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CLINICAL REASONING: CASE REPORT

Dorsal pancreatic agenesis revealed by acute biliary pancreatitis in a postpartum woman with gestational diabetes: a case report

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Summary

Dorsal pancreatic agenesis is an exceptionally rare congenital anomaly involving the partial or complete absence of the pancreatic body and tail. Metabolic dysfunction, including impaired glucose tolerance and insulin-dependent diabetes, is frequently reported; however, the relationship between dorsal pancreatic agenesis and gestational diabetes mellitus has been reported only sporadically.

The objective of this report is to present a rare case of dorsal pancreatic agenesis revealed by acute biliary pancreatitis and recurrent gestational diabetes mellitus and to review the relevant literature concerning its metabolic and clinical implications.

We report the case of a postpartum woman in her early 30s who presented with acute biliary pancreatitis. Imaging revealed complete agenesis of the pancreatic body and tail, with preservation of the pancreatic head derived from the ventral bud. She had a history of recurrent gestational diabetes, raising the possibility that pregnancy-associated insulin resistance had unmasked a compensated beta-cell deficit due to dorsal pancreatic agenesis. The patient underwent an uncomplicated laparoscopic cholecystectomy with symptom resolution.

This case highlights three clinically significant observations: (1) recurrent gestational diabetes mellitus may represent an early clinical clue to reduced pancreatic endocrine reserve in dorsal pancreatic agenesis; (2) acute biliary pancreatitis can incidentally reveal congenital pancreatic anomalies; and (3) structural pancreatic abnormalities should be considered in patients with atypical or recurrent gestational diabetes mellitus. The association between dorsal pancreatic agenesis and gestational diabetes has been reported, albeit rarely, but the interaction between recurrent gestational diabetes mellitus, obesity, and reduced pancreatic endocrine reserve remains insufficiently characterised.

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Introduction

Dorsal pancreatic agenesis is a rare congenital malformation resulting from the absence of the dorsal pancreatic bud derivatives, namely the body, tail, and superior head. Fewer than 120 anatomically confirmed cases have been reported, though incidental discovery has increased with modern cross-sectional imaging [1–6]. Dorsal pancreatic agenesis is typically identified during the evaluation of abdominal pain, pancreatitis, or biliary disease, yet a growing number of cases are detected incidentally [1,3–6]. Endoscopic ultrasound also plays an important role in confirming the absence of dorsal ductal structures when computed tomography (CT) or magnetic resonance imaging (MRI) findings are inconclusive [14].

Dorsal pancreatic agenesis exhibits wide clinical variability. Approximately half of affected individuals develop abnormalities of glucose metabolism, including impaired fasting glucose, insulin-dependent diabetes, and highly labile glycaemia, consistent with loss of islet-rich dorsal parenchyma [2, 4, 6–9]. Severe endocrine presentations such as diabetic ketoacidosis have also been documented [9, 10]. Conversely, some patients remain asymptomatic until a physiological stressor reveals a previously compensated beta-cell deficit.

Pancreatitis is another recognised manifestation; it is typically idiopathic or recurrent and is sometimes associated with ductal anomalies or congenital vascular abnormalities [1, 3, 6, 12]. Dorsal pancreatic agenesis may coexist with congenital disorders such as polysplenia, heterotaxy, or a preduodenal portal vein [13].

Neoplastic complications, including pancreatic ductal adenocarcinoma, neuroendocrine tumours, and chronic calcific pancreatitis, have also been reported in association with dorsal pancreatic agenesis [11, 12].

Despite these findings, the relationship between dorsal pancreatic agenesis and gestational diabetes mellitus has been only sporadically reported [18].

In this report, we describe a rare case of complete dorsal pancreatic agenesis identified during evaluation for acute biliary pancreatitis in a woman with recurrent gestational diabetes and complement this report with a targeted review of the literature to contextualise its clinical and metabolic significance.

Case presentation

Chief complaint

A woman in her early thirties, six months postpartum and breastfeeding, presented with a 48-hour history of persistent epigastric pain radiating to the back, accompanied by vomiting. She denied fever, alcohol consumption, and medication changes.

Past medical history

The patient's medical history included the following:

- Gravida 2, para 2 (G2P2); six months postpartum and breastfeeding.
- Gestational diabetes mellitus, diagnosed during her most recent pregnancy in 2025 (according to the standard International Association of Diabetes and Pregnancy Study Groups criteria adopted by the World Health Organization), managed by the endocrinology and obstetrics teams.
- Possible unrecognised gestational diabetes mellitus in 2015 (no formal oral glucose tolerance test performed at that time, but retrospective history was suggestive of hyperglycaemia during pregnancy).
- Class I obesity (postpartum body mass index (BMI) 31.3 kg/m^2; pre-pregnancy BMI not formally recorded).
- Mild intermittent asthma.
- No known drug allergies.
- No prior pancreatic interventions.
- No known family history of diabetes mellitus or pancreatic disease.
- Occupational history as a homemaker.
- Self-reported Balkan ethnic background.

Clinical findings

Physical examination revealed epigastric and right upper quadrant tenderness without guarding or rigidity. She was afebrile.

Timeline

- Day 0: Onset of epigastric pain
- Day 2: Emergency department presentation
- Day 3: CT scan performed → diagnosis
- Day 3-5: Supportive treatment with intravenous fluids and analgesia
- Day 6: Laparoscopic cholecystectomy after the normalisation of liver biochemistry
- Day 8: Discharged asymptomatic after an uneventful postoperative course
- Day 35: Follow-up visit; clinically stable

Diagnostic assessment

Laboratory investigations

Lipase was markedly elevated at 2678 IU/l [13–60 IU/l]. Liver enzyme levels were significantly abnormal: alanine aminotransferase (ALT) 558 IU/l [7–35 IU/l]; aspartate aminotransferase (AST) 213 IU/l [10–35 IU/l]; gamma-glutamyl transferase (GGT) 484 IU/l [7–40 IU/l]. Total bilirubin was 24 µmol/l [5–21 µmol/l]. C-reactive protein (CRP) was 15 mg/l [0–5 mg/l]. Renal function and complete blood count were normal. Fasting glucose was 3.7 mmol/l [3.7–5.6 mmol/l]. Lipase and liver enzymes normalised rapidly within 48 hours of admission.

Faecal elastase was not formally measured; however, stools were clinically normal with no steatorrhea and preserved nutritional status. This is consistent with previous reports indicating that exocrine insufficiency is less common than endocrine dysfunction in dorsal pancreatic agenesis [4, 6].

Imaging

Contrast-enhanced CT of the abdomen (figures 1 and 2), performed approximately 72 hours after the onset of symptoms, revealed the following:

- Acute pancreatitis confined to the pancreatic head (classified as Balthazar grade C, indicating pancreatic enlargement with mild peripancreatic inflammation, and with a modified CT Severity Index [mCTSI] score of 2, consistent with mild pancreatitis without necrosis).
- Complete agenesis of the pancreatic body and tail, consistent with congenital dorsal pancreatic agenesis.
- Peripancreatic fat stranding near the perirenal fascia.
- Mild inflammatory reaction around the gallbladder, duodenum, and right ureter.
- Hyperdense material within the gallbladder compatible with microlithiasis.
- No biliary duct dilatation, venous thrombosis, or pseudoaneurysm.
- Small hepatic cysts (incidental).

Figure 1: Contrast-enhanced computed tomography (CT) imaging of the abdomen in dorsal pancreatic agenesis. Axial views, arterial phase (cranial to caudal; A1-A4). Panel A1 (uppermost slice) demonstrates the preserved pancreatic head and uncinate process (white arrow; residual ventral-bud-derived parenchyma outlined in orange), which appear morphologically normal without radiological signs of compensatory hypertrophy. Panels A2 and A3 (mid-level slices) show a complete absence of pancreatic parenchyma (hashed red line) anterior to the splenic vein (SV) and the superior mesenteric artery (SMA, red circle), in the expected anatomical location of the dorsal pancreatic body and tail. Panel A4 (most caudal slice) confirms the absence of pancreatic tissue extending toward the splenic hilum, with no ectopic, migratory, or accessory pancreatic tissue identified.

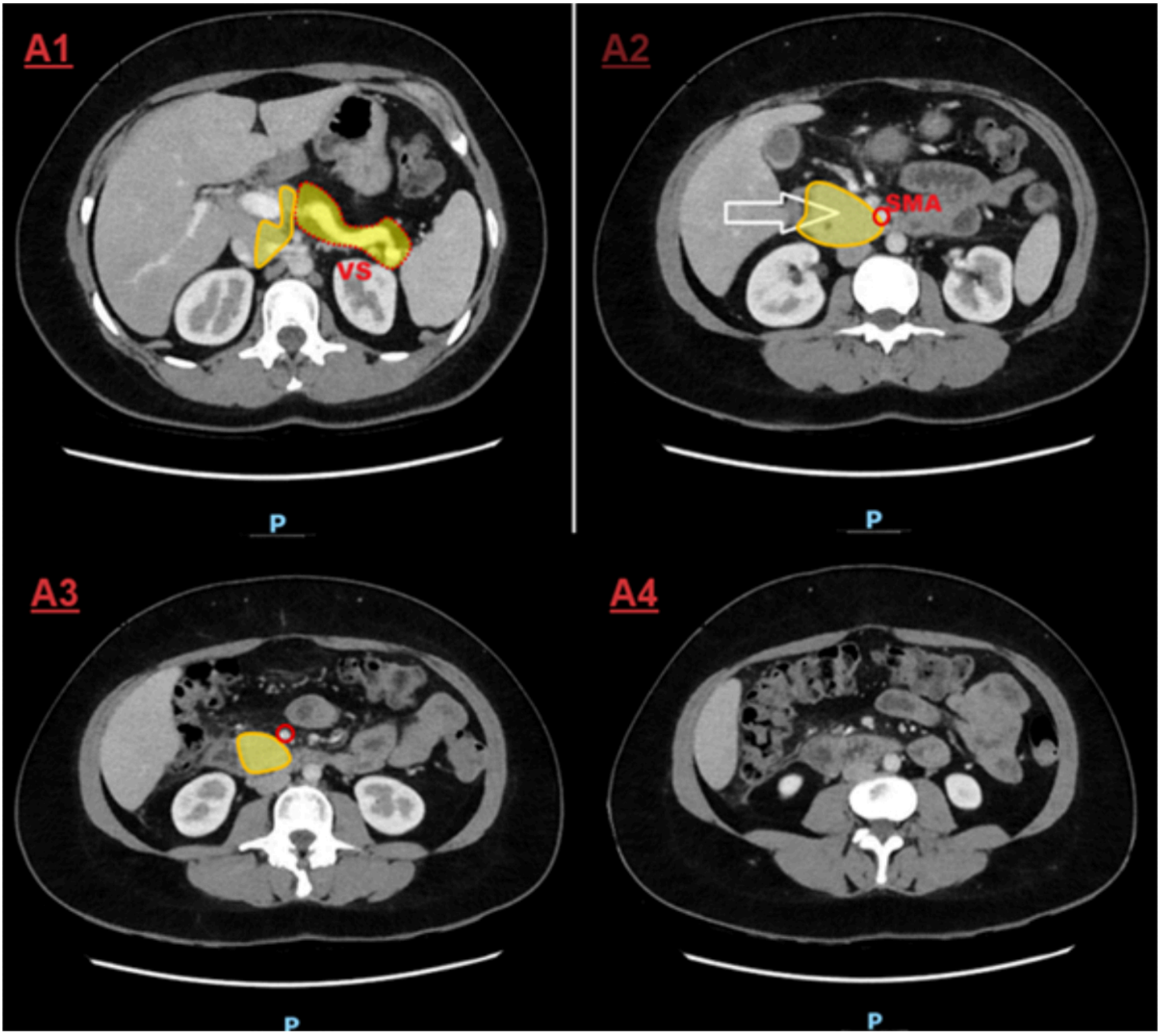
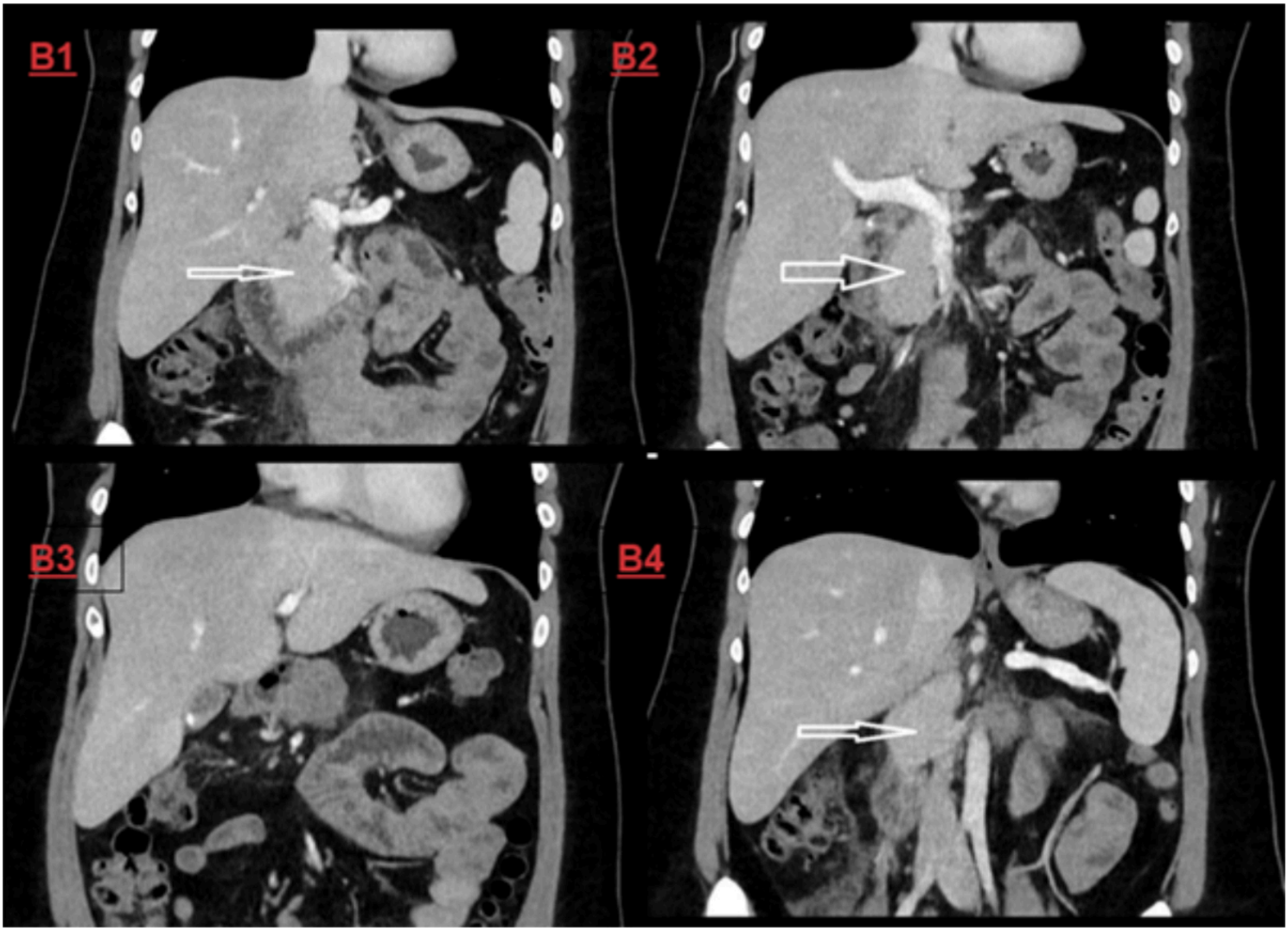


Figure 2: Contrast-enhanced computed tomography (CT) imaging of the abdomen in dorsal pancreatic agenesis. Coronal views, arterial phase (B1-B4). White arrowheads/arrows indicate the pancreatic head (ventral bud derivative), which represents the only residual pancreatic parenchyma. No body or tail structures are visualised; the splenic hilum abuts the stomach without intervening pancreatic tissue. No associated congenital vascular anomalies (e.g. preduodenal portal vein, splenic vein variants) or polysplenia/heterotaxy features are identified, consistent with isolated dorsal pancreatic agenesis.



Diagnostic challenges

A potential diagnostic challenge is distinguishing true dorsal agenesis from pancreatic atrophy or fatty replacement; however, the absence of dorsal parenchyma, along with preserved anatomy and the lack of prior pancreatic disease, supported a congenital origin in this case.

Differential diagnosis

Differential diagnoses included pancreatic lipomatosis, chronic pancreatitis-related atrophy, and pancreatic divisum; these were considered unlikely due to the complete absence of dorsal structures and the absence of prior pancreatic pathology.

Final diagnosis

Complete dorsal pancreatic agenesis with acute biliary pancreatitis.

Prognosis

The long-term prognosis in dorsal pancreatic agenesis is primarily determined by endocrine function. Given the reduced beta-cell mass, this patient may have an increased risk of progression to type 2 diabetes mellitus compared with that of the general population of women with gestational diabetes mellitus. Therefore, long-term metabolic follow-up and counselling regarding future pregnancies are warranted.

Management

The patient received intravenous fluids and analgesia. Due to suspected transient choledocholithiasis and gallbladder microlithiasis, she underwent laparoscopic cholecystectomy after the normalisation of her hepatic tests. Postoperatively, her pain resolved, and her liver enzymes normalised. She was discharged with outpatient follow-up.

Follow-up and outcomes

At follow-up, the patient remained asymptomatic, with normal liver function tests, no evidence of recurrent pancreatitis, and no clinical or biochemical evidence of hyperglycaemia. Long-term glycaemic monitoring was recommended given the reduced pancreatic endocrine reserve.

No adverse or unanticipated events were observed during hospitalisation or at follow-up.

Discussion

In this patient, complete agenesis of the pancreatic body and tail was identified during the evaluation of acute biliary pancreatitis. Although pancreatitis is a recognised manifestation of dorsal pancreatic agenesis, most published cases describe idiopathic or recurrent forms without a clear precipitant [1, 3, 6, 12]. In contrast, the present case was associated with gallstone disease. The reduced ductal and parenchymal reserve inherent to dorsal pancreatic agenesis may predispose patients to more severe inflammation in response to even brief episodes of transient choledocholithiasis.

Understanding the metabolic phenotype of this case requires revisiting normal pancreatic embryology. The dorsal bud forms the superior pancreatic head, body, and tail and contains the majority of the islet cell population [15]. Therefore, the complete absence of the dorsal pancreas represents a substantial lifelong reduction in beta-cell mass. Many patients with dorsal pancreatic agenesis exhibit impaired glucose tolerance or overt diabetes [2, 4, 6–9], and severe presentations, including ketoacidosis, have been described, though these are uncommon [9, 10].

Pregnancy represents a profound insulin-resistant state driven by placental hormones. In this patient, recurrent gestational diabetes mellitus likely reflects, at least in part, the pregnancy-induced unmasking of a previously compensated beta-cell deficiency. Although a direct causal relationship cannot be established from a single observation, this case raises the possibility that recurrent gestational diabetes mellitus could serve as an early clinical manifestation of reduced endocrine reserve in individuals with dorsal pancreatic agenesis. Such an association has been described only rarely in the literature, including in a previous report of gestational diabetes in a woman with dorsal pancreatic agenesis [18]. The observed metabolic phenotype likely reflects an interaction between reduced beta-cell reserve due to dorsal pancreatic agenesis and coexisting insulin resistance related to obesity, rather than a direct causal relationship.

Although this case represents isolated dorsal pancreatic agenesis, congenital anomalies such as polysplenia, heterotaxy, and a preduodenal portal vein have been described [13]. Several rare associations with genetic and syndromic conditions, including mutations in PTF1A and GATA6 [16, 17], have been reported. Genetic testing was not pursued in this case, as the patient had no phenotypic features suggestive of a syndromic disorder.

Dorsal pancreatic agenesis has been associated with pancreatic ductal adenocarcinoma, neuroendocrine tumours, and chronic calcific pancreatitis [11, 12]. Although rare, these associations highlight the need for increased clinical awareness when evaluating patients with dorsal pancreatic agenesis who present with new symptoms.

Compensatory hypertrophy of the residual ventral pancreas has been hypothesised as a potential adaptive mechanism in dorsal pancreatic agenesis [4,11]. In the present case, CT imaging showed no radiological evidence of enlargement of the pancreatic head, which appeared morphologically normal. Whether the absence of compensatory hypertrophy contributes to the endocrine

insufficiency observed in some patients with dorsal pancreatic agenesis, or whether such hypertrophy occurs subclinically and is insufficient to fully compensate for the loss of dorsal islet mass, remains an open question that warrants investigation in larger series.

The clinical relevance of this case lies not in establishing a novel association but in highlighting how physiological stressors such as pregnancy may reveal subclinical endocrine insufficiency in congenital pancreatic anomalies.

This report has several strengths, including detailed imaging confirming complete dorsal pancreatic agenesis and a well-documented clinical course linking the diagnosis to a common clinical presentation. However, several limitations must be acknowledged. As a single case report, causality between dorsal pancreatic agenesis and recurrent gestational diabetes mellitus cannot be established, and the findings may not be generalisable. In addition, long-term metabolic outcomes remain unknown. Importantly, coexisting obesity represents an independent confounder of gestational diabetes mellitus, and the absence of pre-pregnancy metabolic data (including baseline BMI and glucose tolerance) limits the interpretation of the metabolic phenotype attributed to dorsal pancreatic agenesis. In addition, detailed information regarding the management of gestational diabetes during the 2025 pregnancy, including the potential use of insulin therapy, was not fully available, which limits further characterisation of the metabolic phenotype. No genetic analysis was performed, precluding the assessment of monogenic causes of pancreatic agenesis.

Clinical and educational implications

This case highlights several practical lessons. First, dorsal pancreatic agenesis should be considered when the pancreatic body and tail are absent on imaging, and it must be distinguished from more common mimics such as atrophy or lipomatosis. Second, pregnancy may act as a physiological stress test of pancreatic endocrine reserve. In this setting, recurrent or atypical gestational diabetes may prompt consideration of an underlying structural pancreatic abnormality. Third, pancreatitis in dorsal pancreatic agenesis is not necessarily idiopathic and may be triggered by common conditions such as biliary disease. Finally, evaluation should address both endocrine and exocrine pancreatic function, and affected patients may benefit from long-term metabolic follow-up.

Conclusion

Although a causal link cannot be established from a single case, this report illustrates that recurrent gestational diabetes might signal an underlying reduction in pancreatic endocrine reserve due to dorsal pancreatic agenesis. Acute biliary pancreatitis may bring this otherwise silent anomaly to light. Given the potential metabolic consequences, awareness of dorsal pancreatic agenesis is particularly relevant to the care of women of reproductive age.

Patient perspective

The patient expressed reassurance after understanding the benign nature of the anatomical finding but reported concern regarding the risk of future diabetes. Follow-up was arranged.

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