Int Surg 2012;**97**:260–265

Mesenteric Histiocytosis or Only an Inflammatory Infiltration of Histiocytes? A Case Report From a Surgical Point of View

Florian Höhler[1], Karl-Heinz Dietl[1], Karel Novak[1], Joachim Giedl[2], Christian Paetzel[3]

[1]*Department of general, visceral and thoracic surgery, Klinikum Weiden, Weiden i.d. Obpf, Germany*

[2]*Institut of Pathology, Klinikum Weiden, Weiden i.d. Obpf, Germany*

[3]*Institut of Radiology, Klinikum Weiden, Weiden i.d. Obpf, Germany*

The mesenteric sclerosing processes are very rare tumors. There are only a few cases of mesenteric fibromatosis described in literature. A case of mesenteric histiocytosis or a mesenteric infiltration by histocytes as a reactive inflammatory process is not described in the surgical literature. Because of its clinical and macroscopic similarity to a fibromatosis or a reactive inflammatory process and a lack of articles in the literature on mesenteric histiocytosis we concentrated our research in literature on the mesenteric fibromatosis and its differential diagnosis.

Key words: Mesenteric histiocytosis – Mesenteric fibromatosis – Infiltration of histiocytes – Differential diagnosis

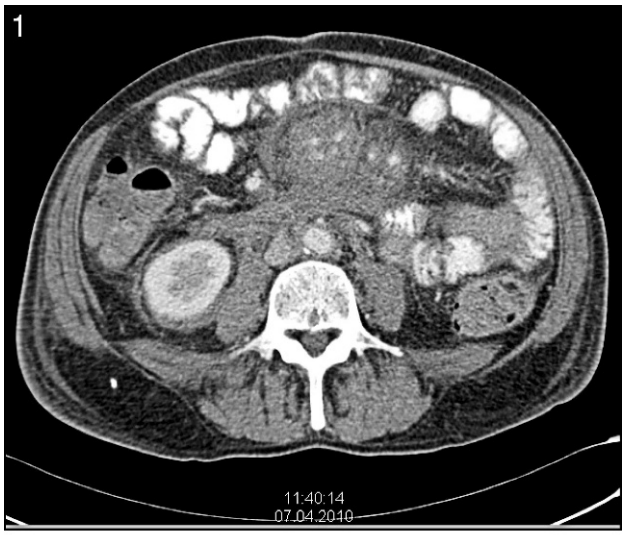
A 65-year-old man suffered from edema of in both legs, dyspnea, vomiting, and diffuse abdominal pain for 2 weeks. He was treated by his family doctor with diuretics and antiemetics without success. The patient was sent into our department for further diagnostics.

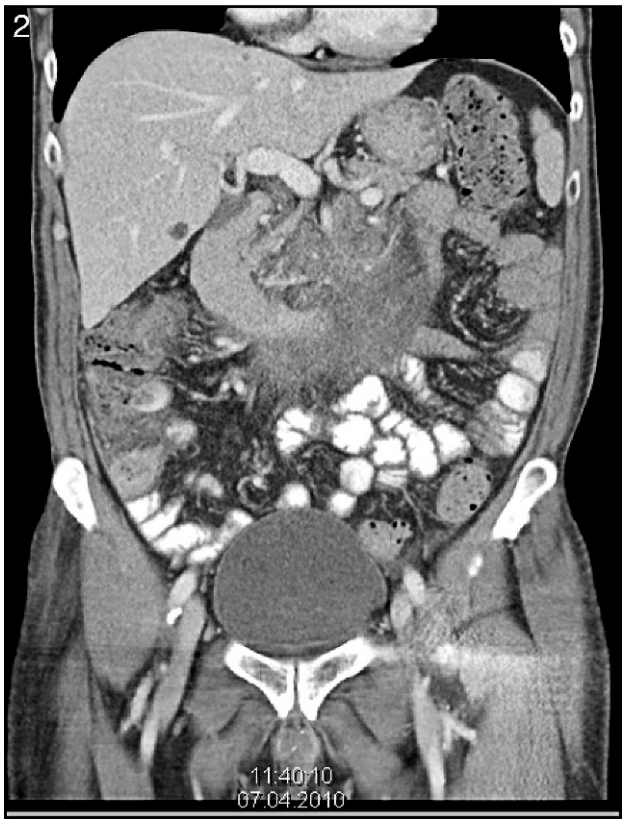
The patient suffered from no other diseases. Except for a hip replacement the patient had not undergone other surgery.

The physical examination revealed a patient in a good general condition—no cyanosis, no dyspnea. He had edema in both legs. The lymph node status was unremarkable. The cardiopulmonary status was normal. There was no abdominal defense. The upper left abdomen was light tender. He had normal peristalsis. The liver and spleen were normal size. There was no throbbing pain over the kidneys. The patient did not have any neurologic deficit.

An electrocardiography showed normal findings. A cardiac ultrasound showed normal function and no valvular defects. There were no pericardial effusion. An X-ray of the chest did not show signs of pulmonary

Reprint requests: K. Novak, MD, Department of general, visceral and thoracic surgery, Klinikum Weiden, Weiden i.d. Obpf, Germany. Tel.: +49 961 303 3002; Fax: +49 961 303 3052; E-mail: karel.novak@kliniken-nordoberpfalz.ag





Figs. 1 and 2 An infiltration of the mesenteric fat with evident injections of vessels under the pancreas near the pars horizontalis of the duodenum. Encasement of the mesenteric vessels. Liver cyst segment VI.

edema. A pleural ultrasound showed a little effusion on both sides.

An abdominal ultrasound demonstrated a first degree pyelectasia on both sides. A calculus was not present on either side. Both kidneys were normal sized. There was a pre-existing renal cyst on the right side. A renal tumor was not present. A liver cyst was also pre-existing. The small intestine and the colon were filled with a lot of liquid. Peristalsis was minimal. A little ascites existed.

A renal scintigraphy described a normal function of both kidneys but a total pyelectasia on the left side and a partial pyelectasia on the right side.

Blood results showed increased leukocytes (14.9/nL) and C-reactive protein (180/mg/L), hypalbuminosis (23.3 g/L) and thrombocytosis (688/nL).

A computerized tomography of the thorax/abdomen/pelvis described a diffuse mesenteric infiltration with obstruction of the mesenteric vessels and the duodenum (Figs. 1 and 2). Based on the results of the computerized tomography the tumor was suspicious of a mesenteric fibrosis. A lymphoma or a malignant tumor of the small intestine was also a possibility (Figs. 1 and 2). The lung was unremarkable. There was a little pleural effusion on both sides.

A laparoscopy followed and a tumor (15 × 15 cm) of the mesentery was detected. Because of the bad overview we switched to a laparotomy for a complete exploration of the abdominal cavity. A solid tumor with starting encasement of the duodenum was palpable. The intestinal wall was thickened. Multiple biopsies for immediate histologic examination during surgery followed. The histologic examination described a fibromatosis, but a malignant tumor could not be excluded. A total resection of the tumor was not possible because of infiltration into neighboring organs. Because of the risk of complete obstruction of the duodenum a retrocolic duodeno-jejunostomy, side-to-end was done. For reconstruction of the small intestine a jejuno-jejunostomy, end-to-side was done. A short segment of the jejunum was also resected and additional biopsies of the mesentery were taken for histologic examination.

The histologic examination of the mesentery described a lymph node with foam cells in the marginal sinus (Figs. 3–5).

The immunohistologic examination described a positive reaction of S-100, vimentin, and CD 68. The reaction on leucocyte common antigen and panzytokeratin was negative. According to our pathologists these results were typical for a histiocytosis. Material was send to another institute of pathology (Prof. Katenkamp, Jena, Germany) and to the institute of hematopathology (Prof. Wacker, Kiel, Germany) for second opinions. In their opinion, the cause of this process can only be found clinically. The final result of the histologic

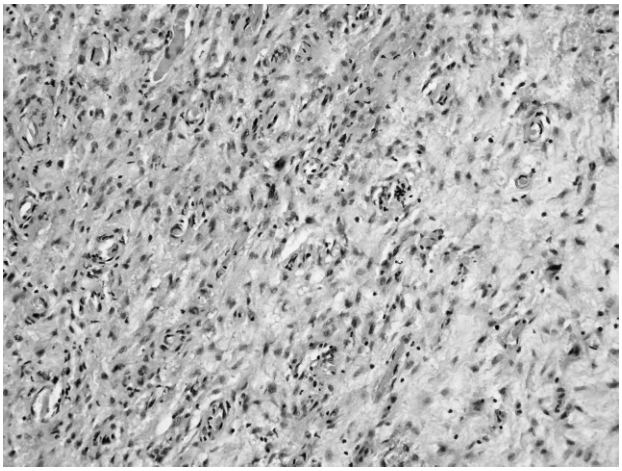


Fig. 3 Mesenteric biopsy with histiocytes and multiple capillaries.

and immunohistochemical examinations described a reactive inflammatory process with dominance of histiocytes. According to the histologic and immunohistologic findings Rosai-Dorfman disease, as a differential diagnosis, could be excluded (Figs. 6, 9 and 10,).

The patient and his histologic results were presented at the interdisciplinary tumor board. A therapy of high dose steroids for 3 months was recommended. We started the therapy immediately with hydrocortisone (1 mg/kg/d).

Postoperative computerized tomography described the tumor with obstruction of the mesenteric vessels and a hypoperfusion of the small intestine. A starting encasement of the duodenum was also described. There was no progress of tumor growth. A little ascites still existed.

A contrast imaging described a slow passage of the proximal small intestine and a gastro-oesophageal reflux.

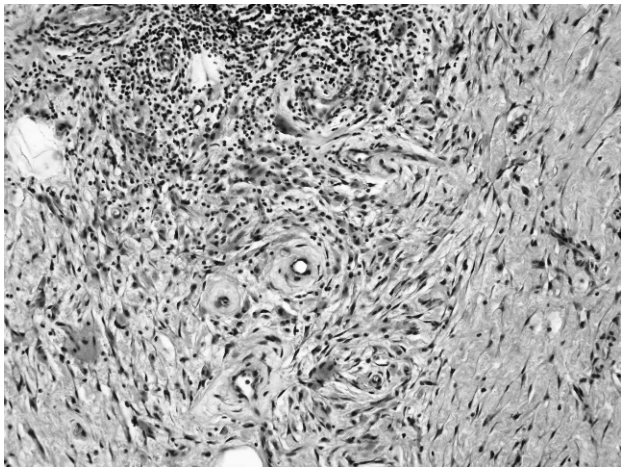


Fig. 4 Mesenteric biopsy with histiocytes, lymphoid follicles, and slight fibromatosis.

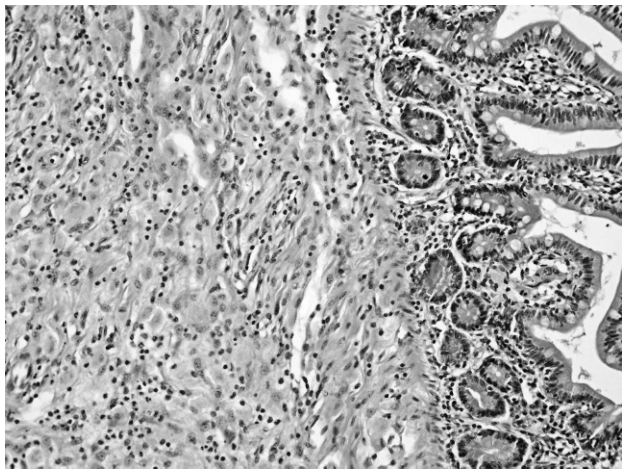


Fig. 5 Small intestine with histiocytes in submucosa.

We reduced the dose of hydrocortisone in a stepwise manner to 10 mg/d. The patient left our department in a good general condition without complaints.

Computerized tomography 3 months postoperatively described no progress of the tumor and recently a small regress of the tumor. Ascites has become smaller. The obstruction of the duodenum could not be found. Pyelectasia at first degree on both sides was still present. The patient was in good general condition without any complaints. The patient was no longer treated with steroids.

Discussion

Kabra *et al*[1] describe the mesenteric fibromatosis as a rare form of fibromatosis. Novák[2] describes this disease together with retroperitoneal fibromatosis, mediastinal fibromatosis, and retroorbital fibromatosis. Singal *et al*[3] mention that a fibromatosis can

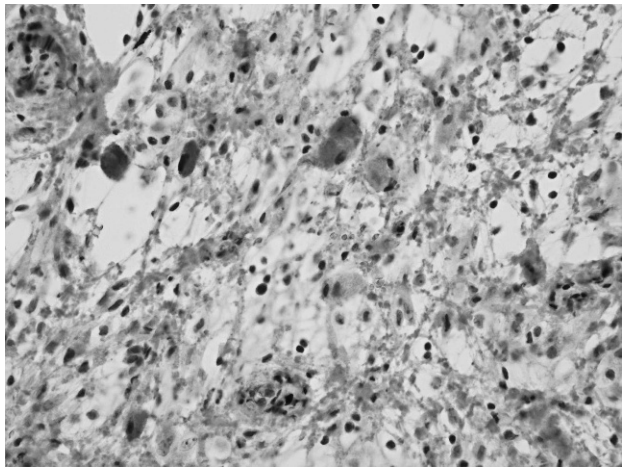


Fig. 6 Immunhistochemy S-100.

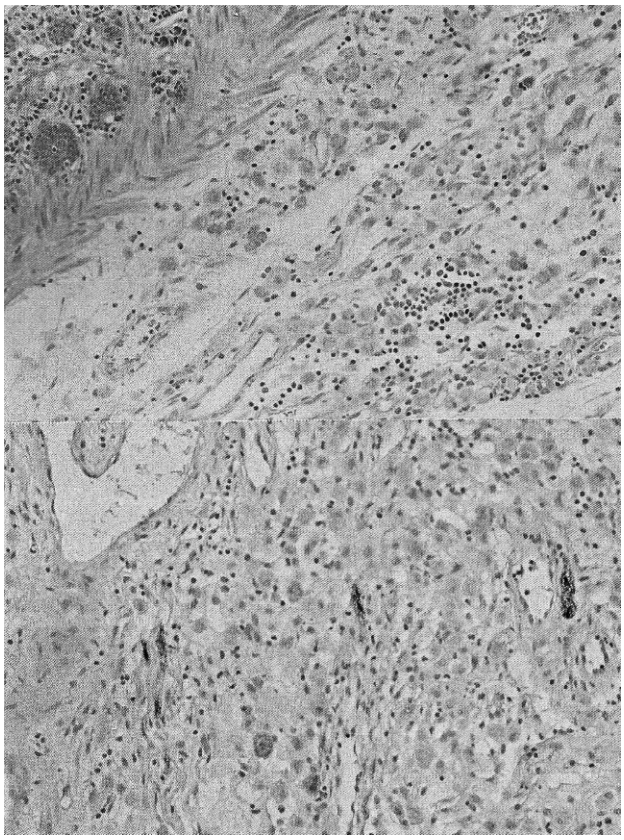


Fig. 7 Immunohistochemistry—S-100-positive histiocyte. Giemsa stain.

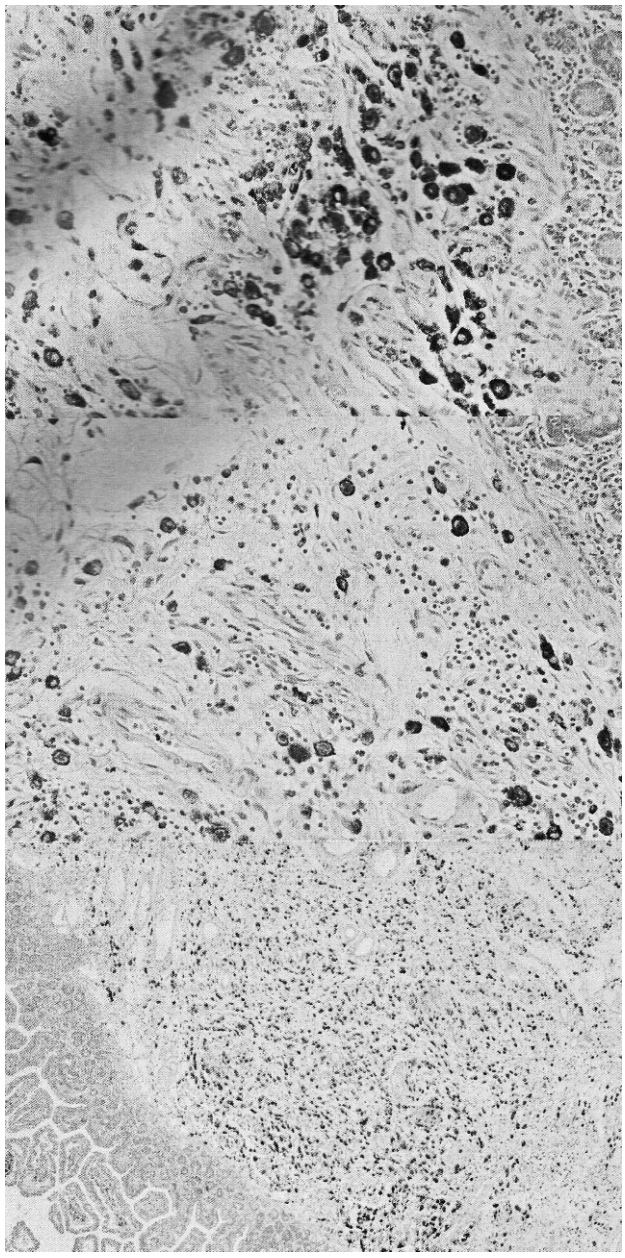
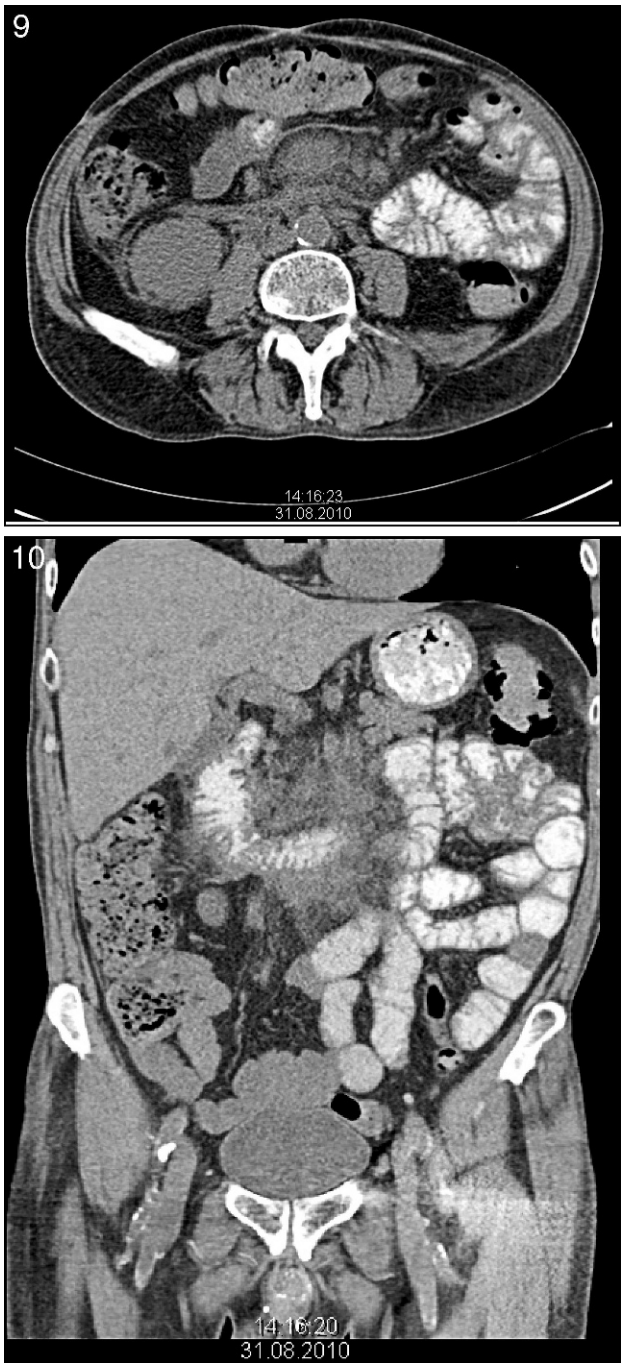


Fig. 8 Immunohistochemistry—CD 68-positive histiocyte.

appear anywhere in the human body. In our case a mesenteric histiocytosis with many capillaries and slight fibromatosis can be described. A case report about mesenteric histiocytosis was not found in literature, but the histopathologic views of mesenteric biopsy and small intestine are different in these images. The mesenteric biopsy appears like sclerosing mesenteritis or reactive inflammatory process as mentioned by Professors Katenkamp and Wacker. But small intestine biopsy includes some infiltration of histiocytes. However, the number of S-100- and CD 68-positive histiocytes in the image (Fig. 6) is not enough to describe the case as a ''histiocytosis.''

According to Singal *et al*,[3] the etiology of fibromatosis is unknown. As Sen *et al*[4] describe, the tumor originates mainly from deep-seated fascial or musculoaponeurotic structures and although the tumor is considered histologically benign, its local behavior is aggressive. Perence *et al*[5] found that mesenteric fibromatosis is characterized by an infiltrative growth. In our patient the tumor showed an infiltration of the jejunum and an obstruction of the duodenum, the mesenteric vessels. and the ureter on both sides. According to Batori *et al*,[6] the tumor can be single or multiple. Colovic *et al*[7] mention that about 10% of the tumors show a very aggressive growth. In our patient a single tumor was found. Growth appeared unaggressive until now.

Tonelli *et al*[8] found that mesenteric fibromatosis can occur isolated, accompanied by a familiar adenomatous polyposis, or Gardner syndrome as an extracolonic manifestation. In our patient neither a familiar adenomatous polyposis nor Gardner syndrome was present.



Figs. 9 and 10 Mesenteric fibromatosis with encasement of the arteria mesenterica superior and duodenum. Little regression of tumor growth. No signs of an occlusion of the duodenum. Extension of the renal pelvis on both sides, especially of the right kidney.

According to Perenze *et al*,[5] the histologic examination is the only diagnostic procedure capable of ruling out a malignant mesenteric neoplasm. In our patient the preoperative computerized tomography described a tumor that was suspicious of a mesenteric fibromatosis. But a statement about its dignity was not possible. Multiple biopsies were taken for histologic examination. The result of the immediate section for histologic examination during surgery could not exclude a malignant tumor. Only the final histologic result described a mesenteric infiltration of histiocytes and excluded a malignant tumor.

Agarwal *et al*[9] described that the Rosai-Dorfman disease is verified by histologic characteristics; a lymphadenopathy that results in an extension of the sinuses. The sinuses show a mass of histiocytes and plasmacytes. The histiocytes show typically an emperipolesis. According to Hsiao *et al*,[10] usually cervical lymphadenopathy is present. An extranodal involvement occurs in up to 40% of cases. The histiocytes are strongly immunoreactive to S-100, CD 68, and CD 14. In our patient a cervical or other lymphadenopathy did not exist. There were a few histiocytes and no plasmacytes in the sinuses. An emperipolesis was not found. The histiocytes were immunoreactive to S-100, vimentin, and CD 68.

Casella *et al*[11] describe surgical therapy as the treatment of choice. Nakai *et al*[12] mentions that in case of mesenteric fibromatosis structures in the neighboring organs, multivisceral resections can be necessary. Perenze *et al*[5] add that it is not always possible to perform a radical removal of the tumor mass because it may have infiltrated important anatomic structures. In our patient an infiltration of the jejunum with obstruction of the duodenum and the mesenteric vessels was present. A complete removal of the tumor was not possible because of the infiltration. A duodeno-jejunal bypass was used because of the risk of a complete occlusion of the duodenum.

Casella *et al*[11] found that local recurrences are frequent. According to Sharma *et al*,[13] the addition of radiation, with or without surgery, does not impact on local control. Tonelli *et al*[8] reports that daily therapy with raloxifene decreases the size of mesenteric fibromatosis. In our patient radiotherapy and a therapy with raloxifene did not take place. Our patient underwent postoperatively a high dose steroid therapy for 3 months, which was reduced step by step. The tumor showed no progress under this therapy. Recently 3 months after surgery a small regress of the tumor was seen in computerized tomography. Many months after imaging we can mention that the infiltration of histiocytes in mesenteric biopsy is not a proliferative lesion but it is reactive. Therefore it should not be described as ''mesenteric histiocytosis'' or fibromatosis. But intraoperatively it is very difficult to make a decision about the essence of the problem. In acute situations

the surgeon needs a working diagnosis as fibromatosis, histiocytosis, or inflammatory infiltration.

In conclusion, mesenteric histiocytosis is not described in surgical literature. Maybe this case report is original but it is not sure. The mesenteric histiocytosis is clinically and macroscopically similar to mesenteric fibromatosis. The tumor is benign but it shows an infiltrative growth. Computerized tomography can detect a mesenteric fibromatosis but cannot give any information about its character. The method of choice to exclude a malignant tumor is a biopsy. An immunohistochemical examination is useful. The therapy of choice is total resection of the tumor. The tumor is no longer resectable because of its advanced growth and infiltration into important anatomic structures. If the tumor is not resectable and an obstruction of the small intestine occurs, an intestinal bypass becomes necessary. A high dose of steroids can restrict tumor growth.

References

1. Kabra V, Chaturvedi P, Pathak KA, de Souza LJ. Mesenteric fibromatosis: a report of three cases and literature review. *Indian J Cancer* 2001;**38**(2–4):133–136
2. Novák K. Mesenteric fibrosis [In Czech]. In: Horký K, ed. *Medical Repetitorium*, Prague, Galén, 2003, **154**
3. Singal AK, Agarwala S, Manna SA, Gupta AK, Bhan MK, Mitra DK. Diffuse intra-abdominal fibromatosis—report of a new entity with review of literature. *Trp Gastroenterol* 2004;**25**(1):28–31
4. Sen E, Gurkan OU, Gulbay BE, Gungor A, Acican T. A case of aggressive fibromatosis of posterior mediastinum. *Tuberk Toraks* 2003;**51**(3):306–309
5. Perence B, Poti O, Margiotta A, Raga D, Marrano N, Minni F. Mesenteric fibromatosis. *Chir Ital* 2005;**57**(6):717–722
6. Batori M, Chatelou E, Mariotta G, Sportelli G, Straniero A, Casella G *et al*. Giant mesenteric fibromatosis. *Eur Rev Med Pharmacol Sci* 2005;**9**(4):223–225
7. Colovic R, Grubor N, Radak V, Micev M, Stojkovic M, Colovic N. Aggressive intraabdominal fibromatosis. *Vojnosanit Pregl* 2006;**63**(9):839–842
8. Tonelli F, Ficari F, Valanzano R, Brandi ML. Treatment of desmoids and mesenteric fibromatosis in familial adenomatous polyposis with raloxifene. *Tumori* 2003;**89**(4):391–396
9. Agarwal A, Pathak S, Gujral S. Sinus histiocytosis with massive lymphadenopathy—a review of seven cases. *Indian J Pathol Microbiol* 2006;**49**(4):509–515
10. Hsiao CH, Tsai TF, Yang TH, Liu CM. Clinicopathologic characteristics of Rosai-Dorfman disease in a medical center in northern Taiwan. *J Formos Med Assoc* 2006;**105**(9):701–707
11. Casella C, Mittempergher F, Baronchelli C, Liserre B. Mesenteric fibromatosis: a case report. *Ann Ital Chir* 2004; **75**(6):705–708
12. Nakai M, Nakamura N, Izumi T. Mesenteric fibromatosis with ureteral stenosis: a case report. *Hinyokika Kiyo* 2003;**49**(5): 253–255
13. Sharma V, Chetty DN, Donde B, Mohiuddin M, Giraud A, Nayler S. Aggressive fibromatosis—impact of prognostic variables on management. *S Afr J Surg* 2006;**44**(1):6–8,10–11