INDEX COPERNICUS INTERNATIONAL

Desmoid tumor of the mesentery in patient after restorative proctocolectomy as a result of familial adenomatous polyposis – case report

Magdalena Krauze-Kalinowska[1], Adam Nałęcz[2], Krzysztof Dębski[3], Marek Dobosz[1]

[1]Department of General and Gastrointestinal Surgery and Nutrition, Copernicus Sp. z o.o., Gdansk, Poland; Head: prof. dr hab. Marek Dobosz
[2]Department of Pathomorphology, Copernicus Sp. z o.o., Gdansk, Poland; Head: dr hab. Ewa Iżycka – Świeszewska
[3]Department of Radiology, Copernicus Sp. z o.o., Gdansk, Poland; Head: dr Maciej Markuszewski

Article history: Received: 30.03.2017 Accepted: 17.02.2018 Published: 30.06.2018

ABSTRACT: Desmoid (desmoid tumor) is a cytologically benign fibrous tumor that originates from musculoskeletal structures of the entire body [1]. The term „desmoid", first introduced by Muller in 1838, derives from the Greek word desmos, which means tendinous [2]. The etiopathogenesis of desmoid is not fully understood, most reports of publications regarding its treatment are based on individual case reports. The prognosis is good, and healing is achieved mainly through surgical excision of the lesion. The aim of the paper is to present a case of a 33-year-old woman with diagnosed familial polyposis in the colon, who has been identified with mesenteric desmoid.

KEYWORDS: desmoid, familial polyposis

AUTHOR'S OBSERVATIONS

The patient, 33 years old, burdened with familial polyposis of the large intestine, following laparoscopic pancolectomy with ileoproctostomy with J-pouch, was admitted to the Department of General Surgery of the Copernicus Hospital in scheduled mode for the removal of the abdominal tumor. In the preoperative abdominal CT scan, the diameter of the umbilical region and lower abdomen was changed to 17 cm, corresponding to the desmoid in relation to the interview and imaging (Figs. 1, 2, 3). On the basis of physical examination and imaging, the patient was qualified for surgical treatment. A laparotomy via median incision was performed, with the diagnosis of a giant tumor in the size of a child's head located in the mesentery of the jejunum, clustered with the bladder, causing partial obstruction of the intestine (Fig. 4). In addition, two smaller lesions the size of a tangerine were found in the mesentery of the ileum, clinging to the intestinal wall; furthermore, the fourth node the size of a hazelnut was located in the retroperitoneal space forward of the gonadal veins. The condition after reconstructive proctocolectomy with the formation of the „J" reservoir - the mesentery of the reservoir is twisted, without intestinal passage disorders. The tumor was dissected with a margin of unchanged mesenteric tissues while maintaining its blood supply, and then resected with a 20-cm long section of the jejunum coating the tumor (Fig. 5). Continuity of the gastrointestinal tract was restored by performing an end-to-end enterointestinal anastomosis with single PDS 4/0 sutures, single-layered. The remaining two desmoidal lesions were removed with fragments of the ileum, a total of 15 cm of intestine. Both resected sites were joined, as was the case with the first change. A small lesion in the retroperitoneal space was also removed. In histopathological and immunohistochemical studies of four tumors with intestinal fragments performed in the Copernicus Pathomorphology Department (study no. PAT-13440/2015), the following reactions were observed: positive nuclear beta-catenin reaction, negative S100, SMA, CD117, desmin, CD34, and Ki67 approx. 2% (Fig. 6, 7). The histological picture confirmed the diagnosis of multifocal mesenteric fibromatosis (desmoid tumor). No complications were obse-

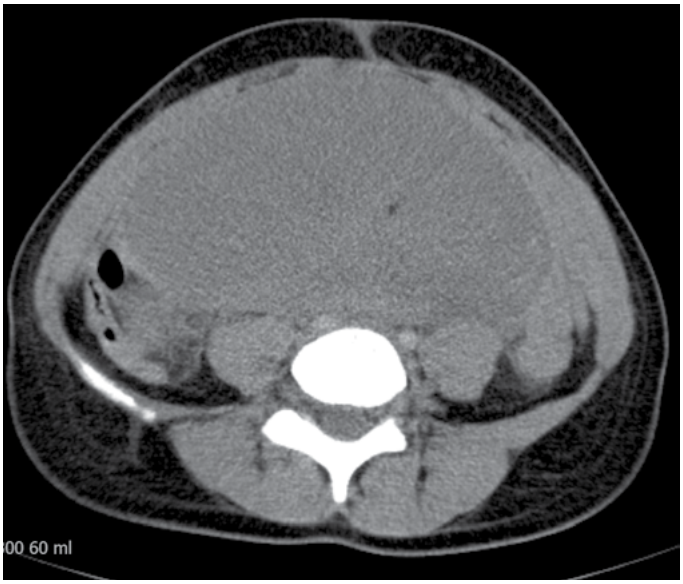

Fig. 1. Computed tomography of the abdominal cavity and small pelvis. Cross section.

rved in the postoperative period. The patient was discharged on the sixth postoperative day in good general condition, with proper healing of the wound. In control imaging examinations (computed tomography), performed for abdominal pain, half a year after hospitalization, a lesion in the abdomen with a diameter of up to 4 cm was found, most likely corresponding to recurrence (Fig. 8). The patient was qualified for surgical treatment once again. Laparotomy was performed with the observation of mesenteric tumor in the mid-jejunum, the size of a tangerine adjacent and fitting closely to the intestinal wall, infiltrating the mesenteric root. The jejunum is distended; however, the cause of sub-ileus was not the tumor, but a small twisting of the intestine running behind the mesentery, which in turn resulted from a previous twist of the intestinal reservoir combined with the rectum stump during the initial operation. The tumor was resected with a 20-cm long section of the jejunum (Fig. 9). Then, as a result of the potential danger of sub-ileus resulting from the passage of the ileum behind the me-

DOI: 10.5604/01.3001.0011.8171

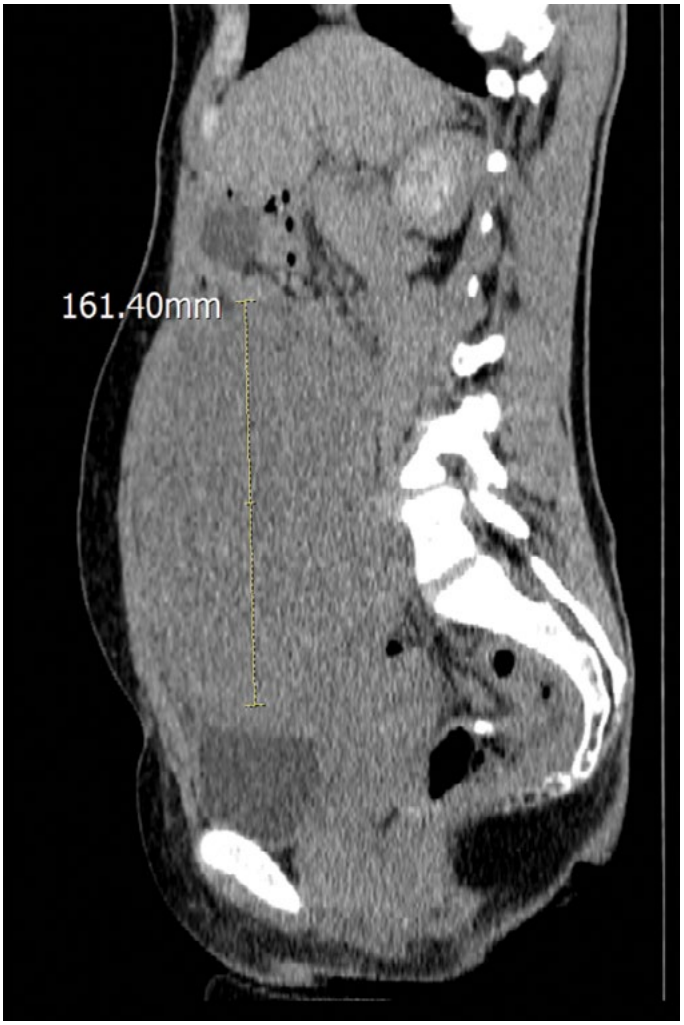

Fig. 2. Computed tomography of the abdominal cavity and small pelvis. Longitudinal-sagittal section.

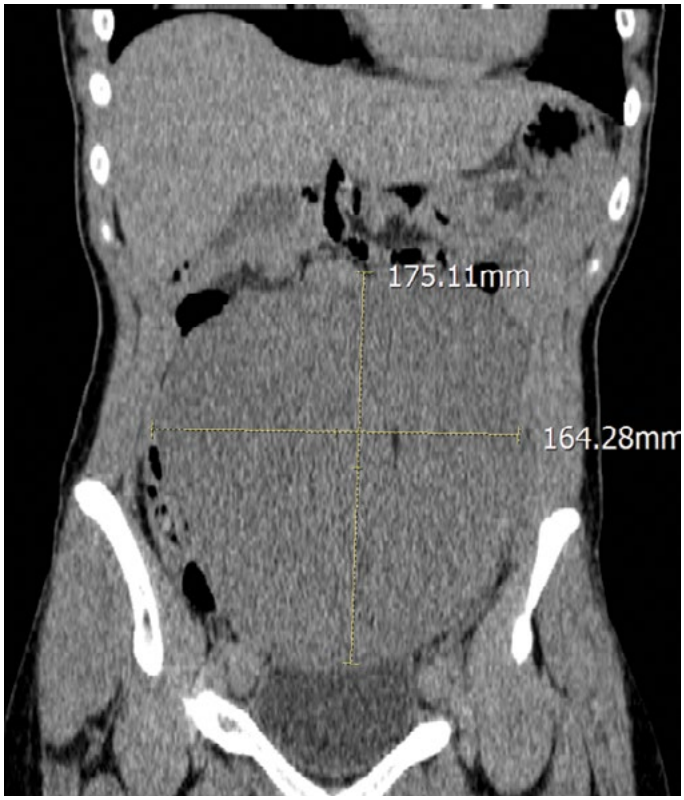

Fig. 3. Computed tomography of the abdominal cavity and small pelvis. Longitudinal-frontal section.

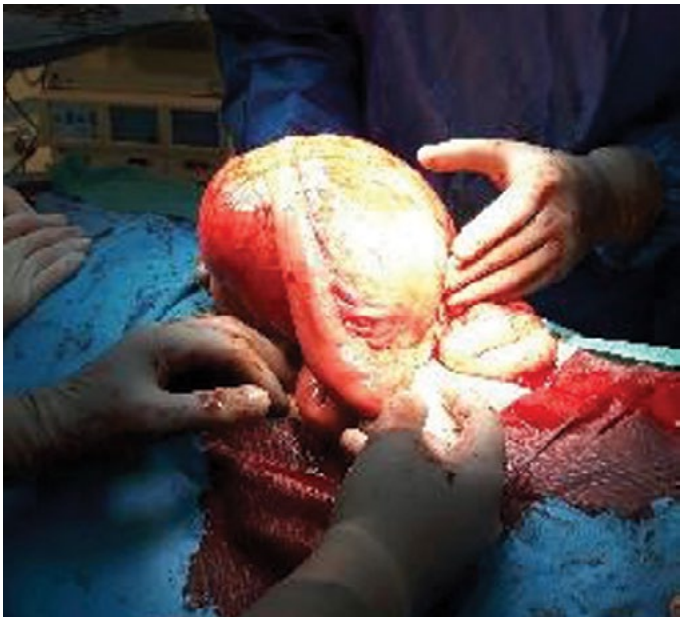

Fig. 4. Tumor in mesentery of the jejunum.

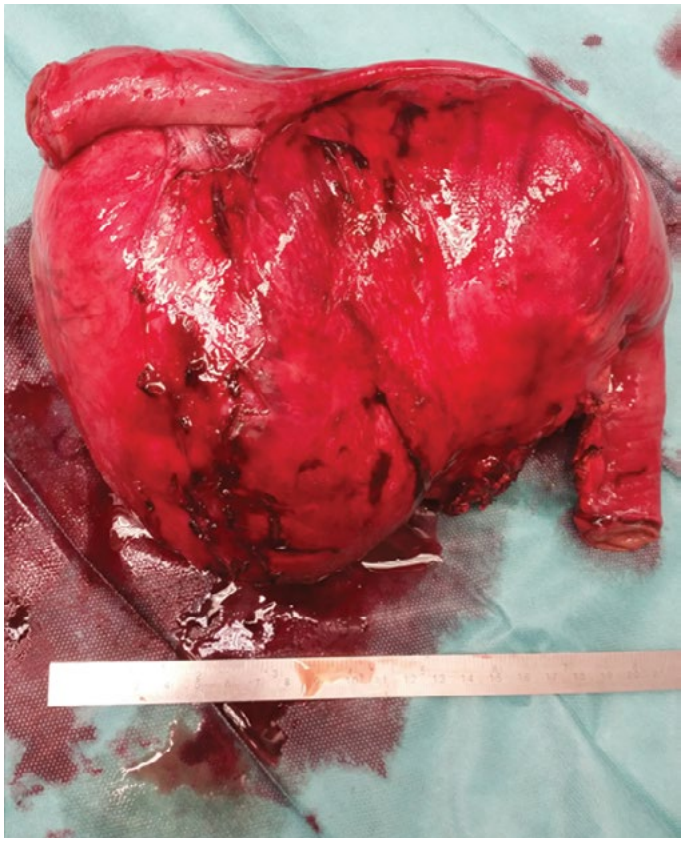

Fig. 5. Tumor with jejunum fragment.

sentery and the pressure that it was causing, a side-to-side anastomosis bypassing the ileum was performed. Postoperative period was uncomplicated. The patient was discharged on the 7th day following the procedure in a good general condition.

DISCUSSION

Desmoid (desmoid tumor) is a cytologically benign fibrous tumor that originates from musculoskeletal structures of the entire

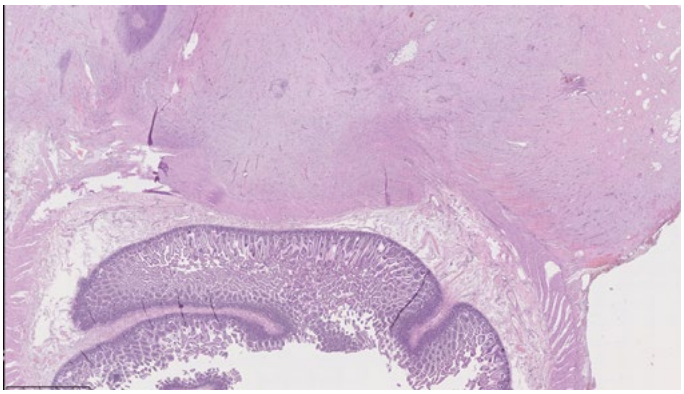

Fig. 6. Areas of tumor with spindle cell fibroblast proliferation.

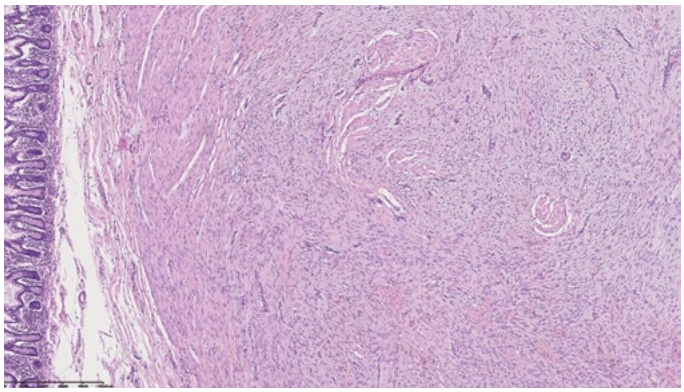

Fig. 7. Areas of tumor with spindle cell fibroblast proliferation.

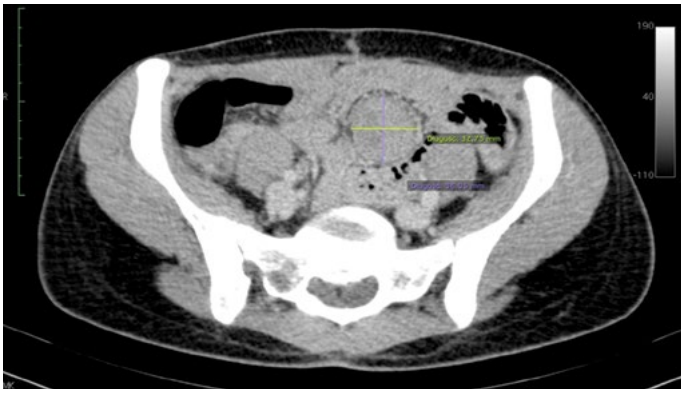

Fig. 8. Computed tomography of the abdominal cavity and small pelvis. Cross section.

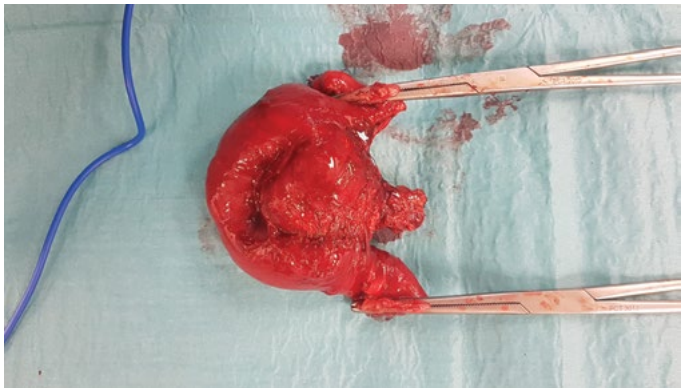

Fig. 9. Tumor with ileum fragment.

body [1]. Desmoid tumors are usually well-differentiated, do not metastasize, however they tend to infiltrate surrounding tissues. Generally, they constitute 0.03% of all cancers [3]. In the case of patients with familial polyposis, their incidence increases to about 13% [4]. Desmoid tumors, depending on the location, can be divided into lesions located in the abdomen, in the abdominal wall and intraabdominal [5]. In patients without familial polyposis, lesions are usually located to the outside of the abdomen, whereas in patients burdened with familial adenomatous polyposis, desmoids are in most cases localized intra-abdominally; they infiltrate the mesentery of the small intestine and occur after at least one surgical operation [6]. Desmoids usually affect young people. In his study of 1999, Church showed an average age of patients amounting to 31 [7]. Fibrous tumor is less common in men with FAP (8%) than in women (13%) [4]. Preoperative diagnostic imaging (computed tomography), presenting the presence of an oval lesion in the abdominal cavity, is not sufficient for an appropriate diagnosis. Due to the history of coexistence of familial polyposis, it can only suggest a fibrous tumor. Clinical diagnosis is based on histopathological and immunohistochemical studies. Macroscopically, desmoid has the form of a cohesive, well-organized tumor with a shiny, whitish, trabecular surface on cross-sections. The microscopic image shows spindle-cellular fibroblast proliferation, without necrotic foci, with moderate cytological atypia, with no presence of abnormal figures of the mitotic division (Fig. 4). The immunohistochemical profile of the lesion includes a positive nuclear reaction to beta-catenin, a negative reaction to the presence of S100 protein, SMA, CD117, desmin, CD34. In differential microscopic diagnosis, the following should be considered: nodular fasciitis, low-grade fibromyxoid sarcoma and fibrosarcoma [8]. Fibrous tumors do not form distant metastases, and the treatment standard is surgical removal with wide margins [9]. However, such treatment does not guarantee full recovery, desmoids have a high tendency to create local recurrences, which is confirmed by this case. There are several publications in the literature describing both neoadjuvant and adjuvanted desmoidal tumors. In 2007, Lev et al. presented in their work that a neoadjuvant approach, consisting of radiotherapy and systemic treatment, may be associated with improved results of patients with desmoid [10]. In 2010, De Camargo, comparing clinical effects of different regimens of fibrous tumors, presented superiority of the use of antiestrogens and patterns containing anthracycline over other chemotherapeutics [11]. Recent reports highlight the role of NSAIDs, including sulindac, in the treatment of desmoids. In 2016, Quast et al. described long-term results of treatment of fibrous tumors with sulindac and high doses of selective estrogen receptor modulators. During the conduct of a single-center study of 134 patients, they proved the therapy not only to be effective, but also showed that it is safe in the majority of patients, both those suffering from sporadic desmoid and desmoid occurring with familial polyps colitis [12]. At the same time, in 2016, Saito et al. conducted a multicenter retrospective cohort study in which they assessed the risk factors for the development of desmoid following proctocolectomy due to FAP in patients of the Japanese population. In their study, they showed that the female gender and the surgical procedure itself are independent risk factors [13]. In April 2017, Trufero et al. summarized all previous publications on available treatments of desmoids [14]. In spite of attempts to include more and more new methods, there are still no clear standards for the treatment of patients with desmoid tumors.

REFERENCES:

1. Mushtaq S, Bushra A. Case report of an intra-abdominal desmoid tumour presenting with bowel perforation. Mcgill J Med 2007 Jul; 10(2): 90-92.
2. 2. Müller J: Veber den Feinern Bau und die Formen der Krankhaftlichen Geschwulste. Berlin: G Reimer; 1838:80.
3. 3. Strassner J, Rascati S, PA-C. A desmoid tumor originating in the abdominal wall. Journal of the American Academy of Physician Assistants 2015 doi 10.1097/01.
4. 4. Szmidt J, Gruca Z, Krawczyk M, Kużdżał J, Lampe P. Podstawy Chirurgii. Medycyna Praktyczna, Kraków 2009; tom 1: 54, 827, tom 2: 1108.
5. 5. Allen P. W. The fibromatoses: A clinicopathologic classification based on 140 cases. Am J Surg Pathol 1977; 1: 255-270.
6. 6. Church JM. Desmoid tumors in familial adenomatous polyposis. Surg Oncol Clin N Am 1994; 3: 435-47.
7. 7. Church JM., McGannon, Ozuner G. The clinical course of intra-abdominal desmoid tumours in patient with familial adenomatous polyposis. Blackwell Science Ltd. Colorectal Disease 1999; I: 168-173.
8. 8. Gattuso P, Reddy VB, David O, Spitz DJ, Haber MH. Differential Diagnosis in Surgical Pathology, 2nd Edition, 2010; 898-900.
9. 9. Kasper B, Baumgarten C, Bonvalot S, Haas R, Haller F, Hohenberger P, Moreau G, van der Graaf W.T.A, Gronchi A on behalf of the Desmoid Working Group Management of sporadic desmoid-type fibromatosis: a European consensus approach based on patients' and professionals' expertise – a Sarcoma Patients EuroNet and European Organisation for Research and Treatment of Cancer/Soft Tissue and Bone Sarcoma Group initiative. European Journal of Cancer 2015; 51: 127–136.
10. 10. Lev D, Kotilingam D, Wei C, Ballo MT, Zagars GK, Pisters PWT, Lazar AA, Patel SR, Benjamin RS, Pollock RE. Optimizing Treatment of Desmoid Tumors. Journal of Clinical Oncology 2007; Vol. 25, Nr 13.
11. 11. VP de Camargo, ML Keohan, DR D'Adamo, CR Antonescu, MF Brennan, S Singer, LS Ahn, RG Maki. Clinical outcomes of systemic therapy for patients with deep fibromatosis (desmoid tumor). Cancer 2010 May 1'116(9):2258-65. Doi: 10.1002/cncr.25089.
12. 12. Quast DR, Schneider R, Burdzik E, Hoppe S, Möslein G. Long-term outcome of sporadic and FAP-associated desmoid tumors treated with high-dose selective estrogen receptor modulators and sulindac: a single-center long--term observational study in 134 patients. Familial Cancer 2016; 15:31-40. Doi 10.1007/s10689-015-9830-z.
13. 13. Saito Y, Hinoi T, Ueno H, Kobayashi H, Konishi T, Ishida F, Yamaguchi T, Inoue Y, Kanemitsu Y, Tomita N, Matsubara N, Komori K, Kotake K, Nagasaka T, Hasegawa H, Koyama M, Ohdan H, Watanabe T, Sugihara K, Ishida H. Risk Factors for the Development of Desmoid Tumor After Colectomy in Patients with Familial Adenomatous Polyposis: Multicenter Retrospective Cohort Study in Japan. Ann Surg Oncol 2016; 23:S559-S565. Doi 10.1245/s10434-016-5380-3.
14. 14. Trufera JM, Bernad IP, Ramón IT, Cubero JH, Cid RP. Desmoid-Type Fibromatosis: Who, When, and How to Treat. Curr. Treat. Options in Oncol. 2017; 18:29. Doi 10.1007/s11864-017-0474-0.

Word count: 1150 Page count: 4 Tables: – Figures: 9 References: 14

DOI: 10.5604/01.3001.0011.8171 Table of content: https://ppch.pl/issue/11207

Corresponding author: Magdalena Krauze-Kalinowska, Department of General and Gastrointestinal Surgery and Nutrition, Copernicus Sp. z o.o., Gdansk, Poland, e-mail: mkrauze@copernicus.gda.pl

Cite this article as: Krauze-Kalinowska M., Nałęcz A., Dębski K., Dobosz M.; Mesenteric desmoid tumor in the patient after restorative proctocolectomy due to familial adenomatous polyposis – a case report; Pol Przegl Chir 2018: 90 (3): 56- 59