Case Report

IgG4-Producing MALT Lymphoma in the Renal Hilum

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Keywords

B cell lymphoma · IgG4-related disease · IgG4-producing B cell lymphoma

Abstract

IgG4-related disease is diagnosed when both the elevation of the serum IgG4 level and invasion of IgG4-positive interstitial cells and sclerosis to a tumor are noted. Some cases have demonstrated malignant disease. In the head and neck lesion in particular, IgG4-producing mucosa-associated lymphoid tissue (MALT) lymphoma has arisen during the treatment of IgG4-related disease. We herein report the first case of IgG4-producing MALT lymphoma during the treatment of IgG4-related disease in the renal hilum. A 79-year-old man was being followed for autoimmune pancreatitis and IgG4-related sclerosing cholangitis. During follow-up, magnetic resonance cholangiopancreatography detected a mass in the renal hilum, so he was referred to our department for a further examination. Positron emission tomography-computed tomography detected a standard uptake of 9.7, and the tumor size was gradually increasing. Due to these findings, laparoscopic nephro-ureterectomy was performed. A pathological examination revealed IgG4-producing marginal zone B cell lymphoma. We herein report a rare case of IgG4-producing B cell lymphoma in the renal hilum.

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Introduction

IgG4-related disease is a chronic inflammatory disease in which both the elevation of the serum IgG4 level and invasion of IgG4-positive interstitial cells and sclerosis to a tumor are noted. Recently, IgG4-related disease has shown an increased incidence in patients with malignant disease [1]. Mucosa-associated lymphoid tissue (MALT) lymphoma develops from MALT as a result of chronic inflammation and tumor originates from B cells. In the head and neck area in particular, IgG4-producing MALT lymphoma has arisen during the treatment of IgG4-related disease, and a correlation between IgG4-related disease and IgG4-producing lymphoma has been suspected [2, 3].

We herein report the first case of IgG4-producing MALT lymphoma arising in the renal hilum during the treatment of IgG4-related autoimmune pancreatitis.

Case Presentation

A 79-year-old man was being followed for autoimmune pancreatitis and IgG4-related sclerosing cholangitis. He was receiving prednisolone, and his disease was well controlled. During follow-up, magnetic resonance cholangiopancreatography detected a mass in the renal hilum, so he was referred to our department for a further examination.

Retrograde pyelo-nephrography showed no defect in the ureter or renal pelvis but did show stenosis from an external tumor. Positron emission tomography-computed tomography (PET-CT) detected a standard uptake value (SUV) of 9.7, and the tumor size was gradually increasing (Fig. 1, Fig. 2). Due to these findings, laparoscopic tumorectomy or nephro-ureterectomy with an operative rapid pathologic diagnosis was planned. A pathological examination revealed IgG4-producing marginal zone B cell lymphoma. During surgery, however, the tumor was found to be strongly adhered to the surrounding tissue and unable to be removed, so nephro-ureterectomy was performed (Fig. 3). A pathological examination revealed IgG4-producing marginal zone B cell lymphoma. The patient remains free from recurrence at 35 months after surgery.

Discussion

This case was one of IgG4-producing MALT lymphoma in the renal hilum during the treatment of IgG4-related autoimmune pancreatitis and IgG4-related sclerosing cholangitis. The differential diagnosis was retroperitoneal tumor, MALT lymphoma, and other malignancies. Some studies have suggested the usefulness of PET-CT. In PET-CT, the maximum SUV (SUV_{max}; cut-off point of 6.23) showed a sensitivity of 85.7% and specificity of 80.3% [4]. The present case showed an SUV_{max} of 9.7 and was suspected of being malignant disease.

The detailed protocol for treating IgG4-related disease has not been established. When steroids have been used to induce remission or as maintenance treatment, cases of recurrence have been reported. Although an increased incidence of malignant disease has been reported, the recommended follow up period and how to evaluate the risk remain unclear. Because our case had IgG4-related disease-based chronic inflammation, an increased risk of MALT lymphoma over general renal-origin MALT lymphoma was suspected. We plan to perform follow up using imaging modalities while controlling his disease with steroids based on the serum IgG4 and IgG levels.

A few cases of renal-origin MALT lymphoma have been reported. In those cases, surgical resection or radiation was performed and showed a favorable outcome [5]. However, in the reported cases of idiopathic orbital inflammation or MALT lymphoma, the incidence of IgG4-positive cases was higher than that of IgG4-negative cases [6]. The present patient underwent nephro-ureterectomy due to the risk of malignant disease. He has been free from recurrence for 35 months since the surgery.

Statement of Ethics

Written informed consent to participate and for publication was obtained from the patient. A copy of the written consent form is available for review from the Editor-in-Chief of this journal.

Disclosure Statement

We declare no conflicts of interest.

Funding Sources

No funding was received.

Author Contributions

K.S., T.K. drafted the manuscript. R.K., J.K., S.S., R.T., H.U., K.C. performed the experiment.

Availability of Data and Material

Due to ethical restrictions, the raw data underlying this paper are available upon request to the corresponding author.

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Fig. 1. Initial (a) and follow-up (b) axial CT findings. Initial (c) and follow-up (d) sagittal CT findings.

Fig. 2. PET-CT.

Fig. 3. Surgical specimen.