Kidney transplantation in a patient with absent right common iliac artery and congenital renal abnormalities

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A B S T R A C T

INTRODUCTION: Congenital atresia of the common and external iliac arteries is a rare vascular anomaly that may be associated with congenital renal or genitourinary malformations. In ESRD patients, its presence may pose potential problems during renal transplantation.

CASE PRESENTATION: We report a rare case of kidney transplantation in a patient with VACTERL syndrome who was found to have absent right common and external iliac arteries during pre-operative imaging. Vascular supply to the right lower limb is derived from an anomalous branch from the left internal iliac artery which takes on a convoluted course across the pelvis. Kidney transplantation was performed successfully with implantation performed on the left side.

DISCUSSION: Isolated cases of congenital iliac artery atresia have been described in association with urological abnormalities but no clear association has yet been established. However, we feel that it may be useful to perform routine angiographic evaluation for ESRD patients with congenital genitourinary abnormalities being planned for kidney transplantation. While most cases of congenital iliac artery anomalies are symptomatic with claudication, some remain asymptomatic with normal physical examination findings. There is some evidence in literature suggesting the usefulness of routine pre-operative CT in a selective group of patients.

CONCLUSION: Kidney transplantation in such cases is safe and we recommend routine pre-operative imaging of patients known to have congenital genitourinary abnormalities. The kidney should be implanted heterotopically to the contralateral side of the vascular anomaly and care must be taken to preserve vascular supply to the lower limbs.

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1. Introduction

Renal transplantation has become the standard of care for end stage renal disease (ESRD), including cases secondary to congenital genitourinary malformations. However, the literature on kidney transplantation in patients with associated congenital malformations of the iliac vessels is limited. Congenital vascular malformations of the ilio-femoral arteries are less common than those in the thoracic and abdominal aorta and usually discovered incidentally or by the presence of chronic lower limb ischemia [1,2]. Congenital hypoplasia or atresia are the most commonly reported malformations of the iliac arteries, and may be associated with a persistent sciotic artery [3,4]. Also, the majority of vascular malformations are unilateral, usually on the right side [1]. We herein report a very rare case of kidney transplantation in a patient with absent right common iliac artery and VACTERL (vertebral anomalies, anal atresia, cardiac defects, tracheo-esophageal fistula/atresia, renal and radial anomalies and limb defects) association.

2. Case presentation

A 24 year old male underwent an elective living-related kidney transplant (LRRT) for ESRD secondary to neurogenic bladder and vesico-ureteric reflux (VUR). He has a background history of VACTERL association with imperforate anus (previous colostomy creation and subsequent closure), sacral hemivertebrae (neuro-
logically normal) and renal/urological defects: (a) bilateral grade 5 VUR with previous bilateral ureteral re-implantation, (b) neurogenic bladder status post-colonic bladder augmentation and Mitrofanoff creation, (c) crossed fused ectopia of the left with right kidney with prior nephrectomy in preparation for LRRT (Table 1).

In view of his multiple congenital anomalies and previous surgeries, computed tomography (CT) of the abdomen and pelvis was done for pre-operative planning. The CT revealed aberrant iliac vasculature – the right common and external iliac arteries are not visualized as the abdominal aorta ends with the left common iliac artery bifurcating into the left internal and external iliac arteries (Fig. 1). A branch arising from the right distal abdominal aorta superior to the bifurcation gives off a lumbar branch before passing posterior to the right psoas muscle into the pelvis. It gives off several pelvic branches before continuing anteriorly along the pelvic side wall to receive a large anomalous branch from the left internal iliac artery, just proximal to the femoral canal. The large anomalous branch, which is the main vascular supply to the right lower limb, demonstrates a convoluted course across the pelvis from left to right. The combined vessel then continued distally as the right common femoral artery (Fig. 2).

Despite the vascular abnormalities detected on radiological imaging, the patient was asymptomatic. He had no evidence of limb ischemia and pulses were symmetrical bilaterally in the lower limbs. He subsequently underwent LRRT with implantation of kidney performed in the left iliac fossa. Intra-operatively, dissection of the left iliac vessels clearly demonstrated the anomalous vessel branching off the left internal iliac artery and this was carefully preserved (Fig. 3). The renal artery and vein were anastomosed end-to-side to the left external iliac artery and vein, respectively, a significant distance distal to the origin of the anomalous vessel. Ureteral anastomosis was performed to the augmented colonic cystoplasty. Good perfusion to the kidney was observed after vascular clamps were released and bilateral dorsalis pedis pulses were palpable intra-operatively. Post-operatively, the patient did not develop any
both demonstrated an absent right common iliac artery with per-
our patient, the cases described by Llauger et al. and Midani et al.
been described sparingly in a few isolated case reports. Similar to
hypoplasia or atresia[3].

isolated cases of congenital iliac artery atresia have previously
been described in association with renal, ureteric, renovascular,
and other pelvic abnormalities [2]. However, no clear associa-
tion between congenital iliac vasculature and renal abnormalities
has yet been established. In particular, congenital atresia of iliac
arteries has not been reported in the presence of crossed fusion
renal ectopia. Nonetheless, it should be recommended that rou-
tine angiographic evaluation be performed for ESRD patients with
congenital genitourinary abnormalities who are being planned for
kidney transplantation. Careful palpation of the femoral arter-
ies should be an important step in clinical examination of the
potential recipient and weak or unequal pulsations should be fur-
ther evaluated by imaging. While most cases of congenital iliac
artery anomalies are symptomatic with claudication, some remain
asymptomatic with normal physical examination findings like
our patient. Pre-operative CT angiography is currently not performed
routinely in patients with normal physical findings. However, a
recent retrospective review by Smith et al. to define the value of
routine CT angiography found that 22.9% of recipients had a mean-
ingful alteration in treatment based on angiographic findings [10].
Furthermore, contrast-induced nephropathy was avoided with the
use of moderate-dose nonionic iodinated contrast and adequate
hydration as there was no change in renal function or need for
emergent dialysis in the pre-dialysis cohort [10]. Although the
cost was substantial, predictors of a study that would alter care
include chronic infection, low patient weight and ventral torso
surgical scarring [10]. We feel that congenital renal abnormalities
should be added to the list of predictive factors, and CT angiog-
raphy with 3D reconstruction may be performed to evaluate for
underlying vascular anomalies that may complicate surgery. If a
similar unilateral vascular abnormality is found, the kidney should
be implanted on the contralateral normal side, with vascular anas-
tomosis performed distal to the origin of anomalous vessel and
careful preservation of the vessel.

As in our patient, the case described by Midani et al. also pos-
possessed a congenital renal abnormality with an absent right kidney
[7]. Isolated cases of congenital iliac artery atresia have previously
been described in association with renal, ureteric, renovascular,
and other pelvic abnormalities [2]. However, no clear associa-
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3. Discussion

Congenital anomalies of the iliofemoral arteries are uncommon,
with only 6 cases reported in a series of 8000 angiograms by Greeb
et al.[5]. Hypoplasia/atroisia of the external iliac artery was the most
commonly observed anomaly, present in 5 out of the 6 cases of this
series [5]. Other reports in the literature include, bilateral aplasia
of internal iliac or external iliac arteries [6,1], agenesis of external
iliac artery [4], congenital absence of a common iliac artery[2,7],
absence of unilateral common and external iliac arteries [8], abnor-
mal external iliac artery arising from the ipsilateral renal artery [9],
and quadriﬁcation of the terminal aorta where all four iliac arter-
ies arose separately from the aorta [5]. A report by Tamisier et al.
classiﬁed congenital malformation of the external iliac artery into
three groups: (1) anomalies in origin or course of the artery, (2)
hypoplasia or atresia with a persistent sciatic artery and (3) isolated
hypoplasia or atresia [3].

Congenital absence/atroisia of the common iliac artery has only
been described sparingly in a few isolated case reports. Similar to
our patient, the cases described by Llauger et al. and Midani et al.
both demonstrated an absent right common iliac artery with per-
fusion to the right lower limb supplied by an anomalous branch
from the left iliac internal iliac artery, which was seen to be continuous
with the right external iliac artery after a convoluted course hori-
izontally across the pelvis. Inferior gluteal and pudendal branches
could be identiﬁed arising from the aberrant vessel before its end-
ing in the right external iliac artery [2,7]. Despite the similarities,
our patient is unique in several ways — such a vascular anomaly, to
our knowledge, has never been described in a patient with VACTERL
association. This is also only the second reported case of kidney
transplantation in an ESRD patient with congenital absence of the
common iliac artery.

Fig. 4. MAG 3 scan depicting perfusion to the transplanted kidney in the left iliac
fossa.

Our experience from this case has shown that it is safe to
perform kidney transplantation in such cases with the appro-
imate pre-operative imaging and surgical planning. There appears
to be no evidence of lower limb ischemia or steal phenomenon
and no compromise of graft function. The presence of an absent
common iliac artery is therefore, not a contraindication to trans-
plantation. However, the presence of such a vascular abnormality
may pose a potential problem in the future with the development
of atherosclerotic disease or thrombosis. These patients should be
kept on close regular follow-up in terms of the vascular status of
their lower limbs.

4. Conclusion

Kidney transplantation in such cases is safe and we recommend
routine pre-operative imaging of patients known to have congen-
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Conflict of interest

The authors of this manuscript have no conflicts of interest to disclose.

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Author contribution

C.M. Tay, T.K. Ng and H.Y. Tiong were part of the surgical team that performed the operation and contributed in designing the report; E.P.Y. Siew reported the pre-operative scans and performed the 3-D reconstruction of images; A. Vathsala has been responsible for the posttransplant care of the patient and is his Principal Renal Transplant physician. C.M. Tay wrote the manuscript; T.K. Ng and H.Y. Tiong helped with revision of the manuscript.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. All patient identifiers have been removed to ensure the patient confidentiality.

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