Interesting Case

Congenital renal arteriovenous malformation presenting as severe hypertension

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Introduction

Hypertensive patients presenting at the extremes of age may have a secondary cause of hypertension. Congenital arteriovenous malformation (AVM) and acquired arteriovenous fistula (AVF) are rare causes of secondary hypertension [1–10]. Acquired AVFs tend to be single linear connecting vessels and comprise 70–80% of renal arteriovenous abnormalities and usually result from trauma, biopsy, surgery, malignancy, or inflammation [1,3]. Congenital AVM is described as crisoid with a knotted, tortuous appearance of numerous feeding vessels, and multiple interconnecting fistulas; however, a variant called ‘angiomatous’ AVM has a single vessel feeding multiple small interconnecting vessels [1,4]. We present a case of hypertension secondary to congenital AVM managed by superselective embolization and a brief review of the literature.

Case

A 17-year-old male was referred for evaluation of hypertension. One year earlier he was found to be hypertensive upon admission for suspected appendicitis and discharged later with the advise of follow-up in the hypertension clinic. Four months later he presented to the local internist with headaches and a blurring of vision and was admitted for management of severe hypertension (blood pressure 220/150 mmHg). Serum creatinine and other biochemistry parameters were normal along with normal abdominal ultrasound. On presentation at our hospital 8 months later, he still had recurrent headaches but denied blurring of vision. He was an active young man with no history of drug abuse or smoking. Family history for hypertension or renal disease was absent. Five years previously he had suffered a blunt abdominal trauma in a road traffic accident but was discharged 24 h after uneventful observation. His antihypertensives included nifedipine, atenolol, prazosin, and furosemide. He claimed to be compliant with medications except salt intake. On examination he was thin, weighing 47.7 kg. Blood pressures in left and right arms were 155/90 and 150/90 mmHg, respectively. He had a loud second heart sound, no abdominal and flank bruits, and fundus examination revealed AV nicking and silver wiring. Laboratory investigations showed normal biochemistry, urinalysis, 24 h urinary excretion of protein and vanillyl mandelic acid, and serum thyroid function tests. Chest X-ray, ultrasound of abdomen, echocardiogram, and renal captopril scan were unrevealing.

Blood pressure remained uncontrollable even after maximizing the three-drug regimen. A renal angiogram of the right kidney revealed a large lower pole congenital AVM, as suggested by its characteristic numerous feeding vessels, multiple arteriovenous connections and early venous filling (Figure 1). After consultation with the vascular and interventional radiology departments, embolization was decided. Renal vein renin levels were obtained before embolization from both sides as well as from the inferior vena cava. Gianturco metallic coil (Cook, Bloomington, IN, USA) and gel foam was embolized in the main feeding vessel of the AVM (Figure 2). Post-embolization he developed severe hypertension requiring i.v. nitroprusside for 36 h. Renal vein renin (RVR) levels did not lateralize to the right side as the ratio was 1.1 : 1 [ > 1.5 : 1] and the renal/systemic renin index did not show contralateral supression. RVR were not obtained post-embolization. DTPA nuclear renal scan post-embolization showed
decreased perfusion in the lower pole of the embolized kidney (Figure 3). One week later, at the time of discharge, he still required captopril 25 mg and nifedipine-retard 20 mg twice daily and atenolol 100 mg every day.

Three months later he presented with dizziness, his blood pressure was normal but a faint systolic bruit was heard at the right lumbar region that later disappeared spontaneously. Repeat angiogram showed a small persisting AVM. His medication at discharge was only captopril 25 mg twice daily. Four months later a repeat angiogram was carried out for embolization of the persisting AVM; however, the AVM clotted before gel foam could be introduced (Figure 4). At last follow-up 6 months later, blood pressure was normal on 12.5 mg captopril twice daily.

Discussion

Congenital AVM is a rare occurrence with only a little more than 200 reported in the literature; 14-27% of arteriovenous abnormalities are congenital and in a review of Japanese patients around 53% were congenital [6-9].

Our patient had suffered a blunt abdominal trauma, which is a known risk factor for AVF formation [3,11]. However, presence of numerous feeding vessels and multiple arteriovenous interconnections in our patient was suggestive of AVM. These congenital vascular anomalies are quite rare and in 72% of cases present as haematuria due to their location in the calyceal or pelvic submucosa especially with the angiomatous variety [2,4,5,12,13]. Our patient, however, never had haematuria. Other presentations may be systolic or diastolic hypertension in 46-50% and 5% may also present as high output cardiac failure [1,2,6,8,10]. The case described here shows a causal association between the AVM and hypertension, responding to embolization of the AVM. Our patient most likely had renin-dependent hypertension yet we were unable to show the lateralization of RVR levels. Others have similarly shown the lack of lateralization of RVR in AVM/AVF except a few [1,14-16]. This may be the result of large amount of blood flowing through the malformation diluting the renin from the ischaemic parenchyma.

Acquired AVFs on the other hand are mostly asymptomatic. In a recent review of 31 AVFs complicating 1042 native kidney biopsies, 12 (39%) were symptomatic and only four (13%) required intervention [3].

Radiological workup of a patient with suspected secondary hypertension may include Doppler sonography, radionuclear renography, and renal angiogram. Colour-coded Doppler sonography (CCD-S) is operator dependant and has been more successful in patients with superficially located transplanted kidneys [3,12]. AVM was not detected in our patient by CCD-S pre- or post-angiographic localization that may be due to the technical inexperience of our centre. Radionuclear renography may show the presence of a large AVM as an increased concentration in the affected area followed by rapid clearance [17]. The AVM in our patient was probably not large enough to show such increased concentration. Computed tomography and

Fig. 1. Congenital AVM in the lower pole of the right kidney. Venous filling is obvious even before the rest of the kidney is visualized.

Fig. 2. A residual small AVM persisting post-embolization of gian turco coil and gel foam.
magnetic resonance angiography was also recently used successfully to localize the AVF [12,18,19]. Nephrectomies, partial or complete, used to be the treatment of AVM [1–3,11]. Embolization during angiography, however, is the currently preferred initial intervention [2,3,20–23]. Surgery on the other hand, is still only recommended for large AVF/AVMs due to the risk of systemic embolization of the injected material [11,19]. Embolization shows promising results in majority of AVMs and AVFs [3,20–23]. In a recent review by Tarif et al., 91% of the AVFs in native kidney biopsies and 67% of AVFs as a result of allograft renal biopsies were successfully embolized [3]. Different materials have been used for embolization including autologous clots, gel foam, wool coils, PVP, detachable silicone balloons, and more recently metallic coils. The AVM in our patient had a major feeding vessel that was successfully embolized using a gianturco coil and gel foam (Figure 2). Takebayashi et al. [20], in a recent review of 30 cases of congenital AVM presenting with haematuria, reported their results of embolizations using absorbable gelatin sponge, absolute alcohol, and polyvinyl alcohol. Twenty-one of the 30 embolizations (69.3%) had complete occlusion and the remaining eight resulted in partial occlusion; the cessation of haematuria was however noted in all cases. None of their patients had hypertension and the plasma renin activity was normal at baseline. Nevertheless, one patient developed renin-dependent hypertension post-embolization. Our patient had an acute rise in blood pressure post-embolization probably due to the renin release from the infarcted area. In the patients described by Takebayashi, the absence of acute rise of blood pressure even with large areas of infarction may be the result of alcohol use for ablation [20]. It is proposed that use of absolute alcohol leads to complete cellular death and total vascular occlusion, eliminating the risk of recanalization and activation of the renin–angiotensin system [20,21].

Therapeutic embolization leads to an infarcted area that continues to secrete renin for some time resulting in sustained hypertension. In our case it took at least 2–3 months before the response was observed. At least 59% of the patients with congenital AVM and hypertension respond to embolization [1]. Successful outcome is defined as a decrease in systolic pressure of 30 mmHg and diastolic pressure of 10 mmHg on the same amount or less antihypertensive therapy. In our patient, requirement of antihypertensives significantly decreased to only 12.5 mg of captopril twice a day post-embolization indicating a successful response to intervention.
In conclusion, AVM is a rare cause of hypertension and embolization is successful in most of the cases. An acute increase in blood pressure may be observed post-embolization and a beneficial response may be delayed by more than 2 months.

**References**

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