A noteworthy rarity: fibrocalculous pancreatic diabetes as an uncommon aetiology of diabetes

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A 31-year-old man presented with osmotic symptoms, severe weight loss (15 kg) over 6 months, and episodic epigastric pain and steatorrhea spanning over three years. There was no history of alcohol exposure, autoimmunity, or familial diabetes. The patient exhibited stocking-type sensory loss and a BMI of 17 kg/m². Investigational findings revealed a fasting blood sugar level of 200 mg/dL, HbA1c of 11%, negative urine ketones, and a low plasma insulin level (< 5 micU/mL). Corrected calcium and plasma triglycerides were within normal limits.

Abdominal X-ray (Figure) suggested fibrocalculous pancreatic diabetes (FCPD). FCPD is characterised by a classic triad of diabetes, exacerbations of abdominal pain, and steatorrhea. It predominantly affects individuals of South Indian or African ancestry and may result from malnutrition, deficiencies in vitamins A and C, toxic cyanide exposure from

chronic Cassava consumption, and genetic predisposition.(1) Notably, FCPD is unusual in its lack of association with ketosis and long-term macrovascular complications. However, patients face an increased risk of pancreatic malignancy and severe malnutrition, impacting prognosis.(1)

The patient was initiated on high doses of premixed insulin and is under the joint care of endocrine and gastroenterology teams for ongoing management.

Consent for publication was obtained from the patient.

References

1. Kumaran S, Unnikrishnan AG. Fibrocalculous pancreatic diabetes. J Diabetes Complications. 2021 Jan;35(1):107627. doi: 10.1016/j.jdiacomp.2020.107627.

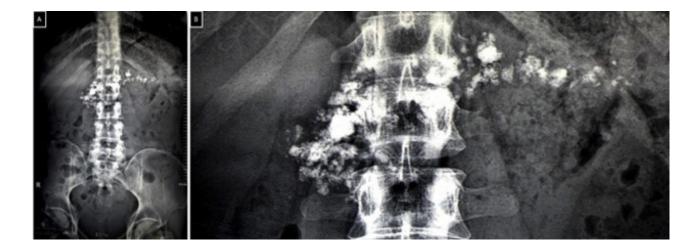


Figure 1 - Panel A and B: Anteroposterior abdominal plain roentgenogram, and magnification of area of interest, depict the "bag of stones" appearance, with multiple large and small, discrete, calcium-containing pancreatic ductal and parenchymal calculi, overlapping L1 and L2 vertebrae.