CASE REPORT

Left renal mass presenting uncommon pattern of extension in a patient with intestinal malrotation

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Abstract A 41-year-old man who had intestinal malrotation was presented with left renal tumor. The tumor extended venous thrombus up to hepatic portion and showed invasion in vessels and lymph nodes of mesocolon, which extended to porta hepatis. Needle biopsy of the renal tumor showed glandular adenocarcinoma. While colon cancer metastasis was possible, adenocarcinoma or glandular differentiation of urothelial carcinoma of the renal pelvis was also likely. The disease progressed rapidly and the patient died in a few months after the initial hospital visit. The very uncommon pattern of disease progression in this case was considered to be associated with intestinal malrotation, which is characterized by unfixed short mesentery, abnormal alignment of mid- and hind-guts, and the lack of normal anatomical structures between peritoneum and retroperitoneum including the ligament of Treitz. This case provides an important implication of intestinal malrotation in disease progression, which may affect clinical decision-making in the extent of surgical resection including lymph node dissection.

Keywords Renal tumor · Venous thrombus · Intestinal malrotation

Introduction

In the normal development, the foregut, midgut, and hindgut herniate out from the abdominal cavity and then undergo 270° counterclockwise rotation around the superior mesenteric vessels. Intestinal malrotation, also known as intestinal non-rotation or incomplete rotation, can be defined as any variation in the normal rotation and fixation of the gastrointestinal tract [1], occurring approximately 1 in 500 live births. The previous studies demonstrated that 80–90% of the cases are diagnosed during the first month of life [2, 3]. A series of 2000 barium enema showed the prevalence of intestinal malrotation was 0.2% [4]. However, recent studies indicated that more cases were diagnosed in adulthoods [5].

We herein report a case of advanced abdominal tumor occurring in an individual with intestinal malrotation. The tumor involving left kidney extended venous thrombus reaching the hepatic portion of inferior vena cava, invaded via mesocolon up to porta hepatis. Our case indicates that abdominal tumors arising in a patient with intestinal malrotation may present uncommon pattern of disease progression and affect our treatment strategy such as the extent of surgical resection and lymph node dissection.

Case report

A 41-year-old man presented to our clinic with upper abdominal discomfort and back pain since 1 week before. The patient reported no medical history of himself or his

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family. Physical examination showed no abnormal findings, while laboratory testing revealed normal complete blood cell counts, except for platelet count of 127 billion/L [normal reference range 150–350 billion/L], and abnormal serum biochemistry including aspartate transaminase (57 IU/L, [normal reference range 12–30 IU/L]), alanine transaminase (51 IU/L [normal reference range 10–42 IU/L]), gamma-glutamyl transpeptidase (138 IU/L [normal reference range 9–54 IU/L]), lactic dehydrogenase (657 IU/L [normal reference range 124–226 IU/L]), and C-reactive protein (2.3 mg/dL [normal reference value, <0.2 mg/dL]).

Several tumor markers were elevated including neuron-specific enolase (23.76 ng/mL [normal reference value, <12 ng/mL]) and cytokeratin 19 fragment (13.8 ng/mL [normal reference value, <2 ng/mL]), while AFP, CEA, CA19-9, and SCC were within normal limit. Soluble interleukin receptor 2 was not assessed unfortunately. Urinalysis and urine cytology showed no abnormal findings.

Contrast-enhanced computed tomography (CECT) scan (Fig. 1) revealed that the patient had midgut malrotation (non-rotation type) as evident by entire small intestine

located right side and colon located left side. CECT also showed a poorly enhanced 5-cm mass in the left kidney, which was accompanied by extensive tumor thrombus extending to the hepatic portion of the inferior vena cava (IVC). The left adrenal gland was also enlarged to 4 cm, suggesting adrenal metastasis. Multiple lymph node swellings were observed in the mesentery, left renal hilum, and porta hepatis. Among them, the largest node was identified in the mesocolon of the descending colon, which was also accompanied by extensive tumor thrombi in the mesenteric veins and portal vein up to porta hepatis. The adipose tissue between the mesenteric mass and the left renal hilum contained numerous strandings, suggesting prominent lymphatic infiltration of the tumor.

No abnormal findings were identified by gastrointestinal endoscopy and colonoscopy. A systemic positron emission tomography scan revealed no other lesion with abnormal uptake.

Histopathologic examination of needle biopsy specimen revealed high-grade adenocarcinoma (AC) with glandular structure (Fig. 2A). The tumor cells were positive for CDX2 (Fig. 2B), partially positive for CK7 (Fig. 2C)

Fig. 1 **A** Coronal image showing intestinal malrotation (non-rotation type) characterized by whole small intestine located *right* side and the colon *left* side of the body. Axial (**B–G**) and coronal (**H, I**) images on contrast-enhanced computed tomography. Multiple swollen lymph nodes and venous tumor thrombi are shown in porta hepatis (white arrowheads in **B** and **H**). Left renal mass (red arrowheads in **E** and **I**) extends vein thrombus in renal vein and hepatic portion of IVC (white arrows in **C, D**, and **I**). Left adrenal gland is occupied with tumor (yellow arrowheads in **C, D**, and **I**). The largest lymph node swelling is identified in the mesocolon (red arrow in **F**) with thickened wall of the corresponding descending colon (yellow arrow in **G**)

Fig. 2 H&E (A) stain and immunostains for CDX2 (B), CK7 (C), CK20 (D), GATA3 (E), and PAX8 (F). Renal tubular epithelium is strongly stained for PAX8. Glandular adenocarcinoma with strong positive stain for CDX2 is consistent with colon cancer

and CK20 (Fig. 2D), and completely negative for GATA3 (Fig. 2E) or PAX8 (Fig. 2F). Although microscopic appearance and immunohistochemical staining pattern strongly suggested metastatic AC from colon cancer, the possibility of AC or urothelial carcinoma (UC) with glandular differentiation originated from renal pelvis could not be completely excluded.

Although we recommended systemic chemotherapy based on the pathological diagnosis, the patient strongly preferred anti-PD-1 antibody treatment and received it elsewhere. The treatment did not show any response and the patients died at 2.5 months after initial symptom manifestation.

Discussion

Non-rotation type of intestinal malrotation is characterized by unfixed, short mesenterium, abnormal alignment of mid- and hind-guts, and the lack of normal anatomical structures between peritoneum and retroperitoneum including the ligament of Treiz. These anatomical anomalies might affect the form of direct invasion or lymphatic spread of the malignant neoplasm arising intra- or retro-peritoneum.

As the primary site of the tumor, colon cancer is likely based on the largest lymph node located in the left mesocolon and the extensive tumor thrombi in the mesenteric veins. Although colonoscopy did not identify any lesion suspicious of colon cancer, the sensitivity of colonoscopy is not perfect in terms of colon cancer detection, since tiny

primary colon cancer might be hidden in the submucosal layer.

Colon cancers often metastasize to mesenteric lymph nodes that can extend those in porta hepatis but seldom involve kidneys on the way. The association between intestinal malrotation and colon carcinogenesis was suggested on the basis of chronic bowel obstruction and consequent inflammation [6]. However, there have been only 15 cases of colon cancer associate with intestinal malrotation reported in the English literature [7]. Although there are a few cases showing unusual pattern of extension [8], none of these cases showed renal involvement, presumably because those cases had relatively early-stage diseases. The frequency of colon cancer invasion or metastasis to the kidney was reported as only 0–2.7% [9–11]. Involvement of the kidney as the first manifestation of primary colon cancer is an even rarer event [9].

Primary AC or UC with glandular differentiation of the renal pelvis is another possible diagnosis on the basis of the largest lesion located on the kidney showing modest enhancement, no capsule-like structure, and highly infiltrating appearance on contrast-enhanced CT scan. Primary pure AC of renal pelvis is extremely rare, whereas UC accounts for less than 10% of primary renal malignancy. Tumor thrombus in the venous system to the level of IVC is extremely rare for renal pelvic urothelial carcinoma [12], whereas it is relatively common for RCC (4–10%) [13]. It is well known that an aggressive renal pelvic cancer frequently invades the renal parenchyma keeping with renal outline (reniform) as the present case showed. Partially positive immunostaining for CK7 also supports urothelium

origin of the tumor. Needle biopsy could incidentally hit a lesion of predominant glandular differentiation of urothelial carcinoma. Renal pelvic cancers often metastasize to retroperitoneal lymph nodes but not to mesenteric or portal hilar nodes. In this particular case, however, anatomical anomalies might result in unusual pattern of disease spread.

Unfortunately, the definitive diagnosis was not obtained since the patient did not have surgical resection or autopsy. In any case, the present case suggested that anatomical variation could cause uncommon pattern of disease spread. It is noteworthy that malignant tumor can spread in an unexpected manner in an individual with intestinal malrotation. Even in earlier stage diseases, extent of surgical resection or lymph node dissection should be carefully considered on an individual basis.

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