CASE REPORT

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Secondary hypertension in childhood resulting from pelviureteric junction obstruction by crossing renal vessel

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ABSTRACT

We present the case of a six-year-old boy with hypertension related to pelviureteric junction obstruction by crossing renal vessel. He had a history of urinary infection, left hydronephrosis and right vesicoureteral reflux in infancy. At the age of three years, there was spontaneous resolution of the right vesicoureteral reflux. At five years of age systemic hypertension was detected. Ultrasonography revealed an atrophic right kidney and isotope renography showed that the right kidney contributed only 9% of total renal function. Faced with an atrophic, poorly functioning right kidney and systemic hypertension, a right nephroureterectomy was performed. During surgery, pelviureteric junction obstruction by an abnormal vessel was detected. After surgery, blood pressure normalised so nephrectomy was both diagnostic and therapeutic.

Pelviureteric junction obstruction by crossing renal vessel is a rare cause of secondary hypertension. Although most cases of pelviureteric junction obstruction are diagnosed in the prenatal period, it sometimes becomes apparent later, particularly if due to crossing renal vessel. Hypertension is an uncommon manifestation and represents advanced disease.

Key-Words:
Hypertension; pelviureteric junction obstruction by crossing renal vessel.

INTRODUCTION

In childhood, hypertension is defined as blood pressure (systolic and/or diastolic) equal to or above the 95th percentile for gender, age and height, on at least three separate occasions[1-4]. In the paediatric age group, it has a prevalence of 2-5%, and is usually secondary to an underlying organic cause. In 60 to 80% of patients, the cause is renal or renovascular[1,2]. The causes of renal hypertension include sequelae of pyelonephritis, glomerulonephritis, reflux nephropathy, hydronephrosis and urinary tract obstruction[2]. Renovascular hypertension is due to anomalies of the renal artery (most often fibromuscular dysplasia) and renal vein thrombosis[1,2]. Other rare aetiologies include cardiovascular, endocrine and neurological causes, obesity and drug administration[1,2]. As in adults, if a cause cannot be found, it is called primary or essential hypertension[1-3]. In older children and adolescents, essential hypertension is on the increase, frequently in association with obesity[1].

CASE REPORT

We present the case of a six-year-old boy, with nonconsanguinous and healthy parents and no relevant family history. The pregnancy was uncomplicated

and obstetric ultrasonography was normal. There was a normal delivery at term, with no issues during the neonatal period. Growth and psychomotor development were appropriate for age. In terms of personal history, there were two urinary infections (at three and 15 months of age) and chickenpox at 15 months of age. Renal ultrasonography performed after the first urinary infection (three months of age) showed mild to moderate left hydronephrosis with normal cortico-medullary differentiation, and a normal right kidney. Direct radionuclide cystography showed active vesicoureteral reflux (VUR) II-III/IV degree on the right. At ten months of age, isotope renography showed symmetrical function (left kidney 47%, right kidney 53%) with no parenchymal lesions. At 12 months of age, a further renogram showed symmetrical function with no obstructive pattern. At three years of age, there was spontaneous resolution of the right VUR, with mild left kidney hydronephrosis persisting.

At five years of age, the patient suffered a fracture of the radius, and during his hospital stay, had blood pressure values consistently above the 95th percentile for age, gender, and stature. Ambulatory blood pressure monitoring over a 24-hour period (ABPM) showed a high percentage of high systolic and diastolic values (above the 95th percentile) constantly (i.e. during both day and night) (Fig. 1). Renal ultrasonography was performed, which revealed renal asymmetry, with the right kidney being smaller than the left (7 cm *vs*. 8.1 cm), and having irregular contours and multiple areas of reduced renal parenchymal thickness, associated with marked pelvicalyceal dilatation; there was mild pelvicalyceal dilatation on the left. Isotope renography showed functional asymmetry (right kidney contributing only 9% of total function) associated with global parenchymal loss of the right kidney; the left kidney appeared normal (Fig. 2). Captopril renography was not conclusive because of the severe damage to the right kidney; though an obstructive pattern on the left was excluded. Renal Doppler ultrasonography showed normal haemodynamic parameters in the abdominal aorta and renal arteries; renal veins were patent. Direct radionuclide cystography confirmed the absence of VUR. Renal function and plasma renin were normal, but serum aldosterone levels were slightly high (38.9 ng/dl).

Faced with an atrophic right kidney, contributing less than 10% of overall function, and systemic hypertension, a right nephroureterectomy was performed. During surgery pelviureteric junction obstruction (PUJO) by an abnormal vessel was detected, with the renal pelvis under tension and a thin ureter. Pathologic study of the right kidney revealed that the renal cortex was reduced in thickness to 3mm and both calyces and renal pelvis were dilated and ureter with no alterations. Microscopic study showed cortical atrophy with haemorrhage, and chronic interstitial inflammation, but a normal ureter. Because of the distorted anatomy, it was not possible to determine if it was an abnormal artery vessel or vein causing the obstruction. After surgery there was normalisation of blood pressure, confirmed by ABPM performed eight months later.

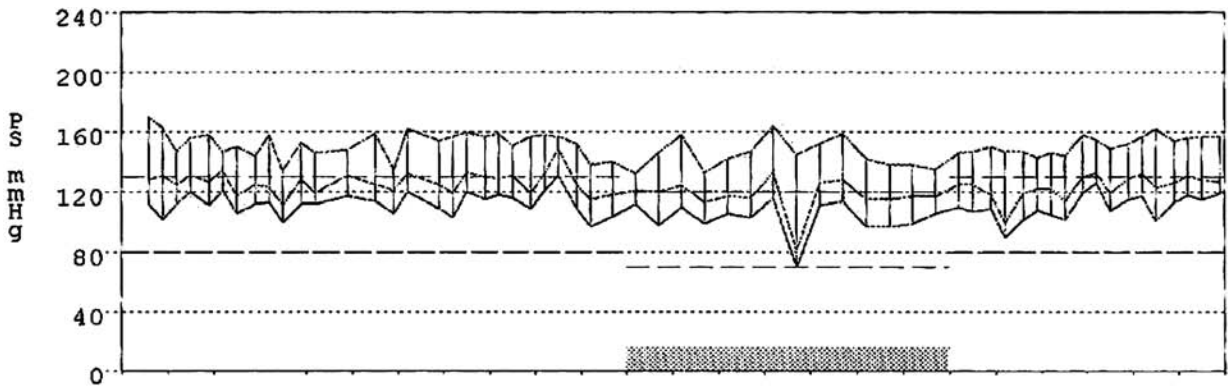

Figure 1

Ambulatory blood pressure monitoring over a 24 hours period (ABPM).

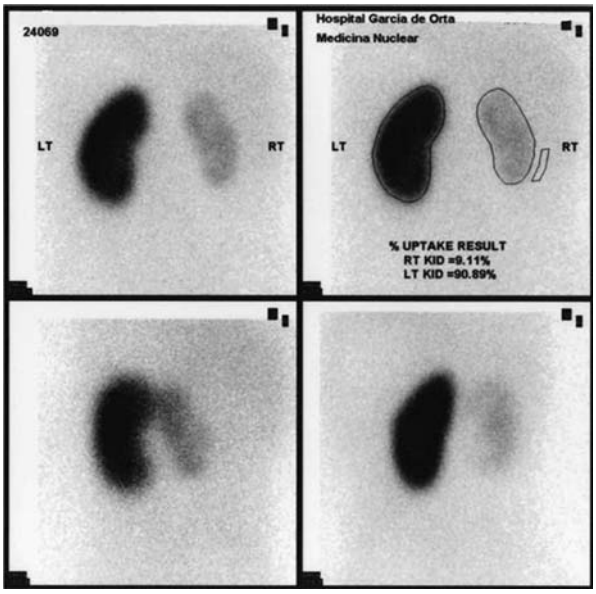

Figure 2
Isotope renography.

DISCUSSION

PUJO is classified as intrinsic or extrinsic, depending on its aetiology. Intrinsic PUJO is caused in the majority of cases by a nonfunctional ureteral segment due to an intrinsic muscular defect; rare causes include ureteral valves and ureteral polyps[5-8].

Extrinsic PUJO, which is less frequent, is usually caused by compression by crossing renal vessel and, rarely, by tumours[5-7]. In up to 90% of cases, PUJO can be diagnosed in the prenatal period by detection of hydronephrosis during obstetric ultrasonography; this more often corresponds to intrinsic PUJO[5,7,8]. More rarely, ureteral obstruction may manifest later, when it is due to crossing renal vessel, ureteral polyp or tumour[5-7].

More than 90% of cases of PUJO by crossing renal vessel are symptomatic[5]. The most common clinic manifestation is renal colic, with abdominal pain, nausea and vomiting, due to the fact that ureteral obstruction is frequently intermittent[5,9]. During symptomatic periods, renal ultrasonography is fundamental to detect acute hydronephrosis, which is normally absent during nonsymptomatic periods[5,9]. It may also present with intermittent haematuria or recurrent urinary tract infections and most rarely with a palpable mass in the flank[5,7-9].

In this type of intermittent obstruction, renal function is usually normal[5]. However, the condition may evolve to a progressive and silent hydronephrosis, with consequent cortical atrophy, and the development of impaired renal function and hypertension[4]. Hypertension can be explained by increased renin production by renal juxtaglomerular cells, caused by kidney destruction by hydronephrosis. Hydronephrosis is caused by extrinsic compression of the urinary tract by the abnormal vessel, which sometimes only becomes apparent after kidney growth and vessel stretching. If the abnormal vessel has an arterial origin and is responsible for the perfusion of a major kidney area, renal hypoperfusion also contributes to juxtaglomerular cell stimulation and hypertension[10,11].

In this particular asymptomatic child, in whom hypertension was accidentally detected, renal/renovascular hypertension is the most probable aetiology. Definitive diagnosis was made during surgery, when an abnormal vessel deemed responsible for the PUJO was visualised. This was a right lumbotomy surgical approach, so it was not possible to compare the renin levels from the right and left renal veins. Although the plasma renin was normal in the peripheral blood, it is expected that the plasma renin level in the right renal vein would have been frankly elevated[9]. Treatment of PUJO caused by crossing real vessel is always surgical; whenever possible, the approach should be a pyeloplasty with urinary tract reconstitution[5,11]. When there is severely reduced function in the affected kidney (usually defined as below 10% of total function) the surgical solution is nephrectomy because the kidney is unlikely to recover after urinary tract reconstruction[5,7].

This case report illustrates a rare form of PUJO caused by crossing renal vessel, and complicated by hypertension. It also highlights the importance of routine blood pressure checking in all children over three years old seen in a medical setting[1-3].

Conflict of interest statement. None declared.

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