Case report
UDC: 616-006.44:616.383:616-076

Primary non-Hodgkin lymphoma of the jejunum associated with mesenteric lipodystrophy: A case report

Dragana PETROVIĆ[1]
Darjana JOVANOVIĆ[1]
Branimir GUDURIĆ[1]
Boris FILIPOVIĆ[2]
Aleksandar MUTIBARIĆ[1]

[1]INSTITUTE OF ONCOLOGY SREMSKA KAMENICA, SREMSKA KAMENICA, SERBIA AND MONTENEGRO
[2] DEPARTMENT OF RADIOLOGY, CLINICAL CENTER NOVI SAD, NOVI SAD, SERBIA AND MONTENEGRO

Mesenteric lipodystrophy is a rare condition characterized by a nonspecific inflammatory process that involves the root of the mesentery in a lipoma-like lesion. We reported a case of sclerosing mesenteritis presenting as recurrent abdominal pain, profound weight loss, dilated cardiomyopathy, malabsorption and a mass on computed tomography scan of the abdomen in a 42 years old man with previous history of lymphoma. Because of the wider differential diagnosis in such cases, the patient underwent an extensive workup culminating in a laparoscopy with biopsy. This case illustrated that mesenteric lipodystrophy should be included in the differential diagnosis of retroperitoneal and mesenteric tumor.

KEY WORDS: *Lipodystromy; Mesentery; Lymphoma; Diagnosis, Differential*

INTRODUCTION

Mesenteric lipodystrophy (ML) is a rare condition of unknown etiology that causes nonspecific inflammation and fibrotic process affecting the fatty tissue of the mesentery. Mesenteric lipodystrophy may present as an acute abdomen or with non-specific upper abdominal symptoms. The causative agents are unknown, although an association with lymphoma has been reported. This case report describes the clinic and pathologic findings of a large lipoma-like tumor of the mesentery in a 42 years old patient with previous history of lymphoma.

CASE REPORT

A 42-year-old man presented with weight loss, intermittent convulsive abdominal pain, weakness, fatigue, and diarrhea. Patient's history commenced in 1987 with a diagnosis of non-Hodgkin lymphoma of the jejunum (diffuse large B cells lymphoma, WF classification) (stage CS I E B), for which he received six cycles of the adjuvant chemotherapy with COPP (cyclophosphamide, vincristine, procarbasine, and prednisone). The complete remission was achieved. Afterwards, he was treated with maintaining therapy, containing vinblastine 10 mg once a month during the next two years. Having the maintaining therapy finished, the patient was followed up. He remained in a good state of health for the next 10 years.

In August 1997, the patient was admitted because of gastrointestinal disorder. His recent illness started with abdominal cramps, the frequent loose, watery stools without protein and carbohydrate, mucus, blood or fat loss, followed by weight loss, weakness, and malaise. He denied any other discomfort. At the time of admission, he was not taking any medicine; physical examination did not reveal any abnormalities except the abdominal hernia. The temperature was 36.4˚C, the pulse was 54, and the respirations were 20. The blood pressure was 120/80 mm Hg. Laboratory values were: hemoglobin 11.1 g/dL, hematocrit 32%, red blood counts 3.41 G/L, white cell counts 5.1 G/L, platelet counts 497 G/L. The values of liver enzymes were normal, as well as values of glucose, serum creatinine, urea, serum alkaline phosphatase, sodium, and potassium. The value of lactate dehydrogenase was slightly increased (11.19 μkat/l; normal range 5.22-10.30 μkat/l). A stool specimen was normal. An electrocardiogram revealed a sinus bradycardia at a rate of 54, with negative T waves in V4-V6. He was HIV negative.

The patient underwent an extensive workup to establish the diagnosis. The ultrasonography of abdomen revealed two rounded hypoechogenic masses in retroperitoneum, suggesting lymphadenomegaly, since the findings of other abdominal organs were normal. Considering these results, CT of abdomen was proposed. The result of abdomen CT was as following: in retroperitoneal space, from pelvis in cranial direction, the hypodense mass is seen displacing the loops of small bowel laterally and caudally.

Address correspondence to:
Dr. Dragana Petrović, Institute of Oncology Sremska Kamenica, 21204 Sremska Kamenica, Institutski put 4, Serbia and Montenegro

The manuscript was received: 22.10.2003
Provisionally accepted: 21.11.2003
Accepted for publication: 19.12.2003

Retroperitoneal lymph nodes were not enlarged significantly. Aorta and vena cava were not affected. The liver was homogeneous in density, slightly enlarged. The lien and pancreas had the normal shape. The results of abdomen CT primarily suggest the presence of retroperitoneal tumor, furthermore, considering differential diagnosis it could be recognized as liposarcoma with predomination of mucus component (Figure 1).

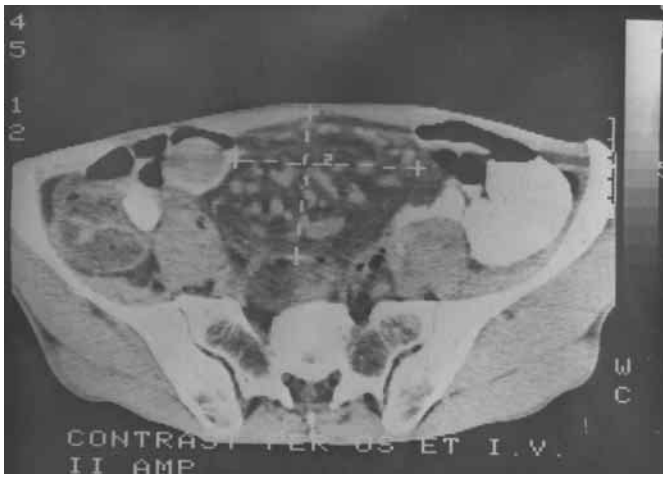

Figure 1. Computed tomography of abdomen shows hypodense mass in retroperitoneum)

The patient was further examined by endoscopy. Colonoscopy could only be done up to lienal flexure, because of the presence of strong spasm and intraperitoneal adhesions. The colon mucous membrane was without any pathological changes. The small-bowel barium study revealed normal passage of barium throughout the loops. The loops of jejunum were dislocated to the left part of abdomen and the loops of ileum to pelvis. Ileum loops were distended and Kerkring' s crypts were elongated. Stenosis, which could be the cause of this dilatation, was not visible. Enteroclysis was suggested. Results of all these examinations could not provide the correct diagnosis, so the patient underwent explorative laparotomy on September 10th 1997. During the exploration of abdomen, relapse of lymphoma was not found and no changes on the jejunum anastomoses were visible. From the root of mesentery, in peritoneal cavity, the loosely fatty, thickened and partly steatonecrotic lesion was found. The tumor did not produce any invasion neither on vascular system nor on small bowel passage (Figure 2). Extempore biopsy of lesion in the mesentery indicated steatonecrosis, and the final result of histological examination confirmed mesenteric lipodystrophy (Figure 3). In the second week of September 1997, the patient was discharged in good condition.

In November 1997, the patient came complaining of frequent, loose and watery stools without mucus and blood, weight loss, and low-grade fever. The patient was treated on outpatient basis with ciprlofloxacine and loperamide ambulatory, which led to clinical improvement. He was admitted to hospital for additional examination. During his hospitalization, a magnetic resonance imaging (MRI) of abdomen was performed, which pointed to hepatomegalia (170 mm at midclavicular line) without pathological changes, dilated intrahepatic bile ducts, gallbladder, renal pelvis system and proximal ureter from left side, v. mesenterica superior, and multifocal round masses diameter of 1 cm in mesentery (Figure 3). Laboratory values were normal. Treatment with ciprofloxacine was started and certain improvement was noted, so he was dismissed by the beginning of December 1997. The patient was regularly controlled.

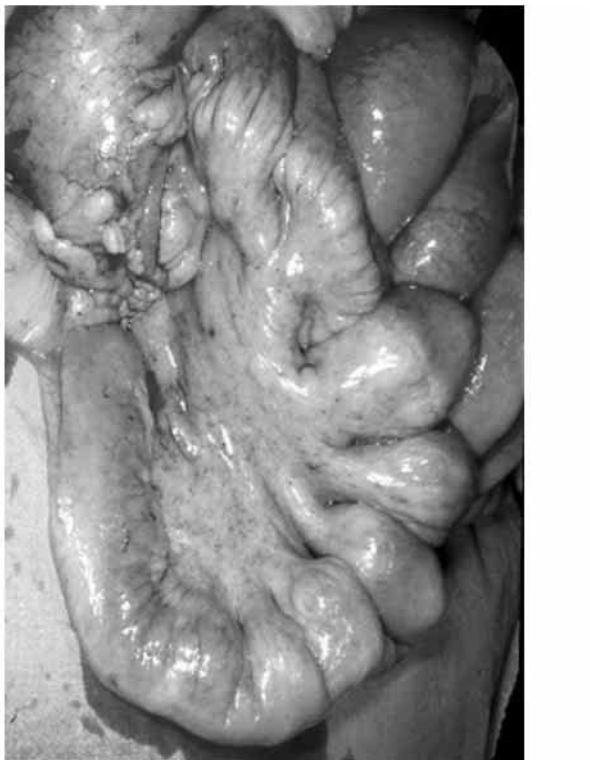

Figure 2. The fatty, steatonecrotic lesions in the mesentery (intraoperative view)

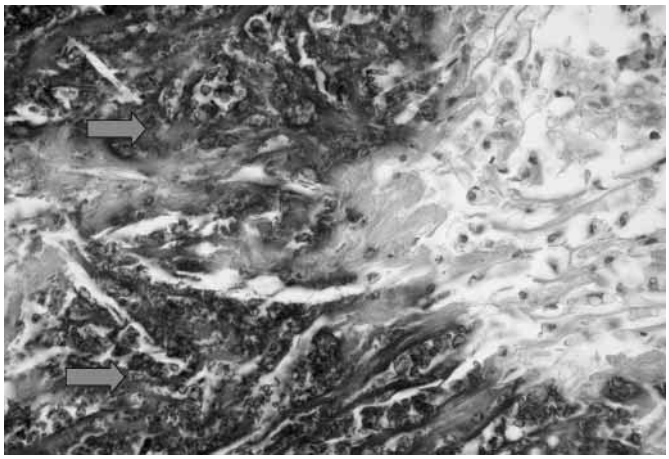

Figure 3. Photomicrograph of histological specimen of mesenteric tissue shows diffuse fat necrosis and calcification (dark purple staining indicated by arrow) (hematoxylin-eosin)

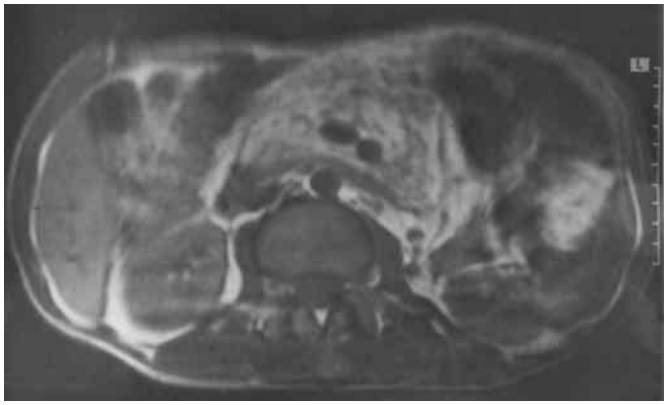

Figure 4. The multiple, small, round lesions in the mesentery (MRI scan of abdomen)

In September 1998, he came to cardiologist because of chest discomfort. Physical examination showed sinus tachycardia (100/min), a grade 2 systolic ejection murmurs was heard at the cardiac apex, arterial pressure 13/9 kPa, as well as signs of chronic heart failure. The chest radiograph demonstrated left ventricular enlargement, signs of pulmonary congestion. The electrocardiogram showed sinus tachycardia, negative T-waves in precordial leads. Echocardiography presented left ventricular enlargement (LVID 8,4) with normal thickened walls, reduced ejection fraction (EF 0.30), and considerably dilated cardiomyopathy was established. Standard therapy with salt restriction, diuretics, digitalis, and nitrates was started. He was being given the considered therapy in prescribed order and committed to controls.

In the autumn of 1999, the patient suffered of gastrointestinal malaise again, which resulted in profound weight loss. He denied any other discomfort. On the reception, patient was of asthenic constitution, with no signs of cardiac decompensation. The mild anemia existed. Serum amylase, liver enzyme and electrolyte values were normal. LDH was slightly increased. The computed tomography of abdomen: on each scan the signs of dilatation of bowel loops (both small and large bowel) were visible, which was caused by the presence of air and secret. Enteroclysis was done, and the result suggested malabsorbtion syndrome.

During the hospitalization, the patient developed the tachyarrhythmia, which was presented with polymorphic ventricular extrasystoles. Mexiletine a 100 mg every 8 h was introduced. By the oral administration of corticosteroid (40 mg/day), diet nutrition, the patient' s condition was improved and he was dismissed. He never came to be controlled and we were informed that he had died.

DISCUSSION

Mesenteric lipodystrophy, also referred as sclerosing mesenteritis, retractile mesenteritis or mesenteric panniculitis, is a rare condition slightly predominant in male population with a peak incidence found in the fifth and sixth decades of life (1,2). The etiology of ML is unknown. Predisposing factors such as infection, trauma, surgery, malignancy, and autoimmune disease have been suggested (3). Presenting features of ML are protean and usually include abdominal pain, weight loss, nausea, vomiting, malaise, diarrhea, gastric outlet obstruction or small bowel obstruction. (2,4,5) Symptoms may be absent, progressive or intermittent. Symptoms often persist for a year or more. Physical examination may disclose an abdominal mass with or without tenderness in approximately half of the patients, whereas 20% of the patients have no abdominal complaints, and the mesenteric mass can be an incidental finding (6,7). Laboratory tests are generally of none help.

The diagnostic criteria of ML include: (1) presence of a single, multiple or diffuse mass-like inflammatory lesions in the mesentery; (2) histological confirmation of fat necrosis and inflammatory reaction in the mesenteric lesions; (3) exclusion of pancreatitis, inflammatory bowel disease, vasculitis and Weber-Christian disease (extra-abdominal fat necrosis) (8). The diagnosis is generally established by laparotomy and biopsy (2,5). The characteristic histological appearance of ML consists of degeneration of fat cells, areas of fatty necrosis, and an occasional infiltration with lipophages, macrophages, plasma cells, neutrophils, proliferative myofibroblasts that form hyaline collagen, lymphocytes, and foreign body giant cells (2,7).

Computed tomography provides the best means of imaging this lesion. CT is useful in defining the extent of ML and may be helpful in planning the treatment (9,10). The typical CT findings include inhomogeneous masses of both fat and soft tissue attenuation at the root of the mesentery and surrounding vessels, without vascular distortion. The findings on CT of the abdomen vary, depending on the stage of the disease and, either the inflammatory or fibrosis component dominates (11). When diffuse inflammation of mesenteric fat is the predominant component, the inflammatory lesions appear as well-defined fatty tumors throughout the small bowel mesentery (11). These fatty tumor-like lesions present inflammatory infiltrates and fibrotic areas associated with mesenteric vessels, which are displaced and surrounded by the involved fat but not invaded (10,11). When fibrosis is the predominant feature, the mesenteric lesions appear as predominantly soft-tissue densities (11). On magnetic resonance imaging (MRI), ML may appear predominantly as a high-intensity tumor, with intensity identical to that of peritoneal fat (12).

The differential diagnosis of ML should include mesenteric fibromatosis, inflammatory pseudotumors, idiopathic retroperitoneal fibrosis, sclerosing malignant lymphoma, malignant primary mesenteric tumor, metastatic neoplasm, and various granuloma-

tous lesions (5,7). The prognosis for ML is generally good, and in most cases, spontaneous remission is the most common outcome. Corticosteroids and immunosuppressive agents have been used successfully in a few aggressive cases (13,14). Surgical intervention may be necessary if obstructive symptoms develop. In the issued case, mesenteric lipodystrophy was manifested in unspecific gastrointestinal symptoms like intermittent abdominal pain, weight loss, nausea, malaise and diarrhea. Physical examination could not reveal any abdominal mass and laboratory tests were not conclusive. Because of the wider differential diagnosis in such cases, the patient underwent an extensive workup. The CT of abdomen only, revealed retroperitoneal tumor imaging as liposarcoma. Respecting the results of examination, diagnosis of lymphoma was excluded as well as other conditions and diseases usually associated with ML. Though preoperative diagnosis was not possible in our case, the patient underwent to explorative laparotomy and the tumor biopsy. Surprisingly histology revealed invasion by sclerosing mesenteritis. The mesenteric lipodystrophy is an extremely rare condition never noted in our medical literature. The certain cause of ML in our case was never found. The development of dilated cardiomyopathy as well as malabsorbtion syndrome could be explained by the evolution of ML. It is supposed these two conditions caused fatal ending.

CONCLUSION

Mesenteric lipodystrophy is a rare condition that can be mistaken for a mesenteric neoplasm based on clinical, radiologic, and gross characteristics. The causative agents are unknown, although an association with lymphoma has been reported. A firm diagnosis can be reached only by histological examination and a number of conditions need to be considered in the differential diagnosis. This case highlights the manifestation of mesenteric lipodystrophy and suggests that it should be included in the differential diagnosis of retroperitoneal and mesenteric tumor.

REFERENCES

1. Harris RJ, van Stolk RU, Church JM, Kavurucu MS. Thoracic mesothelioma associated with abdominal mesenteric panniculitis. Am J Gastroenterol 1994;89:2240-2.
2. Sans M. Varas M. Anglada A, Bachs ME, Navarro S, Brugu J. Mesenteric panniculitis presenting as fever of unknown origin. Am J Gastroenterol 1995;90;1159-61.
3. Bush RW. Hammar SP, Rudolph RH. Sclerosing mesenteritis response to cyclophosphamide. Arch Intern Med 1986;146:503-6.
4. Parra-Davila E. McKenney MG. Mesenteric panniculitis: Case report and literature review. Am Surg 1998;64:768-75.
5. Emory TS, Monihan JM, Carr NJ, Sobin LH. Sclerosing mesenteritis, mesenteric panniculitis and mesenteric lipodystrophy: a single entity? Am J Surg Pathol 1997;21(4):392-8 (Abstract quote).
6. Ornniston MC. Thomson H. Mesenteric panniculitis. Postgrad Med J 1980; 56:67-8.
7. Kelly JK. Hawng WS. Idiopathic retractile (sclerosing) mesenteritis and its differential diagnosis. Am J Surg Pathol 1989; 13:513-521.
8. Hemaidan A, Vanegas F, Alvarez OA, Arroyo MA, Lee M. Mesenteric lipodystrophy with fever of unknown origin and mesenteric calcifications. Southern Med J 1999; 92:513-6.
9. Badiola-Varela CM, Sussman SK, Glickstein MF. Mesenteric panniculitis: findings on CT, MRI, and angiography. Case report. Clin Imaging 1991;15(4):265-7.
10. Sabate JM, Torrubia S, Maideu J, Franquet T, Monill JM, Perez C. Sclerosing mesenteritis: imaging findings in 17 patients. AJR Am J Roentgenol 1999;172(3):625-9 (Abstract quote)
11. Mata JM, Inaraja L, Martin J, Olazabal A, Castilla MT. CT features of mesenteric panniculitis. J Comput Assist Tomogr 1987;11:1221-3.
12. Kawashima A. Fishman EK. Hruban RH. Mesenteric panniculitis presenting as a multilocular cystic mesenteric mass: CT and MR evaluation. Clin Imaging 1993;17:112-6.
13. Genereau T, Bellin MF, Wechsler B, Le TH, Bellanger J, Grellet J, Godeau P. Demonstration of efficacy of combining corticosteroids and colchicine in two patients with idiopathic sclerosing mesenteritis. Dig Dis Sci 1996;41(4):684-8.
14. Bala A, Coderre SP, Johnson DR, Nayak V. Treatment of sclerosing mesenteritis with corticosteroids and azathioprine. Can J Gastroenterol 2001;15(8):533-5.
15. Cosmo P. Borjesson B. Kristensen PB. Retractile mesenteritis. Eur J Surg 1994;160:579-81.