

PERFORATING SCLEROSING MESENTERITIS OF THE COLON MIMICKING A TUMOR: AUTOPSY CASE REPORT AND MINI-REVIEW

G. Gaggero¹, L. Valle², J. Ferro¹, D. Taietti³

¹Pathology Unit, IRCCS Istituto Giannina Gaslini – Genoa, Italy

²Pathology Unit, IRCCS Ospedale Policlinico San Martino – Genoa, Italy

³Pathology Unit, ASST Fatebenefratelli – Sacco, Milan, Italy

Abstract. Aim and subject. *Sclerosing mesenteritis is a rare non-tumoral disease that usually affects the small intestine. Several etiological hypotheses have been proposed (external or surgical trauma, autoimmunity, neoplasia). Clinical case description and main results.* We describe the autopsy case of a 66-year-old woman who presented to hospital unconscious with a palpable abdominal mass. The patient underwent abdominal CT, which revealed a mass involving the transverse colon and mesocolon with non-pathognomonic features, but with a strong suspicion of neoplasia. However, the patient died quickly, before a biopsy could be performed. The only prominent finding at autopsy was a transverse colonic wall mass associated with colic perforation and mucosal hyperplasia. Histology revealed sclerosing mesenteritis after a thorough microscopic and immunohistochemical investigation of possible differential diagnoses. **Conclusions.** There are three types of sclerosing mesenteritis, depending on the fibrous component, and the case described is an example of retractile mesenteritis (preponderant fibrosis). A mini-review of the Pubmed database shows that the different ways of referring to this disease make it difficult to search, but a total of 213 cases appear to be published so far. Overall, the location, clinical presentation and macroscopic findings of our case represent a true rarity.

Key words: forensic pathology, sclerosing mesenteritis, colon, abdominal mass

Corresponding author: Gabriele Gaggero, e-mail: gabriele.gaggero@outlook.it

ORCID: 0000-0001-9098-563X

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INTRODUCTION

An abdominal mass (AM) is an abnormal growth that may cause visible swelling and thus change the shape of the abdomen, or it may be detected only as a palpatory or radiologic finding; AM may be associated with weight gain or a cachectic state [1]. AMs can be caused by: injury, cysts (ovarian, renal, pancreatic), tumors (gastrointestinal, renal, adrenal, hepatic, pancreatic, gynecologic and soft tissue neoplasms, including those

of retroperitoneal origin) or other diseases (Crohn's disease, abdominal aortic aneurysm, pancreatic abscess, diverticulitis, hydronephrosis, hepatomegaly, splenomegaly, IgG4-related diseases, and sclerosing mesenteritis) [2-7].

CLINICAL CASE DESCRIPTION

A 66-year-old Caucasian woman was brought to the hospital unconscious and severely cachectic. The patient's vital signs showed accelerated pul-

sation, arterial hypotension, oliguria and normal O₂ saturation, while preliminary laboratory tests showed an anemic state with low haemoglobin and hematocrit (low B12 and low folate), hypoalbuminemia and elevated C-reactive protein. Preliminary investigations revealed a mesogastric mass on abdominal palpation, which was localized in the transverse colon on computed tomography (CT) and interpreted as neoplastic. Indeed, the CT scan showed focal thickening and retraction of the transverse mesocolon characterised by a dense “fatty stranding” appearance. The affected segment of the transverse colon showed a significant circumferential thickening of the wall and a mass-like intraluminal appearance, which led to a strong initial suspicion of a primary neoplasm of the colon. The mucosal enhancement pattern was homogeneous and followed the contour of the bowel wall, but the mass-like appearance and extent of mesenteric involvement led the radiologist to favor a neoplastic process. There was no evidence of colonic perforation on CT: the absence of free gas or fluid in the peritoneal cavity suggested that the integrity of the bowel wall remained intact despite the significant thickening and mass-like appearance. However, before further evaluation and/or treatment, the patient died within an hour.

Autopsy revealed a greenish peritoneal effusion and a perforated transverse colon, but buffered by loops

of small intestine (Figure 1a). On opening the colon, the perforation corresponded to a substenotic wall/ mucosal thickening of hyperplastic appearance (Figures 1b and 1c); the remaining colon and all other (abdominal, thoracic, and cranial) organs showed no significant macro/microscopic lesions.

Histology of the colic specimens showed fibrotic hypocellular thickening (Figure 1d), also with necrotic foci. Necrosis led to ulceration of the mucosal epithelial surface and mild perivisceritis.

The clinical and radiologic suspicion of a neoplastic lesion was ruled out by microscopy (lacking cellular atypia and/or mitosis) and immunohistochemistry: negativity for cytokeratins, CD117, DOG1, beta-catenin, and ALK excluded carcinoma, gastrointestinal stromal tumor (GIST), fibromatosis, and inflammatory myofibroblastic tumor.

Other hypotheses included idiopathic bowel disease, amyloidosis, endometriosis, IgG4-related disease, and an infectious cause, all of which were inconsistent with the observed microscopic findings (also negative for PAS, Grocott, Ziehl-Neelsen, and Congo red stains).

The final diagnosis was sclerosing mesenteritis (SM) of the colon with bowel perforation and abnormal mucosal hyperplasia associated with intraluminal bulging and bowel subocclusion.

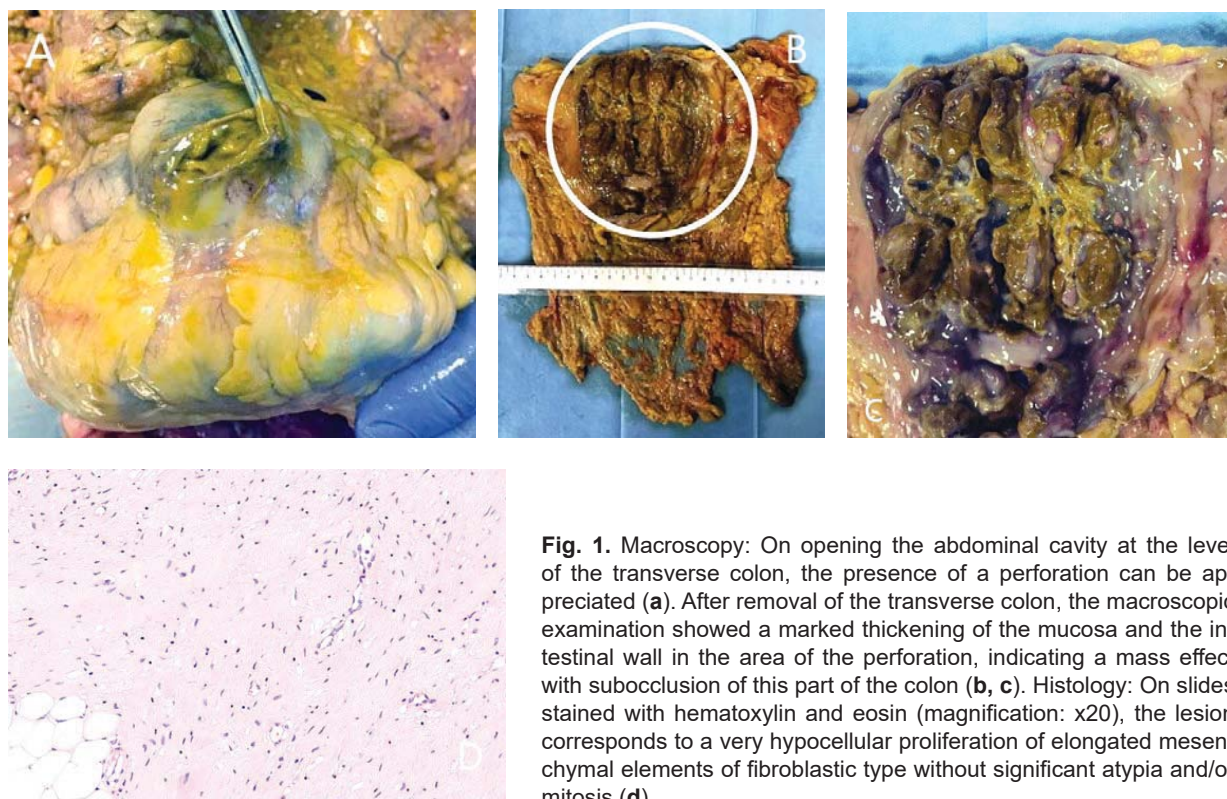


Fig. 1. Macroscopy: On opening the abdominal cavity at the level of the transverse colon, the presence of a perforation can be appreciated (a). After removal of the transverse colon, the macroscopic examination showed a marked thickening of the mucosa and the intestinal wall in the area of the perforation, indicating a mass effect with subocclusion of this part of the colon (b, c). Histology: On slides stained with hematoxylin and eosin (magnification: x20), the lesion corresponds to a very hypocellular proliferation of elongated mesenchymal elements of fibroblastic type without significant atypia and/or mitosis (d).

DISCUSSION

SM is a rare disease (1% prevalence) whose pathogenesis is still partially unknown (hypotheses proposed, but never definitively proven, include: previous abdominal trauma or surgery, autoimmune or infectious diseases, drugs, and malignancy) [8]. SM usually occurs in the mesentery/small bowel: its radiological features have been widely reported, but although radiology (CT) can help in the diagnosis, it is not always definitive, as in our case. On the other hand, there are very few endoscopic descriptions in the literature [9]. SM is rarely associated with perforation and even more rarely observed in the colon [10, 11]. Histologically, although there are no uniform diagnostic criteria worldwide, three microscopic patterns of SM are usually recognized: a) mesenteric lipodystrophy (predominant fat necrosis), b) mesenteric panniculitis (predominant chronic inflammation), c) retractile mesenteritis (predominant fibrosis) [8], and the microscopic findings led to the diagnosis of retractile mesenteritis in our case. A similar fibrotic pattern may be seen in recurrent diverticulosis/diverticulitis with perforation of a diverticulum but without massive peritonitis to produce chronic fibrotic pericolic damage. However, the complete macro/microscopic (and radiological) absence of diverticula rules out this hypothesis.

As a result of these different clinicopathological terminologies, the epidemiological data of SM are not clear in the literature. However, a mini-review of the Pubmed database alone was performed using a broad search string: ("mesenteritis" OR "sclerosing mesenteritis" OR "retractile mesenteritis" OR "mesenteric panniculitis" OR "mesenteric lipodystrophy" OR "mesenteric lipogranuloma" OR "fibrosing mesenteritis"). Selecting the systematic review option from the search results, we were surprised to find that there is only one systematic review on this topic, published in 2017 by Sharma P et al. [12], which describes just 192 established cases. A survey of articles from 2017 to April 2024 (again on Pubmed) reveals only: a monocentric miniseries describing 17 cases [13] and 4 case reports (one of which was fatal, like ours), making a total of 213 published cases [10-15].

SM can be treated pharmacologically (corticosteroids, tamoxifen, thalidomide and azathioprine) [5], but in case of intestinal perforation surgery is unavoidable. However, perforation does not always cause acute symptoms: in fact, in our case, it is conceivable that the colic perforation produced a non-acute clinical condition for a long time, even causing the patient's cachectic state and multiple organ

failure, leading to the unconscious state found at the time of hospitalization. Indeed, the patient presented with a constellation of signs and symptoms consistent with cachexia secondary to malabsorption and/or intestinal leakage. The vital/laboratory signs, although variable and influenced by many factors, provided valuable insights into the pathophysiology of the disease:

- Heart rate: the elevated heart rate observed in this patient is a common finding in cachexia. It is likely to be multifactorial, reflecting increased sympathetic tone, hypovolemia and underlying inflammatory processes;
- Blood pressure: the hypotension observed is consistent with the hypoalbuminemia and fluid loss associated with malabsorption/intestinal leakage. Decreased oncotic pressure and reduced intravascular volume contribute to the development of hypotension;
- Oliguria: reduced urine output indicates dehydration, a common complication of malabsorption/intestinal leakage due to fluid loss from the gastrointestinal tract;
- Oxygen saturation: the normal oxygen saturation in this patient suggests that there is no significant underlying respiratory compromise. However, it is important to note that chronic malnutrition can impair tissue oxygenation at the cellular level;
- Severe anaemia, often associated with malabsorption/intestinal leakage, can reduce the oxygen supply to the brain, causing fatigue, weakness and, in severe cases, loss of consciousness.

CONCLUSIONS

SM is a rare non-neoplastic disease with a still poorly understood etiology and little more than 200 confirmed cases in the literature, which must be distinguished clinically, radiologically and anatomopathologically (macroscopically and microscopically) from many other mass-forming entities in the abdomen. Histological and immunohistochemical analyses allow the exclusion of other pathologies that may be in the differential diagnosis and better define the subtype of SM involved, depending on the degree of fibrosis or inflammation present. Finally, considering that the data available indicate that the most frequent site of involvement in SM is the small intestine and that SM almost never leads to perforation and only exceptionally to death, the clinical case described here represents a true rarity.

Conflict of Interest Statement: The authors declare no conflicts of interest related to this work.

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Ethical statement: Since this was an autopsy case of a homeless person who arrived at the hospital unconscious and died unconscious, and since it was not possible in any way to find his family members, the consent for publication was requested from the Regional Ethics Committee (EC), which gave its approval (Liguria Region EC register number: 675/2021, approved 11/24/2021)

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