Congenital Arteriovenous Fistula of the Horseshoe Kidney with Multiple Hemangiomas

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SUMMARY

Introduction Congenital renal arteriovenous fistulas (AVF) are rare, especially if they are associated with other developmental renal anomalies.

Case Outline A 34-year-old female was hospitalized due to total painless hematuria and bladder tamponade. Excretory urography revealed a horseshoe kidney with normal morphology of pyelocaliceal system and ureters. Aortography and selective renovasography detected a cluster-like vascular formation with multiple arteriovenous fistulas (AVF). Due to a large AVF gauge and poor flow of the efferent vein to the inferior vena cava, a surgical procedure of two renal artery segmentary branches ligation and division was performed. During the operative procedure, the presence of multiple superficial renal hemangiomas was detected.

Conclusion Although selective arterial embolization represents the preferable treatment option, conventional surgery remains favorable alternative in selected cases with large and complex AVF.

Keywords: congenital arteriovenous fistula; horseshoe kidney; multiple hemangiomas; selective arterial embolization; conventional surgery

INTRODUCTION

Congenital arteriovenous fistulas (AVF) of the kidney are rare. Renal AVF, initially described by Varela in 1928, represents a low incidence clinical entity. There are two types of fistulas, congenital and acquired [1]. Acquired fistulas are more frequent, while the congenital account up to 25%, mainly in women after the third or fourth decade [2]. The lesion is located in the upper pole in almost 50% of patients. Since it is often located next to the collecting system, micro- or macroscopic hematuria has been reported in more than 75% of patients [3]. Blood shunting into the venous circulation results in a heterogeneous clinical picture. The clinical presentation depends on the AVF size and duration [4]. Excretory urography may reveal some specific radiological signs such as irregular filling defect in the pelvicalyeal system, hypo- or afunctional segment of the kidney or calyectal distortion or obstruction. However, these typical radiological features are present only in 50% of excretory urograms. Although three-dimensional Doppler ultrasonography and MR angiography are noninvasive and more accurate than excretory urography, selective renal arteriography or digital subtraction angiography represent the most definitive diagnostic methods. Though selective embolization is the most preferable treatment option, other procedures including balloon catheter occlusion, vascular ligation, partial nephrectomy or nephrectomy are still indicated in selected cases [2, 5].

CASE REPORT

A 34-year-old female was hospitalized due to a sudden and abundant painless hematuria occurring for the first time. Excretory urography revealed the horseshoe kidney with a normal morphology of the pyelocaliceal system and ureter (Figure 1).

Lower contrast medium concentration was observed in the lower pole calyces of the right kidney. Since it is often located next to the collecting system, micro- or macroscopic hematuria has been reported in more than 75% of patients [3]. Blood shunting into the venous circulation results in a heterogeneous clinical picture. The clinical presentation depends on the AVF size and duration [4]. Excretory urography may reveal some specific radiological signs such as irregular filling defect in the pelvicalyeal system, hypo- or afunctional segment of the kidney or calyectal distortion or obstruction. However, these typical radiological features are present only in 50% of excretory urograms. Although three-dimensional Doppler ultrasonography and MR angiography are noninvasive and more accurate than excretory urography, selective renal arteriography or digital subtraction angiography represent the most definitive diagnostic methods. Though selective embolization is the most preferable treatment option, other procedures including balloon catheter occlusion, vascular ligation, partial nephrectomy or nephrectomy are still indicated in selected cases [2, 5].

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General renal function was normal and dynamic scintigraphy showed the right kidney hypertrophy to medium extent with impairment of its function.

DISCUSSION

Congenital renal AVF are the result of developmental renal disorders and those related to its vascular formation, and therefore, their association with other developmental renal anomalies may be expected. However, only one case was reported in the literature within our reach; associated congenital AVF with duplicated renal pelvises. Since congenital AVF are rare malformations [6], our patient is considered very interesting on account of three associated anomalies, arcuate kidney with AVF and multiple superficial renal hemangiomas.

Selective renovasography is the method of choice for renal AVF diagnosis [7, 8]. The management of renal AVF is individual and depends on the size and localization of changes as well as on the severity of symptoms. In general, small asymptomatic fistulas do not require any treatment, especially if they are acquired malformations, because their spontaneous restitution is common [9]. Nephrectomy is justified only in cases where conservative surgery is not feasible due to AVF size and localization and if renal function is compromised [10]. Partial nephrectomy has been rarely performed these days and may be considered only in polar and deep intrarenal AVF [9, 11]. Selective arterial embolization is widely accepted as the preferable option in AVF management and is especially favorable for small AVF [6, 8].

As we decided against the supraselective arterial embolization, our choice was surgery due to easier approach to the renal artery of arcuate kidney. Although conventional surgery has limited role now, it remains the favorable alternative in selected cases with large and complex AVFs or following failure of selective arterial embolization.
Конгенитална артериовенска фистула потковичастог бубrega с мултиплим хемангиомима

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КРАТАК САДРЖАЈ
Увод Урођене артериовенске фистуле бубрега су ретке, нарочито уколико су удржане с осталим развојним поре­међајима бубрега.

Приказ болесника Жена стара 34 године примљена је на болничко лечење због талатне безболне хематурије и тампо­наде мокраћне бешке. Интрагенска пивелографија открила је потковичастости бубрег нормалне морфологије пивелокаликсног система и уретера. Ангиографија и селективна ре­новелографија откриле су грозадство васкуларну формацију с вишеструким артериовенским фистулама. Због великих димензија ових фистула и слабог протока крвј из еферентне вене у доњу шупљу вену, урађена је хируршка интервенција подвезивања и пресецања две сегментне гране бубрегне артерије. Током операције откривен су вишеструке пов­­шинских хемангиоми бубрега.

Закључак Иако је селективна артеријска емболизација те­рапија избора, конвенционално хируршко лечење остаје погодна альтернатива код одобраних болесника с великим и комплексним артериовенским фистулама.

Кључне речи: конгенитална артериовенска фистула; пот­­ковичастости бубрег; мултиплим хемангиоми; селективна арте­ријска емболизација; конвенционална хирурзија

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