Case report

A 65-year-old male was evaluated because of umbilical abdominal pain with intermittent diarrhoea and fever accompanied by 12 kg of weight loss. Medical history only noted a one-year history of type 2 diabetes mellitus, for which he used metformin 500 mg once daily. He denied changes in stool colour or melena. Lab tests showed slightly elevated erythrocyte sedimentation rate (ESR) (26 mm/h) and increased liver enzymes (γGT 682 U/l, alkaline phosphatase 317 U/l, aspartate aminotransferase 73 U/l, alanine aminotransferase 253 U/l, bilirubin total 10 μmol/l, and bilirubin direct 3 μmol/l). Abdominal ultrasound and chest radiograph were unremarkable.

CT scan showed mesenteric fat infiltration and lymphadenopathy around the mesenteric vessels (figure 1).

At follow-up visit four weeks later, the patient had fully recovered with no signs of abdominal pain. He gained 5 kg in weight. Liver function and ESR were fully normalised. The patient refused further work-up including biopsy of the process.

What is your diagnosis?

See page 335 for the answer to this photo quiz.

Figure 1. CT scan

Left panel: shows focal infiltration of mesenteric fat with enlarged lymph nodes (white arrows) around the branches of the superior mesenteric artery and superior mesenteric vein. There is no tethering of the nearby bowel.

Right panel: the mesenteric fat shows infiltration (white arrows) slightly more dense compared to normal fat around the liver and retroperitoneal fat.
Sclerosing mesenteritis (synonym mesenterial lipodystrophy) is a rare idiopathic autoimmune disease with fibrosis and inflammation of the intra-abdominal fat (preferential localisation around mesenteric arteries). Differential diagnosis includes lymphoma, constipation, inflammatory bowel disease (Crohn’s disease or ulcerative colitis) or solid tumours (colon cancer or liposarcomas). The prevalence is unknown (but likely to be very low), and diagnosis is made by abdominal CT scan (fat ring sign and tumour pseudocapsule), confirmed by biopsy. It predominantly affects males around 50 to 70 years of age and presents with intermittent abdominal pain, fever, diarrhoea, nausea and dysphagia and subsequent weight loss. In a retrospective study of 98 subjects, 50% of patients presented with a palpable abdominal mass. Interestingly, 41% of these subjects underwent abdominal surgery (e.g. cholecystectomy, appendectomy, hysterectomy or colectomy) in the previous years before developing sclerosing mesenteritis, emphasising the inflammatory basis of the excessive fibroblast growth found in biopsies. Moreover, 50% of these subjects develop either extra-abdominal malignancies (e.g. lymphoma, breast cancer or bronchial carcinoma) or another autoimmune disease such as thyreoiditis or primary sclerosing cholangitis after diagnosis. The prognosis is not well known but in general a benign slowly progressive course is seen, yet self-limiting or a spontaneously resolving disease has been described. Progression of abdominal symptoms may require ‘tailor-made’ therapeutic intervention with prednisone or tamoxifen for a period of six months. In refractory cases, thalidomide had equivocal success. Surgery is not warranted unless sclerosing mesenteritis is dominated by focal intestinal obstruction requiring bypass procedure of the bowel with leaving primary panniculitis in place.

REFERENCES