

High blood pressure due to aortic coarctation and renal artery stenosis in a teenager with type 1 neurofibromatosis

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SUMMARY

Hypertension affect about 1% of patients with neurofibromatosis type 1 (NF1). Major causes are concomitant pheochromocytoma in adults and renovascular hypertension in children. In most cases, NF1 is associated with renal artery stenosis, smooth cell proliferation and adventitial fibrosis. We describe a 16 year old girl with hypertension complicating NF1 secondary to severe coarctation of abdominal aorta and tight stenosis of right renal artery, a very uncommon case. She was first diagnosed when she was 3-years-old and managed with antihypertensive drugs (atenolol, hidralazine and nifedipine); she experienced progressive uncontrollable hypertension but no symptoms, thus she was admitted to repeat studies. Laboratory evaluation (including creatinine, serum electrolytes, urinalysis, urine catecholamines and creatinine clearance) was normal. Percutaneous transfemoral magnetic resonance angiography disclosed severe coarctation of abdominal aorta, functional occlusion of superior mesenteric artery and tight stenosis of right renal artery with poststenotic dilatation. Patient underwent surgery with aorto-aortic by-pass and right kidney artery reimplantation. Periodical controls confirmed no hypertension, even four years after surgery and normal flow patterns in Doppler ultrasonography. Patients with NF1 must be screened for pheochromocytoma and renovascular hypertension. If hypertension appears, careful management is mandatory, as periodical follow-up even after surgery, since the long-term recurrence rate of renovascular lesions is not well established.

Key words: Neurofibromatosis. Renovascular hypertension. Coarctation. Renal artery stenosis.

RESUMEN

La frecuencia estimada de la hipertensión arterial en pacientes con neurofibromatosis tipo 1 (NF1) es de aproximadamente un 1%, habitualmente secundaria a feocromocitomas y estenosis de las arterias renales. La coartación de aorta asociada en estos pacientes es una causa infrecuente de hipertensión. Presentamos el caso de una paciente con NF1 que padece hipertensión desde los 3 años de vida, mal controlada con tratamiento farmacológico. Antecedentes familiares de NF1 (madre). A la exploración física presentaba múltiples manchas café con leche, efélides axilares y presión arterial por encima de percentil 95 para edad y altura. Las pruebas de laboratorio (creatinina sérica, electrolitos, catecolaminas urinarias y aclaramiento de creatinina) fueron normales. La monitorización ambulatoria de presión arterial reveló hipertensión diurna con valores nocturnos normales. La ecografía Doppler renal mostró un patrón anormal de flujo en arteria renal derecha compatible con estenosis y la angiografía magnética: coartación severa de aorta abdominal, con oclusión funcional de arteria mesentérica superior y estenosis moderada de arteria renal derecha con dilatación postestenótica. Ante estos hallazgos, la paciente fue intervenida quirúrgicamente, con realización de by-pass aorto-aórtico y reimplantación de arteria renal derecha. Durante los cuatro meses siguientes a la intervención recibió tratamiento hipotensor con nifedipino oral que se suspendió ante la buena evolución clínica. Los controles periódicos en la consulta durante los cuatro años siguientes a la cirugía fueron satisfactorios, con presión arterial normal y función renal conservada. **Comentarios:** El control de la presión arterial en pacientes con NF1 es aconsejable para detectar casos de hipertensión arterial que, en muchas ocasiones, va a requerir tratamiento quirúrgico.

Palabras clave: Neurofibromatosis. Hipertensión renovascular. Coartación. Estenosis de arteria renal. Enfermedad renal vascular.

INTRODUCTION

The incidence of high blood pressure in patients with NF1 is around 1%,¹ mainly associated to pheochromocytomas in adulthood. During childhood, most cases are secondary to renal artery stenosis and rarely to coarctation of the abdominal aorta.²

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Figure 1. Arteriography image showing severe coarctation of the abdominal aorta, functional occlusion of the superior mesenteric artery and severe stenosis of the right renal artery with post-stenosis dilatation.

We present a 16-year old female with NF1, who had been controlled since she was 3 year old because of hypertension secondary to abdominal coarctation, which was angiographically confirmed. She had no symptoms but the blood pressure could not be controlled with drugs (atenolol, hydralazine, nifedipine).

On physical exam multiple «café au lait» spots could be seen, as well as freckles in axillary areas, with no cutaneous neurofibromas. A murmur could be heard on right superior quadrant of the abdomen, and peripheral pulses were palpable and symmetrical. Neurological examination was normal. During the follow-up, the blood pressure measurements were between 150/110 and 140/100 mmHg. These values were always over the 95th percentile for sex, age and height.

Further investigations: renal function and urine catecholamines levels were within the normal range. The funduscopic examination revealed grade I hypertensive retinopathy. The echocardiography showed mild left ventricle hypertrophy. The cranial MRI disclosed a glioma involving the optic chiasm and nerve. An ambulatory blood pressure monitoring was performed: During the day high blood pressure was detected with nocturnal *dip*. On renal Doppler ultrasound the renal flow was diminished in the right kidney, which suggested renal artery stenosis. The abdominal angioresonance showed severe coarctation of the abdominal aorta with a lumen reduction of 85% and right renal artery stenosis. The

same could be seen on percutaneous femoral angiography (fig. 1).

With these findings surgical intervention was decided consisting on aorto-aortic bypass and right renal artery reimplantation. A control angiography was performed that confirmed the reestablishment of the circulatory pattern (fig. 2). Pharmacological therapy was required associated to oral nifedipine during four months. Four years after the intervention renal function was normal, Doppler ultrasounds performed during the follow-up showed preserved flows, and the arterial pressure was normal.

The frequency of high blood pressure in patients with NF1 is around 1% and can be due to renal artery stenosis, pheochromocytomas, coarctation of the abdominal aorta, or cerebral tumors.¹ During childhood, the most frequent cause is renal artery stenosis, while in adults it is pheochromocytoma.³ The association between renal artery stenosis and aortic coarctation in patients with neurofibromatosis is exceptional⁴. In fact, there are approximately ten cases reported in the literature.⁵⁻⁷

In spite of the diagnostic advances (ABPM, renal Doppler ultrasound, angio-MRI) conventional angiography still is the main diagnostic investigation, especially in those cases that will be susceptible to surgical therapy. The management of these patients must be individualized, according to the underlying condition and age. The surgical treatment is successful in 80% of the cases. For this reason it is currently the preferred therapeutic option, and also taking into account that in children the results of the angioplasty are not satisfactory.⁸ Anyway, the follow-up of the patients in the intermediate and long term is very important, as we do not know the relapse rate of these lesions.

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