This pathological anatomy revealed a case of gastric plexiform fibromyxoma, an angio-myxoid plexiform myofibroblastic tumor which is a benign tumor that has recently been defined as a multinodular myxoid tumor with a peculiar plexiform growth pattern, myxoid stroma, prominent vasculature, and spindle cells with myofibroblastic differentiation.

This type of tumor was recently characterized by Takahashi et al.[1] and Miettinen et al.,[2] and 60 cases of plexiform fibromyxoma including the present case have been reported so far.[3] According to previous reports, this tumor can occur at any age range, 7–75 years, typically middle age, and has a roughly equal gender distribution. The clinical presentation is generally similar to that of GISTs, with hematemesis being the most common symptom.[4] In some rare cases, pyloric obstruction with weight loss may be noted. This tumor is predicted to exhibit benign biological behavior and there have been no reported cases of local recurrence or distant metastases after resection with a margin of normal tissue.

In conclusion, the current article reports a case of plexiform fibromyxoma, a rare mesenchymal gastric neoplasm that requires distinction from the others gastric mesenchymal tumors. Because of this, it is very important we know its symptoms, endoscopic and radiological images and its immunohistochemical in order to make an better diagnosis and treatment.

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Anti-ammonia treatment-responsive myoclonus as initial presentation of acquired hepatocerebral degeneration

Mioclono que respondió al tratamiento antiamonio como presentación inicial de la degeneración hepatocerebral adquirida

Acquired hepatocerebral degeneration (AHD) is an often debilitating neurological disorder characterized by a variety of movement disorders in the setting of chronic liver disease (CLD) and portosystemic shunt. AHD was first described by van Woerkem in 1914.[1] Cirrhosis-related parkinsonism is the core manifestation in AHD and has been described to have a prevalence of 4.2%.[2] Reports of myoclonus in patients with AHD are scarce[3] and they always present along with other clinical manifestations, most often parkinsonism and cerebellar signs. Liver transplantation is presently the only therapeutic option that has been shown to ameliorate and even reverse neurological deterioration.[4] Here, we describe the case of a 57-year-old woman with an unusual presentation of AHD who responded well to treatment with anti-ammonia therapy.

A 57-year-old woman was admitted to the emergency department because of altered mental status and abnormal upper-limbs movements. She had a history of primary biliary cirrhosis and reported no alcohol consumption. Viral markers for HIV, hepatitis B and C were all negative. She also had a history of esophageal varices and had a previous diagnosis of splenorenal shunt, which was detected by a contrast-enhanced abdominal computed tomography scan. Her regular medication included propranolol and ursodeoxycholic acid. In the past she has had several episodes of acute hepatic encephalopathy (AHE); these episodes have been successfully treated, in the outpatient setting, with oral L-ornithine-L-aspartate (LOLA) and oral lactulose.

On admission our patient had an altered mental status. On physical exam, there was no jaundice, telangiectases, ascites, superficial collateral abdominal veins or hepatosplenomegaly. She had sudden, brief, shock-like jerks of her upper limbs consistent with myoclonus. Hyperreflexia was also found in the same limbs, and Babinski sign was elicited bilaterally. Kayser–Fleischer rings were not seen on ophthalmologic examination. She initially received lactose enemas as treatment for AHE, with improvement

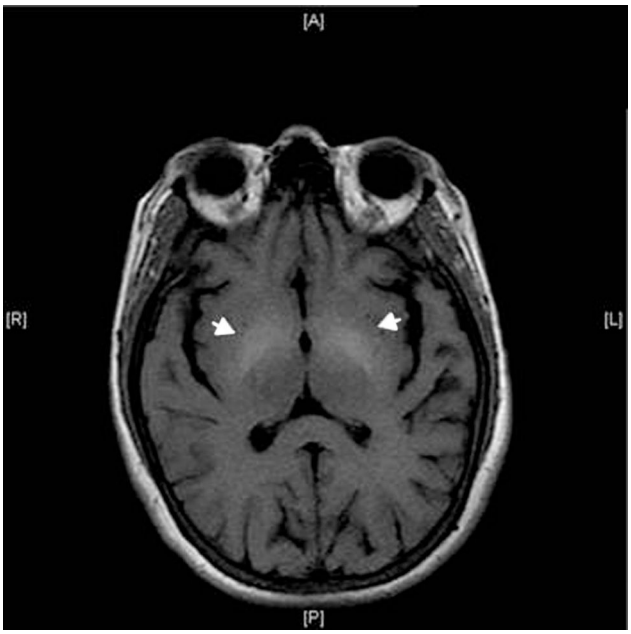

Figure 1 Axial T1-weighted brain magnetic resonance imaging. Symmetrical hyperintensities in lentiform nuclei (arrows).

in her mental status. However, there was no change in her myoclonus. A brain magnetic resonance imaging (MRI) scan revealed bilateral symmetrical hyperintensities in lentiform nuclei on T1-weighted images (Fig. 1). She had a mild thrombocytopenia (148 K/mm^3) but neither anemia nor leukocytosis was found. She was euglycemic, and renal function was normal. Serum electrolytes and coagulation tests were non-contributory. She was not hypoalbuminemic but had a mild hyperbilirubinemia (total: 2.2 mg/dL; conjugated: 0.6 mg/dL); other liver function tests were as follow: serum glutamic–oxaloacetic transaminase 88 U/L (10–42 U/L), serum glutamate-pyruvate transaminase 42 U/L (10–42 U/L), alkaline phosphatase 134 U/L (38–126 U/L).

Based on MRI findings, we diagnosed AHD and ammonia-lowering therapy was continued to treat her myoclonus, since liver transplantation was not a practical possibility in our setting. The patient showed a mild improvement during her hospital stay. When her mental status normalized, after one week of treatment, she was discharged on oral lactulose (20 g thrice daily), rifaximin (400 mg twice daily) and LOLA (6 g thrice daily). She was then evaluated weekly for tolerability and adherence. One month later she presented to the office with complete resolution of her myoclonus, and we decided to continue the same treatment. After 18 months of follow up, the patient continues with ammonia-lowering therapy and has not had any recurrence of movement disorders or encephalopathy.

CLD may be associated with a wide variety of motor and neuropsychiatric manifestations as a result of the diversion of portal blood flow to the systemic circulation, presumably through toxic effects of chemical substances that cross the blood–brain barrier. AHD is a chronic encephalopathy characterized by cognitive impairment, parkinsonism, and other movement disorders besides myoclonus, including ataxia, chorea, and dystonia.[5,6] Pathophysiological mechanisms are poorly understood and many factors are probably involved. Ammonia and manganese are both candidate substances for chronic neurological dysfunction in liver diseases, and more recently it has been demonstrated that there is a pathologically decreased striatal dopamine D2 receptor availability and decreased dopamine transporter availability in the pathogenesis of cirrhosis-related parkinsonism.[7] Brain MRI typically showed high intensity signal in the lenticular nuclei on T1-weighted images, which differentiates this entity from AHE.[8]

Although chronic AHD can present with almost any kind of movement disorder, myoclonus is an extremely rare symptom that has been very seldom reported.[6] At least one other case has been described in the literature, involving a patient with Budd-Chiari syndrome and T1 hyperintensities in pallidum and substantia nigra.[9] In CLD and portosystemic shunt, ammonia toxicity and manganese accumulation in the mitochondria of glial cells in the pallidum and other basal ganglia structures may lead to disruption of energy metabolism and the characteristic imaging findings.[5,7,10] In general, dopaminergic treatment is ineffective, manganese chelation unavailable, and anti-ammonia therapy often without any substantial benefit, leaving liver transplantation as the most reasonable alternative.[2–5,7] In our case, anti-ammonia treatment proved successful, suggesting that this therapy should be attempted in settings where liver transplantation is not possible.

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Small bowel obstruction secondary to distal migration of the intestinal probe in a patient with an endoscopic gastrostomy for continuous carbidopa-levodopa infusion

Obstrucción de intestino delgado secundaria a migración distal de la sonda intestinal en una paciente con gastrostomía endoscópica para la infusión continua de carbidopa-levodopa

Levodopa is the gold standard treatment for Parkinson's disease (PD). However, due to its short plasma half-life, requiring repeated doses, long-term levodopa use (usually within 5–10 years after initiating treatment) is associated with motor fluctuations and dyskinesias. Impaired gastric emptying is also common in PD patients and leads to unpredictable responses to oral medication. These facts induce disability and lower quality of life.[1] Therapeutic alternatives for patients with advanced PD include deep brain stimulation, continuous subcutaneous apomorphine infusion and intrajejunal levodopa-carbidopa infusion. Continuous intraduodenal/intrajejeunal infusion of a levodopa-carbidopa (L-C) gel (Duodopa®, Abbvie) – which contains levodopa 20 mg/ml plus carbidopa 5 mg/ml – improves motor fluctuations[2,3] (*off* time and severity and dyskinesia), non-motor symptoms,[4] autonomy for basic activities[5] and quality of life.[6] This therapy is performed by placing a specific percutaneous endoscopic gastrostomy (PEG) kit (Frecka® PEG gastric set) that contains inside a 9Fr duodenal pigtail (Freka® CH9 intestinal tube) probe. This technique requires cooperation between the neurologist that sets the indication for this treatment and does a close surveillance of the patient, and the Gastroenterologist/Endoscopist that places the PEG probe and performs the surveillance and replacements of the device.

A 76 year-old woman with advanced PD complicated with motor fluctuations required Duodopa® administration to control her motor symptoms. Therefore, a PEG-Duodopa was placed, with Propofol sedation, after informing the patient and her family about the benefits and the possible risks related to the procedure. Thirty minutes before the PEG-duodopa allocation, the patient received antibiotic prophylaxis with Cefazolin 2 g. It was performed without any immediate complications and lack of complications was verified by immediate endoscopic control, as usual. The patient was discharged from hospital 2 days after the procedure, with marked improvement of the motor symptoms, since PEG-duodopa was used for the first time 24 h after its allocation. After eleven days, the patient was admitted to the emergency room because of abdominal distension with pain and vomiting, which was highly suggestive of an intestinal obstruction. Plain abdominal X-ray showed the duodenal probe tip of the Freka® CH9 Intestinal Tube projected in right iliac fossa and absence of pneumoperitoneum. Severe abdominal pain persisted despite administration of analgesics, so an urgent abdominal CT was indicated to discard any other complication. It confirmed the presence of the pigtail probe coiled in the ileum due to distal migration, conditioning severe retrograde small bowel dilation (Fig. 1). Since the patient had no response to conservative treatment, emergency surgery was indicated, performing an ileostomy, extraction of the probe and primary suture without associated complications. Ten days after surgery, after discussing

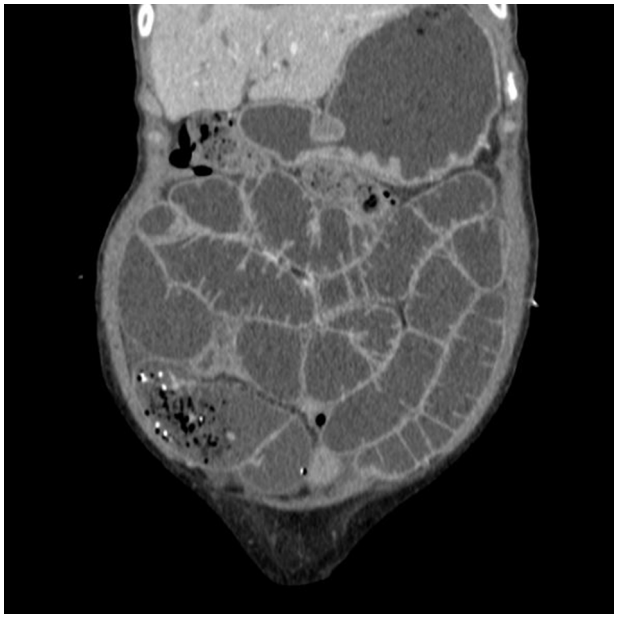

Figure 1 TC. Pigtail probe coiled in distal ileum due to migration, conditioning severe retrograde small bowel dilation.