We describe herein an extramedullary plasma cytoma (EMP) in the retroperitoneal space, an uncommon manifestation of plasma cell tumor occurred in a rare site. Only a few cases have been reported in a search of English-language literature. In this report, the imaging appearances of the tumor in both computed tomography (CT) and sonography will be presented. With relevant laboratory data and histologic examination obtained from ultrasound-guided biopsy, a definitive diagnosis of EMP was made.

CASE REPORT

An 85-year-old male patient, who had a history of hypertension and diabetes mellitus, came to our emergency department with chief complaints of fever, nausea and vomiting for 2 days. Physical examination revealed fever up to 38.8°C and laboratory studies disclosed elevated serum CRP level up to 12.37 mg/dl, high percentage of segmented WBC without leukocytosis, elevated serum amylase and lipase levels of 185 U/L and 318 U/L respectively. Computed tomography of abdomen demonstrated a large infiltrative soft tissue mass, which was homogeneous in appearance with a tiny calcified spot, in the right perirenal and pararenal spaces (Fig. 1a). It was barely distinguishable to the right kidney on non-contrast enhanced images. After administration of intravenous contrast, the soft tissue mass was homogeneously enhanced and it invaded the upper pole of the right kidney, which was well demonstrated on the coronal multiplanar reformatted image (Fig. 1b). Right renal artery and vein were not involved and no hydronephrosis or retroperitoneal lymphadenopathy was found. Based on the imaging findings of this computed tomography of abdomen, the differential diagnoses included lymphoma, infiltrative renal cell carcinoma and unliquefied retroperitoneal abscess.
An ultrasound examination for ultrasound-guided biopsy revealed a homogeneous and hyper-echoic mass circumscribing the right kidney in the perirenal and pararenal spaces (Fig. 2a). Foci of blood-flow signal were found inside the soft tissue mass on the color Doppler image (Fig. 2b).

Microscopic examinations and specific immunocytochemical staining disclosed mixed mature lymphocytes and plasma cells (Fig. 3a) with predominant kappa light chains (Fig. 3b). In addition,

Figure 1. a. Unenhanced CT of abdomen showed a homogeneous and infiltrative mass lesion with a calcified spot (white arrow) in the right upper retroperitoneum. b. Contrast-enhanced CT of abdomen in coronal multiplanar reformation showed the infiltrative mass with homogeneous enhancement invading the upper pole of right kidney (black arrow).

Figure 2. a. A homogeneous hyperechoic mass lesion infiltrated the perirenal space, adjacent right kidney and the liver, and b. increased flow signals were shown in the mass lesion on the color Doppler sonography.
serum protein electrophoresis reported high peak over gamma globulin region. Bence Jones protein was not detected in the urine. A series of conventional radiography for detailed skeletal survey and bone scan yielded no evidence of other bony lesions. These associate findings ruled out the possibility of multiple myeloma. On the basis of these findings, a diagnosis of retroperitoneal plasmacytoma was then established.

DISCUSSION

Plasmacytomas are tumors of plasma cells or B lymphocytes with variable maturity, identical to those seen in multiple myeloma histologically. They mostly occur in bone as a solitary lesion (solitary plasmacytoma of bone) or as multiple lesions (multiple myeloma). If they arise outside of bone in soft tissues, they are designated as EMP [1]. Approximately 95% of plasma cell tumors are presented as multiple myeloma and solitary plasmacytoma of bone and only less than 5% are EMPs. Besides, most of EMPs develop in the head and neck, especially in the mucous membranes of upper respiratory tract, followed by gastrointestinal tract [3]. Rarely, plasmacytomas are found in the peri- and pararenal spaces of retroperitoneum [3, 4, 5].

The image findings of EMP can be solitary or infiltrative in appearance, with or without regional lymph node involvement. Renal venous involvement leading to thrombosis and hemorrhage had been reported in two cases [6, 7]. Obstructive jaundice and thrombosis of the inferior vena cava occurred as a result of a solitary EMP in the retroperitoneum was also reported in one case [2]. In our case, it was an infiltrative and isodense soft tissue mass in the right upper perirenal space, invading right kidney and adjacent liver. The right renal artery and vein were encased without thrombus formation. On the sonographic images, the lesion was hyperechoic to adjacent renal parenchyma and there was blood flow signal within the lesion in color Doppler image. No enlarged lymph node was identified in the peritoneal or retroperitoneal spaces. Calcified foci in the plasma cell tumors were reported, which was considered as a calcified amyloid deposition [8]. We also found a small calcification in our case. However, none of these image characteristics, as we have described in this case, is specific or diagnostic to EMP, since it may also be found in other retroperitoneal infiltrative lesions.

Based on image findings, the differential diagnoses of an infiltrative and a homogeneous mass lesion in the retroperitoneum include a group of lymphoproliferative disease such as lymphoma, leukemia and plasmacytoma, infiltrative renal cell carcinoma, retroperitoneal sarcoma and metastasis [9]. In addition, other non-neoplastic conditions such as xanthogranulomatous reaction, hematoma, retroperitoneal phlegmon, and abscess formation are also included. In our case, the first three differential diagnoses for lymphoproliferative disease are most

Figure 3. a. Microscopic view of the retroperitoneal tumor demonstrated mixed mature lymphocytes and plasma cells (hematoxylin and eosin, x400). b. Immunocytochemistry stain showed predominant kappa light chain in the tumor.
likely, while it is difficult to tell them apart solely by image findings. To make a more reliable diagnosis ahead of pathologic result, associate laboratory data and other imaging studies such as skeletal survey and bone scan play an important role in excluding other systemic diseases.

Clinically, the criteria of diagnosing EMP includes (1) less than 5% plasma cell in a bone marrow biopsy, (2) no Bence Jones protein in urine, (3) normal skeletal survey and (4) a biopsy proven plasma cell tumor in an extramedullary site. The presence of a high peak of monoclonal band in a serum protein electrophoresis used to implicate a systemic disease. However, it cannot exclude a patient from being diagnosed with EMP, since presence of serum monoclonal component has been reported in 14-25% of cases [10]. On the other hand, hyperamylasemia in the EMP had been cited in one case report, but it also can be seen in several kinds of epithelial and nonepithelial tumors, including multiple myeloma and retroperitoneal plasmacytoma [11]. Therefore it didn’t help in excluding other differentials, even though we found elevated serum amylase in our case.

Since the tumor is highly radiosensitive, radiotherapy, with or without surgery, is the treatment of choice. In addition, chemotherapy has also been used in initial tumor control or as a treatment when systemic progression occur [9]. The size of plasmacytoma has been reported to be an important prognostic factor. In general, it has a tendency to recur locally and even spreads out as systemic disease without adequate treatment.

In summary, we report a rare case of primary EMP in the retroperitoneum. To distinguish an infiltrative renal lesion is usually difficult based on image manifestations. Subsequent patient management may range from antibiotic treatment to nephrectomy, radiotherapy or even systemic chemotherapy. Despite its rarity, primary EMP should be included in the differential diagnosis of infiltrative retroperitoneal tumor. The final diagnosis cannot be made without combination of laboratory studies, radiologic and pathologic examinations.

REFERENCES
後腹腔漿質細胞瘤的電腦斷層以及超音波的影像學表現：病例報告

賴彥君1  王家槐2,3  王信凱2,3  沈書慧2,3  周宜宏2,3  張政彥2,3

亞東醫院  影像醫學科1
台北榮民總醫院  放射線部2
國立陽明大學  醫學院3

本篇病例報告主要描述一個位於後腹腔腫外的漿質細胞瘤的影像學表現，漿質細胞瘤罕見生長於此，在英文的文獻裡只有少數的病例報告。在這個病例中，我們描述了這個腫瘤的電腦斷層以及超音波的影像學表現，加上充分的實驗室數據以及經超音波穿刺得到的組織學檢查，腫外漿質細胞瘤得以確診。