

CASE REPORTS

PERFORATED GIST OF MECKEL'S DIVERTICULUM

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Meckel diverticulum is the most common congenital abnormality of gastrointestinal track (2-4%), however complications occur rarely (4-16%). We describe a case of 63- years old female presenting simultaneously two serious complications of Meckel diverticulum. Operated patient was diagnosed with perforated tumor of Meckel diverticulum. Segmental resection of small bowel including tumor was performed. Pathology examination revealed gastrointestinal stromal tumor (GIST) in Meckel diverticulum. No significant malignancy risk factors were found (low mitotic count). Consequently, computed tomography periodic surveillance was implemented.

We report the possibility of simultaneous presentation of two serious complications of Meckel diverticulum. Tumors of Meckel diverticulum may mimic other abdominal pathologies and thus, they should be considered in differential diagnosis of abdominal tumors.

Key words: GIST, Meckel diverticulum, perforation

Meckel's diverticulum is the most frequently occurring congenital defect of the gastrointestinal tract (2-4%) (1). However, complications develop in 4-16% of cases only. Ymaguchi et al. has reported 600 cases of Meckel's diverticulum, with complications found in 287 patients, among whom 36.5% manifested obstruction symptoms, with obstruction caused by intussusception in 13.7% of them, 12.7% suffered from mucositis with perforation, 11.8% – from haemorrhaging, 3.2% were diagnosed with diverticular neoplasm, and 1.7% patients – with a fistula (2). Meckel's diverticulum perforation usually accounts for 7.3-14% of all complications associated with this defect (3, 4). In most cases, the cause of perforation is ulceration of ectopic gastric tissue (5), while in extremely rare cases – tumours. In the past 30 years, there have been reported only 14 cases of Meckel's diverticulum perforation associated with tumour presence, and in 3 of them it was gastrointestinal stromal tumour (GIST) (4). This paper presents the first case report of

GIST causing Meckel's diverticulum perforation in Poland.

CASE REPORT

A female patient, aged 63, was referred to a hospital due to abdominal tumour. The patient reported atypical hypogastric pain, intensifying for the past several days, and lack of appetite with concomitant fever up to 38.5°C. The outpatient ultrasound examination revealed a tumour of heterogeneous echogenicity, of 12 x 10 cm, localised above the uterus, and a small amount of fluid in the lesser pelvis. The picture was interpreted as a dermoid ovarian cyst. In history, the patient did not report any previous health complaints. The laboratory tests performed on admittal revealed elevated inflammatory markers (WBCs: 12.9 K/ μ l, CRP: 118.8 mg/l, ESR: 92.4 mm/h). On physical examination, extensive abdominal pain was found, most intense at the region of

detectable hypogastric tumour, mainly on the right side. No symptoms of peritonitis were observed.

The patient was qualified for surgery. Perioperatively, there was found an extensive tumour of 14 cm in diameter, originating from Meckel's diverticular wall, localised in the ileum, approx. 80 cm before the ileocaecal valve (fig. 1). The diverticular stalk was narrow (1.5 cm), of 3 cm in length. The tumour was localised in the lesser pelvis, posteriorly to the uterus. Tumour perforation was found, of 2 cm, near its apex, covered by a fragment of greater omentum. Small amount of turbid fluid was present in the rectouterine pouch. Segmental small bowel resection was performed, with Meckel's diverticulectomy and tumorectomy, with 10-cm margins in the intestine which was subsequently anastomosed.

Postoperatively, the specimen was cut and an encapsulated lesion with haemorrhagic gelatinous contents, necrotic tissues and pus inside was found. The tumour wall had the form of solid whitish infiltration, with extensive vascularisation, of 2 cm in thickness at the base. The postoperative course was uncomplicated. The patient was discharged home in a good general condition on day 7 post surgery.

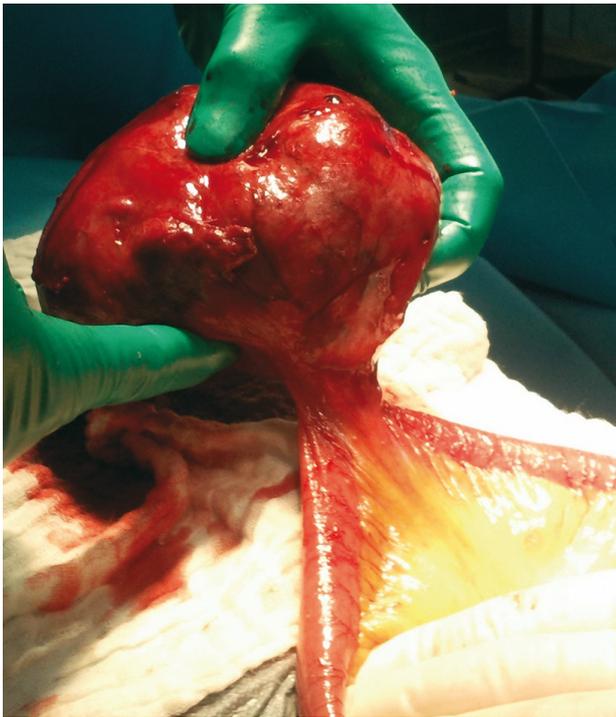


Fig. 1. GIST of Meckel's diverticulum. Perforation site is located on the other side of the tumor

The results of histopathological examination describe gastrointestinal stromal tumour (GIST) of Meckel's diverticulum (fig. 2). Mitotic activity: 0/50 HPF. The neoplastic cells exhibited positive immunorexpression of CD117 (+++) and CD34(-) (fig. 3). The lesion was excised whole. A single lymph node with inflammatory follicular proliferation was found in the mesenteric stump. The GIST advancement was assessed as pT3N0, stage II as per the stage grouping for small-intestinal GIST, WHO 2010, AFIP prognostic group 3a.

Due to the tumour size and GIST perforation found perioperatively, the patient remains under permanent oncological supervision for the purposes of assessing the need for potential

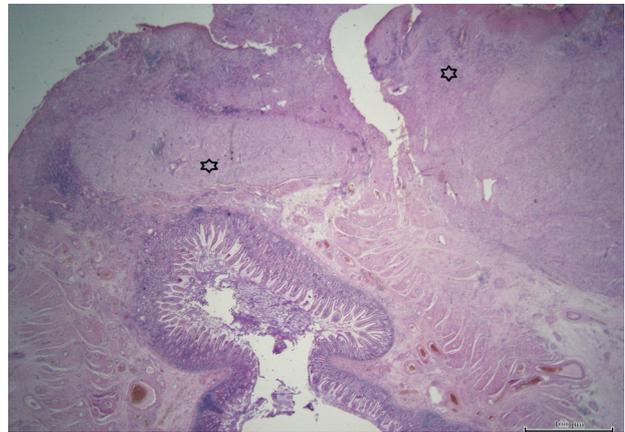


Fig. 2. Tumor mass adjacent to the Meckel's diverticular mucosa (tumour zone is indicated by asterisks). (Specimen prepared and diagnosis made at the Zakład Opieki Zdrowotnej Olympus Consilio facility in Łódź)

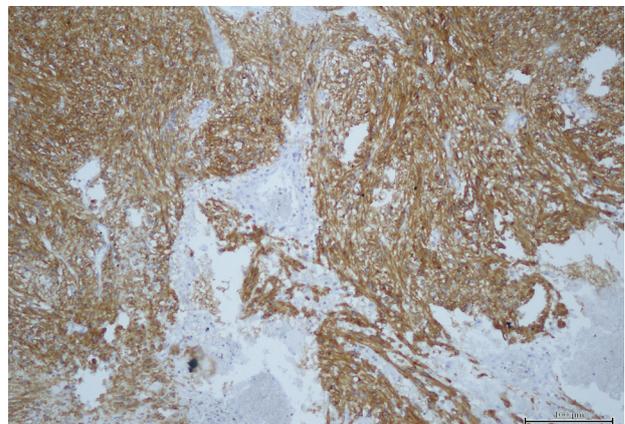


Fig. 3. CD117 – brown staining indicates the presence of GIST-specific protein in tumor cells. (Specimen prepared and diagnosis made at the Zakład Opieki Zdrowotnej Olympus Consilio facility in Łódź)

adjuvant treatment. A follow-up abdominal computed tomography scan was performed three weeks post surgery, and did not reveal the presence of distant metastases or other pathologies. The results of a subsequent CT scan, performed six months post surgery, were analogical. The patient remains in a good general condition, with no signs of disease recurrence or dissemination.

DISCUSSION

Only 10% of Meckel's diverticulum cases are diagnosed preoperatively (5). This is because the condition may mimic other complaints, such as the peptic ulcer disease, biliary colic and diverticulosis. However, most often, appendicitis is incorrectly suspected (6). Tumours are reported in only 0.5-3.2% of Meckel's diverticulum patients (5). The literature review performed by Hager has determined that in the majority of cases the lesions are of carcinoid nature (31.5%), while less often: leiomyosarcoma (25.5%), adenocarcinoma (11.4%) and leiomyoma (9.4%) (4). Malignant lesions have been found in 77% of Meckel's diverticulum patients. The symptoms accompanying Meckel's diverticulum are present three times more often in males than in females, and usually manifest at the age of approx. 30 (7).

GIST-type lesions account for 0.1-3% of all gastrointestinal neoplasms (8). The most common symptoms associated with the presence of GIST focus localised in the small bowel are mainly pain (74%), incorrect body weight (72%), gastrointestinal haemorrhaging (44%), mechanical intestinal obstruction (44%), body weight loss (16%), fever or abscess (14%) and dysuric symptoms (12%). The perforation of GIST lesion and the resulting peritonitis occur in 8% of tumours of this type localised in the small intestine (9, 4). The tumour growth leads to the development of central necrosis inside the lesion, and in consequence to the creation of a fistula between the diverticular lumen and the tumour body (10). Less commonly, a fistula with the peritoneal cavity is created.

The preoperative diagnosis of Meckel's diverticular tumour is hindered due to the limited symptoms accompanying this condition. The use of radiological examinations with contrast agent administration, endoscopic and ultrasound examinations, computed tomogra-

phy or mesenteric angiography may prove useful in diagnosing such lesions (6). However, the making of correct diagnosis preoperatively is rare. An abdominal ultrasound examination can reveal the presence of a tumour, while a computed tomography scan can show an asymmetrical focal thickening of intestinal wall, similarly as in the case of inflammatory lesions in the bowel (e.g. Crohn's disease or acute appendicitis) (11). Angiography can visualise the wall hypervascularisation and the system of vessels supplying blood (4). Similarly as in the cases of other small bowel tumours, the preoperative diagnosis of GIST at this localisation is difficult and is usually made during laparotomy and confirmed by histopathology on the excised material (12).

In cases where no metastatic foci are found in preoperative examinations or during surgery, it is not possible to determine definitely the tumour nature (10). Therefore, it should be treated as a potentially malignant lesion, and the tumour should be excised with an intestinal margin of at least 10 cm and with accompanying lymphadenectomy.

Mrowiec et al., in the performed multivariate analysis, distinguished the following three factors indicative of poor prognosis in GIST: coexistence with other neoplasms, male gender and high mitotic index (at least 10 mitoses per 50 HPF) (13). However, currently, the proven and generally accepted risk factors include the tumour size and mitotic index. Gastric GIST up to 5 cm and small bowel GIST up to 2 cm, of low mitotic index and no necrosis, are usually benign. Tumours exceeding 5 cm and with low number of mitoses are considered to be of uncertain prognosis. On the other hand, large lesions, over 10 cm in diameter, may cause recurrence and metastases, and if having high mitotic index they are always classified as highly malignant (8).

Other risk factors are also taken into account. The tumour localisation in the small intestine is considered by some authors an important and unfavourable marker. Better therapeutic results are observed for lesions localised in the stomach. An additional unfavourable prognostic factor suggested by publications is a complication in the form of perforation or haemorrhaging from the tumour, requiring emergency surgery.

In the case presented in this paper, there was found a considerable-size tumour with

necrosis, which according to some authors might be an additional risk factor (14). Moreover, the lesion perforation could create further risk of increased likelihood of neoplastic cell dissemination to the peritoneal cavity. The overall 5-year survival in this disease is 46-60% for locally advanced lesions, 24% for perforated lesions, and 0% for lesions with multiple foci and distant metastases at diagnosis (8). The prognosis is dependent on the extent of performed resection. In total resection, survival reaches 42% over a 5-year follow up, while it is only 8% if total resection cannot be performed (9).

Due to the lack of GIST sensitivity to standard chemotherapy and radiation therapy, radical surgical excision of the lesions remains the treatment of choice in this disease. GIST-type lesions exhibit overexpression of c-kit tyrosine kinase receptor (KIT), also known as CD117. GIST is characterised by the presence of mutations in the KIT proto-oncogene (15). Imatinib, as a tyrosine kinase inhibitor, has potent action against the tumour tissue. The treatment with imatinib is registered for GIST

dissemination or non-resectional lesions. The use of imatinib may also be considered in patients at high risk of disease recurrence, who have undergone radical lesion resection. However, adjuvant treatment with imatinib remains controversial and no unambiguous treatment duration or drug dosage dependent on molecular tests have been established to date.

Due to the presence of malignancy risk factors, the patient remains under permanent oncological supervision for the purposes of assessing the potential need for application of adjuvant treatment based on imatinib.

CONCLUSIONS

Over the past 30 years, three cases of perforation of a histologically-confirmed gastrointestinal stromal tumour of Meckel's diverticulum have been reported. Meckel's diverticular tumours may mimic other pathologies of the abdomen, therefore they should be taken into consideration during the differential diagnostics of abdominal tumours.

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