Case Report

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Dissection of Iliac Artery in a Patient With Autosomal Dominant Polycystic Kidney Disease

A Case Report

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Abstract

Autosomal dominant polycystic kidney disease (ADPKD) is a risk factor for several cardiovascular disorders such as intracranial aneurysm or aortic dissection, preferentially occurring at the thoracic or abdominal level. A 47-year-old man suffering from ADPKD had renal transplantation. Sixteen hours after surgery, he presented with left leg pain. Clinical and ultrasound examination revealed thrombosis of the external left iliac artery. Therefore, we decided to perform intra-arterial angiography to evaluate the possibility of an endovascular treatment. Aortofemorography showed an obstruction of the external left iliac artery that was found during emergency surgery, consecutive to a dissection, which occurred following the surgery for kidney transplantation. The resected segment of the dissected vessel was analyzed by histology. Collagen fibers organization and density in the adventitia and smooth muscle cells density in the media were similar in the dissected and a normal artery from a healthy donor. By contrast, an almost complete disappearance and fragmentation of elastic lamellae were observed in the media of the dissected artery, most likely responsible for the weakening of the arterial wall and its dissection. Association between ADPKD and single dissection of the iliac artery has been rarely reported. Relationship between inactivation of polycystin/PKD genes and elastic fibers degradation through elevated TGFβ signaling and matrix metalloproteinase 2 (MMP2) elastolytic activity, as recently reported in ADPKD, would be worth investigating.

Key Words

Autosomal dominant polycystic kidney disease · Arterial dissection · Elastic lamellae

Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is the most frequent genetic renal disorder with an incidence of 1/1000. It is characterized by the progressive formation of fluid-filled cysts in the kidney leading to early onset renal failure. Several cardiovascular disorders have been associated with ADPKD, hypertension being the most common problem [1], while other major complications include intracranial and aortic aneurysms and dissections (for review, see [2]). Here, we report a case of left iliac artery dissection in a patient with ADPKD.

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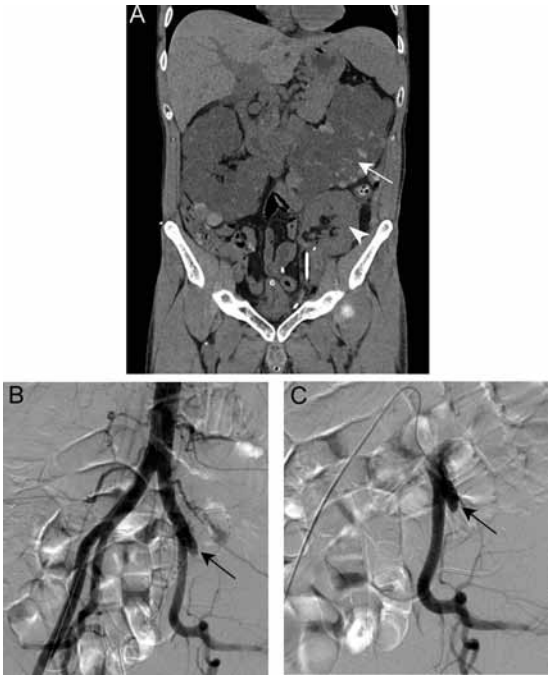

Figure 1. Imaging of the ADPKD patient. (A) CT-scan showing the polycystic kidney (**arrow**) and the transplanted kidney (arrowhead). (B) and (C) Aorto-femorography showing the occlusion of the left external iliac artery (**arrow**).

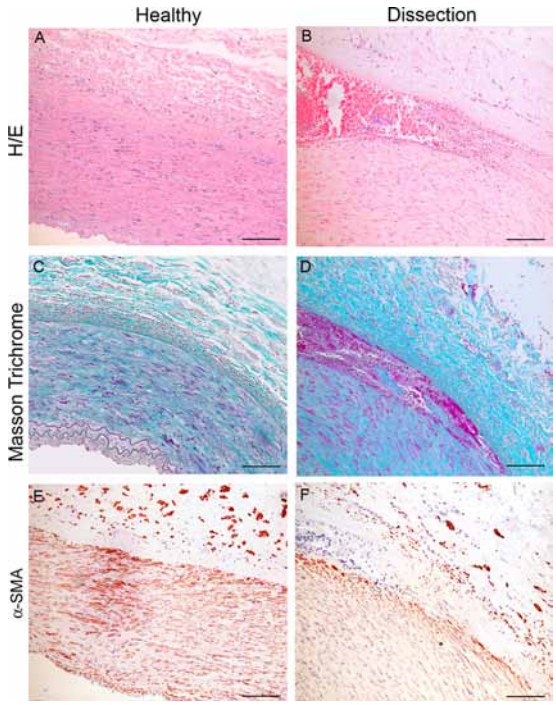

Figure 2. General organization of the wall of the dissected iliac artery. (A) and (B): hematoxylin/eosin staining. (C) and (D): Masson trichrome staining. (E) and (F): α-smooth muscle actin immunohistochemistry performed on a healthy iliac artery (A, C, E) and the dissected segment of the iliac artery of the ADPKD patient (B, D, F). Bar = 100 μm.

Case Report

A 47-year-old man presenting with familial polycystic kidney disease (ADPKD) underwent surgery for kidney transplantation. During surgery, any macroscopic anomalies were observed at the level of close iliac artery. However, approximately 16 hours after surgery, the patient presented pain at the level of the left leg. Clinical examination revealed absence of left femoral pulses. Ultrasound examination confirmed thrombosis of the aorto-iliac access. Computed tomography (CT) angiography revealed the absence of perfusion at the level of the transplanted kidney and confirmed the thrombosis of the left iliac artery (Fig. 1A). Therefore, intra-arterial angiography was performed to decide which treatment could be performed. An endovascular treatment was not possible because of the complete thrombosis of the external left iliac artery with suspected dissection (Fig. 1B and 1C). The patient underwent emergency surgery, during which a localized hematoma was observed at the level of the arterial anastomosis with extension to the external left iliac artery. The diagnosis of dissection was made after reopening of the anastomosis. Because of the fragile aspect of the arterial tissues, we performed a large resection of the dissected segment of the iliac artery that was replaced by an arterial prosthesis. The donor renal artery was reimplanted on the prosthesis. Pieces of the dissected segment were fixed for histological analysis. This study was approved by the hospital-university ethics committee.

The general organization of the iliac artery was analyzed by hematoxylin/eosin (H/E; Fig. 2A and 2B) while a Masson trichrome staining allowed to evaluate the fibrillar collagen framework (Fig. 2C and 2D) and an immunostaining with anti-α-smooth muscle actin (α-SMA) showed smooth muscle cells (Fig. 2E and 2F). The H/E staining clearly showed a large blood infiltrate between the adventitia and the media (Fig. 2B). As compared to a healthy iliac artery, no difference was observed in the collagen framework or in the smooth muscle cells organization at the level of the dissected wall. However, an orcein staining showed a striking rarefaction and disruption of the elastic lamellae in the media of the dissected iliac artery (Fig. 3B and at

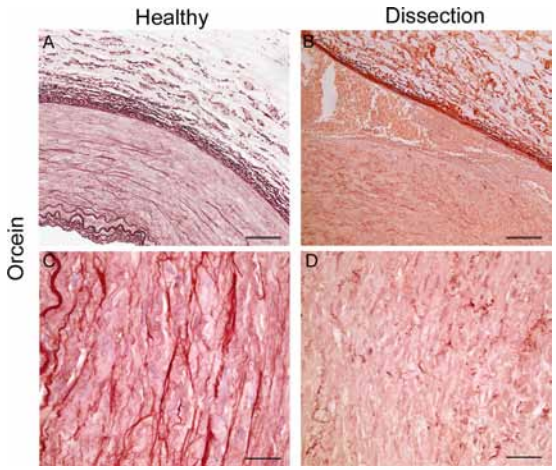

Figure 3. Visualization of elastic fibers by orcein staining of a healthy iliac artery (A) and (C) and the dissected segment of the iliac artery of the ADPKD patient (B) and (D). Bar = 100 μm in (A) and (B) and 25 μm in (C) and (D).

higher magnification Fig. 3D) as compared to a healthy artery (Fig. 3A and 3C). The dissection occurred at the junction of the external elastic lamella and the media. No inflammatory cells were found in the media or in the adventitia.

Discussion

Spontaneous dissection of the iliac artery is an extremely rare event and may occur as a complication of traumatic injuries or systemic disorders such as Marfan syndrome and α-1 antitrypsin deficiency [3,4]. ADPKD has been associated with a large number of cardiovascular disorders. Intracranial aneurysms occur with a 10% incidence in ADPKD patients [5]. Aortic dissection, which usually occurs at the thoracic level, is a rare complication of ADPKD. In this report, we describe the first case of a spontaneous iliac artery dissection associated with ADPKD without any other apparently affected blood vessel. This genetic disease is caused by mutations in the PKD1 or PKD2 genes encoding for, respectively, polycystin 1 and 2. These proteins are expressed by vascular smooth muscle cells (VSMC) and are involved in Ca^{2+} homeostasis and in cell interactions with their surrounding extracellular matrix that are critical in maintaining the integrity of the media and regulating the VSMC phenotype [2,6]. The density of VSMC and the expression of α-SMA in the media of the dissected wall appeared both normal, which suggests that elastic fibers rarefaction is not related to VSMC apoptosis. Mutations in PKD genes could, however, result in defective interactions between VSMC and the surrounding matrix, which, in turn, would lead to modification of the VSMC pattern of protein expression and to degradation of the elastic tissue. Recently, elevated TGFβ signaling was observed in an advanced stage of ADPKD, coinciding with increased levels of target genes of the TGFβ pathway such as matrix metalloproteinase 2 [7], known for its elastolytic activity and its activation in abdominal aortic aneurysms [8].

In this case report, we observed a potential association between ADPKD and iliac dissection that would need to be validated on a large series of patients.

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