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# Autoimmune pancreatitis in pediatrics, a rare disease☆

## Pancreatitis autoinmune en pediatría, una enfermedad poco frecuente

Autoimmune pancreatitis (AIP) is a systemic fibroinflammatory disease that affects the pancreas and other organs[1]. Prevalence in the pediatric population is low and the most frequent clinical presentation is abdominal pain, obstructive jaundice, and weight loss[2].

We present herein the case of a 15-year-old adolescent girl, with an unremarkable past medical history, who was seen by her family physician for chronic, intermittent, colicky abdominal pain of moderate intensity, located in the right upper quadrant, accompanied by nausea of one-month progression. Ultrasound imaging identified multiple stones, measuring 2–3 mm, that occupied 70% of the gallbladder lumen, for which she was referred to the pediatric surgery department. She underwent laparoscopic cholecystectomy, revealing a fibroinflammatory process with an epiploic mass. The patient presented with postoperative intense abdominal epigastric pain, with vomiting and elevated pancreatic enzymes (amylase 1,006 U/l and lipase 1,271 U/l), and a computed tomography scan showed edema of the pancreas (Balthazar B). Magnetic resonance cholangiography revealed no evidence of stones and endoscopic retrograde cholangiopancreatography showed no evidence of stones in the pancreatic duct. The patient also presented with biloma that merited drainage performed by an interventional radiologist. The patient continued to receive parenteral nutrition and antibiotics, and lastly, was discharged with no apparent complications. One month later, she again arrived at the emergency department due to abdominal pain and vomiting. Laboratory test results revealed elevated pancreatic enzymes (amylase 1,248 U/l and lipase 3671 U/l), and an abdominal computed tomography scan showed pancreatic edema and a single peripancreatic collection (Balthazar D). Given the progression time, a new pancreatitis event was diagnosed. A recurrent pancreatitis approach was started, with a new magnetic resonance cholangiography study showing the single finding of segmental irregularity of the pancreatic duct (Fig. 1). Due to the suspicion of AIP, immunoglobulin subclasses were ordered, resulting in IgG4 levels of 221 mg/dl (normal value: <140 mg/dl) and negative antinuclear antibodies. The patient was started on immunosuppressive therapy with prednisone (prednisone 0.6 mg/kg/day). At follow-up at more than 6 months, she presented with clinical improvement and has developed no other events of pancreatitis.

Recurrent acute pancreatitis is characterized by the presentation of 2 episodes of acute pancreatitis, with complete pain resolution and pancreatic enzyme normalization between the two episodes. Recurrent acute pancreatitis occurs in 15–20% of patients after a first event of the disease, and even though AIP is rare in the pediatric population, it should be considered an etiologic possibility[3].

Two forms of AIP are currently recognized. Type 1 AIP is a multisystemic disease that affects the pancreas (IgG4-related systemic disease) and type 2 AIP appears to be an autoimmune disease that specifically affects the pancreas[4]. In pediatrics, the study of AIP is limited to case series, and so its prevalence is unknown. Our patient presented with abdominal pain, which is found in up to 91% of children with AIP, but she did not have obstructive jaundice, which is reported in 42%[2], signifying that clinical presentation can be heterogeneous. Gallstones are not seen in AIP, and a study that evaluated biliary tract involvement in adults with AIP reported the presence of bile duct stricture and the thickening of the bile duct wall and gallbladder wall as the most frequent findings[5]. Likewise, only 22% of children are likely to have elevated IgG4 levels[2]. That finding guided the diagnosis of our patient, in addition to the magnetic resonance images of the pancreas. She presented with the remission of pain after the administration of the steroid, thus meeting 3 of the criteria for the diagnosis of AIP, according to the International Consensus Diagnostic Criteria for Autoimmune Pancreatitis[6], which includes one or more of the following 5 characteristics:

1. Imaging studies: (a) pancreatic parenchyma (on computed tomography or magnetic resonance imaging) and

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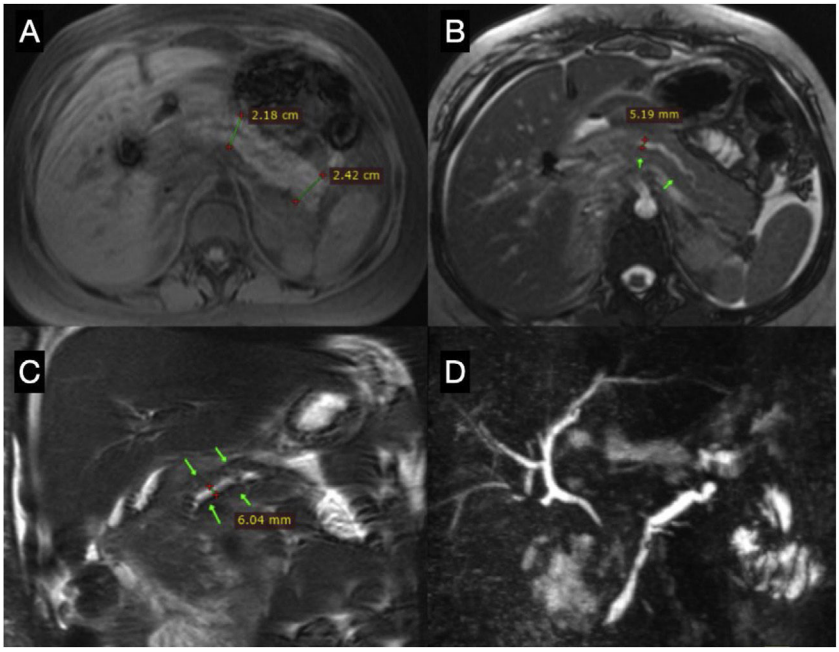


**Figure 1** Magnetic resonance cholangiography. (A) T1-weighted axial view, showing the enlarged body and tail of the pancreas. (B) T2-weighted axial view, showing the body of the pancreas, with irregular dilation of the main pancreatic duct. (C) T2-weighted coronal view, showing the dilation of the main pancreatic duct. (D) MIP reconstruction of the cholangiopancreatography, showing irregular dilation of the main pancreatic duct and normal intrahepatic bile ducts.

(b) pancreatic duct (endoscopic retrograde cholangiopancreatography or magnetic resonance cholangiography).
2. Serology (IgG4, IgG, and antinuclear antibodies).
3. Involvement of other organs.
4. Histopathology of the pancreas.
5. Response to steroid therapy.

Due to previous abdominal interventions and the clinical, imaging, and biochemical characteristics consistent with AIP, we decided to forego pancreatic biopsy in our patient, in addition to the fact that endoscopic ultrasound imaging is not available at our center.

In conclusion, AIP should be studied in children with recurrent acute pancreatitis. The clinical manifestations can be heterogeneous. Even though the frequency of the disease in the pediatric population is low, multicenter studies are needed to characterize the clinical presentation, diagnosis, and progression of AIP in children.

## Ethical considerations

The authors obtained a written statement of informed consent from the patient described in this article and the document is in the possession of the corresponding author. The present work meets the current bioethical research norms, and because of the type of publication, does not require approval from the hospital's ethics committee. The authors declare that this article contains no personal information that could identify the patient.

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## Conflict of interest

The authors declare that there is no conflict of interest.

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# Primary intestinal follicular lymphoma: A case report☆

## Linfoma folicular primario intestinal: reporte de un caso

The most common extranodal location of lymphoma is the gastrointestinal tract. The most frequent lymphomas are the large B-cell type, followed by MALT-type lymphoma.[1] Primary intestinal follicular lymphoma is rare, occurring in 1–3% of all B-cell non-Hodgkin's lymphomas.[2]

We present herein the case of a 44-year-old man who presented with 6-month progression of epigastric pain, associated with nausea and a 5 kg weight loss. Laboratory tests showed 8,170 cells/mm$^3$ with 22% lymphocytes, Hb 14.7 g/dl, LDH 165 IU/l, and beta 2 microglobulin 1.2 mg/l. Endoscopy revealed a multifocal nodular pattern in the duodenum (Fig. 1a). Lower endoscopy showed a nodular pattern and an ulcer in the distal ileum (Fig. 1b), as well as an irregular mucosal area in the cecum. Tomography revealed mesenteric and retroperitoneal adenopathies and thickness of the cecal wall (Fig. 2a). The biopsy result was a structurally atypical lymphoid infiltrate with a nodular pattern (Fig. 2b). Immunohistochemistry was positive for CD20, CD10, BCL2 (Fig. 2c), and BCL6 (heterogeneous). Follicular dendritic cells were identified by CD23. Cyclin D1, CD3, and CD5 were negative, Ki-67 was 30%, and histiocytes were identified by CD68. With those findings, the diagnosis of primary intestinal follicular lymphoma was made. The final staging was II2 E (colon). The patient received 6 sessions of chemotherapy (R-CHOP), and at one year of follow-up, had favorable progression, with clinical, endoscopic, and imaging remission. We evaluated the duodenum and the distal ileum, and both appeared normal after one year of follow-up with chemotherapy (Figs. 2d and e).

Most patients with this type of lymphoma are adults, with an average 50 years of age. Both men and women are equally affected. They are frequently asymptomatic (43%), but some patients have abdominal pain (28%), nausea and vomiting (8%), and gastrointestinal bleeding (6%). The most frequent endoscopic presentation of follicular lymphoma is the nodular pattern.[3–5] For the Lugano classification, imaging studies, such as computed tomography (CT) or positron emission tomography (PET), are necessary.[6] The differential diagnosis includes lymphoid hyperplasia and duodenal xanthomas. The decrease in the vascular network and the presence of irregular vessels in the magnification are suggestive of lymphoma.[7] The presence of an ulcer in the terminal ileum, as well as retroperitoneal adenopathies, can be suggestive of tuberculosis in endemic areas.[8]

If the patient manifests symptoms, treatment should be started, varying in accordance with the histologic and clinical grade. If the disease is disseminated or has poor prognostic factors, systemic treatment with chemotherapy combined with rituximab is indicated.[9]

We reported the present case, given that it is a rarity, together with the fact that there is no consensus on treatment.

In conclusion, primary intestinal follicular lymphoma is a unique lymphoproliferative entity. The disease course is

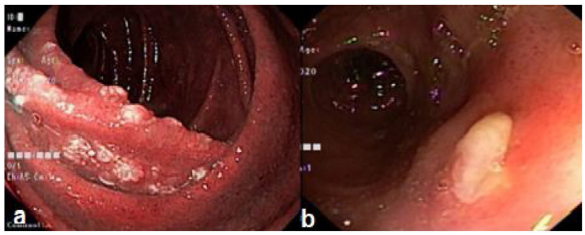


**Figure 1** (a) Duodenum with whitish nodular mucosa. (b) Ulcer in the distal ileum.

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