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Ectopic Kidney and Intestinal Anomalies in a Patient with Pancreatic Pseudocyst

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Abstract
Developmental anomalies involving the urinary system coexisting with that of the gastrointestinal tract are rare. This case reports a middle-aged woman presenting with progressive abdominal distension and discomfort. Radiological investigations showed a huge pancreatic pseudocyst and crossed left renal ectopia. Abnormality of the parietal peritoneum was noted at surgery where the duodenum and the ascending and descending colon were completely intraperitoneal with mesentery and mobile. The appendix was also found to be absent. She had successful surgery with internal drainage of the pseudocyst. Attention should be given to this coexisting clinical scenario in patients with ectopic kidney.

Keywords: Ectopic kidney, crossed renal ectopia, congenital intestinal anomalies, pancreatic pseudocyst, agenesis vermiform appendix

Introduction

The upper urinary tract is the most affected with congenital anomalies, accounting for 3 to 11% of congenital anomalies in the population [1,2]. Congenital anomalies leading to renal malposition can result in crossed renal ectopia whereby the kidney is located on the opposite side in relation to its embryonic position. This is thought to result from abnormal crossing over of the embryonic metanephric blastema and ureteric button from its migration from the pelvis either due to the abnormal position of the umbilical artery or from excessive bending of the embryo. Despite this position, the ureter is still inserted into the normal side of its embryonic origin. As seen in this patient, both kidneys were separate and were located on the right side of the spine. Normally, the duodenum (apart from its first part) and the ascending and descending colon are relatively fixed to the posterior abdominal wall by the parietal peritoneum during embryonic development. They are therefore retroperitoneal abdominal structures. This case also presented gastrointestinal anomalies involving the parietal peritoneum in which the duodenum and colon were completely intraperitoneal with a mesentery and the appendix was found to be absent. Added to this conundrum was the development of a pancreatic pseudocyst causing pressure symptoms to surrounding structures.
This case reports a rare complex scenario of clinical conditions that included left crossed renal ectopia, duodenal and colonic attachment anomalies, vermiform appendix agenesis and a pancreatic pseudocyst coexisting and diagnosed in an adult female.

Case Presentation

A 36-year-old female presented with progressive abdominal distension for 13 years associated with vague abdominal pain. She reported no other complaints, nor any previous abdominal surgery. Physical examination revealed a grossly distended abdomen with fluid. No other significant clinical finding. An abdominal CT scan showed a huge multiloculated cystic mass measuring 20cm x 18cmx 29cm. The mass showed few peripheral calcifications and was inseparable from the pancreas posteriorly. It exerted mass effect on the spleen, adjacent intestine, and two kidneys on the right (Figure 1). An intravenous urogram (IVU) showed that the right kidney was correctly placed, but the left kidney was found below the right kidney with hydronephrosis and kinked left ureter running across the midline to the left ureteric orifice (Figures 2 and 3). The renal function test was normal.

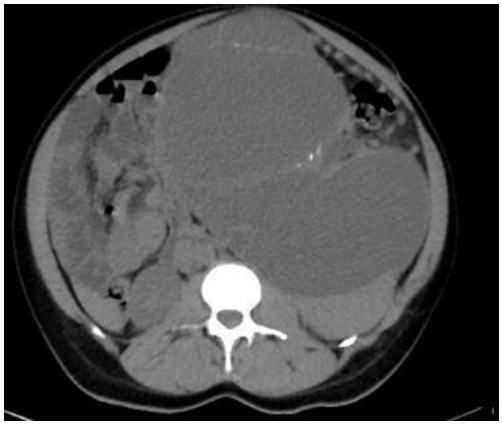


Fig 1: Abdominal CT scan showing huge pancreatic pseudocyst with two kidneys by its right side.

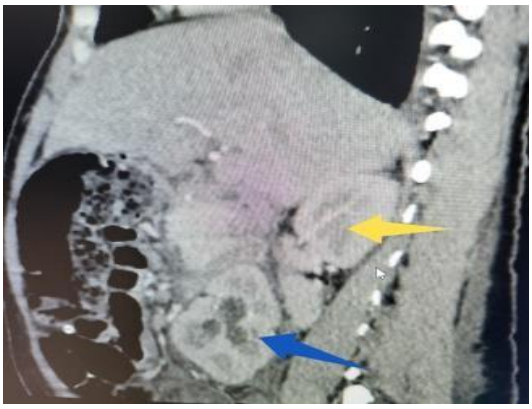


Fig 2: IVU showing ectopic left hydronephrotic kidney.

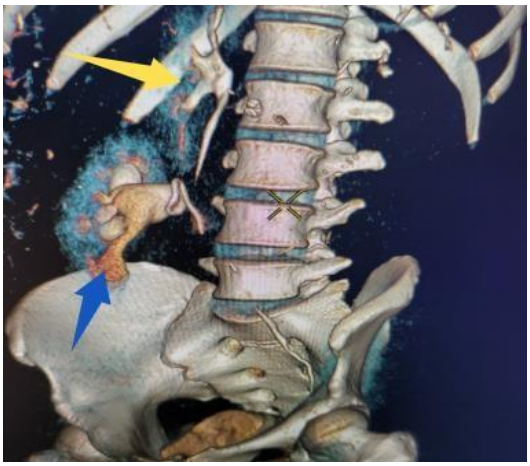


Fig 3: Three-dimensional reconstruction of IVU showing the ectopic left hydronephrotic kidney (blue arrow) below the normal right kidney (yellow arrow).

She was prepared and subsequently had a laparotomy. Findings at surgery included a huge pancreatic pseudocyst with thickened walls containing 3 litres of turbid fluid and necrotic debris. The stomach was found to be normal. The entire duodenum and colon were completely intraperitoneal with mesentery and mobile. The appendix was missing (Figure 4). The left lumbar region was empty with no kidney, but the right kidney was intact and in the normal position. The left kidney was found in the right side of the abdomen attached to the right side of the pseudocyst wall (Figure 5). She had a side-to-side cystojejunostomy for internal drainage of the pseudocyst fluid. Peritoneal lavage with warm saline was performed and wound was closed over an active drain. She had post-operative hypokalemia that was corrected and bilious vomiting that settled. She was discharged after 4 weeks on admission.

Histology of the pseudocyst wall showed fibrous tissue interspersed with goblet cells and cells that contained elongated chromatin.

Post-operative investigations including CT intravenous urogram and renal scintigraphy were scheduled but she was lost to follow-up.

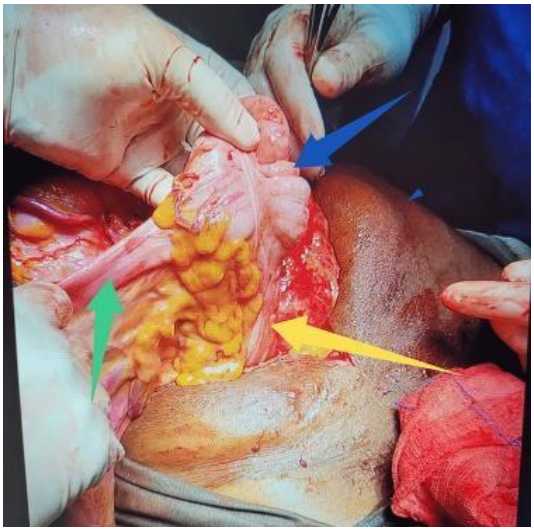


Fig 4: Colon anomalies showing mobile ascending colon on its mesentery (yellow arrow), caecum without the appendix, and terminal ileum (green arrow)

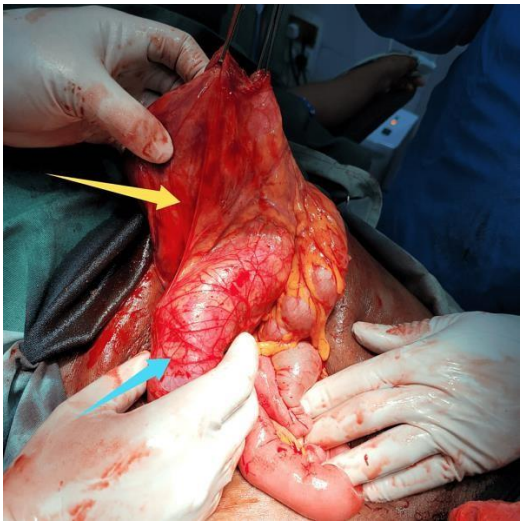


Fig 5: Crossed renal ectopia with ectopic left kidney (blue arrow) attached to the right side of the pancreatic pseudocyst wall (yellow arrow), which had already been drained

Discussion

Ectopic kidneys occur in 1 in 1,000 life births with only 10% of cases diagnosed [3]. Crossed renal ectopia is also rare, affecting 7.5 of 10,000 life births [4]. Left-to-right crossed renal ectopia is two to three times more common than right-to-left crossed renal ectopia [5]. This anomaly, first described by Pannorlus [6], may be asymptomatic and diagnosed accidentally at abdominal ultrasound. If symptomatic, the commonest complaint is usually abdominal pain. However, ectopic kidneys are prone to complications like upper tract

obstruction, renal stone formation, urinary tract infection, hypertension, renal failure, and tumors. Disorders of the urinary tract have been associated with ectopic kidneys. Renal cell carcinoma and Wilms tumor have been reported [7,8]. Taslim BB *et al*. reported nephrolithiasis in crossed renal ectopia [9]. Modi *et al*. Gupta *et al*. and Ali *et al*. have all reported kidney stones occurring in children with renal ectopia [10-12]. Renal ectopia has also been associated with urethral stricture, bilateral cryptorchidism, and hypospadias [13].

Approximately two-thirds of ectopic kidneys are associated with congenital malformations of other systems such as the central nervous system, skeletal, cardiovascular and gastrointestinal systems. A rare syndrome involving the circulatory and musculoskeletal systems with crossed fused renal ectopia called Thrombocytopenia and Absent Radius (TAR) syndrome has been described by Ahmad [14]. Chhakchhuak and Agarwal in India described other rare solitary pelvic ectopic kidney and limb anomalies as rare variant of the Acrorenal syndrome.The kidney had nephrolithiasis with hydronephrosis, while oligodactyly was found in both hands and syndactyly in both feet [15].

Other rare acrorenal syndromes described include Acrorenal-Ocular syndrome characterized by optic nerve coloboma, hand and renal anomalies, Townes-Brock syndrome characterized by hearing loss, thumb anomalies and imperforate anus in addition to renal anomalies.

This case describes mesenteric, duodenal and colonic anomalies, failure of the appendix to develop and pancreatic pseudocyst coexisting with crossed renal ectopia. This is a combination of abnormalities not previously described.

For this patient, the pseudocyst may have developed from pancreatitis due to abnormality in the insertion of the pancreatic duct to the mobile duodenum causing pancreatic ductal obstruction. Hydronephrosis observed in the ectopic kidney was likely due to pressure effect on the crossing left ureter to the right by the pseudocyst, causing partial obstruction. It could also have been due to ureteric kinking. This is expected to resolve with internal drainage of the compressing pseudocyst. She is also at an increased risk of developing large bowel obstruction from colonic volvulus because of redundant mobile colon. She

has been counselled that appendicitis is not on the list of diagnosis if she ever develops lower abdominal pains because she does not have an appendix.

Conclusion

This case reports that ectopic kidney can be diagnosed at adulthood incidentally. It also highlights its association with pancreatic pseudocyst, abnormalities in duodenum and colon embryonic development, and agenesis of the appendix. This serves to raise clinical awareness on the coexistence of these conditions in clinical practice.

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