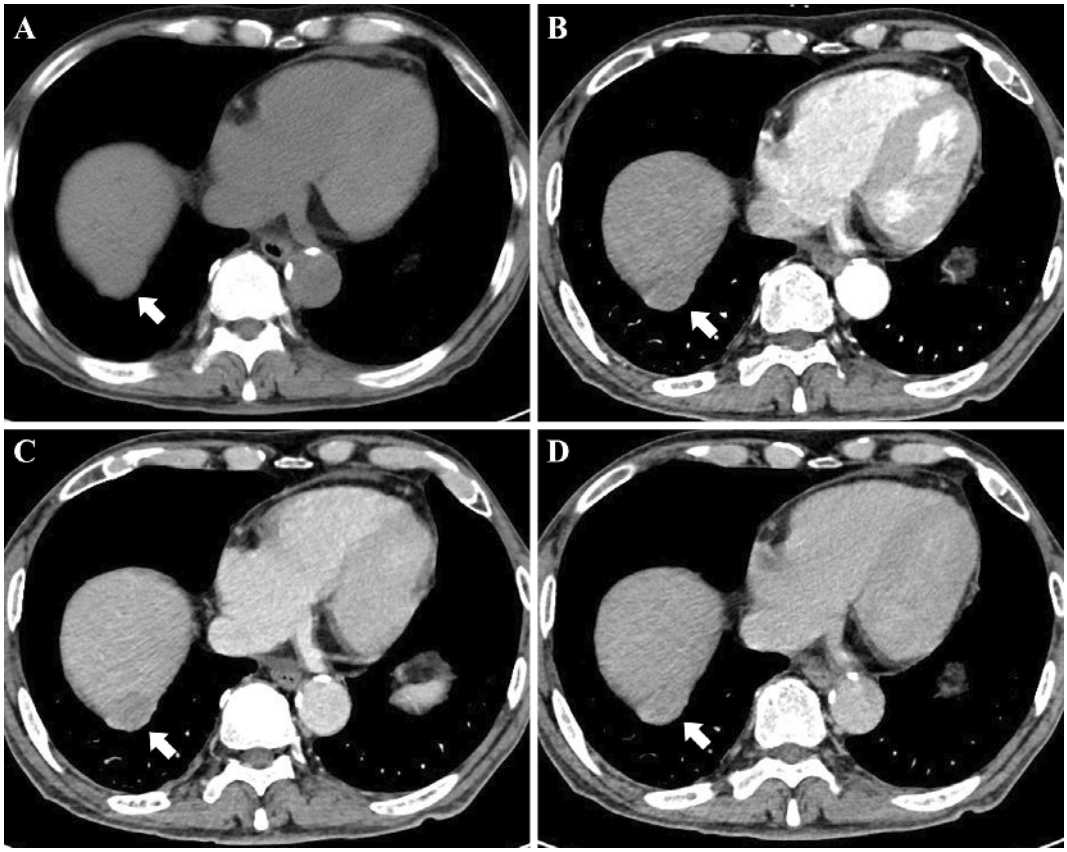

Figure 3. Dynamic CT findings of the tumor in 2017. The tumor was slightly enhanced in the early phase and showed prolonged enhancement in the portal and equilibrium phases. (A) Plain. (B) Early phase. (C) Portal phase. (D) Equilibrium phase. CT: computed tomography

nosed with definitive type 1 AIP and followed up without treatment because he did not exhibit any symptoms.

In 2017, follow-up magnetic resonance imaging revealed a 25-mm tumor on the surface of segment 8 in the liver. The tumor was slightly enhanced in the early phase and showed prolonged enhancement in the equilibrium phase on dynamic contrast-enhanced CT (Fig. 3). The serum IgG4 level remained elevated (409 mg/dL). PET/CT revealed a high ^{18}F-FDG uptake in the hepatic tumor lesion (SUV_{max} 3.4), right submandibular gland (SUV_{max} 4.7), bilateral hilar lymph nodes (SUV_{max} 3.2), and remnant pancreatic tail (SUV_{max} 5.5) (Fig. 4). Based on these findings, we considered this tumor to be liver metastasis of the PanNET, IgG4-related hepatic IPT, or other hepatic tumor.

A percutaneous tumor biopsy was very difficult and risky to perform because the tumor was located on the surface of the liver adjacent to the diaphragm. We planned to perform laparoscopic partial hepatectomy to confirm the diagnosis. However, the intraoperative findings showed that the tumor was located not on the liver surface but in the diaphragm. We therefore performed laparoscopic partial resection of the diaphragm (Fig. 5). Histopathologically, marked lymphoplasmacytic infiltration and storiform fibrosis were found (Fig. 6A). An immunohistochemistry assessment showed the infiltration of CD38-, IgG-, and IgG4-positive plasma cells. More than 10 IgG4-positive plasma cells per HPF and a ratio of IgG4-/IgG-positive cells of more than 40% were observed (Fig. 6B-D). The patient was diagnosed with an IgG4-related diaphragmatic IPT based on the mass formation in the diaphragm, elevated serum IgG4 level, and surgical histopathological findings, which all fulfilled the Comprehensive Diagnostic Criteria for IgG4-RDs in Japan (9). The patient was followed up without treatment because he had no symptoms.

One year after surgery, CT showed left ureteral wall thickness with hydronephrosis, which was considered a manifestation of the IgG4-RD and can caused renal dysfunction. We initiated the administration of oral corticosteroids at a dose of 30 mg/day (0.6 mg/kg/day), which was tapered at 2.5 mg/week until reaching a dose of 20 mg/day. Subsequently, corticosteroids were gradually reduced by 2.5 mg every 4 weeks. Seven months after starting treatment, the dose was reduced to 5 mg/day, and maintenance therapy has been continued at the same dose thus far. After corticosteroid treatment, the ureteral wall thickness and swelling in the remnant pancreatic tail improved, and the serum IgG4 level decreased to normal.

Discussion

IgG4-RDs were proposed as a new entity of systemic disease with multiple organ involvement by Kamisawa et al. in

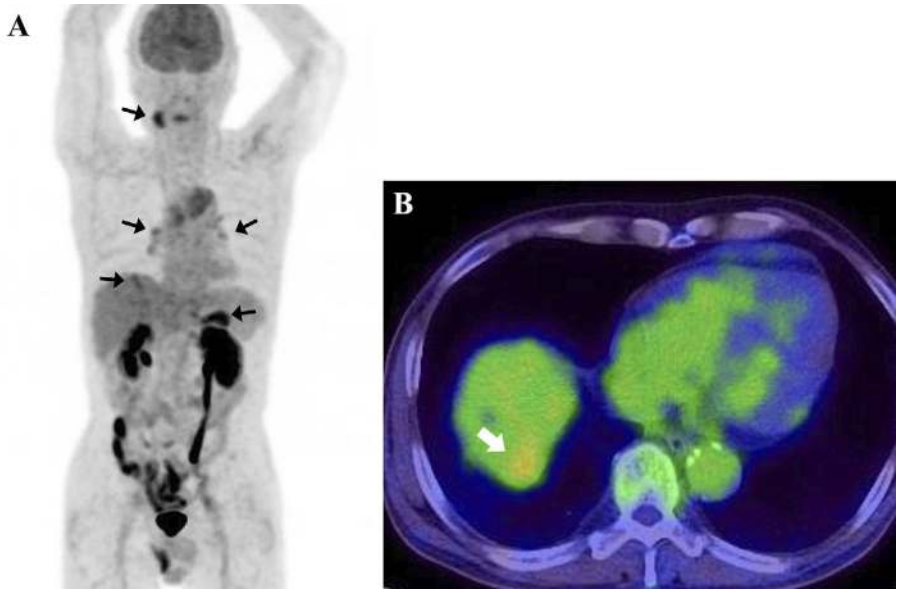

Figure 4. **^{18}F-FDG PET/CT findings in 2017. (A) Maximum-intensity projection image showed an abnormal ^{18}F-FDG uptake in the hepatic tumor lesion (SUV_{max} 3.4), right submandibular gland (SUV_{max} 4.7), bilateral hilar lymph nodes (SUV_{max} 3.2), and remnant pancreatic tail (SUV_{max} 5.5) (the abnormal uptake is indicated by arrows). (B) Hepatic tumor lesion on PET/CT axial image (white arrow). ^{18}F-FDG: ^{18}F-fluorodeoxyglucose, CT: computed tomography, PET: positron emission tomography, SUV_{max}: maximum standardized uptake value**

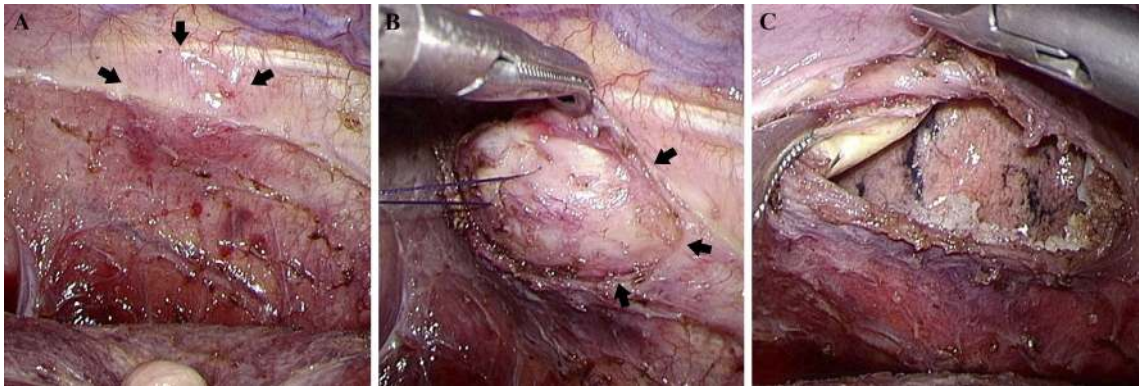

Figure 5. **Intraoperative findings. (A, B) The tumor was located in the diaphragm (arrows). (C) Laparoscopic partial resection of the diaphragm was performed.**

2003 (10). IgG4-RDs can affect various organs in the body. Miyabe et al. conducted a pooled analysis of five large series of IgG4-RDs (11). The most common organ manifestation was in the pancreas, which accounted for about 45% of the patients. The lacrimal and salivary glands were involved in 25% of patients, the bile ducts in 20%, and the retroperitoneum, lungs, and kidneys in 20% each. Other organ involvements included the prostate, sinuses, aorta, liver, gallbladder, thyroid, pleura, mediastinal fibrosis, skin, mesentery, paraspinal region, pituitary, meninges, pericardium, testes, and colon. To our knowledge, no report of an IgG4-RD involving the diaphragm has yet been published.

The present patient showed an IgG4-related IPT that occurred in the diaphragm during follow-up of AIP. Recently, some IPT cases were described as part of the spectrum of IgG4-RDs (6). IPTs are characterized by tumor-like mass lesions showing variable amounts of fibrosis with polymorphous inflammatory infiltrates, including lymphocytes, plasma cells, and myofibroblastic spindle cells (5). Chougule and Bal (6) reviewed 83 cases of IgG4-related IPTs reported in 40 articles. The most common sites of involvement were the lungs (22 cases), liver (11 cases), orbit (8 cases), central nervous system (8 cases), kidneys (8 cases), and ureter (7 cases). Other less common sites included the breasts, stomach, pituitary, mediastinum, urinary bladder, lymph nodes, oral cavity, adrenal gland, and testes; there were no cases with involvement of the diaphragm. Interestingly, Hoer et al. (12) reported that the histological findings in a five-year-old boy who presented with an IPT in the diaphragm in 1999 showed fibroblasts arranged in a storiform growth pat-

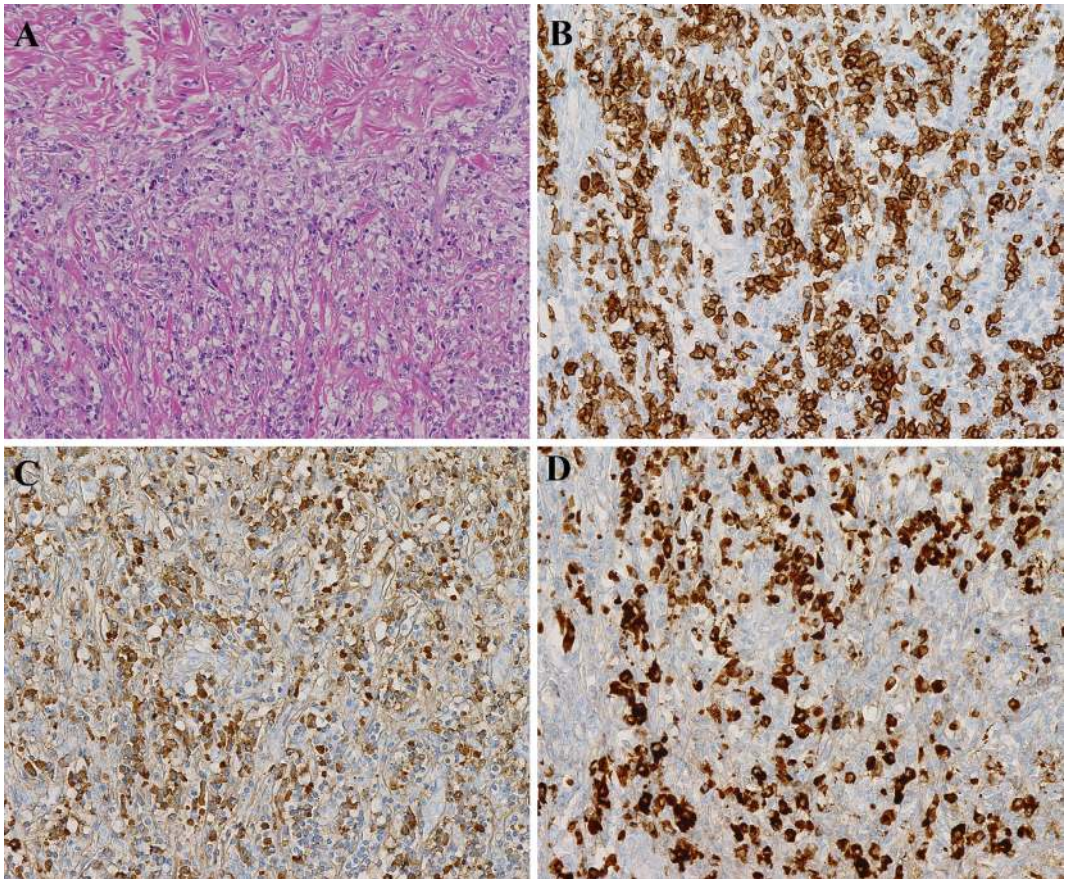

Figure 6. Histological findings of the diaphragmatic tumor. (A) The histological findings revealed dense lymphoplasmacytic infiltration and storiform fibrosis (Hematoxylin and Eosin staining, magnification ×100). Immunohistochemistry demonstrated (B) CD38-, (C) IgG-, and (D) IgG4-positive plasma cells (magnification ×200). More than 10 IgG4-positive plasma cells per HPF and a ratio of IgG4-/IgG-positive cells of more than 40% were observed. CD38: cluster of differentiation 38, HPF: high-power field, Ig: immunoglobulin

tern and perivascular infiltration of plasma cells, as well as some lymphocytes, which suggested the possibility of an IgG4-related diaphragmatic IPT. However, immunohistochemical and serum-based assessments of IgG4 were not performed because the concept of IgG4-RDs had not yet been established.

It is difficult to differentiate IgG4-RDs from malignancies and other IgG4-RD mimics. The clinical, laboratory, and imaging findings are often insufficient to exclude malignancies (13, 14). In particular, IgG4-related IPTs are difficult to differentiate from malignancies due to the similarity of the imaging findings (5, 6). Moon et al. (15) suggested a short-term steroid trial and responses to steroids as useful diagnostic tools for AIP. However, malignant lymphoma may be improved by steroids alone, and there have been scattered reports of IgG4-RD accompanied by malignant lesions (9, 16-18). In these cases, a steroid trial may lead to a misdiagnosis or delayed treatment of malignancy. According to the International Consensus Diagnostic Criteria for AIP (8), a steroid trial, despite being included as a diagnostic criterion, should be carefully conducted only after a negative workup for cancer has been conducted, including EUS-FNA. An international consensus guidance statement on the management and treatment of IgG4-RDs (14) strongly recommended that diagnostic confirmation by a biopsy be conducted to exclude malignancies and other IgG4-RD mimics.

We reviewed 19 IgG4-related IPTs that occurred at uncommon sites (fewer than 3 cases reported) (Table) (19-36). The lesions were located at the breast in three cases, epidural in two cases, mediastinum in two cases, paratestis in two cases, urethra in two cases, and oral cavity, trachea, adrenal gland, bladder, rectum, abdominal wall, pericardium, uterine in one case each. All cases were diagnosed by histological findings, and 15 of the 19 cases (78.9%) were diagnosed by excision specimens. Furthermore, IgG4-related IPTs were not considered as a differential diagnosis before the biopsy or surgery in any case. These case reports suggest that the diagnosis of IgG4-raleted IPTs, especially in rare locations, based solely on clinical, laboratory, and imaging findings is extremely difficult. A histological examination should thus be performed in order to diagnose IgG4-related IPTs and exclude malignancies.

In our case, we considered an IgG4-related IPT as a dif-

Table. **IgG4-related Inflammatory Pseudotumors Located at Uncommon Sites.**

Case	Reference	Location	Age/ Sex	Symptom	Serum IgG4 level (mg/dL)	Other organ involvement	Diagnosis method	Treatment
1	(19)	Breast	46/F	Induration	185	None	Excision biopsy	Resection
2	(20)	Breast	66/F	Lump	n/a	None	Surgical specimen	Resection
3	(20)	Breast	45/F	Lump	n/a	None	Excision biopsy	Resection
4	(21)	Epidural	57/F	Dorso-lumbar pain, paraparesis	66.2	None	Surgical specimen	Resection and corticosteroids
5	(22)	Epidural	50/M	Back pain, paraplegia	n/a	None	Needle biopsy	Resection and corticosteroids
6	(23)	Mediastinum	70/F	Dyspnea	n/a	None	Surgical specimen	Resection
7	(24)	Mediastinum	44/M	No symptoms	n/a	None	Surgical specimen	Resection
8	(25)	Paratestis	67/M	Painless scrotal mass	n/a	Pancreas, retroperitoneal	Surgical specimen	Resection
9	(26)	Paratestis	41/M	Painless scrotal mass	n/a	None	Surgical specimen	Resection
10	(27)	Urethra	72/F	Dysuria	n/a	Pancreas, eyelid	Needle biopsy	Corticosteroids
11	(28)	Urethra	75/F	Urinary retention	n/a	Pancreas	Needle biopsy	Corticosteroids and azathioprine
12	(29)	Oral cavity	65/M	Maxillary alveolar swelling	n/a	Lung	Surgical specimen	Resection
13	(30)	Trachea	22/F	Stridulous breathing	n/a	None	Excision biopsy	Resection and corticosteroids
14	(31)	Adrenal gland	41/F	Abdominal pain	n/a	None	Surgical specimen	Resection
15	(32)	Bladder	72/F	Hematuria	n/a	None	Surgical specimen	Resection
16	(33)	Rectum	28/F	Constipation, Anal discomfort	n/a	None	Excision biopsy	Resection and corticosteroids
17	(34)	Abdominal wall	54/M	Abdominal pain	43.5	None	Excision biopsy	Resection
18	(35)	Pericardium	75/F	No symptoms	358	None	Surgical biopsy	Observation
19	(36)	Uterine	39/F	No symptoms	671	None	Surgical specimen	Resection

ferential diagnosis, but a percutaneous tumor biopsy was very difficult and risky to perform because of the location of the lesion. In addition, liver metastasis of the PanNET could not be ruled out, so we performed surgery to establish a definitive diagnosis rather than implementing a diagnostic steroid trial.

Corticosteroids are the first-line agents for remission induction of IgG4-RDs, and most IgG4-RDs respond well (14, 37). Corticosteroid treatment is recommended in symptomatic patients, considering the side effects (38). However, it is difficult to achieve complete remission because relapse is not uncommon. Relapse rates have been reported to range from 46-90% during tapering and after withdrawal of corticosteroids (37). Although the selection of patients who should receive maintenance therapy remains unclear, maintenance therapy with corticosteroids following remission reduces the rate of relapse (37). A Japanese multicenter randomized controlled study in 49 patients with AIP showed a significantly lower relapse rate (23.3%) in patients who received low dose corticosteroid therapy for 3 years than in those who discontinued the therapy at 26 weeks (57.9%) (39). For some patients, corticosteroid-sparing immunomodulators and rituximab are used to avoid cumulative toxicity of corticosteroids (3, 37). Rituximab, a B-cell-depleting agent, might be the first alternative option for patients in whom corticosteroid treatment is risky and have a history of multiple relapses. Carruthers et al. (40) reported a prospective open-label trial using rituximab in 30 patients with IgG4-RD, 47% of whom were in complete remission at 6 months without corticosteroid treatment. In this case, the patient responded well to corticosteroids and has remained in remission with maintenance therapy of low-dose corticosteroids.

To our knowledge, this was the first case of an IgG4-

related diaphragmatic IPT. This case suggests that IgG4-RDs can affect any organ in the body. We should consider IgG4-related IPTs as a differential diagnosis when mass lesions are detected in patients with a definitive or suspected diagnosis of an IgG4-RD.

The authors state that they have no Conflict of Interest (COI).

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