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PHOTO QUIZ

Cavitating lymph node syndrome

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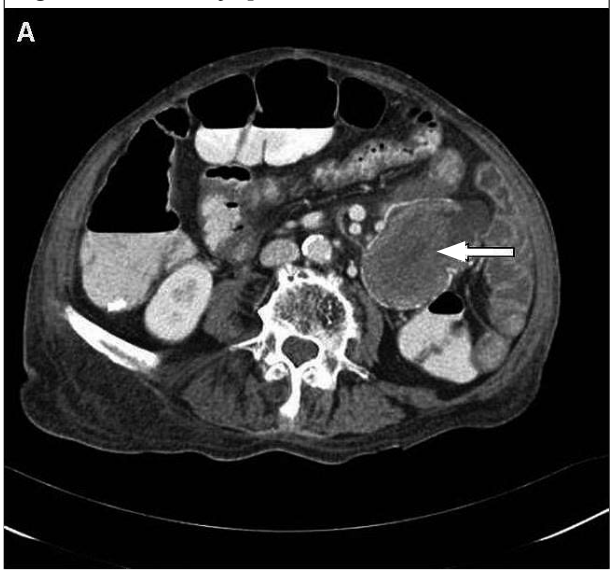
CASE REPORT

A 73-year-old woman with refractory coeliac disease presented with progressive fatigue, weight loss, steatorrhoea, and a solid abdominal mass on physical examination. Duodenal biopsies showed persistent villous atrophy with morphologically and immunophenotypically abnormal (CD3+/CD4-/CD5-/CD8-) intraepithelial lymphocytes suggestive of an enteropathy-type T-cell lymphoma. Computerised tomography demonstrated hyposplenism and a large cystic tumour of the mesentery (*figure 1*, arrow). Multiple imaging-guided and surgical biopsies of the tumour showed necrotic debris with haematin pigment.

Figure 1. *CT-scan of cystic mesenterial mass*



WHAT IS YOUR DIAGNOSIS?

See page 404 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (ON PAGE 403)

CAVITATING LYMPH NODE SYNDROME

DIAGNOSIS

The patient died of abdominal sepsis and intractable diarrhoea. Autopsy revealed enteropathy-type T-cell lymphoma of the duodenum, mesentery, and endometrium with ischaemic changes and bacterial invasion of the bowel. The cystic tumour of the mesentery measured 13 cm and consisted of multiple egg-like structures (*figure 2*) with a calcified shell and fatty necrotic content. Microscopy showed variably necrotic tissue with haematin pigment inside and lymph node remnants outside the tumour, compatible with lymph node cavitation. Cavitating lymph node syndrome was first described by Hemet *et al.*[1] in 1969. It is thought to be a rare but pathognomonic complication of (mostly) refractory coeliac disease, and hence associated with poor outcome.[2] Lymph node cavitation has received increasing attention. Anecdotal reports have already been published in which lymph node cavitation was the initial presenting symptom of coeliac disease. Both the aetiology and pathogenesis are unknown. Possibly, lymph nodes undergo haemorrhagic infarction due to chronic immune stimulation, or the consequence of complement-mediated endothelial damage by immune complex deposition. Other explanations are enlargement of mesenteric lymph nodes due to lymphatic obstruction or an exuberant accumulation of chyle in the specialised mesenteric lymph nodes leading to cavitation. This might explain why mesenteric lymph nodes are exclusively involved. Both CT and MRI investigations are highly characteristic and show the presence of multilocular masses with a diameter of 2 to 7 cm with accumulation of fluids and/or fatty material.[3,4] Radiological recognition of this rare but distinctive lesion might have prevented unnecessary invasive diagnostic procedures in our patient. Early recognition is also important in non-refractory patients as regression of the lesion has been reported after a strict gluten-free diet.

Figure 2. *Macroscopy of cystic mesenterial mass*

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4. Huppert BJ, Farrell MA, Kawashima A, Murray JA. Diagnosis of cavitating mesenteric lymph node syndrome in celiac disease using MRI. AJR Am J Roentgenol. 2004;183:1375-7.