CASE REPORT

UDC: 617.55::[616.34-007.272-02:616.383
DOI: 10.2298/VSP120118039B

Sclerosing mesenteritis as a rare cause of upper ileus

Sklerozirajući mezenteritis kao redak uzrok visokog ileusa

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Abstract

Introduction. Sclerosing mesenteritis is a rare pathological entity characterized by non-specific tumor-like expansion in mesentery. Accurate diagnosis of this disease is rarely made preoperatively. Surgery takes place in diagnosis, as well in treatment of the disease. We presented a case of sclerosing mesenteritis that affected the final portions of duodenum and initial part of jejunum with clinical picture of upper gastrointestinal obstruction. **Case report.** A 46-year-old man without previous medical history was presented with vomiting and loss of weight in the last 6 months. Due to suspicion of parapancreatic tumor by CT examination and clinical presentation of the disease, the patient underwent laparotomy. A mass infiltrated mesenteric root, initial part of superior mesenteric artery, the fourth duodenum portion and the ligament of Treitz, while the stomach and duodenum were dilatated. The intraoperative biopsy indicated a benign process. The mass was reduced with desobstruction of the duodenum. Definitively, histopathological finding showed fibromatosis in different phases of activity. Postoperative course passed without complications. The patient continued to receive an immunosuppressive drug therapy. After a 6-month treatment the patient showed no gastrointestinal problems. **Conclusion.** Sclerosing mesenteritis that affects the duodenum and the proximal part of the jejunum with subacute upper gastrointestinal obstruction is an extremely rare condition. In the presented case a surgical procedure was necessary for marking the diagnosis and treatment as well.

Key words:
panniculitis, peritoneal; intestinal obstruction; diagnosis; surgical procedures, operative; treatment outcome.

Apstrakt

Uvod. Sklerozirajući mezenteritis je retki patološki entitet koji karakteriše nespecifičnu infiltracija u predelu mezenterijuma, nalik tumoru. Tačna dijagnoza ove bolsti retko se postavlja preoperativno. U cilju dijagnoze, kao i terapije, hirurgija zauzima značajno mesto. Prikazali smo bolesnika sa sklerozirajućim mezenteritisom sa zahvatanjem završnih delova dvanaestopalačnog creva i početnog dela jejunuma koji je imao kliničku sliku opstrukcije gornjeg gastrointestinalnog trakta. **Prikaz bolesnika.** Prethodno zdrav muškarac, star 46 godina, javio se zbog povraćanja i gubitka telesne mase u poslednjih šest meseci. Klinička slika i nalaz na CT pregledu abdomena, izazvali su sumnju na parapankreasni tumor, te je bolesnik operisan. Nađena je masa koja je infiltrisala koren mezenterijuma, početne delove gornje mezenterične arterije, četvrtu porciju dvanaestopalačnog creva i Treitz-ov ligament, sa dilatacijom dvanaestopalačnog creva i želuca. Intraoperativna biopsija promene ukazivala je na to da se radilo o benignom procesu. Masa je redukovana sa dezopstrukcijom dvanaestopalačnog creva. Definitivni histopatološki nalaz pokazao je da se radilo o fibromatozi u različitim fazama aktivnosti. Postoperativni tok protekao je uredno, bez komplikacija. Nastavljena je imunosupresivna terapija, a na kontrolnom pregledu nakon šest meseci bolesnik nije imao gastrointestinalne tegobe. **Zaključak.** Sklerozirajući mezenteritis sa zahvatanjem dvanaestopalačnog creva i proksimalnog dela jejunuma sa subakutnom opstrukcijom gornjeg gastrointestinalnog trakta je izuzetno retko stanje. Kod prikazanog bolesnika hirurška intervencija bila je neophodna za postavljanje dijagnoze, ali i kao terapijska procedura.

Ključne reči:
paniculitis, peritonealni; creva, opstrukcija; dijagnoza; hirurgija, operativne procedure; lečenje, ishod.

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Introduction

Sclerosing mesenteritis (SM) (or retractile mesenteritis) is a rare, benign and chronic fibrosing disease with inflammatory etiology of unknown origin, which affects the mesentery of the small bowel. Rarely, the mesentery of the transversal colon, peripancreatic region, omentum, retroperitoneum or the pelvic region can be affected, as well [1]. Three different histopathological changes are described in this process which include fat tissue necrosis, chronic nonspecific inflammation and fibrosis [2, 3]. Due to a very different course of the disease many different names for SM were used such as: mesenteric lipodistrophy, retractile or liposclerotic mesenteritis, mesenteritic Weber-Christian disease, xantogranulomatosis mesentiritis, mesenteric lypogranuloma and system nodular panniculitis [3]. If inflammation or fat tissue necrosis are predominant features, the disease is considered to be mesenteric panniculitis; if otherwise fibrosis with retraction is predominant feature the disease is called retractile mesentiritis. However, the presence of fibrosis in any degree, makes SM the most accurate term in a large number of cases [3, 4].

The disease more commonly affects middle-aged male adults [1]. Due to different atypical and nonspecific manifestations of the disease (abdominal pain, loss of weight, intestinal obstruction, fever, chylous ascites, palpable abdominal mass, constipation or diarrhea), preoperative diagnosis of SM is difficult in most cases [3–7]. Diagnosing this disease is complicated, posing a great problem and a challenge for radiologists, gastroenterologists and surgeons, even for pathologists who encounter this disease very rarely, with only 300 cases described in the literature so far [8]. In order to avoid misdiagnosis, one should think about this disease, even so the finall diagnosis demands biopsy and histopathological examination. With an no clearly defined treatment modalities of SM, surgery can take place in diagnosis, as well as in treatment of the disease [1, 3].

We reported a patient presented with an upper bowel obstruction caused by a retroperitoneal mass, which seemed to be pancreatic or parapancreatic tumor, and turned out to be SM on histopathological examination.

Case report

A 46-year-old male was admitted to our institution with vomiting and weight loss (7 kg for a month and 15 kg in a 6-month period). Problems started in the last six months with dyspepsia, anorexia and occasional pain in epigastrium, which in time grew to be stronger. For instance, vomiting was deteriorating and led to anorexia in the last l5 days. During admission the patient had light pain in the epigastrium with a palpable mass in that region. All laboratory parameters including tumor markers were in physiological ranges, except for the proteins whose value was 53 g/L.

Esophagogastroduodenoscopy (EGDS) showed dilatation of the bulb and the second duodenal portion with extraluminal compression in the region of the second duodenal knee, which was almost completely narrowing it's lumen. Endoscope ultrasound (EUS) showed heterogeneous echo change with hypoechogenic fields 4 cm in diameter in the region of Treitz ligament next to the duodenal wall which was infiltrated. X-ray of gastroduoduodenum showed almost complete obstruction of the lumen of the third and the fourth duodenal portion with the preserved mucosa relief and a significant dilatation of the proximal duodenal segment and moderate gastrectasis (Figure 1). Multislice computed tomography (MSCT) of the abdomen showed tumor formation bordered by the wall of the third and the fourth duodenal portion extended to the ligament of Treitz with thickening of the bowel wall, enlarged lymph nodes around superior mesenteric artery (SMA) and dilatation of the proximal segment of the duodenum and the pylorus (Figure 2).

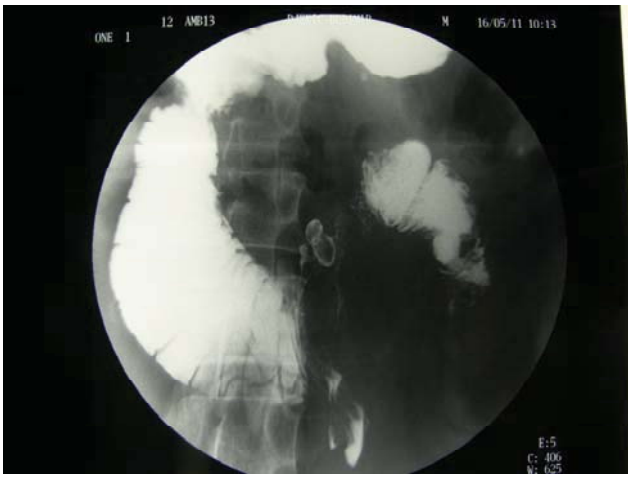

Fig. 1 – X-ray of gastroduodenum – almost a complete obstruction of the duodenal lumen with proximal duodenal dilatation

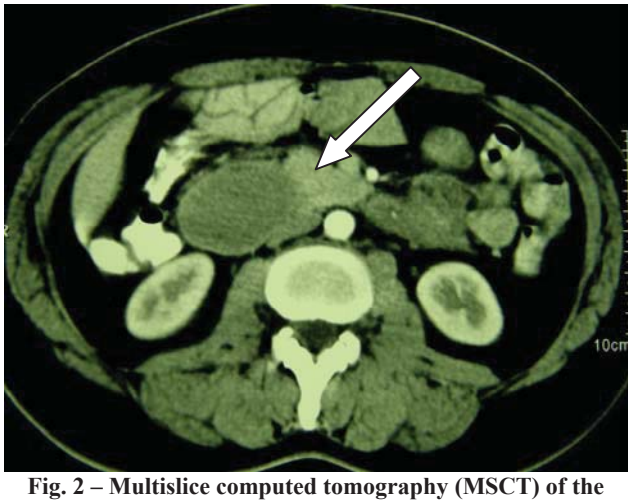

Fig. 2 – Multislice computed tomography (MSCT) of the abdomen – a tumor formation in pancreatic and parapancreatic region with dilatation of the second duodenal portion

The patient was presented on a meeting of gastroenterologists, surgeons and radiologists. They decided that surgical treatment should be applied, since the tumor formation which led to almost complete obstruction of the duodenum

and ileus, was the cause of the patient's condition. After short preparations, the patient underwent laparotomy, and retroperitoneal tumor was found with a remarkably firm consistency, unclearly bordered, which infiltrated mesenteric root, initial part of SMA, the fourth duodenal portion and the ligament of Treitz. The duodenum and almost a complete stomach were dilatated. A tumor formation was retracting proximal part of jejunum and pulled initial part of the descending colon. Using the transgastrocolic approach, we performed meticulous preparation of the mesenteric root and SMA with preparation and dissection of tumor tissue. During dissection a couple of excision bioptats were taken for the intraoperative histopathological examination and showed no malign cells. The operation was finished with a significant tumor reduction and duodenal desobstruction. Postoperative course was with no complications. The patient was introduced to *per os* food intake on the 3rd postoperative day with intact intestinal passage. The definitive histopathological finding showed fibromatosis in different phases of activity with a small degree of fat necrosis (Figures 3–5).

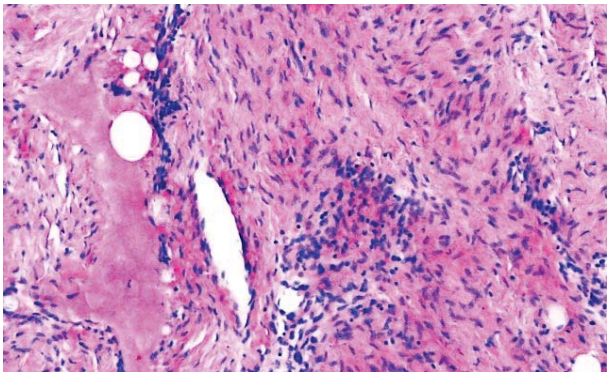

Fig. 3 – Proliferation of the cellular connective tissue with vascular compartments and bar of mononuclear inflammatory infiltrate (HE, ×40)

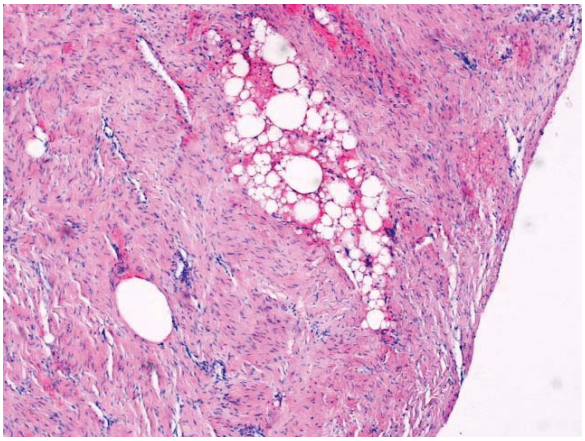

Fig. 4 – The islands of a mature fat tissue with fields of steatonecrosis surrounded with the cellular and acellular connective tissue (HE, ×10)

Seven days after the surgery MSCT angiography of the abdomen showed a significantly lower level of fibrotic mass compared with the period before the operation, without duodenal obstruction (Figure 6) and the normal trunk of SMA (Figure 7). After a full recovery of the patient, the same physicians, including pathologists, decided to start treatment with oral methotrexate and prednisone. After a six-month treatment the patient had no gastrointestinal problems. The next examination with radiological assessment was scheduled in 6 months.

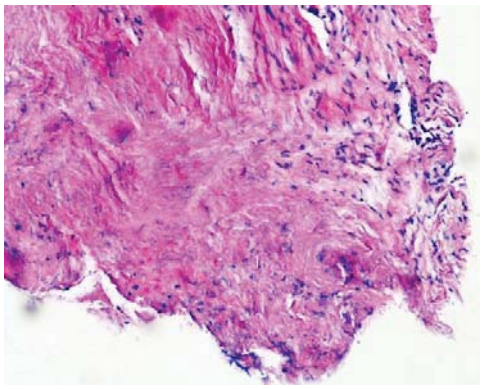

Fig. 5 – Proliferation of the acellular connective tissue, dezmoid like (HE, ×40)

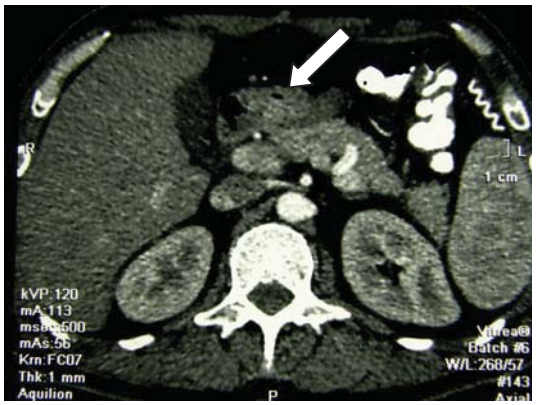

Fig. 6 – Postoperative multislice computed tomography (MSCT) – lower level of a fibrotic mass without duodenal dilatation

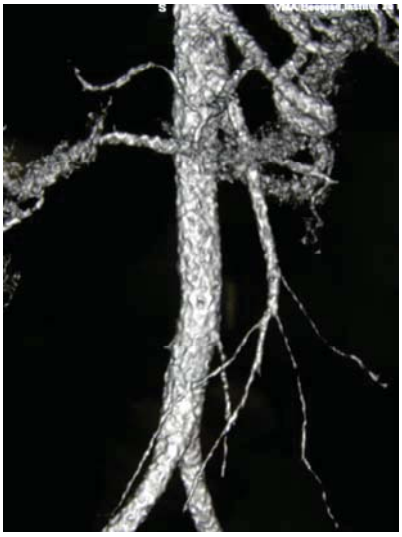

Fig. 7 – Postoperative multislice computed tomography (MSCT) angiography – the normal trunk of superior mesenteric artery

Discussion

SM presents a rare disease of unknown etiology firstly described by Jura [9] in 1921. Until today the literature has described about 300 cases [8]. It presents nonspecific tumor-like expansion in the mesentery, characterized with a different grade of chronic inflammation, fibrosis and fat necrosis [2, 3]. Various names were used to describe this disease focusing on histopathological features: fibrosis (retractile mesenteritis), inflammation (mesenteric panniculitis) or fat tissue necrosis (mesenteric lipodistrophy). It was acknowledged that these states represent only different histological types of the same clinical entity known as SM, which is being used as the most appropriate term for describing this disease [10].

SM can be associated with an autoimmune diseases, mesenteric ischemia, cancers (especially lymphoma), tuberculosis, lymphadenitis, previous abdominal surgeries and trauma [5]. It is by two times more common in men, white race, while very rare in children [11, 12].

Preoperative diagnosis of SM is usually difficult and rarely exact [3, 5–8]. The most common symptoms are abdominal pain and vomiting, while constipation, diarrhea and hematochezia are less common [5, 6, 13]. In around 50% of patients there is palpable abdominal mass[14]. Symptoms of acute or subacute upper obstruction of the small intestine are rare, but nevertheless described in the literature [5]. Laboratory findings are usually normal, although normocytic anemia may be present [15]. Radiological examinations can be helpful in reaching the correct diagnosis. X-rays with contrast can show different stages and levels of obstruction of the intestinal tract. In case of fibrosis predominance, the level of obstruction is in the region of jejunum, ileum, left or right colon, very rarely in the duodenal region [14]. Lumen obstruction is mainly partial and very rarely complete or almost complete [16–18]. Preservation of the bowel mucosa during the contrast examinations is crucial in differentiation between SM and cancers [13, 18–20]. MSCT can show tumor mass in the mesentery with fat tissue density, which surrounds blood vessels and suppresses the bowel without any signs of it's infiltration [15, 17]. If the fibrosis is predominant in the mesentery, MSCT scan may suggest a malign tumor of the connective tissue or pancreatic or parapancreatic tumor [11], just like in our patient. CT scan is an important diagnostic procedure when the localization of the disease is in the region of small bowel mesentery. Two CT findings are considered more specific for the diagnosis of SM [12]: a) the presence of the tumour pseudo-capsule, which is a hyperattenuated stripe surrounding the mass in the mesentery of the small bowel (this is seen in 60% of cases); b) the "fat ring" – sign of hypodense fatty halo surrounding mesenteric nodules and vessels. This is seen in up to 75% of cases. It should be emphasized that if the mass is localized in the region of intestinal loops closer to mesenteric side, it suggests SM. On the other hand, if the mass is located in the peripancreatic region, cancer of pancreas or the tumor of non-pancreatic origin is more probable [1, 19]. Differentiation of SM and sarcoma presents a great diagnostic problem. In that case an open surgical or laparoscopy exploration with biopsy should be done [19–22]. Only in few so far described cases the diagnosis of SM was made before the surgery [3, 5–8, 10, 23]. However, histopathological finding is necessary to confirm diagnosis [16].

There is no specific treatment or the treatment protocols for SM. In the study from the Mayo Clinic which included 92 patients with any form of SM and different treatment modalities, the results suggested that symptomatic patients might benefit from medical therapy, particularly tamoxifen and prednisone combination treatment. Only 10% of patients respond to surgery alone, and 20% to additional medical treatment after surgery [1]. Also, there have been reports on the response to antibiotics [24], irradiation [25], and cyclophosphamide [26]. Some literature data indicated that SM may regress spontaneously after laparotomy (especially in cases of fat necrosis predominance) [4, 27]. If chronic inflammation predominates various immunosuppressive therapies are suggested [1, 26]. In cases of bowel obstruction, partial resection, bypass, and colostomy may be necessary [10]. It is suggested to reserve surgery for unsuccessful medical treatment and complications of SM [28], but in cases when the diagnosis cannot be established with certainty and/or when a patient's clinical condition requires emergency treatment, surgery is indicated. The complications of SM requiring surgery are: shortening and retraction of the mesentery with compression of the mesenteric blood vessels, followed or not with intestinal ischemia and/or partial or complete intestinal obstruction [13, 19, 20]. Frequently, surgery is required for excision biopsy, but compression of the vessels will limit any further dissection of the mesentery for exposure or resection [27, 29].

In the available medical literature, in the Medline database, there is no case of SM with duodenal obstruction and clinical presentation of acute and/or subacute upper ileus [1, 5, 6, 8, 14, 18].

Our patient had no known exposure to toxic agents and did not use any medications. Abdominal trauma or previous abdominal surgery could not be implicated. Also, our patient had no previous history of any disease. Surgery was necessary to resolve duodenal obstruction and to confirm SM. In the same act the reduction of fibrotic tissue without resection of the gut was performed. Due to inconclusive preopertive and intraoperative diagnosis, and a significant reduction of fibrotic tissue followed by duodenal desobstruction, there was no need to do gastroenterostomy and expose the patient to additional risks. A postoperative prednisone and methotrexate administration resulted in the complete resolution of symptoms and pathological clinical findings.

Conclusion

SM can affect any part of the small intestine and colon, including the retroperitoneum in the form of different inflammatory diseases and abdominal tumors, with diverse clinical pictures. Etiology and pathogenesis are unknown. There are still no clear criteria in making certain diagnosis preoperatively or the defined treatment protocols for different forms of SM. It seems that the only certain are states in

which surgery is the primary option. Perennial follow-ups of treated patients, are necessary to answer to at least a few questions: Is surgery without resection of the small intestine sufficient for some forms of the disease? In which cases only medical treatment should be applied? What forms of SM demands specific type of therapy?

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Received on January 18, 2012.
Revised on March 14, 2012.
Accepted on April 10, 2012.
OnLine-First, October, 2012.