classification and management strategies. J Am Coll Surg. 2014;219:704–12.
5. Lawrence C, Howell DA, Stefan AM, Conklin DE, Lukens FJ, Martin RF, et al. Disconnected pancreatic tail syndrome: potential for endoscopic therapy and results of long-term follow-up. Gastrointest Endosc. 2008;67:673–9.
6. Solanki R, Koganti SB, Bheerappa N, Sastry DA. Disconnected duct syndrome: refractory inflammatory external pancreatic fistula following percutaneous drainage of an infected peripancreatic fluid collection. A case report and review of the literature. JOP. 2011;12:177–80.
7. Varadarajulu S, Wilcox CM. Endoscopic placement of permanent indwelling transmural stents in disconnected pancreatic duct syndrome: does benefit outweigh the risks? Gastrointest Endosc. 2011;74:1408–12.
8. Ramia JM, Fabregat J, Pérez-Miranda M, Figueras J. Síndrome del duto pancreático desconectado. Cir Esp. 2014;92:4–10.

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Bilateral Intrahepatic Pancreatic Pseudocyst☆

Seudoquiste pancreático intrahepático bilateral

Pancreatic pseudocysts are a frequent complication of pancreatitis. The majority are situated in the head or body of the pancreas, but 20% can be extra pancreatic (pleura, mediastinum, pelvis, spleen). Hepatic localization is exceptional, and only 35 cases have been reported in the literature, only 7 of which had bilobar involvement.

We present the case of a 59-year-old woman with a history of chronic alcoholism and recent hospitalization for re-exacerbated chronic pancreatitis. CT detected multiple calcifications in the head of the pancreas and intra-pancreatic pseudocysts as well as secondary pancreatic ascites. The patient was managed conservatively with a positive response to medical treatment.

The patient came to the emergency room one month after discharge, once again presenting symptoms of epigastric abdominal pain that irradiated toward the back and had been progressing over the previous three days. Emergency work-up showed evidence of leukocytosis (14 000/mcl) with neutrophilia and hyperamylasemia (614 IU/L), but no alterations in the hepatic profile. Abdominal physical examination showed signs of ascites. Diagnostic paracentesis defined amylase levels in the ascitic fluid of 14 778 IU/L. Based on a diagnostic impression of re-exacerbated chronic pancreatitis and pancreatic ascites, parenteral nutrition, diuretics and octreotide were commenced.

A second CT scan showed ascites, coarse calcifications in the head of the pancreas with retrograde dilatation of the pancreatic duct, and 2 pseudocysts in the body measuring 12 mm×11 mm and 22 mm×16 mm, which were connected by a tubular formation with a large bilobar hepatic lesion that was cystic and multiseptated, measuring 13 cm×10 cm. Hypertrophy of the posterior hepatic segments was also detected (Figs. 1 and 2).

In the context of chronic pancreatitis with pancreatic ascites, pancreatic pseudocysts are considered the probable origin of hepatic lesions, although *a priori* other causes cannot be ruled out, such as metastasis. Therefore, ultrasound-guided diagnostic fine-needle aspiration was used. The amylase levels in the fluid was 49 180 IU/L, and the pathology study was negative for malignancy, with normal hepatocytes.

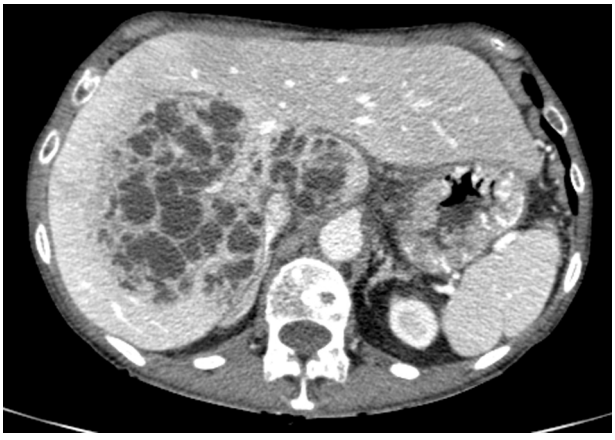

Fig. 1 – Axial CT scan: multiseptated bilobar hepatic cystic lesion.

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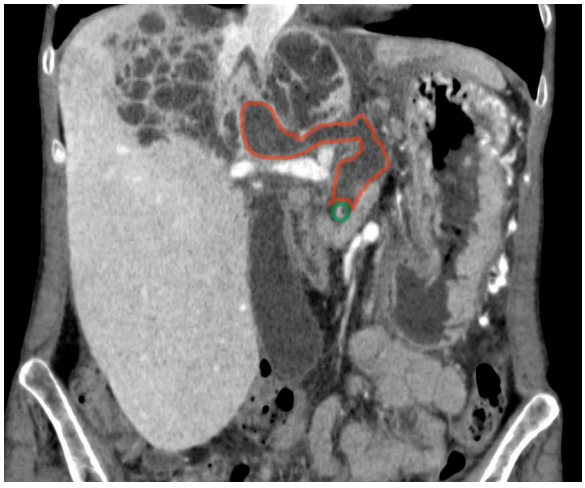

Fig. 2 – Coronal CT scan: fistula communicating the pancreatic duct with a hepatic cyst lesion, marking the perimeter; intra-pancreatic lithiasis marked by a circle.

With the diagnosis of intrahepatic pancreatic pseudocysts connected to the pancreatic duct, endoscopic retrograde cholangiopancreatography (ERCP) was ordered with papillotomy for drainage. Biliary sludge was found along with 2 calculi in the main pancreatic duct, which could not be cannulated. After this test, the patient presented improved symptoms and lab work, with a later CT scan that revealed several pancreatic pseudocysts, one of which was at the *porta hepatis*, connected to the main pancreatic duct. The intrahepatic pseudocyst and ascites were completely resolved, so the patient was discharged from the hospital.

Outpatient CT scan detected a decrease in the pancreatic pseudocyst with a large calcification in the head of the pancreas and persistent dilatation of the Wirsung duct, which could not be cannulated in the previous ERCP, so a surgical intervention was scheduled. The pseudocyst was drained with a longitudinal opening along the pancreatic duct. Several calculi were found, and we located the pseudocyst drainage orifice. Surgery was completed with a Roux-en-Y longitudinal pancreatojejunostomy, using the Partington–Rochelle technique.

Postoperative hospital stay was 15 days. The patient was administered 2 units of packed red blood cells due to postoperative anemia, with no other complications.

After 6 years of follow-up, the patient remains asymptomatic, with good endocrine and exocrine pancreatic function.

Intrahepatic pancreatic pseudocysts are more frequent in men (3:1) of middle age, associated with symptoms of acute pancreatitis (75%), and as a complication of exacerbated alcoholic chronic pancreatitis (50%).[1] Generally, they are multiple, more frequent in the left hepatic lobe and subcapsular, and their connection to the pancreas is the hepatogastric ligament, by which the pancreatic liquid discharged into the omental bursa reaches the liver.[1–4] Their localization in the right liver is less frequent, where the hepatoduodenal ligament would be the route of access, forming predominantly intraparenchymal pseudocysts.[1–3,5–9] Bilobar localization, as in our case, has been described in the literature in only 7 cases.[1,5,9]

Clinical symptoms are similar to those of pancreatitis,[1,2,5] involving epigastric pain and the palpation of a mass or hepatomegaly in some cases. The analytical findings usually reveal elevated serum amylase and lipase levels, although transaminase levels remain within normal ranges.[1,2,5]

In cases of acute pancreatitis, some type of imaging technique (generally CT) is usually sufficient to diagnose hepatic pseudocyst, as it shows abnormalities in the pancreas and intrahepatic hypodense cystic lesions.[1,9] Other causes should be contemplated in the differential diagnosis by CT scan, including neoplasms.[2,5]

The aspirated intracystic fluid is used to determine amylase levels and thereby confirm the suspected diagnosis, which is especially useful when tomography shows no evidence of pancreatic alterations. Furthermore, a study of cellularity is able to rule out neoplasms.[1,2,5,7,9]

No consensus has been reached to establish clinical guidelines for the management of intrahepatic pancreatic pseudocysts.[1–3,5] Although about half of pseudocysts disappear spontaneously, those located in the liver are generally treated with percutaneous, surgical or endoscopic drainage according to their communication with the pancreatic duct and the existence of duct lesions.[3,5] However, a small number of cases resolve spontaneously.[1,2,4,5] While the need for drainage is accepted when there are symptoms caused by compression or complications (rupture, infection or hemorrhage),[2] no clear criteria have been established for its indication, although percutaneous drainage is extensively used as it provides diagnostic confirmation and treatment.[1,5]

REFERENCES

1. Fethi G, Ayoub Z, Yosri S, Nabil N, Chedli D. Pancreatic pseudocysts located in the liver: a systematic review of literature. Tunis Med. 2009;87:801–4.
2. Kibria R, Akram S, Ali SA. Successful endoscopic transpapillary management of intrahepatic pancreatic pseudocyst. JOP. 2010;11:41–4.
3. Bhasin DK, Rana SS, Chandail VS, Nanda M, Nadkarni N, Sinha SK, et al. An intra-hepatic pancreatic pseudocyst successfully treated by endoscopic transpapillary drainage alone. JOP. 2005;6:593–7.
4. Al Ani R, Ramadan K, Abu-Zidan FM. Intrahepatic pancreatic pseudocyst. N Z Med J. 2009;122:75–7.
5. Les I, Córdoba J, Vargas V, Guarner L, R Boyé R, Pineda V. Pseudoquiste pancreático de localización hepática. Rev Esp Enferm Dig. 2006;98:616–20.
6. Ancel D, Lefebvre M, Peyrin-Biroulet L, Chone L, Sido A, Regent D, et al. Pancreatic pseudocysts of the right hepatic lobe during acute biliary pancreatitis. Gastroenterol Clin Biol. 2005;29:743–5.
7. Balzan S, Kianmanesh R, Farges O, Sauvanet A, O'Toole D, Levy P, et al. Right intrahepatic pseudocyst following acute pancreatitis: an unusual location after acute pancreatitis. J Hepatobil Pancreat Surg. 2005;12:135–7.
8. Martínez-Sanz N, González-Valverde FM, Vicente-Ruiz M, Pastor-Pérez P, Ruiz-Marín M, Albarracín-Marín-Blázquez A. Intrahepatic pancreatic pseudocyst: a case report. Rev Esp Enferm Dig. 2015;107:249–50.

9. Baydar B, Cantürk F, Alper E, Aslan F, Akpınar Z, Cengız O, et al. Intrahepatic localization of pancreatic pseudocyst: a case report. Turk J Gastroenterol. 2013;24:447–9.

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Spontaneous Rupture of a Splenic Aneurysm in Classic Ehlers–Danlos Syndrome☆

Rotura espontánea de aneurisma de arteria esplénica en el síndrome de Ehlers–Danlos tipo clásico

Ehlers–Danlos syndrome (EDS) is a group of rare hereditary disorders (1:5000 newborns) that are characterized by cutaneous hyperextensibility, joint hypermobility and connective tissue fragility. Within the forms of presentation, aneurysmal rupture is rare; while it is a characteristic feature in the vascular variation, it is exceptional in other types.[1] We present the case of a 38-year-old woman diagnosed with classical EDS as a result of hemoperitoneum secondary to the rupture of a giant aneurysm of the splenic artery.

The patient came to the emergency room with abdominal pain and vomiting that was not associated with any previous trauma. Her medical history included renal failure and autoimmune hypothyroidism. Upon arrival to the ER, the patient was hemodynamically unstable. Physical examination detected a distended abdomen that was painful to palpation, associated with splenomegaly. Lab work showed hematocrit 10% and hemoglobin 3.3 g/dl. Abdominal CT scan (Fig. 1) and angiography (Fig. 2) revealed a large aneurysm 4 cm from the origin of the splenic artery and 2 pseudoaneurysms located in the splenic hilum. The largest was embolized with thrombin and metallic coils. In spite of the interventionist treatment, the patient remained hemodynamically unstable, so we decided to operate. We discovered a hemoperitoneum of more than 2 l with splenomegaly (approximately 30 cm) and a pseudoaneurysm over the body of the pancreas that was partially ruptured at the inferior pole of approximately 10 cm. Hilar dissection was conducted with transfixing ligatures and a standard splenectomy was performed. The patient progressed favorably and was discharged on the tenth day post-op.

The patient presented leptosomic appearance and a marfanoid phenotype, so she was referred to the medical genetics department. Genetic studies detected a mutation in the COL5A1 gene (c.1588G>A) that, at the protein level, changes the glycine in position 530 for serine (p.Gly530ser).

According to Villefranche, EDS is classified into 6 main subtypes. The classical type, the type associated with hypermobility and the vascular type are the most common, while kyphoscoliosis, arthrochalasis and the dermatosparaxis types are less common conditions. Most EDS forms correspond with mutations in genes coding for collagen chains or enzymes involved in the biosynthesis of these proteins.[1] In the classic type (around 50% of cases), the defect is caused by mutations in the genes coding for V alpha-1 (COL5A1) or alpha-2 (COL5A2) collagen.[2,3] When EDS is suspected, we should review the patients' medical history for complications, such as frequent hematomas, cervical insufficiency, anal prolapse in childhood, Premature rupture of membranes, vaginal lacerations, low lung capacity or murmurs.[3,4] The frequency and

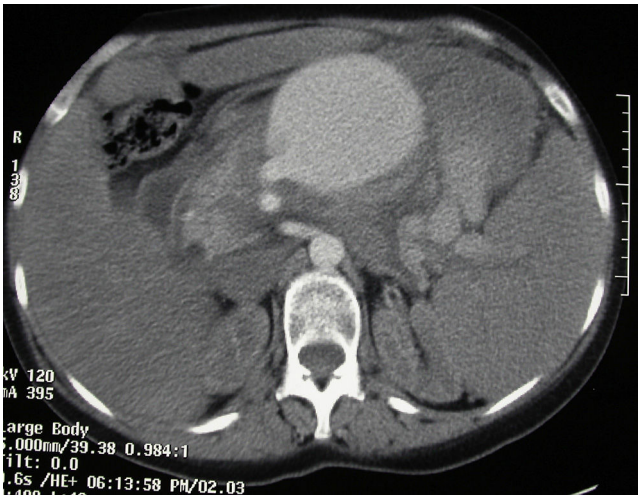

Fig. 1 – Aneurysm of the splenic artery on abdominal CT scan.

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