Duodenal gastrointestinal stromal tumor mimicking recurrent renal carcinoma: Case report

Nükseden böbrek tümörünü taklit eden duedonum gastrointestinal stromal tümörü: Vaka sunumu

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Abstract

Duodenal gastrointestinal stromal tumors (GISTs) are seen very rare and small part of GISTs which has variable clinical symptoms. We present an asymptomatic duodenal GIST mimicking recurrence of mucinous tubular and spindle cell carcinoma (MTSCC) of kidney in radiological imaging studies.

Key Words: Duodenum, GIST, renal carcinoma, recurrence, imatinib

Özet

Duedonum gastrointestinal stromal tümörü (GIST), belirsiz klinik semptomları olan ve tüm GIST'in az bir kısmını oluşturan çok nadir görülen bir tümördür. Radyolojik görüntülemede böbreğin musinöz tubüler ve iğsi hücreli tümörünün rekürrensini taklit eden asemptomatik duedonum GIST'i olan vakayı sunduk.

Anahtar Kelimeler: Duedonum, GIST, böbrek tümörü, rekürrens, imatinib

Introduction

Mucinous tubular and spindle cell carcinoma (MTSCC) is a rare unique polymorphous tumor of kidney. In the WHO 2004 classification, MTSCC was recognized as a distinct variant of renal cell carcinoma in first time and the tumor indicate differentiation from distal nephron segments (1). Diagnosis is confirmed by describing interconnecting tubular and spindled cells with low-grade nuclei and mucinous stroma in pathological specimen (2). Survey of MTSCC is excellent and only rare report of lymph node or metastases have been reported (3).

Gastrointestinal tumors (GIST) are defined as spindle cell tumors with a CD-117 expression and account only %1-3 of all gastrointestinal tumors (4). There is no specific symptom for GIST and imaging modalities are the most common tools to achieve diagnosis (5). Treatment of GIST depends on surgical resection of tumor if possible. Effectiveness of limited resection and pancreaticoduodenectomy is described well in literature (6-7). Also benefits of imatinib are well defined (8).

In this report, we aim to present asymptomatic duodenal GIST mimicking recurrence of MTSCC in radiological imaging studies.

Case Report

A 57-year-old male patient -with a history of right radical nephrectomy- admitted hospital for routine follow up. Right radical nephrectomy was performed 14 months ago for solid lesion in 6.5x5.5x6.0 cm diameters. Final pathology was mucinous tubular and spindle cell carcinoma. Surgical margins were tumor free and patient did not receive any further treatment. There were no pathological findings in physical examination. Complete blood count and biochemical tests including creatinine level and electrolytes were in normal range. Urine analyses and chest computer tomography had showed no abnormalities. Contrast-enhanced abdominal computer tomography revealed a solid lesion with 4.5x3.9x6.0 cm diameter in right retroperitoneal area **(Figure)**. The mass was close with duodenum and vena cava inferior and mass was evaluated as a local recurrence of renal cell carcinoma.

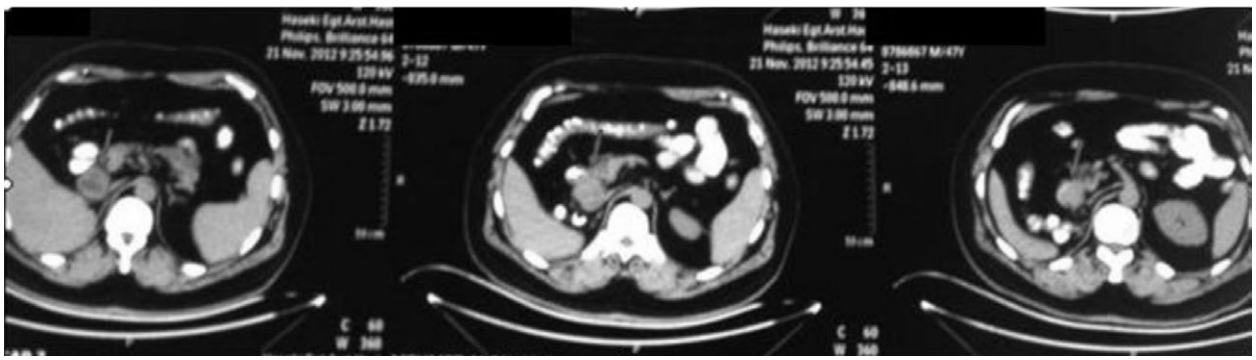

Figure: Computed tomography view of duodenal GIST.

Exploration was indicated in urooncology board. Operation was performed with right flank incision. During the operation, invasion of mass into the second part of the duodenum was detected. Mass was removed with anterior wall of the second part of the duodenum. There was no involvement to the papilla of Vater. Operation time was 140 minutes and estimated blood loss was 110 cc. Oral feeding was stopped for 5 days and parenteral nutrition was given. Post-operative period was uneventfully and patient was discharged 7th day after the operation. Final pathology was duodenal gastrointestinal stromal tumor in 5x4x5 cm in size with positive surgical margin. In microscopic examination, mitotic count was 5 mitosis/50 high power fields, spindle cell was positive and Ki-67 labeling index was observed 5%. In immunohistochemical examination CD-117(c-kit), CD-34, S100, SMA, vimentin were positive and desmin was negative. After medical oncology consultation, the patient was received 400 mg/day Imatinib therapy.

Discussion

Gastrointestinal stromal tumors are the most common mesenchymal tumor in human digestive system and originate from Cajal cells, which are located in muscular part of gastrointestinal organs as an interstitial cell (9). GISTs occur anywhere in gastrointestinal system, however are the most commonly seen in the stomach (%50-60) , small bowel (%25), colon (%10) and esophagus (%5-10) respectively. Duodenal GISTs are very rare and occur 3-5% of all cases (10). Expression of CD 117 protein (c-kit) is an important evidence to separate GIST from other mesenchymal tumors such as leiomyomas, myoblastomas and sarcomas (11).

Different anatomical location, growth (intramural or extramural) and size of the tumor lead variable complaints and there is no 'sine qua non' symptom or physical examination finding to identify GIST. According to the literature gastrointestinal bleeding, abdominal pain and intestinal obstruction are the most common symptoms (12-13). Some of GISTs are frequently diagnosed incidentally such as our patient, due to being asymptomatic, especially for the small tumors or when the tumor grows into extraluminal area. Also anorexia, fatigue, back pain and jaundice were described. If the common bile duct is compressed, GISTs in the second part of duodenum are more likely to become symptomatic, but in this case, the patient was asymptomatic with moderate size GIST in second part of the duodenum. (13-14)

Gastrointestinal endoscopy and radiologic examinations have essential role for detection of GIST. Endoscopy is mostly performed to identify the bleeding in gastrointestinal system and physician must be alert for GIST if submucosal swelling with mucosal ulceration is detected. Also it is often possible to obtain histological samples for the diagnosis (15). Computer tomography and magnetic resonance imaging are useful for investigate local extension of tumor and detection of metastasis (16). The absence of histological examination is the lack of both modalities. Although endoscopic ultrasound-guided fine needle aspiration and CT guided biopsy have been used for definitive diagnosis both procedures has potential risk for bleeding, tumor seeding and tumor rupture (17). In our case, mass was evaluated as a recurrence of renal cancer.

The aim of the surgical treatment for duodenal GIST is achieve clear resection margins (R0 resection) while avoiding tumor cell dissemination owing to fragility of

tumor.

The choice of surgical procedure is depend on size, location and extent of tumor. Routine lymph node dissection is not recommended owing to rarity of lymph node involvement (18).The commonest surgical treatments are pancreaticoduodenectomy (PD), segmental resection, wide resection and Whipple's surgery. PD is the best surgical option for periampullary duaedonal GISTs and large tumours of first and second part of the duedonom, which may be inadequately resected without pancreas. In the absence of involvement of the papilla of Vater, limited resection may be reasonable for duodenal GISTs. Limited resection procedures such as wedge resection, Roux-Y duodeno-jejunostomy and segmental duodenectomy provide reliable oncological results with better quality of life, less complication and shorter hospital stay. Also limited resection has benefits as continuity of gastrointestinal tract and preservation of pancreas (19-20).

Imatinib mesylate (IM) is a tyrosine kinase inhibitor and have important role in the management of GIST. In advanced tumors, IM has been advocated as a neoadjuvant therapy to obtain downstage of tumor, and prevent more extensive surgery (21). Also IM is helpful to complete resection of tumor and decrease the morbidity of resection. Further different studies have demonstrated that adjuvant IM treatment after resection of localized GIST significantly increase recurrence free survival with especially in high risk duodenal GIST (22). The optimal dose of adjuvant therapy is 400 mg/daily. If the tumor has KIT-exon-9-mutation, increased dose of IM 800mg/daily should be required. The other debatable area is optimal duration of IM. More recently, a phase III trial compared 12 months versus 36 months of IM therapy after surgery with a high risk of GIST recurrence. A significantly difference in improved recurrence free survival and overall survival of GIST patients who are at high risk of recurrence between 3 years and 1 year of IM group (23). Because of these results, National Comprehensive Cancer Network (NCCN) guidelines made a revision and now recommend at least 36 months adjuvant IM treatment in patients with high risk tumors (24). In this case, patient had microscopically margin-positive resection and IM treatment has started 400mg/daily.

In conclusion, retroperitoneal masses after radical nephrectomy are mostly considered as renal tumor recurrence; however tumors of the gastrointestinal tract should be kept in mind as GIST. Radiological imaging studies are often insufficient and exploration is mandatory for definitive diagnosis.

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