A 60-year-old woman with a history of cerebellar hemangioblastoma surgery and recently diagnosed diabetes mellitus sought medical attention for abdominal pain and a mass at the level of the epigastrium. Abdominal ultrasound revealed multiple pancreatic cysts. A consequently performed abdominal MR scan identified a strikingly enlarged pancreatic gland extending into the pelvis and multiple heterogeneous cystic lesions in its interior distributed in the head, body, and tail (Figs. 1 and 2). Echoendoscopic-guided FNA was performed (Fig. 3) and the cytology study was consistent with serous cystadenoma (Fig. 4). With the suspicion of von Hippel-Lindau syndrome, a genetic study was ordered that was positive for the 3p25-p26 mutation. During follow-up, endocrine pancreatic insufficiency progressed requiring insulin therapy and the patient developed diarrhea secondary to exocrine pancreatic insufficiency, needing enzyme supplements. The appearance of exocrine pancreatic insufficiency in von Hippel-Lindau disease is rare. In the present case, the important pancreatic involvement and its cystic transformation led to exocrine and endocrine insufficiency.
An uncommon cause of epigastralgia

Figure 2  The same information as Figure 1.

Figure 3  Endosonographic image showing multiple cystic pancreatic lesions that guided the FNA.

Figure 4  The cytology image obtained through the FNA of one of the cystic lesions that showed glycogen-rich cuboid epithelial cells consistent with serous cystadenoma diagnosis.

Ethical responsibilities

Protection of persons and animals. The authors declare that no experiments were performed on humans or animals for this study.

Data confidentiality. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

Conflict of interest

The authors declare that there is no conflict of interest.