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Case Report

A case of dorsal agenesis of pancreas associated with unilateral renal agenesis, unicornuate uterus, and ovarian ectopia: A brief review and learning points

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Abstract

Dorsal agenesis of the pancreas is a rare entity, with about 100 cases reported. It can be overlooked on ultrasound due to the non visualization of the body and tail of the pancreas.This is due to overlying gas in the stomach, which offers a poor acoustic window and obscures visualization. Renal agenesis and Mullerian duct anomalies are uncommon associations of dorsal agenesis of the pancreas because of the separate embryological origin of the pancreas and genitourinary organs. Here, we present a case of a 17-year-old patient who had dorsal agenesis of the pancreas, associated with unilateral renal agenesis, unicornuate uterus, and ectopic ovary. We describe the anomalies and discuss the radiological differential diagnosis and potential pitfalls. We provide a brief review of the literature with few radiological teaching points and possible genetic implications of the case.

Key words: Dorsal agenesis of pancreas; ectopic ovary; unicornuate uterus; unilateral renal agenesis

Introduction

Dorsal agenesis of the pancreas is a rare case, with about 100 cases being reported so far.[1] It may be associated with other congenital anomalies in the genitourinary system. Comprehensive multimodality radiological imaging is required to diagnose all the abnormalities accurately.

CaseReport

A 17-year-old girl presented with intermittent, noncyclical pain in the right lower abdomen for a few months. On clinical examination, the patient had focal tenderness in right iliac fossa at McBurney's point with no signs of generalized peritonitis, and a provisional clinical diagnosis of subacute appendicitis was made. An initial ultrasound scan, done at a peripheral center, failed to localize the appendix, and there was no localized free fluid in the right iliac fossa. The right kidney was absent in the renal fossa and pelvis, and a small, well-circumscribed oval structure with multiple peripheral hypoechogenicities was noted over the right iliac muscle lateral to external iliac artery close to the

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deep inguinal ring [Figure 1]. This was initially interpreted as the right ectopic kidney in the peripheral center, but our center raised the suspicion that it was the right ovary in an unusual location instead of a dysplastic ectopic right kidney as it resembled the ovary morphologically more than the kidney.

Contrast-enhanced computed tomography (CECT) whole abdomen on a 16 slice CT scanner (Philips) with arterial and portal venous phases was subsequently acquired in our center. Routine magnetic resonance imaging (MRI) of the whole abdomen was done in 1.5 T MRI (Philips) with additional Magnetic resonance cholangiopancreatography (MRCP) sequence, and gradient recalled echo (GRE) sequence of the thorax for further confirmation of our findings.

On contrast-enhanced CT, the right kidney was confirmed to be absent in the renal fossa and pelvis. The complete absence of the right kidney in the abdomen [Figure 2A] was associated with the absence of the right renal artery [Figure 2B]. The thoracic location of the right kidney was excluded by acquiring GRE images of MRI chest (images not shown).

The body and tail of the pancreas were not visualized in the CECT scan. Stomach and small bowel loops were noted abutting the splenic vein representing the "dependent stomach sign" and "dependent intestine sign," respectively [Figure 3A and B]. MRCP scan [Figure 3C] showed the duct of Wirsung with a short course and complete absence of duct of Santorini, confirming the diagnosis of dorsal agenesis of the pancreas. There was no peripancreatic fat stranding or any imaging evidence of pancreatitis.

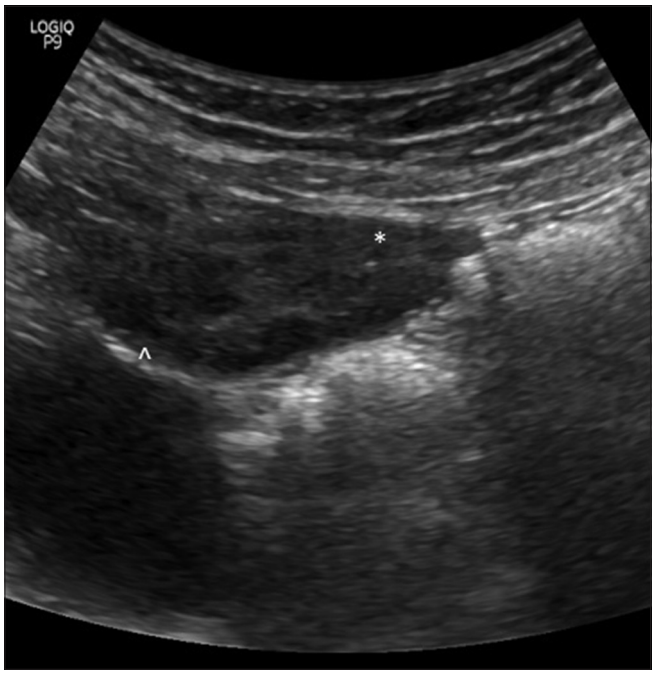

Figure 1: Transabdominal ultrasound showing ovary (*) on right iliac muscle (^). It has multiple peripheral hypoechogenities

The small, well-circumscribed, oval structure adjoining the right iliac muscle did not show contrast uptake as expected of the kidney and had no ureter associated with it. It showed multiple internal peripheral hypodensities [Figure 4A] and was suspected to be right ectopic ovary because the superior extent of the structure was above the level of the pelvic brim. MRI confirmed the presence of right ectopic ovary adjoining the right iliac muscle with numerous hyperintense follicles on T2-weighted MR imaging [Figure 4B] and no additional right ovary in the right adnexa.

In addition to the above findings, there was the presence of an unicornuate uterus in the pelvis with complete agenesis of the right horn [Figure 5] and nonvisualization of the right uterine artery [Figure 2B].

Hence, our final imaging diagnosis was a case of dorsal agenesis of the pancreas with unilateral renal agenesis, unicornuate uterus, and ectopic location of the right ovary.

Discussion

At the time of writing this case report, limited data is available on dorsal agenesis of the pancreas. The first case dates back to 1911, which was reported based on autopsy. From 1911 to 2015, only 106 such cases have been reported.[1]

Dorsal agenesis of the pancreas results from the failure of embryological development of dorsal pancreatic bud to form the body and tail of the pancreas. It is often associated with upper abdominal pain. Transabdominal ultrasound is the first imaging modality to evaluate epigastric and upper abdominal pain. However, dorsal agenesis of the pancreas can be overlooked on ultrasound study as the body and tail of the pancreas may not well be visualized

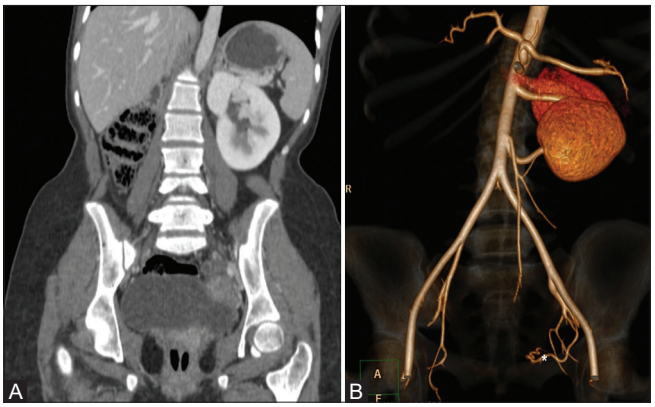

Figure 2 (A and B): (A) Coronal reconstruction of CECT abdomen showing nonvisualization of the right kidney. (B) CT angiogram with 3D reconstruction shows the absence of the right renal artery. It also depicts the nonvisualization of the right uterine artery, whereas the left uterine artery is marked with (*)

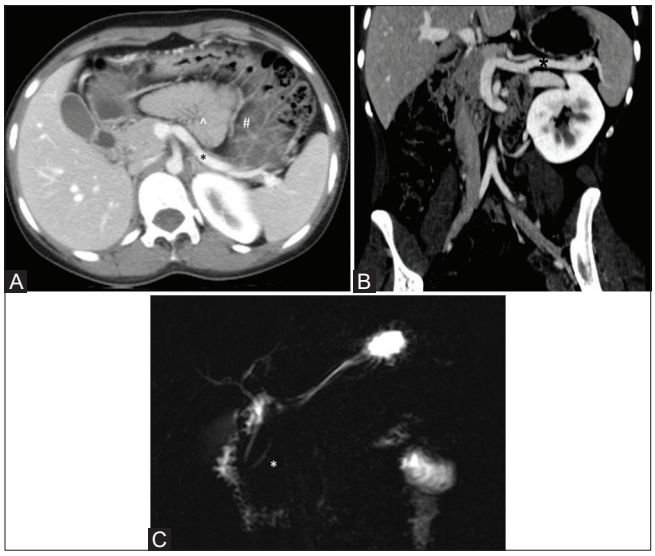

Figure 3 (A-C): (A) CECT abdomen shows absent body and tail of pancreas with small bowel (^) and stomach (#) abutting splenic vein (*).(B) Curved MPRwith reconstruction obtained along the splenic vein (*) shows an absence of body and tail of the pancreas.(C) MRCP sequence shows a short duct of Wirsung (*) and complete absence of duct of Santorini

due to overlying stomach gas. The problem can be solved by making the patient have water just before the scan to displace air from the stomach. On ultrasound, the dorsal agenesis of the pancreas can also be misdiagnosed as a pancreatic mass, because hyperechoic retroperitoneal fat or echogenic bowel in the expected location of the body and tail of the pancreas may be mistaken as a pancreatic mass. In those cases, dorsal agenesis of the pancreas can be correctly suspected by noting the presence of a very sharp interface between normal pancreas head and echogenic fat and observing the peristaltic motion of bowel. MRCP is required for the definitive diagnosis of dorsal agenesis of the pancreas. MRCP demonstrates the absence of the dorsal part of the main pancreatic duct to differentiate it from the more common condition of pancreatic atrophy due to chronic pancreatitis where pancreatic body and tail may not be seen but the main pancreatic duct is visualized.[2] On cross-sectional imaging, the two other signs which lead to the correct diagnosis of dorsal agenesis of the pancreas are dependent stomach sign and dependent intestine sign. They describe the location of the stomach and small bowel in close contact with the splenic vein, respectively. The differential diagnosis for nonvisualization of the body and tail of the pancreas also include pancreatic lipomatosis and pancreatic atrophy. However, in those cases, there would be fat between splenic vein and bowel, preventing close contact of the splenic vein with bowel. In the reported case, there was no evidence of other pathologies associated with short pancreas like preduodenal portal vein and left-sided isomerism.[3]

Unilateral renal agenesis is defined as the congenital absence of a single kidney. It usually leads to compensatory

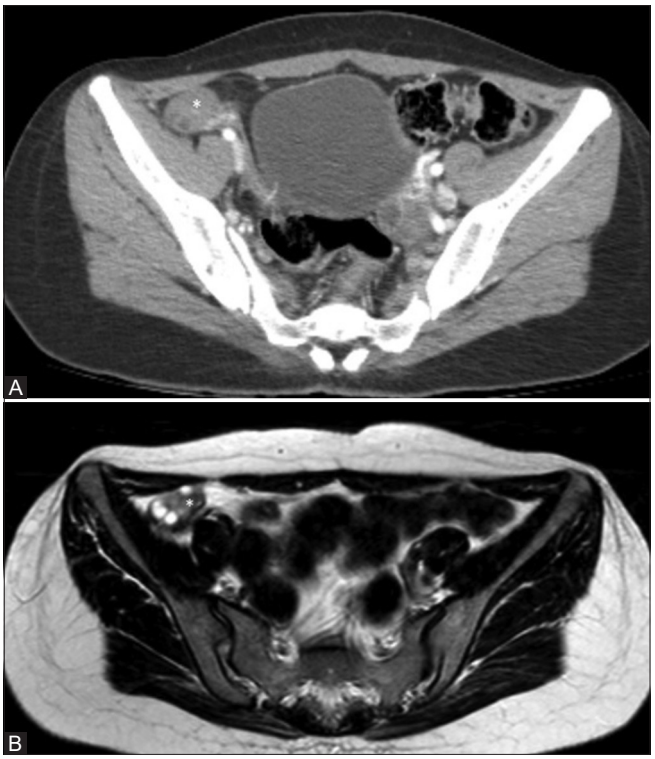

Figure 4 (A and B): (A) CECT axial section showing the location of the right ovary (*) lateral to right external iliac artery close to the deep inguinal ring.(B) T2-weighted MRI axial image showing T2 hyperintense follicles in the right ovary (*)

hypertrophy of contralateral kidney. It is also associated with the failure of the development of ipsilateral ureter, the hemitrigone or the presence of a maldeveloped ipsilateral ureteric stump. It may be associated with ipsilateral adrenal agenesis or adrenal flattening and other urogenital anomalies.[4] On imaging, unilateral renal agenesis has to be differentiated from other causes of solitary kidney like postnatal involution of multicystic dysplastic kidney and hydronephrotic kidney. In the case of unilateral renal agenesis, the position of splenic and hepatic flexures will be more medial than expected, but in the case of postnatal involution of multicystic dysplastic kidneys, the bowel position will be normal. On ultrasound, the small ectopic kidney maybe missed and maybe wrongly diagnosed as unilateral renal agenesis. Hence, for confirmation of the diagnosis of unilateral renal agenesis, either cross-sectional imaging of the thorax and abdomen has to be done or a 99m Technetium DMSA (dimercaptosuccinic acid) scan needs to be performed to exclude the presence of kidney.[5]

It is estimated that uterine anomalies occur in 1 in 500 females, and 43% of them have associated unilateral renal agenesis.[5] The two most common uterine abnormalities associated with unilateral renal agenesis are true unicornuate uterus and bicornuate uterus with a rudimentary ipsilateral horn on the same side as that of renal agenesis.[5] Clinically bicornuate uterus with a rudimentary

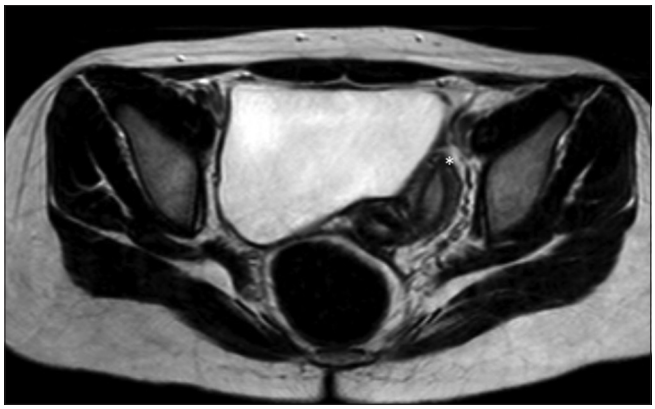

Figure 5: T2-weighted axial MRI image showing the single left horn (*) of unicornuate uterus projecting toward the left side of the pelvis

horn is more symptomatic than true unicornuate uterus and, hence, needs to be differentiated from each other on imaging. At hysterosalpingography, the unicornuate uterus will be small, deviated to one side, and have a fusiform or "banana" shape. However, hysterosalpingography may fail to demonstrate the presence of additional rudimentary horn and thereby fail to differentiate the true unicornuate uterus from the bicornuate uterus with rudimentary horn. Sonography detects the presence of additional rudimentary horn better and is best achieved when the scan is done in the second half of the menstrual cycle when endometrium inside the rudimentary horn, if present, is hyperechoic. MRI with coronal and axial oblique sequences parallel and perpendicular to the long axis of uterus is the gold standard radiological investigation to detect unicornuate uterus. MRI can differentiate it from the bicornuate uterus and other Mullerian duct anomalies with high sensitivity and specificity.[6] Our patient had true unicornuate uterus without any rudimentary horn.

Allen *et al*. have described two anatomical references to define ectopic ovaries.[7] Ectopic ovaries are defined by their presence above the pelvic brim with the upper limit of the normal location of the superior-most part of the ovary being defined by an imaginary inclined plane joining the superior surface of pubis and sacral promontory. A second method of defining an ectopic ovary is, if it is located above, a horizontal plane at the level of bifurcation of the common iliac artery into the external iliac artery and internal iliac artery.[7] The concept of the ectopic ovary was comprehensively detailed by Lachman and Berman[8] who had found the three main causes of ectopic ovary: post-surgical change, post-inflammatory change, and true congenital. The ectopic right ovary is clinically important and has been subsequently described as mimics of acute appendicitis in the published literature. Several cases of ectopic ovaries have been described to have caused menstrual irregularities and abdominal pain.[9] It has also been associated with renal agenesis because both pathologies result from defective development of urogenital ridge. Kollia *et al*. have previously published a case of the ectopic right ovary with right renal agenesis and have outlined the related embryological development of kidney and ovary.[10] Ectopic ovaries have high incidences in females with an absent uterus or unicornuate uterus, with incidences up to 20% and 42%, respectively.[11] High association has noted between Mullerian anomalies and ectopic ovary in several other studies.[7,12] On imaging, whenever an ovary is suspected in an ectopic location, a meticulous search for the detection of another ovary in the usual location needs to be done as multiple ovaries may be present on the same side. Our young patient had no previous surgery or previous complaint of similar pain in the right iliac fossa. Hence, it can be inferred that she has an ectopic right ovary due to congenital cause. In addition, there was no ovary detected in the right adnexal region in our patient.

It is of paramount importance to know that other congenital anomalies occur in association with dorsal agenesis of the pancreas. These include the ectopic spleen, bowel malrotation, heterotaxy syndrome, left-sided gallbladder, choledochal cyst, pancreatic anomalies, and bile duct anomalies, and cardiac anomalies.[13] However, the association of genitourinary anomalies with dorsal agenesis of the pancreas has not been common and is also counterintuitive because the pancreas is of endodermal origin and genitourinary structures like kidney or uterus are of mesodermal origin. In 2015, Lal *et al*. have described a case with dorsal agenesis of the pancreas, pancake kidney, and bicornuate uterus.[14] Until today, to the best of our knowledge, only three cases have been reported having both renal agenesis and dorsal agenesis of the pancreas. The first case was discussed at the Endocrinology Society's 95th Annual Meeting in San Francisco in 2013 by Lee *et al*.[15] Bagul has described the second case of pancreatic dorsal agenesis and unilateral renal agenesis in 2016, where the patient had presented with nonlocalized abdominal pain.[16] Moreira *et al*., in 2017, described the third case presenting with microalbuminuria.[15] With four similar cases, including our case getting reported in a relatively short span of 6 years, radiologists need to be vigilant about a possible association between dorsal agenesis of the pancreas and genitourinary anomalies, despite the different embryological origin.

The theory of possible existence of an unidentified syndrome consisting of dorsal pancreatic agenesis and genitourinary anomalies also gets its credibility from evidence at the level of molecular biology or mice genetic experiments. Several genes like TCF2/HNF1-beta, homeobox genes, and hedgehog genes have been found to have a dual role in the development of pancreas and kidney.[17-19]

Conclusion

Our case, though very rare, has some learning points. The radiologists, especially the ultrasonologists, need to be

aware of the existence of the entities—ectopic ovary and dorsal agenesis of the pancreas. Pain related to ectopic ovary located in right iliac fossa may be misdiagnosed as subacute appendicitis. Ectopic ovary, especially if enlarged, may mimic a dysplastic ectopic kidney on ultrasound, especially when the ipsilateral kidney in the renal fossa is not visualized. Dorsal agenesis of the pancreas may be unsuspectingly missed on ultrasound if the body and tail of the pancreas are not evaluated with precision in every routine case. Based on recent case reports and evidence from molecular biology, there is an increasing possibility of a syndromic association between dorsal agenesis of the pancreas and genitourinary anomalies and awareness from radiologists will help toward detection of more cases and identification of a possible new syndrome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Cienfuegos JA, Rotellar F, Salguero J, Benito A, Solórzano JL, Sangro B. Agenesis of the dorsal pancreas: Systematic review of a clinical challenge. Rev Esp Enferm Dig 2016;108:479-84.
2. Pasaoglu L, Vural M, Hatipoglu HG, Terekliogluu G, Koparal S. Agenesis of the dorsal pancreas. World J Gastroenterol 2008;14:2915.
3. Kim S-H, Cho Y-H, Kim H-Y. Preduodenal portal vein: A 3-case series demonstrating varied presentations in infants. J Korean Surg Soc 2013;85:195-7.
4. Mishra A. Renal agenesis: Report of an interesting case. Br JRadiol 2007;80:e167-9.
5. Shapiro E, Goldfarb DA, Ritchey ML. The congenital and acquired solitary kidney. Rev Urol 2003;5:2.
6. Khati NJ, Frazier AA, Brindle KA. The unicornuate uterus and its variants: Clinical presentation, imaging findings, and associated complications. J Ultrasound Med 2012;31:319-31.
7. Allen JW, Cardall S, Kittijarukhajorn M, Siegel CL. Incidence of ovarian maldescent in women with mullerian duct anomalies: Evaluation by MRI. AJR Am J Roentgenol 2012;198:W381-5.
8. Lachman MF, Berman M. The ectopic ovary. A case report and review of the literature. Arch PatholLab Med1991;115:233-5.
9. Cohen J, Holzman M. A giant ectopic ovary. J Laparoendosc Adv Surg Tech 2001;11:31-5.
10. Kollia P, Kounoudes C, Veloudis G, Giannakou N, Gourgiotis S. True ectopic ovary in the right iliac fossa mimicking acute appendicitis and associated with ipsilateral renal agenesis. J Obstetr Gynaecol Res 2014;40:858-61.
11. Ombelet W, Verswijvel G, de Jonge E. Ectopic ovary and unicornuate uterus. New Engl J Med 2003;348:667-8.
12. Kanter J, Jarrard M, Gavrilova-Jordan L. Management of ectopic ovary in the setting of mullerian agenesis [35P]. Obstetr Gynecol 2017;129:S172.
13. Guimarães AB, Guimarães CA, Manso JEF. Agenesis or pseudoagenesis of the dorsal pancreas. Rev ColBras Cir 2015;42:352-5.
14. Lal A, Pavunesan SK, Mahalingam H, Garg M, Sinha SK, Khandelwal N. A triad of complete dorsal pancreatic agenesis, pancake kidney and bicornuate uterus. An association or an incidental finding: First case in literature. JOP 2015;16:189-91.
15. Moreira A, Carvalho A, Portugal I, Jesus JM. Complete dorsal pancreatic agenesis and unilateral renal agenesis. Radiol Case Rep 2018;13:68-71.
16. Bagul M TP. Complete dorsal pancreatic agenesis and unilateral renal agenesis: First case report and review of literature. Int J Anat Radiol Surg 2016;5:RC1-3.
17. Body-Bechou D, Loget P, D'Herve D, Le Fiblec B, Grebille A-G, Le Guern H, *et al*. TCF2/HNF-1beta mutations: 3 cases of fetal severe pancreatic agenesis or hypoplasia and multicystic renal dysplasia. Prenatal Diagn2014;34:90-3.
18. Rauchman M. The role of homeobox genes in kidney development. Curr Opin Nephrol Hypertens 2000;9:37-42.
19. Yu J, Carroll TJ, McMahon AP. Sonic hedgehog regulates proliferation and differentiation of mesenchymal cells in the mouse metanephric kidney. Development 2002;129:5301-12.