Gastrointestinal stromal tumor of Meckel’s diverticulum: A rare cause of intestinal volvulus

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Meckel’s diverticulum is the most common congenital abnormality of the gastrointestinal tract. Most cases are asymptomatic; however, when symptomatic, it is often misdiagnosed at presentation. Common complications presenting in adults include bleeding, obstruction, diverticulitis, and perforation. Tumors within a Meckel’s diverticulum are rare. Herein, we present a gastrointestinal stromal tumor arising from the Meckel’s diverticulum that led to intestinal obstruction by volvulus.

Key words: Meckel’s diverticulum, volvulus, stromal tumor

INTRODUCTION

Meckel’s diverticulum (MD) is the most common congenital abnormality of the gastrointestinal tract. Most cases are asymptomatic; however, when symptomatic, it is often misdiagnosed at presentation. Common complications presenting in adults include bleeding, obstruction, diverticulitis, and perforation (1,2). Tumors within a MD are rare. Herein, we present a gastrointestinal stromal tumor (GIST) arising from the MD that led to intestinal obstruction by volvulus.

CASE REPORT

A 53-year-old female patient with no known health problems presented with abdominal distention, nausea and vomiting. On the physical examination, a distended abdomen with diffuse abdominal tenderness and rebound indicating acute abdomen was detected. An abdominal ultrasonography revealed a heterogeneous, solid mass measuring 14x12x10 cm located in the mid plain of the pubic region adjacent to the uterine fundus. In addition to those findings, dilated intestinal loops indicating a mechanical intestinal obstruction were detected. Abdominal computed tomography (CT) showed whirl sign signifying small bowel volvulus due to contiguous tumor with distention (Figure 1). A heterogeneous solid mass (14x12 cm) at the iliac bifurcation level, localized between the poste-
rior wall of the abdomen and vertebra corpus, was found (Figure 2). Additionally, the CT showed diffuse distended and dilated bowel loops. The patient was operated due to mechanical bowel obstruction leading to acute abdomen under emergency conditions. At operation, a lobule tumor mass arising from the MD was found, leading to volvulus of the small intestine. After decompression, a segmental small bowel resection including the tumor was made.

According to the pathologic examination, the tumor cells were spindle and 1 mitosis was present in 50 high-power fields. All margins were negative and the tumor had a margin to sub mucosa–serous membrane. CD117 (90%) was diffuse positive and S–100, smooth muscle actin (SMA), and CD34 were negative. The results of Ki–67 index were below 1%. The diagnosis was established as GIST of the MD. The postoperative period was uneventful, and she has been followed for two years without any local recurrence or metastases.

DISCUSSION

Meckel’s diverticulum (MD) is a congenital abnormality that arises at the site of the vitelline duct, which in the embryo connects the primitive gut to the yolk sac. If this fails to obliterate by the seventh week of gestation, congenital defects can persist that include umbilical sinus, omphalomesenteric fistula, enterocyst, fibrous band, and most commonly, MD. The diverticulum is a true diverticulum containing all layers of the intestinal wall and most commonly arises from the antimesenteric aspect of the ileum, proximal to the ileocecal valve. It has an independent blood supply from a remnant of the vitelline artery, a branch of the superior mesenteric artery (1-3).

The diagnosis of a MD is often incidental at laparotomy or laparoscopy and its prevalence is only approximated at 2%. In autopsy studies, 0.14%-4.5% of cadavers contained a MD (1). In a large series of 1476 cases at the Mayo Clinic, Park et al. (2) reported the most common presentations of symptomatic MD in adults to be bleeding (38%), obstruction (34%), diverticulitis (28%), and perforation (10%). Most tumors in MD are found incidentally intraoperatively.

Primary neoplasms are found in about 1% of all MDs; only a minority are malignant. Carcinoid tumors are the most common (33%-44%), followed by leiomyosarcoma (18-25%) and adenocarcinoma (12-16%). Tumors are reported to occur in 0.5% and 3.2% of symptomatic MDs, and of these, 12% of tumors are GISTs (3,4).

GIST accounts for 0.1% and 3% of all gastrointestinal neoplasms, most commonly occurring in the stomach or small bowel (5). Small bowel GISTs have a range of presenting features, including abdominal pain, an abdominal mass, gastrointestinal bleeding, small bowel obstruction, weight loss, fever, abscess, or perforation (6,7). Most symptomatic patients present with tumors larger than 5 cm in maximal dimension (4). As in our case, radiographic presentations of GISTs are usually large,
typically are exophytic in relation to the bowel, and may demonstrate necrosis and/or hemorrhage; lymphatic spread is uncommon (7).

Surgical resection is the mainstay of GIST. In unresectable and/or metastatic tumors, CD117-positive GISTs are treated with imatinib mesylate (8,9). In our case, since the tumor was completely resected and there was no recurrence during the follow-up period, we did not need to use that adjuvant treatment.

Microscopically, GIST cell morphology is usually spindle-shaped (70%), but some GISTs consist of rounded cells (epithelioid type, 20%) or a mixture, but they can also be pleomorphic (8). Most (95%) GISTs stain positively in immunostaining for the KIT protein (the CD117 antigen, an epitope of the KIT tyrosine kinase), exhibiting a diffuse, local or mixed staining pattern (10). CD34 expression is not specific for GIST, because it can also be noted in desmoid tumors, and approximately 70%-80% of GIST lesions are positive for CD34 (10). GISTs may stain positively for SMA (30–40%), but they are usually negative (95%) for S-100 (a neural cell marker) and for desmin (98%, an intermediate filament protein typical of muscle) on immunostaining.

The most reliable prognostic factors are the size of the primary tumor and the mitotic index, which measures proliferative activity of the cells (9,10). Other prognostic factors are specific histologic subtypes (epithelioid vs. spindle cell), the degree of cellular pleomorphism and age of the patient (4). Tumor Ki-67 antigen expression in immunohistochemistry may be comparable or superior to the mitotic count in prognostication (4).

In conclusion, tumors arising from MD are uncommon. When no radiological assistance is provided, the complaints of the patient presenting with intraabdominal mass will not assist the surgeon. Despite being uncommon, MD and tumors should be considered in the differential diagnosis of intestinal obstruction and intraabdominal masses.

REFERENCES