Case Report
A retroperitoneal gastrointestinal stromal tumor (GIST) with inguinal lymph node metastasis: a case report

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Abstract: Computed tomography (CT) scan of a patient showed a 20*15*12 cm retroperitoneal tumor with left inguinal lymph node metastasis. A surgical operation was performed to remove the tumor and metastatic lymph nodes. Postoperative histological examination showed a retroperitoneal GIST with inguinal lymph node metastasis. The patient started to take imatinib 400 mg/d. There were no signs of tumor recurrence at a follow-up period for 15 months.

Keywords: Lymph node metastases, retroperitoneal tumor, GIST

Introduction
Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumours of the gastrointestinal tract. At first diagnosis, about 10% of GISTS present with metastatic disease [1]. The main routes of metastasis in GISTs are transperitoneal dissemination and haematogenous spread to the liver, but involvement of regional or peripheral lymph nodes is rare [2]. Herein, we report a case of retroperitoneal GIST with inguinal lymph node metastases.

Case report
A 45-year-old male complained of early satiety, which persisted for 1 month. Then, he was admitted to our hospital. Gastroscopy demonstrated nothing special. Computed tomography (CT) scan showed a 20*15*12 cm retroperitoneal tumor with left inguinal lymph node metastasis (Figure 1). A surgical operation was performed to remove the tumor and metastatic lymph nodes.

At operation, a huge, thick-walled solid retroperitoneal tumor almost filled the left abdominal cavity. The tumor was well-demarcated from the surrounding organs (pancreatic tail, left spleen, sigmoid colon, bladder), which were displaced but not involved with the tumor. Furthermore, the tumor had no relationship with the adjacent major vessels. The enlarged left inguinal lymph node measured 3 cm, with an irregular shape. Therefore, retroperitoneal tumor dissection and en bloc inguinal lymph node dissection were performed.

Histopathological examination of the tumor showed an epithelioid GIST. In addition, the inguinal lymph node shared the same cell structure with the tumor. Immunohistochemically, all the cells were positive for CD117 (+), ki-67 (+), SMA(+) and DOG-1 (+); negative for CD34, desmin and S-100 (Figure 2). The patient was diagnosed as high grade EGIST due to the presence of lymph nodes metastasis, large tumor size, unfavorable histopathological features (high mitotic index). Mutations in KIT exons and PDGFRA exons were evaluated in the sample, but, nothing special was found. Therefore, he accepted adjuvant imatinib treatment (400 mg, daily). His condition remained stable for a 15-months’ follow-up.

Discussion
GISTs have been documented in all parts of the gastrointestinal tract: especially in the stomach (60% to 70%) and small intestine (25% to
EGIST with inguinal lymph node metastasis

Figure 1. Computed tomography (CT) scan showed an 20*15*12 cm retroperitoneal tumor (A) with an enlarged left inguinal lymph node (B).

Figure 2. H&E staining of the retroperitoneal GIST (A), the corresponding inguinal lymph node metastasis (B). Immunohistochemistry indicated that the cells were positive for c-Kit/CD117 (C), Dog-1 (D), Ki-67 (E), SMA (F).

35%), with rare occurrence in the colon and rectum (5%), esophagus (<2%) and appendix, gallbladder and pancreas [3]. Although rare, GISTs are of clinical relevance because in at least 10-30% of cases are malignant [4]. Some GISTs, so-called Extragastrintestinal Stromal Tumor (EGISTs), primarily in the omentum, mesentery or retroperitoneum, are unrelated to the tubular gastrointestinal tract. EGISTs are rare. Ruiz-Tovar J et al. [5] revealed in a multivariate analysis that retroperitoneum location is a bad prognostic factor. The poorer prognosis of patients with EGISTs than that of patients with GISTs is believed to the fact that EGISTs are frequently accompanied by adverse prognostic factors, such as high proliferative indices, large and distant metastases, and lymph node involvement [6].

The two main routes of GISTs metastases are intraperitoneal dissemination and haematogenous spread to the liver. Lymph nodes and extra-abdominal sites are rarely involved [7]. Furthermore, the distinct pattern of lymph nodes and extra-abdominal sites metastasis of the neoplasm is unclear. Although routine lymphadenectomy is not recommended for GIST treatment, some studies suggested that lymph node dissection should be considered in patients with any suspicion of nodal metastasis
Successful treatment of GISTs requires assessment of the extent and progression of disease. For patients with isolated resectable GIST, surgical resection with negative microscopic margin (R0 resection) remains the only chance for cure. However, without any further treatment, at least 50% of patients develop tumor recurrence. Published evidence from phase II and III multicenter trials supports the use of the tyrosine kinase inhibitor (TKI) as an adjuvant treatment in patients with a moderate to high risk of recurrence [9], which establishes that TKI prolongs recurrence-free survival and improves overall survival compared to historical controls.

Most of the reported GISTs with lymphatic spread occurred in intraabdominal nodes [8, 9]. GISTs with synchronous extraabdominal inguinal lymph node metastasis are rare. To our knowledge, only one case was reported. Vassos N et al. [10] described the case of GIST with isolated synchronous inguinal lymph node metastasis. As shown in our case, we performed an en bloc inguinal lymph node dissection because of the preoperative awareness of the lymph node metastases. According to the National Institutes of Health risk classification for GISTs, the presented tumor belonged to the high risk group. After the operation, the patient received adjuvant imatinib treatment (400 mg, daily). Postoperative CT scan was performed every 3 months for a fifteen months' follow-up, which showed no evidence of tumor recurrence. The patient needs a long follow-up period.

Disclosure of conflict of interest

None.

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