

Pancreatic stones, abdominal groans, and encephalopathic overtones, presented by a Type 3c Diabetes and Familial Hypocalciuric Hypercalcemia duet



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How presenting with diabetic ketoacidosis...

24-year old insulin-dependent diabetic female presented with DKA secondary to ascending cholangitis

- Acute encephalopathy
- Right upper quadrant pain
- Oliguria

Labs and imaging:

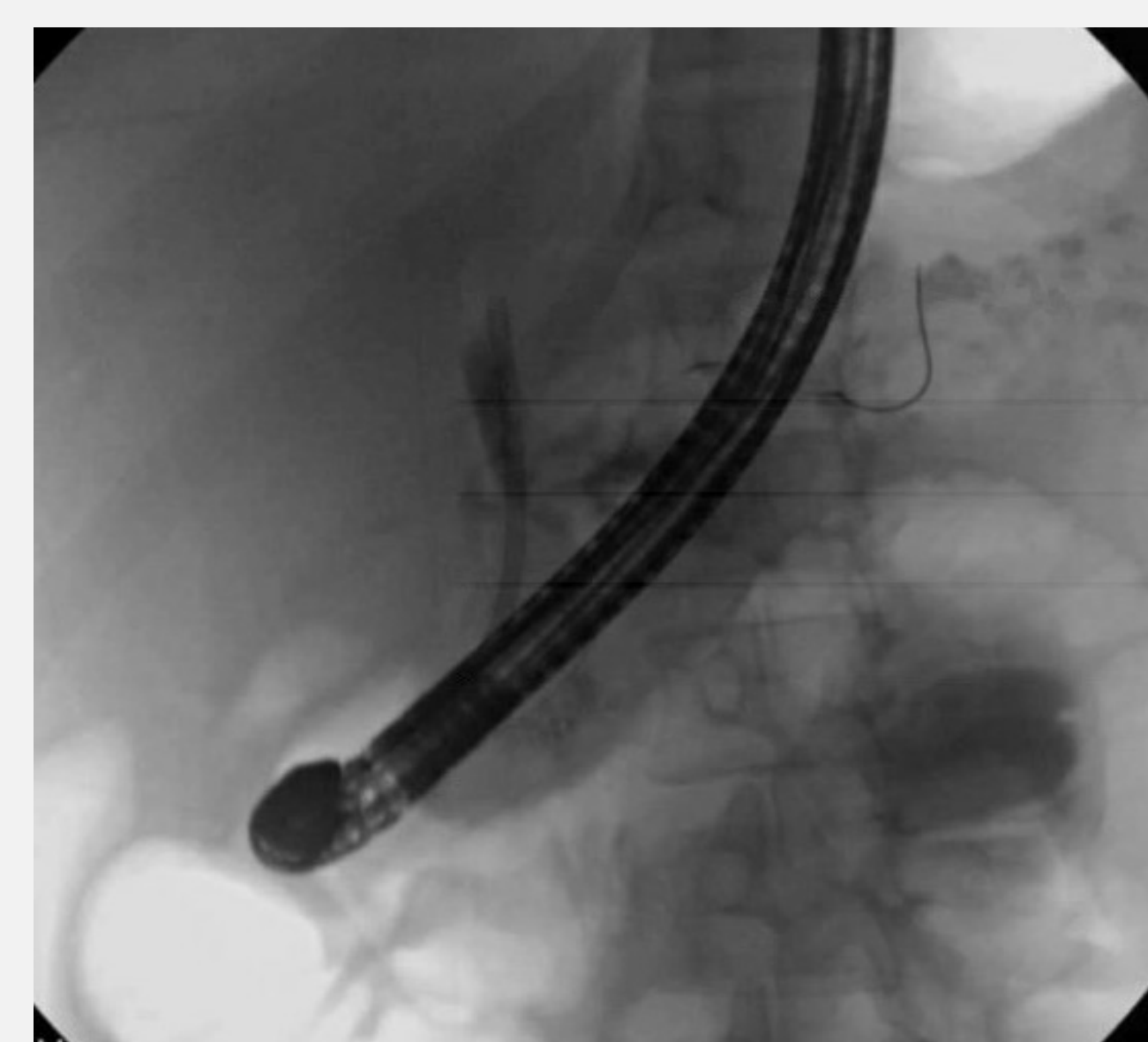
- High ionized calcium of 1.83
- Imaging showed dilated common bile duct (CBD) without choledocholithiasis and pancreatic calcifications consistent with chronic pancreatitis



Endoscopic retrograde cholangiopancreatography (ERCP) and cholangiogram revealed:

- Large stone protruding through major papilla
- Ventral pancreatic duct (PD) with sludge and multiple stones
- Dilated bile and hepatic ducts without filling defects

Sphincterotomy was performed and plastic stents were placed in ventral PD and CBD.



Evaluation of hypercalcemia revealed fractional excretion of calcium <1%, diagnostic of Familial Hypocalciuric Hypercalcemia. Family history revealed maternal family with recurrent acute pancreatitis.

Patient met criteria for type 3c diabetes with negative anti-glutamic acid antibodies.

In this case, FHH was the genetic etiology of chronic pancreatitis, complicated by pancreatogenic IDDM, evidenced by the clinical history, laboratory testing, and extensive radiographic findings.

FHH is a rare and typically benign cause of hypercalcemia, characterized by inactivating mutations of the calcium-sensing receptor (CASR). The role of CASR in pathogenesis of acute to chronic pancreatitis has been debated in different clinical reviews and should be further studied.

There should be high suspicion for complications of chronic pancreatitis when patients have clinical findings of any endocrine or exocrine insufficiency. Our case also highlights the need for clinicians to evaluate unknown causes of acute recurrent and chronic pancreatitis.

...results in a new diagnosis of Familial Hypocalciuric Hypercalcemia