中文題目：胰臟神經內分泌瘤肝脾轉移患者呈現高血胺昏迷一例報告

英文題目：A rare presentation of hyperammonemia encephalopathy in a patient with pancreatic neuroendocrine tumor with liver/spleen metastases - a case report

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Introduction:

We report here a rare presentation of hyperammonemia encephalopathy in a patient of pancreatic neuroendocrine tumor with liver and spleen metastases, whose liver function is still in Child-Pough class A . The developing of hyperammonemia is probably due to tumor porta-systemic shunting instead of disease progression. this rare presentation should be kept in mind in treating the neuroendocrine tumors.

Case report:

A 63 years old female came to our ER due to chronic diarrhea for 2 months and body weight loss 4 kg. The admission examination unveil she is a victim of pancreatic neuroendocrine tumor gr 2, stage IV, with liver and spleen metastases. The neuroendocrine tumor is secretory type , with severe symptoms of diarrhea (>10 times/day) , urine 5-HIAA 4.18mg/day (2-8 mg/day), chromogranin 285.9 ng/mL (19.4-98.1 ng/mL); but her liver function is good : GOT 23 IU/L, GPT 23 IU/L , T.Bilirubin 0.4mg/dl , albumin 3.6 gm/dL , PT 9.9 sec , INR 0.92 , HBsAg (-) , Anti-HBs (+) , anti-HCV (-); while ammonia is the only data abnormally elevated : 46 umol/L (9-33). Because there were no encephalopathy and no ascites , her Child-Pough score is 5 , class A. Her other lab data are normal , including WBC 5.74 x109/L , Hb 12.8 g/dL , Platelets 163 x109/L, normal renal function, and normal stool & urine examinations.

After confirmation of the diagnosis , we give her everolimus 10 mg/day. On day 24 of using everolimus , she was sent to our ER due to nausea vomiting , and consciousness disturbance. Laboratory data revealed glucose 205 mg/dl , Ca14.3 mg/dl , K 3.4 mmol/L, Na137 mmol/L , GOT/GPT 42/37 IU/L , Albumin 4.3 mg/dL, T. Bilirubin 0.8 mg/dL , BUN/Cr 39/1.31 mg/dL, ammonia was checked and was elevated up to 103 mmol/L. The Glasgow coma scale was E3V4M5.

Under the impression of hyperammonemia encephalopathy , we treat her with lactulose , and in the meantime start the treatment of octreotide (30 mg) .

Her ammonia recovered soon(fig.3), coma scale returned to E4V5M6 and the diarrhea also completely recovered to once/day one week later.

The patient now receiving everolimus and octreotide therapy, the clinical condition is stable now.

Discussion:

The hyperammonemia or hyperammonemia encephalopathy is a rare event in neuroendocrine tumor patients, especially when the patient had no cirrhosis of liver. Several case reports in literature had attributed the cause to portal-systemic shunt (1,2). In our case, This patient had multiple small liver metastases, but the spleen hilum vessels and gastric vessels are heavily engorged (fig.1,2), indicating there might be a porta-systemic shunt, causing hyperammonemia. The patient later developed encephalopathy , we figure that the target therapy drug work out slowly , and the drug might causing patient loss appetites, dehydration and slowly developed the encephalopathy as ammonia gradually elevated. Afterward, the combination therapy worked, the tumor being controlled , ammonia level returns normal.

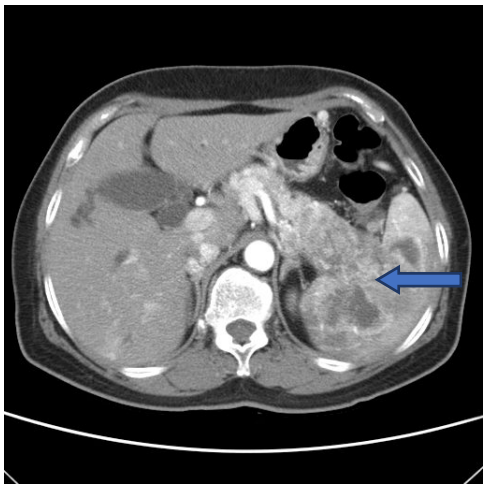

Fig 1. Splenic hila tumor , vessels

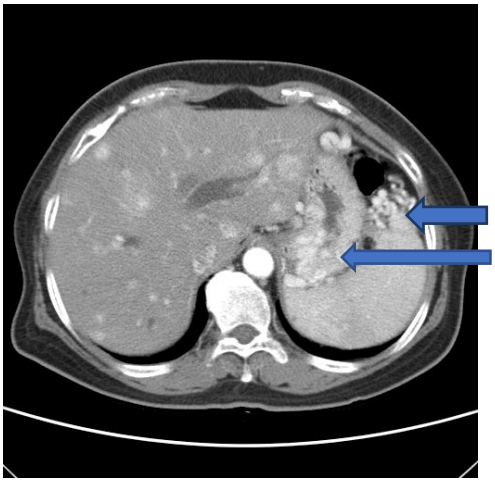

Fig 2 Engorged vessels

Fig 3 Ammonia changes

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