Rare Case of Associated Congenital Anomalies—Ectopic Left Kidney with Pelvic Presentation and Descending Thoracic Aorta Coarctation

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We present the case of a patient with extremely rare associated congenital anomalies: thoracic aorta coarctation and left ectopic kidney with pelvic presentation. A 45-year-old male patient was admitted to our institution for multidetector computed tomography (MDCT) angiography. On admission, he complained of intermittent claudication after 10 meters of walking, frequent chest pain, and hypertension crisis. Femoral pulse was absent in the left leg. At 14 years of age, he underwent insertion of a 10-mm Dacron tubular graft at the descending aorta level for isthmic coarctation. MDCT arteriography revealed significant stenosis (>90%) at the proximal anastomosis site of a previous graft in the form of a floating thrombus (pseudocoarctation–re-coarctation) as well as an ectopic pelvic left kidney with the left renal and accessory renal artery arising from the left common iliac artery. One year ago we attempted percutaneous angioplasty of the aforementioned in-graft stenosis. At the time, the procedure was partly successful and the patient had been doing well, but all of the aforementioned symptoms reappeared and CT angiography showed no signs of improvement. Therefore, ascending-to-descending aortic bypass was done with an 18-mm Dacron tubular graft with preserved postoperative renal function and palpable peripheral pulses. MDCT angiography showed normal postoperative findings and the patient was doing well. In the case presented, MDCT angiography played a significant role in this rare aortic anomaly detection with incidental discovery of a rare developmental kidney complication.

Of all congenital anomalies, urinary tract malformations account for 3%, whereas approximately 40% of all urinary tract anomalies are attributed to ectopic variations.[1–3] Ectopic kidney is a rare anomaly with a frequency of 1:500 to 1:1100 and solitary pelvic kidney 1:22,000, whereas having one normal and one pelvic kidney has a frequency of 1:3000.[4] Usually, these anomalies are associated with vertebral column, spinal cord, genital tract, and lower gastrointestinal tract defects.[1] On the other hand, aortic coarctation is also an exceptionally rare disease, which occurs in young patients and may be seen in 1 of 10,000 individuals, representing around 5–8% of all congenital heart defects.[5] Herein we describe the case of a patient with extremely rare associated congenital anomalies—thoracic aorta

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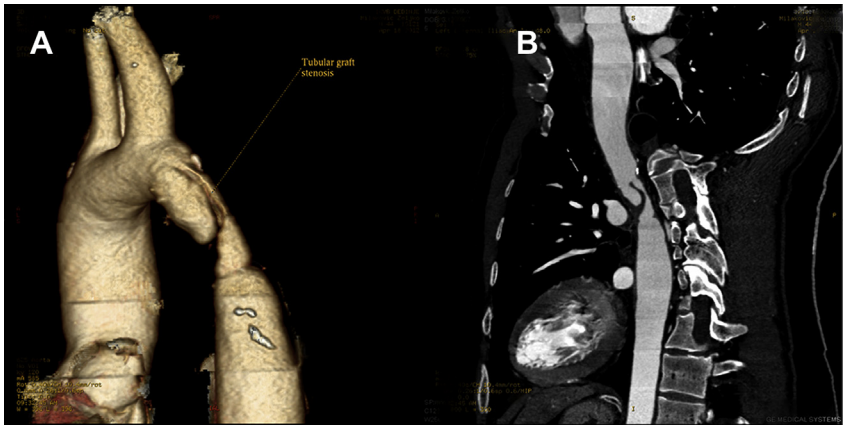

Fig. 1. MDCT arteriography. Significant (>90%) graft stenosis with floating thrombus (pseudocoarctation, re-coarctation) is shown. **(A)** Three-dimensional view. **(B)** Maximum intensity projection (MIP) view.

coarctation and left ectopic kidney with pelvic presentation.

CASE REPORT

A 45-year-old male patient was admitted to our institution for multidetector computed tomography (MDCT) angiography. On admission, he complained on intermittent claudication after 10 meters, frequent chest pain, and hypertension crisis. At 14 years of age, he underwent interpositioning of a 10-mm Dacron tubular graft at the descending aorta level for isthmic coarctation. Clinical evaluation revealed an absence of femoral pulse of the left leg, and arterial tension was 220/140 mm Hg. Laboratory findings were within referent values (urea 5.6 mmol/L, creatinine 113 mmol/L, serum potassium 4.3 mmol/L). Ankle brachial indexes were 0.38 on the right leg and 0 on the left.

Three years earlier he had a stroke resulting in left side weakness, and 10 days prior to admission he had a transient ischemic attack with dysphasia. Brain CT done prior to admission showed old ischemic lesions of the right temporal lobe but no ischemic lesion on the left side.

Ultrasonography of carotid arteries did not reveal significant stenosis. Echocardiography showed regular findings as well (ejection fraction 60% without the disorder in the wall kinetics). Eight years ago he underwent stomach surgery for bleeding ulcer; the risk factors included hyperlipidemia, smoking, and hypertension.

CT angiography revealed significant stenosis (>90%) at the site of the proximal anastomosis of a previous graft in the form of a floating thrombus (pseudocoarctation–re-coarctation) (Fig. 1). Surprisingly, it also revealed another exceedingly rare anomaly: left ectopic kidney with pelvic presentation (Fig. 2).

The left renal artery arose from the left common iliac artery as well as left accessory renal artery, whereas, for the right kidney, two polar arteries were noted (Fig. 2). Our first treatment option was percutaneous angioplasty (PTA) of in-graft stenosis and this procedure was done 1 year prior to this admission.

The procedure was partly successful at the time and the patient had been doing well; however, all of the aforementioned symptoms reappeared and CT angiography showed no signs of improvement. Therefore, we indicated surgical treatment with careful monitoring of intra- and postoperative renal function. Ascending-to-descending aortic bypass was done with an 18-mm Dacron tubular graft.

After standard median sternotomy, the ascending aorta was totally mobilized, and arterial cannulation was performed just below the innominate artery. After cardioplegic arrest, the posterior pericardium was exposed and opened. The descending aorta was mobilized below the coarctation and, at this level, distal end-to-side anastomosis with an 18-mm Dacron tubular graft was established. The graft was then tunneled with a slight angulation behind the inferior vena cava and anterior to the inferior pulmonary vein. Proximal end-to-side anastomosis was created on the lateral wall of the ascending aorta with 4-0 polypropylene suture (Fig. 3).

The postoperative course was uneventful with preserved renal function and palpable peripheral pulses of the lower limbs. Ankle brachial indexes were 1.0 in both legs. MDCT arteriography showed regular findings (Fig. 4) and the patient was doing well. There were no episodes of hypertension crisis or claudication.

DISCUSSION

Kidney pelvic ectopia occurs when kidneys normally start to develop in the pelvis but fail to migrate to their normal anatomic position in the upper abdomen. Factors that could influence this development anomaly are teratogens, with the ureteric bud not meeting with the nephrogenic blastema or genetic factors.[6,7] Ectopia could be pelvic, abdominal, thoracic, iliac, or anywhere along the path of their usual ascent.

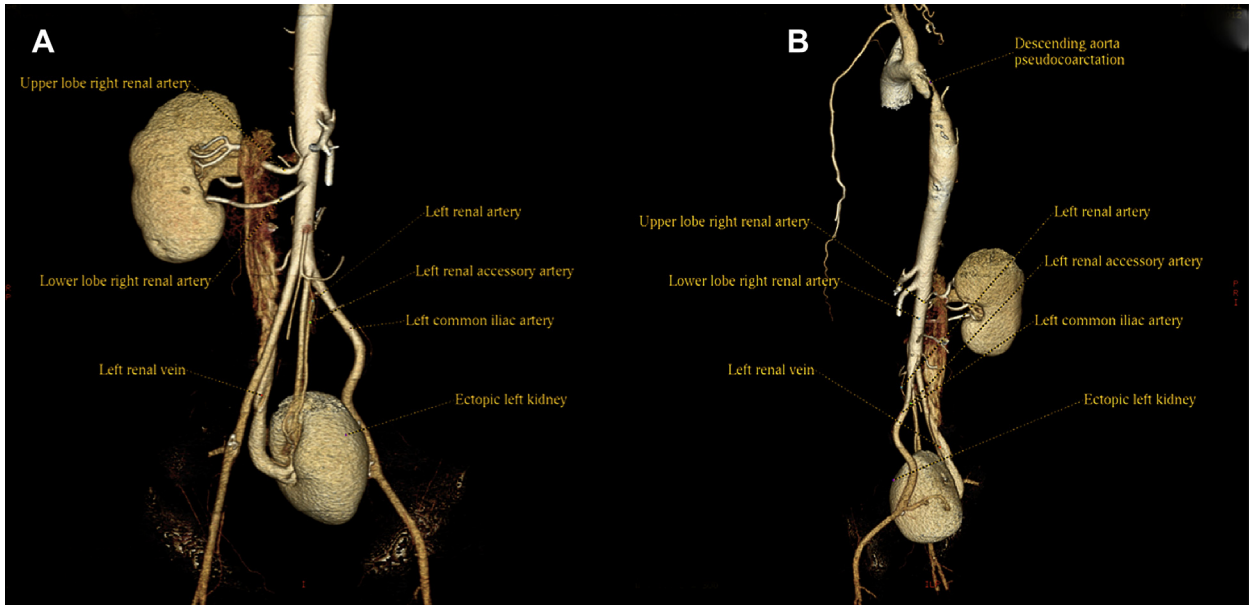

Fig. 2. MDCT arteriography. Ectopic left kidney with pelvic presentation **(A)** and with descending thoracic aorta coarctation **(B)**.

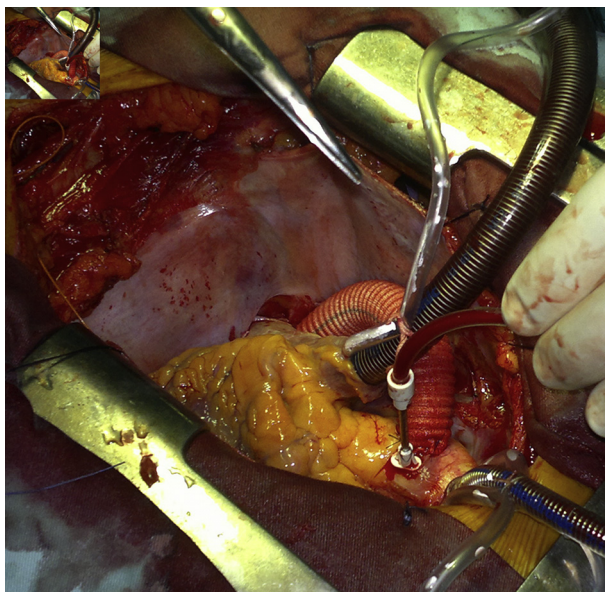

Fig. 3. Intraoperative findings after completed ascending-to-descending aortic bypass.

Aortic coarctation, on the other hand, is defined as a congenital constricted aortic segment that could be of the preductal (infantile) type or postductal (adult) type, depending on whether the coarctation segment is proximal or distal to the ductus arteriosus.[5] Hypoplasia of the portion of the aorta between the origin of the left subclavian artery and ductus arteriosus–isthmus of the aorta is present in most patients with thoracic coarctation. Aortic coarctation is usually associated with other congenital heart anomalies, but the association with left ectopic kidney seen in our case has not been described previously. Bolen et al. described the case of associated aortic pseudocoarctation and crossed fused ectopic kidney.[8] Pseudocoarctation was defined as a congenital abnormality consisting of an elongated aortic arch with a kink at the level of the aortic isthmus; however, significant flow obstruction was not associated with this anomaly.

Two theories have been described to explain possible mechanisms of aortic coarctation occurrence.[9,10] Hemodynamic theory emphasizes the importance of an abnormal preductal flow, or abnormal angle between the ductus and aorta, which increases right-to-left ductal flow, decreases isthmic flow, and, after ductus closure, initiates development of coarctation.[9] Another theory is that abnormal extension of ductal tissue into the aorta (ectopic ductal tissue)[10] creates the coarctation shelf and, with ductal closure, influences the development of aortic obstruction. This ectopic coarctation theory could be linked to the left kidney ectopic anomaly seen in our case.

It is not clear whether these two rare anomalies in our case report are isolated or a result of the same cause—congenital anomaly of ectopic tissue; however, a possible association deserves consideration. As shown in this case, descending aortic coarctation itself is very serious disease, which, if left untreated,

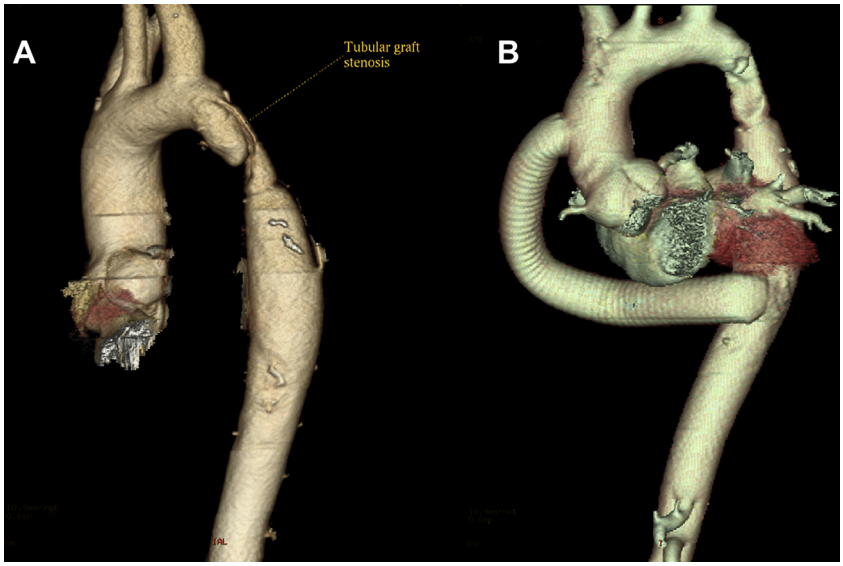

Fig. 4. MDCT angiography. Descending aorta before the procedure **(A)** and after ascending-to-descending aortic bypass **(B)**.

could cause severe lower extremity ischemia, aneurysmal degeneration, aortic wall injury and consequent dissection, kidney failure, or embolic complications. Bearing in mind stroke with left-sided weakness that the patient had 3 years prior to admission, retrograde embolization from the descending aortic lesion was considered, although it seemed unlikely that left-sided weakness could be the result of embolization of the described lesion. On the other hand, transitory dysphasia, which the patient had 10 days prior to admission, seemed very possible.

Because of the specificity and localization of the lesion, an adequate treatment option was a true challenge. Thoracic endovascular aortic repair (TEVAR) was discussed as an option; however, with the previous attempt at angioplasty taken into account, when highly calcified and fibrous characteristics of the lesion were verified, we concluded that the TEVAR procedure would have increased the risk of aortic injury and rupture with possible expansion of the endoprosthesis. Likewise, taking into consideration that this is a re-coarctation, we assumed that ''redo'' surgery would be accompanied by an increased risk of postoperative complications.

In the case presented, an elegant and feasible solution in the form of ascending-to-descending aortic bypass showed satisfactory outcome. As described in previous studies,[11,12] this approach may be performed with low postoperative morbidity and mortality with excellent technical success.[11,12] Indications for this extra-anatomic bypass were coarctation or re-coarctation with associated cardiac problems or complex coarctation or re-coarctation with anticipated difficulties with direct anatomic repair. In series of 18 patients[11] and 17 patients[12] consecutive, surgically treated patients, there were no lethal outcomes, strokes, or paraplegia and no graft-related complications in the postoperative period. In the present case, this surgical approach showed a satisfactory outcome as well.

MDCT angiography proved to be very reliable and useful procedure. For cases in which additional revascularization procedures are required after descending aorta coarctation surgical treatment or percutaneous angioplasty, information on congenital kidney anomalies offered by MDCT angiography would be tremendously helpful.

If ischemic or hemorrhagic complications occur after descending aorta surgical treatment or percutaneous angioplasty, knowledge of the presence of an atypical left renal artery from the left common iliac artery could prevent renal failure in cases of emergency iliac artery clamping, and may contribute to changes in preoperative planning.

In conclusion, MDCT angiography plays a prominent role in rare aortic anomaly detection as well as incidental discovery of rare developmental kidney anomalies. These findings could be of substantial importance if additional revascularization procedures are required after descending aorta coarctation surgical treatment and percutaneous angioplasty.

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