Case Report

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A Case Report of Dorsal Pancreatic Agenesis and Uterus Didelphys in a Young Woman: A Seldom-Reported Association

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Running Title Dorsal Pancreatic Agenesis and Uterus Didelphys

ABSTRACT

Agenesis of the dorsal pancreas is a rare congenital anomaly that often remains asymptomatic and is frequently discovered incidentally during imaging for other conditions. Its association with Müllerian anomalies, such as uterus didelphys, suggests that both conditions may stem from similar embryological disruptions during early fetal development. This report aims to raise awareness of Agenesis of the Dorsal Pancreas (ADP) and its potential associations with Müllerian anomalies. By presenting a detailed case study, we seek to address the diagnostic challenges faced by healthcare professionals and the implications for patient management, emphasizing the importance of early detection and appropriate intervention.

We present a case involving a 24-year-old Iranian female who presented with a 10-day history of abdominal pain and vomiting. Initial ultrasound imaging revealed moderate hydroureteronephrosis in the right kidney, prompting further investigation. A contrast-enhanced CT scan was performed, which not only confirmed ureteropelvic junction obstruction but also unexpectedly revealed dorsal pancreatic agenesis. In addition to these findings, the patient was diagnosed with uterus didelphys, highlighting a significant developmental anomaly.

This case underscores the necessity for increased awareness of ADP and its associations with Müllerian anomalies among healthcare providers. Early and accurate diagnosis through advanced imaging modalities significantly improves patient outcomes and facilitates better management strategies for associated complications.

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Introduction

Agenesis of the dorsal pancreas (ADP) is a seldom-seen congenital abnormality where the body and tail of the pancreas are absent. While ADP may be incidentally detected when examining conditions like diabetes mellitus, pancreatitis, and pancreatic insufficiency, it often remains asymptomatic. The current literature reports fewer than 120 documented cases of ADP, emphasizing the extraordinary infrequency of this anatomical anomaly [1].

Müllerian anomalies encompass a diverse set of developmental abnormalities found in up to 5% of the general population. These anomalies are more prevalent in individuals with infertility (8.0%) and those with a history of pregnancy loss (13.3%), with the highest occurrence seen in individuals with a history of both conditions (24.5%) [2].

In our case, we present a 24-year-old female who sought medical attention for abdominal pain and vomiting. During the diagnostic imaging process, it was incidentally revealed that the patient had Agenesis of the Dorsal Pancreas (ADP).

Case Presentation

A 24-year-old Iranian female visited a urologist, reporting a 10-day onset of abdominal pain and non-bloody, non-bilious vomiting. She had been asymptomatic and without any medical issues from birth until the onset of symptoms, showing normal growth and development during this period. The urologist requested an abdominopelvic ultrasound for her. The ultrasound revealed moderate to severe hydroureteronephrosis in the right pelvicalyceal system. Regarding the pancreas, it was mentioned that it was not assessable due to gas shadow, and the pelvic space could not be evaluated due to the bladder being empty. Subsequently, a CECT scan was requested for her. The CT scan showed severe pelvicalyceal dilation in the right kidney associated with cortical parenchymal thinning and a normal ureter, which is in favor of UPJO (Figure 1). Additionally, evidence of dorsal pancreatic agenesis was seen (Figure 2). In the pelvic cavity, two uterus bodies and two cervices were noted, which is suggestive of uterus didelphys (Figures 3 and 4).

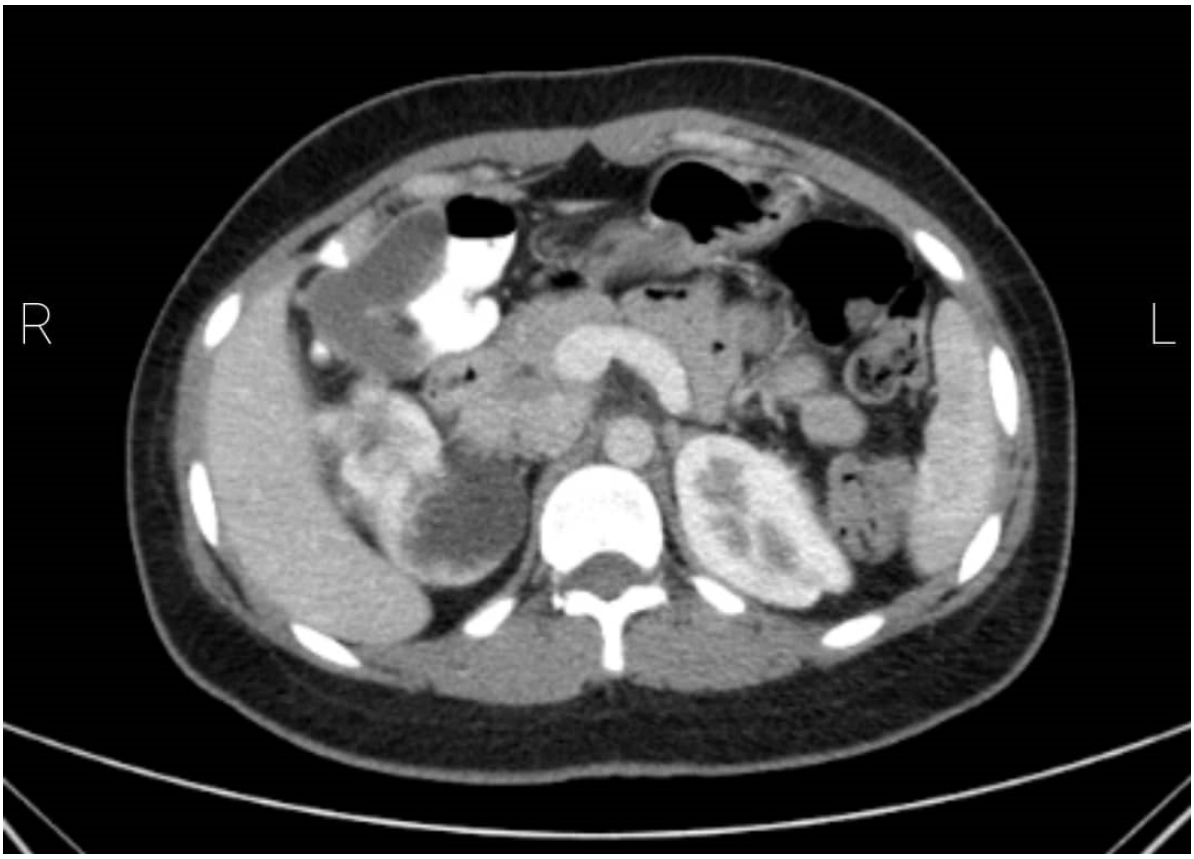

Fig. 1. Axial abdominopelvic CECT scan reveals severe pelvicalyceal dilation in right kidney associated with cortical parenchymal thinning and normal ureter which is in favor of UPJO. Also, evidence of dorsal pancreatic agenesis without vascular or GI loop malrotation is seen

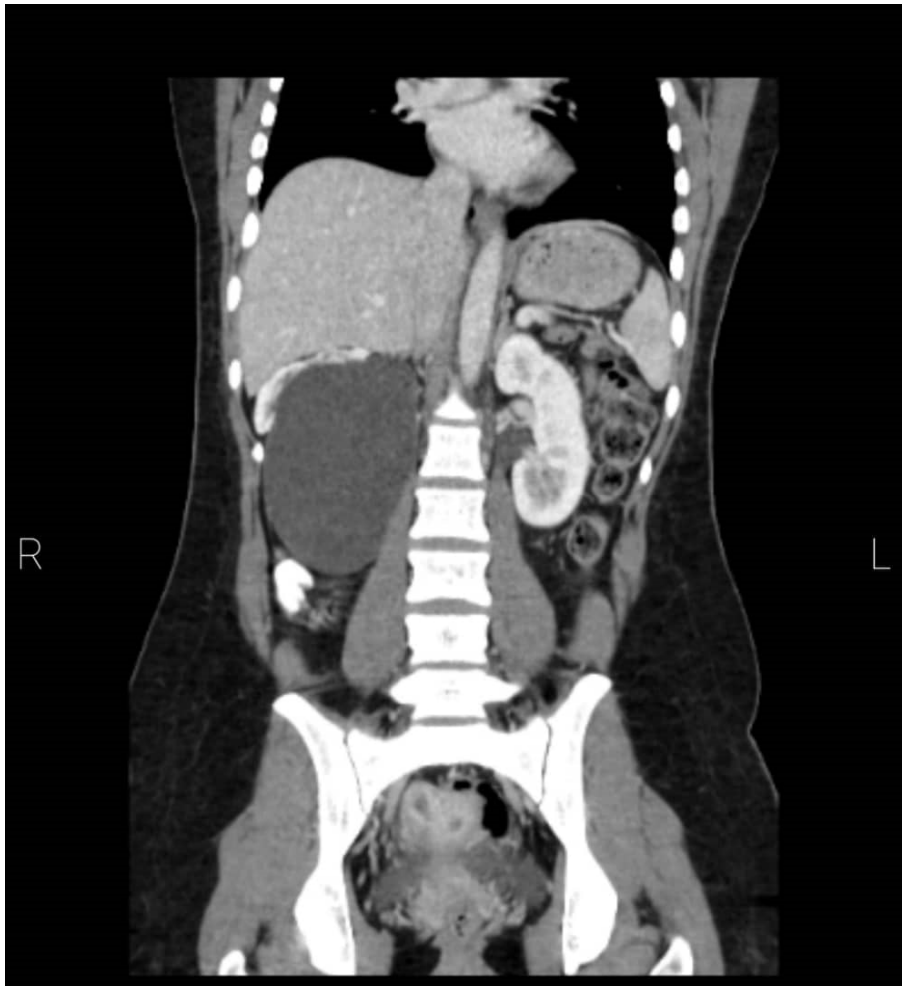

Fig. 2. Coronal view of abdominopelvic CECT scan reveals severe pelvicalyceal dilation in right kidney associated with cortical parenchymal thinning and normal ureter which is in favor of UPJO. Also, evidence of dorsal pancreatic agenesis without vascular or GI loop malrotation is seen

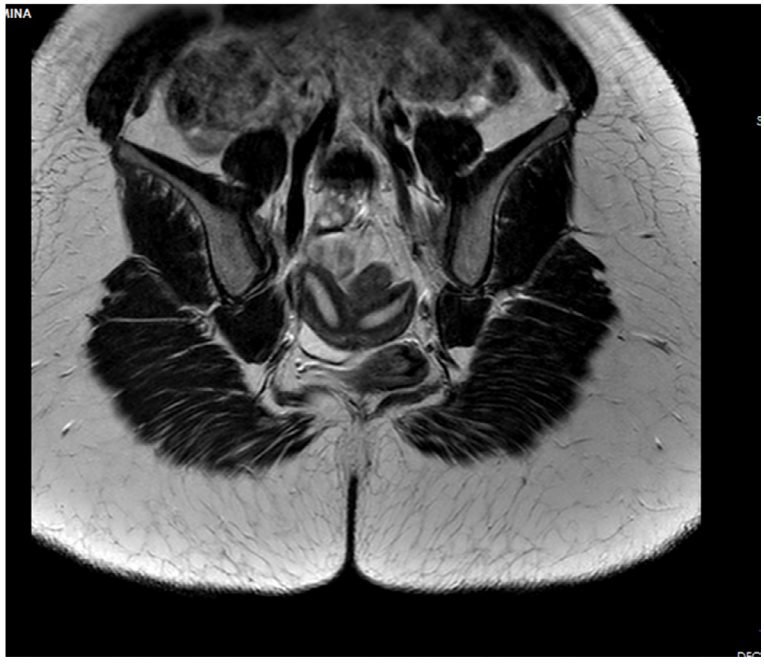

Fig. 3. Axial oblique T2-weighted TSE pelvic MRI reveals two separate uterine horns

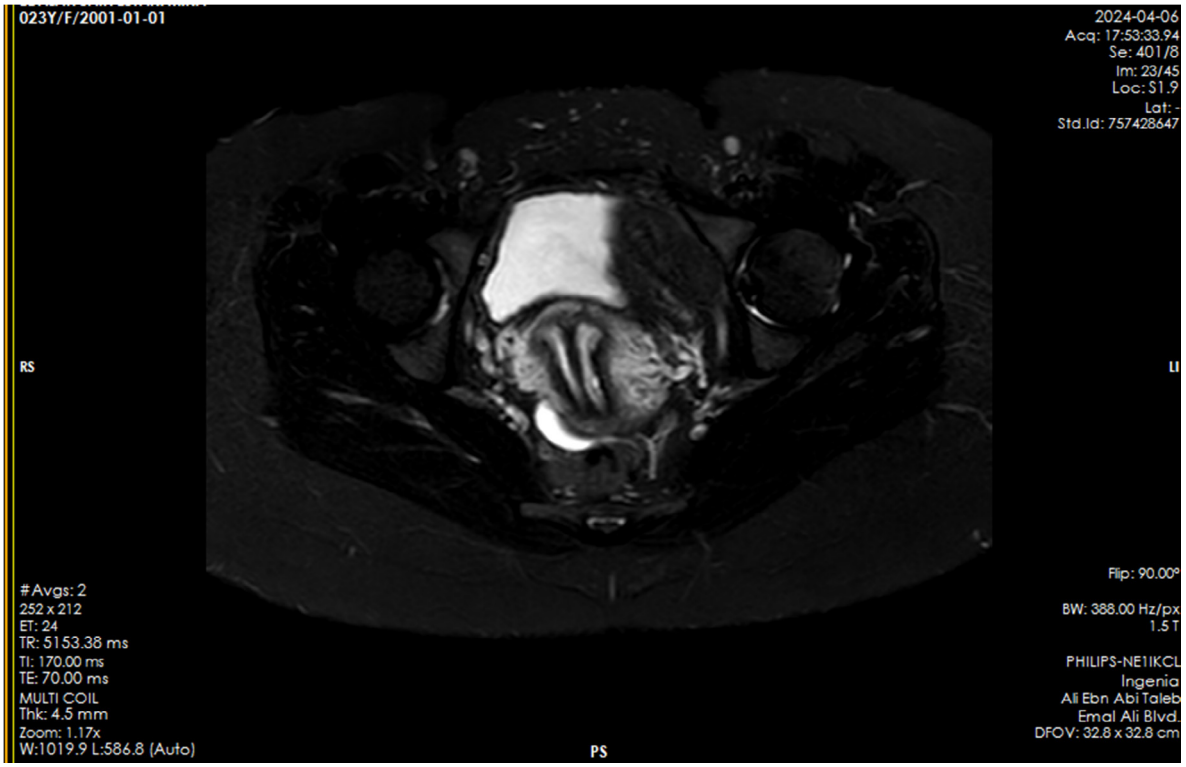

Fig. 4. Axial T2-weighted STIR pelvic MRI reveals two separate uterine horns and two separated cervices as didelphys uterus

Discussion

Dorsal pancreatic agenesis is an extremely rare congenital anomaly. The first case of dorsal pancreatic agenesis was reported as an autopsy finding in 1911 [3], and there have been around 120 cases reported in the literature to date. The pancreas originates from the ventral and dorsal endodermal outgrowths of the duodenum. By the 6th to 7th week of gestation, these ventral and dorsal components merge to create the primary pancreatic duct. Around the 7th week of gestation, the ventral outgrowth shifts posteriorly to unite with the dorsal outgrowth, culminating in the formation of the fully developed pancreatic gland [4]. Developmental failures during embryogenesis can disrupt the normal formation and function of the pancreas, resulting in conditions like ADP. In cases of partial ADP, the individual may have a smaller or underdeveloped dorsal pancreas, which can impact the production and secretion of pancreatic enzymes and hormones. Complete ADP, on the other hand, can result in more severe consequences as the dorsal pancreas is entirely missing. This can lead to issues with digestion, absorption of nutrients, and regulation of blood sugar levels due to the deficiency in pancreatic enzymes and hormones [5, 6]. ADP may be accompanied by other developmental abnormalities, such as intestinal malrotation, polysplenia, coarctation of the aorta, heterotaxia, and tetralogy of Fallot [7], and only two cases have been reported in the literature of its association with uterine anomaly [8, 9]. One of them reported the association of agenesis with a unicornuate uterus, and another reported the association of agenesis with a bicornuate uterus.

The combination of genitourinary anomalies with dorsal agenesis of the pancreas is rare and exciting, as the pancreas originates from the endoderm, while genitourinary structures like the uterus derive from the mesoderm. Müllerian duct anomalies are uncommon, impacting around 1% of women overall and approximately 3% of women experiencing fertility issues [10]. These congenital abnormalities typically stem from either a lack of development (failure of formation) or a failure of the Müllerian ducts to fuse. Patients with congenital uterine anomalies face a greater risk of accompanying anomalies compared to others, such as renal, skeletal, or abdominal wall issues. Renal anomalies are the most common among these, with a prevalence of 20%-30% in patients with Müllerian defects [11].

Despite the different embryological origin, the credibility of the theory proposing an unrecognized syndrome involving dorsal pancreatic agenesis and genitourinary anomalies is supported by evidence from molecular biology and genetic experiments in mice. Genes such as TCF2/HNF1-beta have been identified to play a dual role in the development

of both the pancreas and uterus [12]. Continued research, case documentation, and collaborative efforts will be essential in unraveling the underlying mechanisms, genetic factors, and clinical implications of these rare conditions to provide optimal care and outcomes for affected individuals.

Conclusion

In this case, the incidental discovery of ADP during an imaging study for abdominal symptoms revealed not only the absence of the dorsal pancreas but also a unique association with Müllerian anomalies, specifically uterus didelphys. The coexistence of ADP with Müllerian anomalies underscores the intricate interplay between distinct organ systems and the potential for rare syndromes involving multiple developmental defects. The rarity of dorsal pancreas agenesis, compounded by its association with Müllerian anomalies, presents a diagnostic and management challenge requiring a multidisciplinary approach. Collaborative efforts among healthcare professionals from gastroenterology, radiology, urology, and gynecology are vital for comprehensive evaluation and effective management of such complex cases.

Author Contributions

Ali goodarzi: Writing - Original Draft, Writing – Review and editing

Leila Ostovar: Investigation, Writing – Review and editing

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors declare that they have no competeing interest.

Data availability statement

Data is available from the corresponding author upon reasonable request via email.

Ethical statement

We confirm that this work was conducted in accordance with the Declaration of Helsinki and that all participants provided informed consent.

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