LETTER

Recurrent pancreatitis : it can get on your nerves! A rare case of B-cell lymphoma presenting as recurrent pancreatitis and multiple cranial neuropathy

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To the Editor,

To the editor we present the case of a 57-year old man with no relevant medical history beside an episode of acute edematous pancreatitis three months earlier. The etiology was assumed to be alcoholic due to the abscence of gallstones, dyslipidemia or hyperparathyroidism and the granted regular alcohol intake. Two months after this episode he was admitted at the Neurology Department with a left peripheral facial nerve paresis without any significant lesions on CT or gadolinium enhanced MRI. After initation of corticosteroids the patient recovered and got discharged.

On month later he presented at the Emergency Department with acute epigastric pain without fever. He also reported vertigo, fatigue and a significant weight loss of 17 kg since discharge. There was no itching or night sweating. Biochemistry showed an elevated CRP (63 mg/l) and white blood cell count (12.8 10^9/mm^3), disturbed liver enzymes with a normal bilirubin and an elevated lipase (1365 U/l). Ethanol, triglycerides and LDH were normal.

The abdominal complaints regressed on supportive care whilst the vertigo got worse. On abdominal CT a focal zone of pancreatitis at the tail of the pancreas with scarce necrosis and infiltration of the peripancreatic fat was shown.

In the presumption of a benign paroxysmal positional vertigo several liberating manoeuvers were unsuccessfully attempted after which an isolated paresis of the left superior vestibular nerve was diagnosed by a vestibular evoked myogenic potential (VEMP). Shortly after, the cranial neuropathy started to extend with the involvement of the n. mentalis, presenting itself by chin numbness, and of the contralateral right facial nerve.

In the search of a possible relationship between recurrent pancreatitis and multiple cranial neuropathies, the differential diagnosis included neurosarcoidosis, a neurotropic virus, Whipple's disease and amyloidosis. We performed a lumbar puncture and a gastroscopy with at random gastric and duodenal biopsies.

Eventually, the diagnosis of diffuse large B-cell NHL (germinal center phenotype) was stated on a CD-20 stain of duodenal biopsies and was confirmed on cytological examination of the CNS liquid, pointing to associated leptomeningeal invasion with malignant lymphomatous cells. Bone marrow biopsy and bone scintigraphy turned out to be negative. The patient was referred to the hematology ward and consequently treated by intravenous and intrathecal chemotherapy. After 6 cycles an autologous stem cell transplantation was performed.

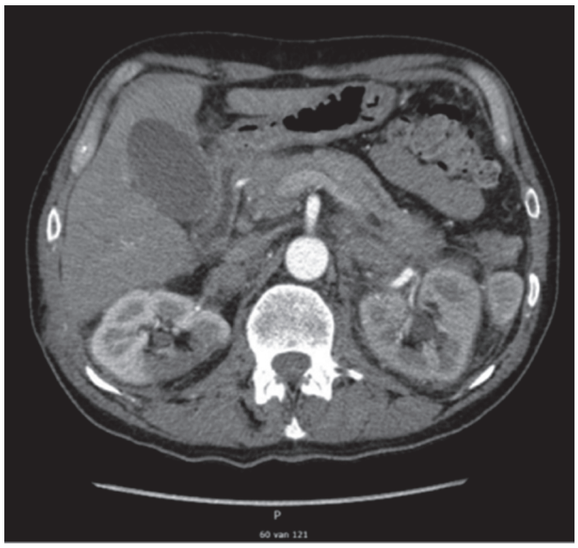

Fig. 1. — CT Abdomen 48h after admission.

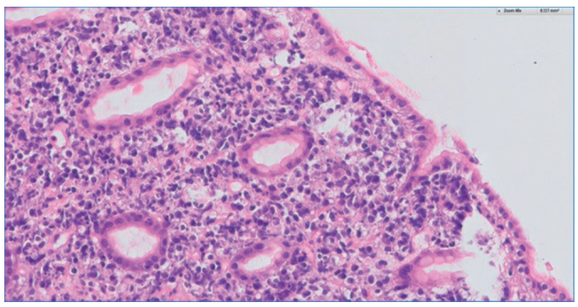

Fig. 2. — CD-20 staining on duodenal biopsy

To the best of our knowledge, this is the first case of large B-cell lymphoma presenting with synchronous pancreatic and leptomeningeal involvement.

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About 50% of all Non-Hodgkin's lymphoma are primary extranodal, of which 30 % is a diffuse large B-cell lymphoma. Only 0,2-2% of patients with NHL have pancreatic involvement at presentation with diffuse large B-cell lymphoma being the dominant histological subtype. (1) Clinical presentation is quite variable and fever and night sweats are uncommon features. To date, only 10 cases of pancreatic B-cell lymphoma presenting as acute pancreatitis have been described in the literature. (1, 2).

Approximately 5 to 10% of patients with systemic NHL will have secondary central nerve system lymphoma, mostly presenting later in the disease course. Only 3 cases of Non-Hodgkin's lymphoma were found presenting with multiple cranial nerve deficits, two B-cell lymphomas (3,4) and one T-cell lymphoma (5). Neurologic signs and symptoms at presentation may be highly diverse and there seems to be a predilection for the cranial nerves III, IV, VI and VII. (3)

This case shows that recurrent pancreatitis is a diagnostic challenge and the combination with weight loss and cranial neuropathy should raise suspicion of malignant invasion of a lymphoma.

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