Castleman’s disease is a rare benign lymphoproliferative disorder that frequently affects lymph nodes of the mediastinal thorax and the neck. It very rarely affects the renal sinus. We report a case of Castleman’s disease arising in the renal sinus in a 64-year-old man. The patient visited the hospital with the chief complaint of hematuria. Abdominal computed tomography revealed a homogeneous mass in the sinus of the left kidney, radiologically interpreted as a malignant urothelial tumor. Subsequently, nephroureterectomy was performed, after which microscopic examination of the specimen revealed a diffuse lymphoproliferative lesion with reactive lymphoid follicles of various sizes and prominent plasma cell infiltration of interfollicular spaces, highlighted by immunohistochemical staining for CD138. The lesion was diagnosed as Castleman’s disease of the plasma cell type. Although preoperative diagnosis of Castleman’s disease is difficult and the incidence is exceedingly rare, it should be considered in the differential diagnosis of renal sinus tumors.

Key Words: Giant lymph node hyperplasia; Plasma cell type; Renal sinus; Urothelial malignancy; Kidney

CASE REPORT

A 64-year-old man visited our hospital with the chief complaint of microscopic hematuria for the past 2 months. Laboratory tests were unremarkable and urine cytology was normal. Abdominal computed tomography (CT) revealed a homogeneous mass in the sinus of the left kidney, radiologically interpreted as a malignant urothelial tumor. Subsequently, nephroureterectomy was performed, after which microscopic examination of the specimen revealed a diffuse lymphoproliferative lesion with reactive lymphoid follicles of various sizes and prominent plasma cell infiltration of interfollicular spaces, highlighted by immunohistochemical staining for CD138. The lesion was diagnosed as Castleman’s disease of the plasma cell type. Although preoperative diagnosis of Castleman’s disease is difficult and the incidence is exceedingly rare, it should be considered in the differential diagnosis of renal sinus tumors.
renal pelvis, calyces and ureter were intact without abnormal urothelial changes. Microscopically, the tumor occupying the renal sinus contained widely scattered, hyperplastic lymphoid follicles, characteristic of a lymphoproliferative lesion. The folli-

Fig. 1. Abdominal computed tomography reveals a homogeneous infiltrative mass in the left renal sinus with relative low attenuation compared to the renal parenchyma (arrows).

Fig. 2. Gross findings. The resected kidney contains a 4 × 2.5 cm gray-whitish infiltrative pelvic mass (arrows) with marked thickening of the walls of the proximal ureter and major calyces.

Fig. 3. Histopathologic findings. (A) The pelvic tumor is a benign lymphoproliferative lesion with widely scattered, hyperplastic lymphoid follicles. The pelvic urothelium is intact. (B) The renal calyces also contain a submucosal lymphoproliferative lesion. (C) Marked plasma cell infiltration in interfollicular spaces. (D) The plasma cells are positive for CD138 upon immunostaining.
cles varied in size and showed polarized germinal centers; moreover, interfollicular spaces were markedly infiltrated by mature plasma cells, highlighted by CD138 immunostaining (Fig. 3). Immunostaining for κ and λ immunoglobulin light chains indicated that the plasma cells were polyclonal in origin. Immunostaining for human herpes virus 8 (HHV-8) was performed, the results of which were negative. These histological and immunohistochemical findings were compatible with a diagnosis of Castleman’s disease of the plasma cell type. Three months after the operation, there was no evidence of disease recurrence on follow-up abdominal CT.

DISCUSSION

The occurrence of Castleman’s disease in the renal sinus is exceedingly rare. To the best of our knowledge, only a few cases have been reported. The clinicopathological characteristics of the reported cases, involving six males and one female, are summarized in Table 1. Although Castleman’s disease generally shows no gender preference, cases involving the renal sinus appeared to show a male predominance. The median age for Castleman’s disease is in the fourth decade. However, the mean age of the cases involving the renal sinus was 65 years. The main presenting symptoms included abdominal pain, weight loss, anorexia, and microscopic hematuria. Three of the cases were clinically unicentric and four were multicentric. Five cases were of the plasma cell type, one of the hyaline vascular type and one of mixed type. The patient in the present case, a 64-year-old male, presented with symptoms of microscopic hematuria, and his disease was histologically classified as plasma cell type and clinically as unicentric type.

Although the exact pathophysiology of Castleman’s disease is unknown, recent reports suggest that HHV-8 infection may stimulate B lymphocytes to induce interleukin 6 (IL-6) production in the mantle zone; IL-6 overproduction has been shown to be associated with the systemic manifestations of Castleman’s disease, especially in cases of multicentric disease. In our case, immunostaining for HHV-8 was performed, but the result was negative.

Preoperative diagnostic imaging methods are not useful in differentiating Castleman’s disease arising in the renal sinus from other diseases because of a lack of tumor-specific imaging features. Previously, Nishie et al. described the CT and magnetic resonance imaging features of three cases of Castleman’s disease involving the renal sinus. They reported that the renal sinus lesions formed in Castleman’s disease can appear as homogeneous masses with mild enhancement, such as is often observed in malignant lymphomas. Thus, it is difficult, on the basis of radiological findings, to differentiate Castleman’s disease from malignant lymphomas infiltrating the renal sinus. Other differential diagnoses may include invasive urothelial carcinoma, granulomatous diseases, sarcomas, and metastases. Consequently, pathological evaluation is currently the only method of identifying Castleman’s disease of the renal sinus.

Here we have described an extremely rare case of unicentric Castleman’s disease of the plasma cell type arising in the renal sinus. Although preoperative diagnosis of Castleman’s disease is difficult and accurate diagnosis is only possible by histopathologic evaluation, it should be considered in the differential diagnosis of renal sinus tumors.

Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

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