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Diffuse acinar cystic transformation of the pancreas-a rare case in a young male patient

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Keywords: Polycystic pancreas. Cystic neoplasm of the pancreas.

Dear Editor:
A 17-year-old male patient presented to our clinic with an ultrasound report from an outside hospital indicating pancreatic enlargement. He had suffered from intermittent abdominal pain and discomfort for more than three years but had never sought medical attention. He also had a history of diabetes mellitus (T2DM) with poor glycemic control, with a fasting glucose level as high as 336.79 mg/dL. The initial MRI scan revealed a diffusely enlarged pancreas, with multiple small cystic lesions showing slightly hyperintense T1 and hyperintense T2 signals within the pancreatic parenchyma and enhancement of the cystic walls (Fig.1A, B). After about
eight months of follow-up, a repeat MRI scan showed that some of the cysts in the pancreatic head and the uncinate process had changed in shape and size compared to the previous scan, and the main pancreatic duct was not well visualized, with local cystic duct-like structures (Fig.1C, D). The patient underwent surgery, and the intraoperative findings showed that the pancreas was enlarged but did not invade the inferior vena cava, and no enlarged lymph nodes were detected. The cut surface of the pancreatic tissue was gray-white, gray-red, honeycombed, soft, and contained clear fluid in some areas (Fig.1E). Representative photomicrographs of histologic sections were shown (Fig.1F, 100× objective, HE). Finally, the diagnosis of ACT (Acinar Cystic Transformation, ACT) was made.

Discussion:
ACT is a rare cystic lesion of the pancreas originating from cystic dilation of normal acinar structures that can occur in any part of the pancreas and can be unilocular or multilocular, the latter may involve the whole pancreas diffusely. The exact pathogenesis of ACT is still unclear, but it has been increasingly reported in young adults in recent years. The disease may present with non-specific clinical symptoms such as abdominal pain, indigestion, and abdominal mass, but most cases are discovered incidentally. ACT, as a benign cystic lesion, is characterized by typical acinar cell clusters lining the cysts, whether unilocular or multilocular, and showing prominent acinar cell differentiation, without obvious cellular atypia, mitosis, necrosis, or infiltrative growth\[1\]. CT and MRI examinations reveal a hypovascular cystic mass with thin septa, no enhancement of the capsule, and no vascular encasement or peripancreatic invasion. It needs to be distinguished from other cystic lesions (such as pseudocysts) of the pancreas and sometimes requires the exclusion of possible coexisting neoplastic lesions (such as IPMN)\[2\]. In this multilocular ACT case, the cysts' shape and size changed dynamically over time during the follow-up, and a local cystic duct-like structure was formed. It was difficult to rule out the possibility of IPMN, so surgery was performed. In addition, considering the diffuse lesions of the pancreas in this patient, the secretory function of the pancreatic islets has been impaired to a certain extent, and now more inclined to T3cDM. When
encountering pancreatic cystic lesions suspected of ACT clinically, it is recommended to follow up on the lesions, monitor the changes of the cysts and pancreatic ducts dynamically to confirm the diagnosis or rule out other lesions, and obtain a definitive diagnosis by pathology and immunohistochemistry when necessary[3].

REFERENCES

Fig. 1

A. Axial T2-weighted fat-suppressed image of the first MRI examination;

B. MRCP image of the first MRI examination;

C. Axial T1-weighted enhanced image of the first MRI examination;

D. Axial T1-weighted enhanced image of the follow-up MRI examination after eight months;
E. Gross appearance of pancreatic tissue during surgery;

F. Histopathologic image of the ACT(HE,100×).