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Case Report

Internal Medicine Section

Complete Agenesis of the Dorsal Pancreas Presenting as Uncontrolled Hyperglycaemia: A Case Report

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ABSTRACT

Agenesis of the Dorsal Pancreas (ADP) is a rare anatomical manifestation of the pancreas, characterised by the total or partial absence of the dorsal pancreatic regions, body, and tail. The anatomy of the pancreas missing the pancreatic tail is considered partial agenesis, while the absence of the body and tail, with the presence of only the pancreatic head, is considered to be complete dorsal agenesis of the pancreas. Here the authors present the case of 20 years old male patient with uncontrolled blood sugars and recent onset of blurred vision. Diagnostic imaging, including Ultrasonography (USG) and Computed Tomography (CT), revealed the pancreatic head, but the body and tail were not seen. Using CT, the patient's condition was identified as complete ADP. Treatment involved initiating basal bolus insulin therapy, leading to glycaemic control. The patient also underwent cataract surgery for diabetic-related complications. This case underscores the importance of considering congenital pancreatic anomalies in young adults and highlights the utility of advanced imaging techniques in confirming the diagnosis of agenesis of the dorsal pancreas. Early recognition and intervention are crucial for optimising patient outcomes. Further research is needed to enhance understanding of ADP pathophysiology and develop effective management strategies.

Keywords: Absence of pancreatic regions, Type 3c diabetes, Uncontrolled blood sugars

CASE REPORT

A 20-year-old male patient, lean, non-addict, was referred to the diabetes outpatient department for glycemic control and fitness for cataract surgery from the Department of Ophthalmology due to blurring of vision in both eyes, with the left eye being more impacted than the right eye.

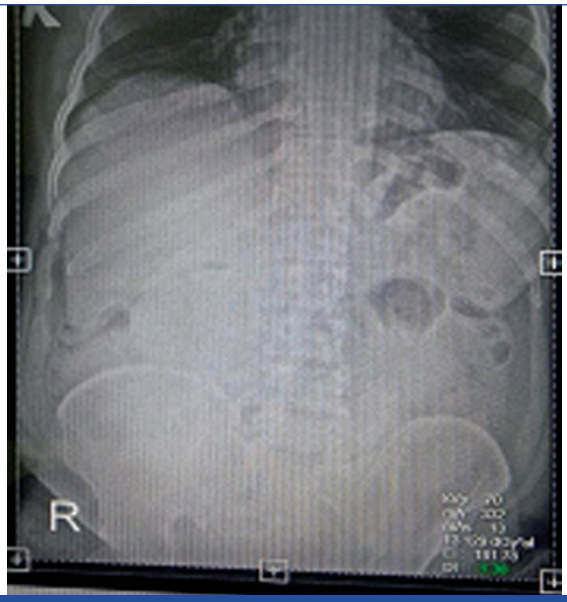
His blood panels showed a Fasting Blood Sugar (FBS) of 333 mg/dL, Post-Lunch Blood Sugar (PLBS) of 533 mg/dL, and glycated hemoglobin (HbA1c) of 13.9%. The patient was a known case of diabetes for two years. There were no complaints of abdominal pain, nausea, vomiting, or altered bowel complaints. In the past, the patient denied being admitted to the hospital for the management of diabetes. There was no family history of diabetes. At the diagnosis of diabetes mellitus in September 2021, the patient was started on a combination of sulfonylurea and metformin 1/500 mg once a day by his family physician, which was subsequently up-titrated to twice a day and three times a day because of uncontrolled blood sugars along with the addition of a dipeptidyl peptidase-4 inhibitor 50 mg twice daily and pioglitazone 15 mg once a day.

On examination, the patient had a Body Mass Index (BMI) of 15.6 kg/m^2. The patient's blood pressure was 110/70 mmHg in the right arm supine position, pulse rate was 84 beats per minute (bpm) regular, respiratory rate was 16 per minute, saturation level was 99% on room air, and he was afebrile. There was no goiter or bony abnormalities seen. All his peripheral pulses were well felt. The systemic examination was essentially normal. The dilated fundus examination did not reveal any diabetic retinopathy or diabetic macular oedema. The biochemical investigations of the patient have been summarised in [Table/Fig-1]. The X-ray chest was normal, and X-ray abdomen standing did not show any pancreatic calcifications [Table/Fig-2]. The patient had a normal 2-D echocardiography. The axial view of the ultrasonography showed spleno-portal confluence with absence pancreatic tissue. A Contrast-enhanced Computed Tomography (CECT) of the abdomen showed non-visualisation of the neck, body, and tail of the pancreas with a normal head and uncinate process [Table/Fig-3], suggestive of complete dorsal

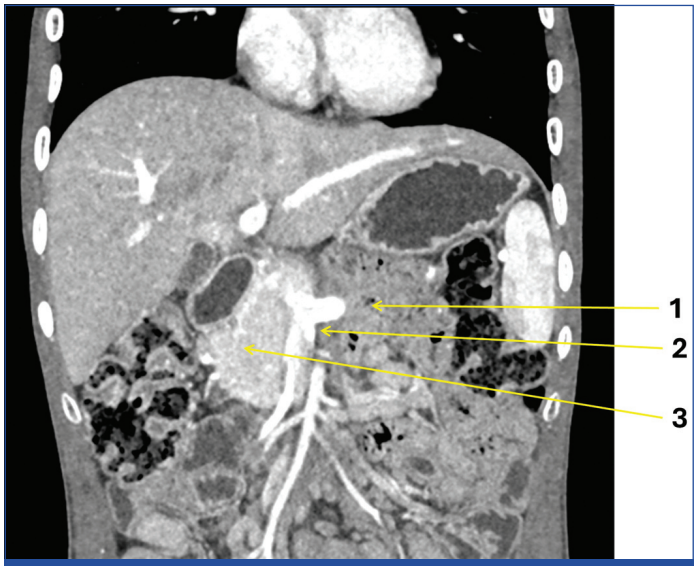
Test	Observed value	Normal range
Haemoglobin	14.5	12.5-18 g/dL
WBC	5440	4000-11000/mm^3
Platelets	172000	150000-450000
FBS	333	70-99 mg/dL
PPBS	533	70-140 mg/dL
HbA1c	13.9	<5.5%
Serum Na	136	135-155 mEq/L
Serum K	4.2	3.5-5.5 mEq/L
Serum Cl	105	96-106 mEq/L
S. Creatinine	0.7	0.4-1.2 mg/dL
SGOT	54	5-40
SGPT	66	5-40
Bilirubin total	1.0	0-1.3 mg/dL
Bilirubin direct	0.2	0-0.3 mg/dL
Alkaline phosphatase	125	25-190
S. Proteins	6.5	6-8.3 g/dL
Albumin	3.9	3.2-5.2 g/dL
Globulin	2.60	1.5-3.8 g/dL
S. Amylase	85	0-90 U/L
Calcium	9.6	9.0-11.0 mg/dL
Vitamin D3 level	27	20-40 ng/mL
TSH	2.97	0.4-5.3 µlU/mL
T. Cholesterol	133	<200 mg/dL
High density lipoprotein	40	>40 mg/dL
Low density lipoprotein	83	<130 mg/dL
Triglyceride	118	<150 mg/dL
C-Peptide	0.6	0.78-5.19 ng/mL
S. Ketone	Absent	Negative <0.6 mmol/L
Urine ketone	Negative	Negative
GAD-65 antibody	0.64	Negative <10 IUmL

ICA	Negative	Negative
IA-2	4.30	Negative <28 U/mL
ZnT8	3.63	Non-reactive <10 AU/mL
Insulin antibody	4.10	Negative <10U/mL

[Table/Fig-1]: Biochemical investigations of the patient.
WBC: White blood cell count; FBS: Fasting blood sugar; PPBS: Post prandial blood sugar; HbA1c: Glycated haemoglobin; AST: Aspartate aminotransferase; ALT: Alanine aminotransferase; TSH: Thyroid stimulating hormone; GAD: Glutamic acid decarboxylase; ICA: Islet cell antibody; ZnT8: Zinc transporter 8 antibody; IA-2: Islet antigen 2 antibody



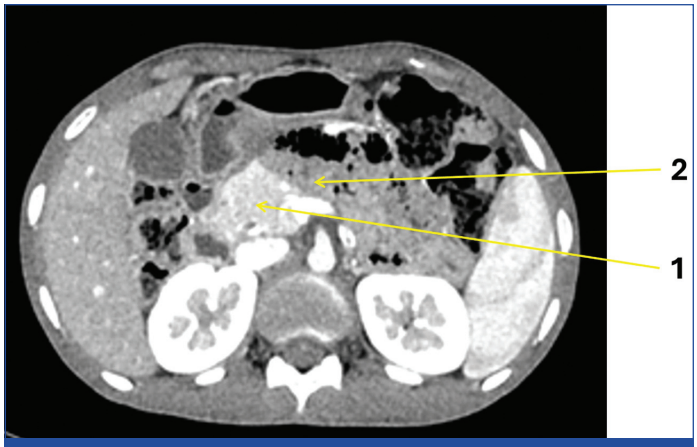
[Table/Fig-2]: X-ray abdomen.



[Table/Fig-3]: Contrast Enhanced Computed Tomography(CECT) of the abdomen: non-visualisation of body and tail of pancreas.
1- Absent body and tail of the pancreas; 2- Confluence of splenic vein and superior mesenteric artery; 3- Head and uncinate process of pancreas

agenesis of the pancreas. Small bowel loops inhabited the area of the body and tail [Table/Fig-4].

The patient was referred to the gastroenterology department and was advised to do a Fecal Elastase 1 test. However, the patient was not willing to have any further radiological or blood investigations due to financial constraints. The patient was started on a basal-bolus regime of insulin comprising three doses of short-acting insulin prior to each meal and long-acting insulin before bedtime. His blood glucose values were brought under control over a period of three months with an A1c of 7.9%. He underwent left eye cataract surgery and subsequently the right eye as well. A Mixed-Meal Tolerance Test (MMTT) was conducted after blood sugar control, and the C-peptide value was 0.6 ng/mL. MMTT is the most sensitive measure of endogenous insulin secretion. The



[Table/Fig-4]: CECT of the abdomen showing small bowel loops occupying the region of the pancreatic body and tail.
1- Head and uncinate process of pancreas; 2- Absent body and tail of the pancreas

MMTT value of 0.6 ng/mL showed inadequate beta-cell reserve. As the patient was moving away to his hometown, he was referred to another healthcare practitioner in his hometown. No follow-up has been conducted since the referral.

DISCUSSION

The presented case highlights several key clinical and diagnostic challenges associated with ADP, a rare congenital disorder. This condition is characterised by the absence or underdevelopment of the dorsal pancreatic bud during embryogenesis, leading to pancreatic insufficiency and potential complications such as diabetes mellitus [1]. The patient's age of 20 years at the time of presentation is notably younger compared to the majority of reported cases of ADP, which typically occurs in older individuals [2]. While previous studies have predominantly documented cases in adults over 30 years of age [1,2], the case underscores the importance of considering congenital pancreatic anomalies even in young adults. The development of cataracts in the patient, a complication typically associated with poorly controlled diabetes mellitus, adds a unique dimension to this case.

While the exact mechanism underlying cataract formation in individuals with pancreatic anomalies remains unclear, it is plausible that metabolic disturbances resulting from diabetes mellitus, compounded by pancreatic insufficiency due to ADP, contributed to the development of cataract. The diagnostic evaluation revealed several key findings consistent with complete ADP. Ultrasonography demonstrated a spleno-portal confluence with absent pancreatic tissue, confirming the absence of the dorsal pancreas. Additionally, radiological imaging, including X-ray abdomen, did not show any pancreatic calcifications, further supporting the diagnosis of ADP. These findings align closely with previous reports documenting similar radiological features in patients with this rare congenital anomaly [3,4].

In the current case study, the patient presented with uncontrolled blood sugars, prompting diagnostic imaging to assess the pancreatic anatomy. While USG is commonly utilised as the primary imaging modality, it may not always provide sufficient detail for diagnosing ADP. Three-dimensional reconstruction CT is considered a more effective tool for accurately diagnosing ADP due to its superior visualisation capabilities [5]. Although stomach or intestinal filling in the distal pancreatic bed is suggestive of dorsal pancreas absence, confirmation typically requires imaging modalities such as Magnetic Resonance Cholangiopancreatography (MRCP) or Endoscopic Retrograde Cholangiopancreatography (ERCP). MRCP offers a non-invasive approach to confirming ADP, while ERCP is considered the gold standard for comprehensive assessment of the pancreatic and biliary tree [6]. CT imaging revealed small bowel loops occupying the region of the pancreatic body, providing definitive evidence for

dorsal agenesis of the pancreas. This diagnostic finding enabled the confirmation of ADP, contributing to the patient's clinical management and treatment plan for uncontrolled blood sugars. The treatment approach adopted in this case, involving the initiation of a basal-bolus regime of insulin therapy, aligns with previous studies addressing the management of complete ADP [7,8]. Despite the rarity of this condition, limited data suggest that insulin therapy is the cornerstone of management for patients with pancreatic endocrine insufficiency resulting from ADP [8]. Early diagnosis of complete ADP is paramount for initiating appropriate management strategies and mitigating associated complications. Given the potential implications for pancreatic function and systemic health, timely recognition and intervention are crucial for optimising patient outcomes.

CONCLUSION(S)

The report presents a unique case of a young adult with hyperglycaemia non-responsive to oral anti-glycaemic agents, with cataracts in both eyes and no family history or genetic associations. A rationalised approach that heightened the suspicion of a pancreatic anomaly supported by the advent of imaging modalities paved the way for the confirmation of complete dorsal agenesis in the dorsal tail and body regions of the pancreas. The strategic findings and timely insulin treatment facilitated the achievement of glycemic control.

Authors' contribution: All authors made individual contributions to the authorship. AK was involved in the diagnosis and management of the manuscript of the subject. KS was responsible for drafting the manuscript, scientific contribution and manuscript submission. VJ, RR, DM were involved in diagnosis. All authors reviewed and approved the final draft.

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