6. Azad AF, Radulovic S. Pathogenic rickettsiae as bioterrorism agents. Ann N Y Acad Sci. 2003;990:734–8.
7. Oristrell-Salvà J, Sampere M, Amengual MJ, Font B, Segura F. Levels in mediterranean spotted fever. Eur J Clin Microb Infect Dis. 2004;23:417–8.
8. Cardeñosa N, Sanfeliu I, Segura F. Diagnóstico microbiológico de las rickettsiosis. Enferm Infecc Microbiol Clin. 1997;15:32–7.

Marta Magaz Martínez[a,*], Antonio Ramos[b], Jose Masegosa[a], Luis Abreu[a]

[a] *Aparato Digestivo, Hospital Universitario Puerta de Hierro-Majadahonda, Majadahonda, Madrid, Spain*
[b] *Unidad de Infecciosas, Hospital Universitario Puerta de Hierro-Majadahonda, Majadahonda, Madrid, Spain*

[*] Corresponding author.
E-mail address: martamagazm@gmail.com
(M. Magaz Martínez).
2444-3824/

Plexiform fibromyxoma, a rare mesenchymal gastric tumor

Fibromixoma plexiforme, un tumor gástrico mesenquimal poco frecuente

Plexiform angiomyxoid myofibroblastic tumor, also known as plexiform fibromyxoma, is a novel rare entity of gastric mesenchymal tumors, typical of gastric antrum, and commonly causing mucosal ulceration with upper gastrointestinal bleeding and anemia, and effectively treated by complete surgical resection usually accomplished by distal gastrectomy.

We report one recent patient from our center meeting clinical and histopathologic criteria compatible with plexiform fibromyxoma.

A 37-year-old male patient with no history of interest was admitted with upper gastrointestinal bleeding with hemodynamic instability. A total of three esophago-gastroduodenoscopies were performed and we could observe a five centimeter-violaceous lesion with antral location. At first it seemed like a blood clot, but later it was checked a greater consistency which was different than a blood clot and more similar to an antral vascular neoformation (Fig. 1)

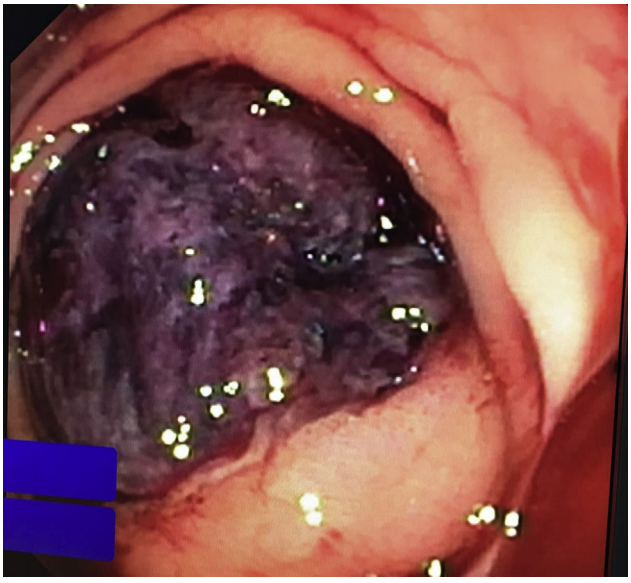

Figure 1 Endoscopic antral lesion.

Abdominal ultrasonography and abdominal contrast-enhanced computed tomography reported an irregular rounded heterogeneous lesion in gastric antrum and acute bleeding areas suggestive of hematoma, with a size of $5.8 \times 4 \times 5$ cm. However they could not check an underlying lesion or associated thickened gastric wall. After a new episode of hemodynamic instability, the patient underwent emergency surgical intervention with an antrectomy (Fig. 2). Postoperative period was uneventful and the patient was discharged on postoperative day 9.

Histopathological examination revealed partial dense collagenous matrices and networks of fine capillary-caliber blood vessels. The tumor demonstrated lobular or fused nodular growth of spindle cells without atypical cytology, with abundant alcian blue-positive myxoid extracellular matrix. Hematoxylin and eosin staining showed lobulated or fused multinodular growth. Immunohistochemically it demonstrated an expression of muscle specific actin, desmin, and immunoexpression of CD10, and it was negative for CD31, CD34, VIII Factor, S100, Ckit, DOG1, HHV8, ALK, MDM2, CD23.

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.

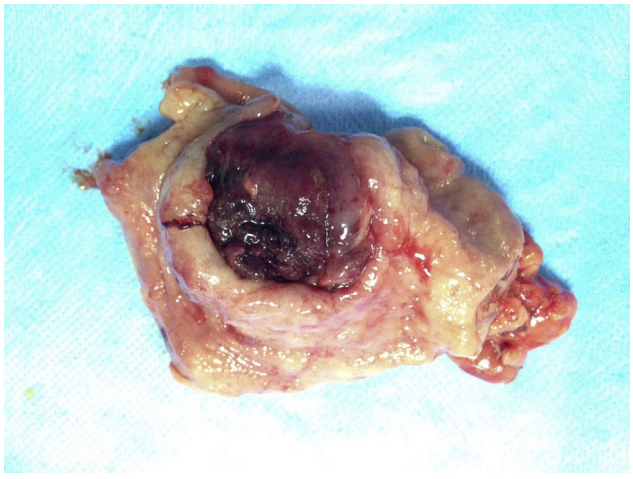

Figure 2 Surgical piece.

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.

References

1. Takahashi Y, Shimizu S, Ishida T, Aita K, Toida S, Fukusato T, et al. Plexiform angiomyxoid myofibroblastic tumor of the stomach. Am J Surg Pathol. 2007;31:724–8.
2. Miettinen M, Makhlouf HR, Sobin LH, Lasota J. Plexiform fibromyxoma: a distinctive benign gastric antral neoplasm not to be confused with a myxoid gist. Am J Surg Pathol. 2009;33:1624–32.
3. Quero G, Musarra T, Carrato A, Fici M, Martini M, Dei Tos AP, et al. Unusual focal keratin expression in plexiform angiomyxoid myofibroblastic tumor: a case report and review of the literature. Medicine (Baltimore). 2016:95.
4. Vallejo-Benítez AM, Suárez-Aguado J, Mora-Cabezas M, Aramendi T, Gimeno M, Salas J, et al. Fibromixoma plexiforme del antro gástrico: descripción de dos casos y revisión de la literatura. Rev Esp Patol. 2013;46:93–100.

Pedro Luis Gonzalez-Cordero[a,*], Daniel Vara-Brenes[a], María del Carmen Pecero-Hormigo[b], Jose María Mateos-Rodríguez[a], Javier Molina-Infante[a], Yvana Anavy Martínez Mateo[c], Miguel Fernández-Bermejo[a]

[a] *Department of Gastroenterology, Hospital San Pedro de Alcantara, Caceres, Spain*
[b] *Internal Medicine, Hospital San Pedro de Alcantara, Caceres, Spain*
[c] *General Surgery, Hospital San Pedro de Alcantara, Caceres, Spain*

* Corresponding author.
E-mail address: pelu7_8@hotmail.com (P.L. Gonzalez-Cordero).
2444-3824/

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 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;