中文題目：濾泡樹突細胞肉瘤---一個以腹部腫瘤表現的罕見個案

英文題目：Follicular dendritic cell sarcoma --- A rare case of presenting as an abdominal tumor

作　　者：陳乃慈[1]，梁世昕[1,2]，吳俊鋒[1,3]

服務單位：[1]佛教慈濟醫療財團法人大林慈濟醫院內科部，[2]佛教慈濟醫療財團法人大林慈濟醫院 血液腫瘤科，[3]佛教慈濟醫療財團法人大林慈濟醫院 新陳代謝科

Background:

Follicular dendritic cell sarcoma (FDCS) is a neoplasm that arises from follicular dendritic cells. In most cases, the major sites affected are the lymph nodes. In fewer than one-third of cases, FDCS can also be found in extranodal sites and the abdominal region is thought to be one of the relative preferred sites of extranodal FDCS. Clinically, they often mimic a wide variety of other abdominal tumors, and correct preoperative diagnosis is often a challenging task.

Case presentation:

This 65-year-old housewife presented with the chief complaints of epigastric pain and lower sternal discomfort for one month. She reported intermittent fullness and heartburn sensation for more than one months. She also recorded her body weight loss of about 8 Kilogram in one year. She accepted upper GI endoscopy first and gastric ulcer was noted. She received a proton pump inhibitor as treatment but she didn't have symptoms relief. Abdominal sonography revealed a tumor besides right lobe of liver about 10 centimeter in size. Therefore, abdominal CT was arranged but revealed a large tumor of right adrenal gland up to 11.1 cm in size, Hounsfield unit:23.5 and heterogeneous enhancement with venous invasion to inferior vena cava.

We performed preoperative evaluation for functionally active assessment. Laboratory examination revealed serum cortisol: 1.17ug/dl in 1 mg dexamethasone test, 24 hrs urinary free cortisol:178.8 ug/day, aldosterone:98.54 pg/mL, PRA <0.07 ng/ml/hr, aldosterone to renin ratio (ARR):1.4; catecholamine fraction in urine including norepinephrine:33.0 ug/24hrs, epinephrine <3.0 ug/24hrs, dopamine: 543.4 ug/24hrs and urinary VMA:3.54 mg/24 hrs. The laboratory data also revealed DHEA-S:64.6 ug/dl and testosterone:0.15 ng/ml. According to the results of the examination, the tumor was nonfunctional. Because of high risk of adrenal malignancy, she decided to undergo right adrenalectomy, right nephrectomy and IVC thrombectomy and repair with renal vein patch graft.

The pathological results revealed follicular dendritic cell sarcoma, which was unexpected in the beginning. Microscopically, the sections of tumor show epithelioid to spindle tumor cells with eosinophilic cytoplasm. Combined with the positive results of the immunohistochemistry study, the tumor was confirmed as follicular dendritic cells sarcoma. The staging was pT4N1M0 p-stage IIIB. She received adjuvant radiotherapy as 4500 cGy in 25 fractions from March to April, 2021 at first. However, multiple masses in bilateral lungs were considered metastases in May 2021. As the result, she received first-line palliative doxorubicin and ifosfamide for three courses, but treatment was discontinued due to septic shock and pneumonia developed one week after the third cycle of chemotherapy. After a shared-decision making process, we decided to use paclitaxel monotherapy as the second-line treatment and the regimen is still administered currently.

Conclusion:

Follicular dendritic cell sarcoma is a rare tumor that typically arises within lymph nodes but can also occur extranodal. It typically presents as a painless slow-growing mass with no associated symptoms. If present, symptoms are more common among patients with liver or abdominal FDCS. The histologic appearance and typical immunohistochemical staining pattern are often sufficiently sensitive and specific to help make the diagnosis. Because the diagnosis of FDCS is usually unsuspected, proper plan (surgical resection or diagnostic biopsy) to provide sufficient tissue is necessary to obtain correct diagnosis.