Novel treatment (new drug/intervention; established drug/procedure in new situation)

Hypertension, secondary to a renal artery aneurysm, treated by ex vivo aneurysm repair and autotransplantation

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Summary

Hypertension is becoming a more common problem in childhood and adolescence. About 5–10% of paediatric patients with hypertension have underlying renovascular disease. Although renal artery aneurysms (RAAs) are an uncommon cause of disease of the renal vessels, they are recognised as a cause of hypertension. We describe a 15-year-old man with symptomatic hypertension who after extensive investigation was found to have a right RAA. Our patient responded to conservative management with two antihypertensive agents, but as a young sports enthusiast, he was keen to pursue more definitive options. Complex anatomy of the aneurysm precluded the use of endovascular treatment. Surgical options were explored, and after counselling, our patient underwent a nephrectomy, ex vivo aneurysm repair and autotransplantation with which his hypertension resolved. Our case highlights the difficulty of diagnosing RAAs in hypertensive patients and that, in carefully selected patients, invasive surgical intervention of RAAs is a viable treatment option.

BACKGROUND

Hypertension is becoming more prevalent among paediatric patients.^{1 2} These younger patients are unlikely to experience the more serious sequelae of sustained hypertension until later in life but they can develop end organ damage including left ventricular hypertrophy (LVH), changes in retinal vasculature and subtle cognitive dysfunction.^{1 2} Hypertension in this age group frequently has identifiable causes making thorough investigation essential in these patients.¹ About 5–10% of paediatric patients with hypertension have underlying renovascular disease (RVD).^{3 4} Renal artery aneurysms (RAAs) are a rare cause of disease of the renal vessels and they have been associated with hypertension.⁵ Making a diagnosis of RVD can be difficult and often requires invasive investigations which need careful consideration in the paediatric population. Our patient's case highlights the difficulties in diagnosing RVD in hypertensive patients and the difficult management decisions that ensue in this age group.

CASE PRESENTATION

A 15-year-old man presented to his general practitioner with a history of central chest pain and frontal headaches. He also had a history of mild asthma but otherwise was fit and well and a keen rugby player. Specifically, there was no history of abdominal trauma or recurrent urinary tract infection (UTI) and no family history of collagen disorders, vasculitides or neurocutaneous syndromes. Clinical examination including fundoscopy was unremarkable except for the finding of hypertension with an initial recording of 170/110 mm Hg.

INVESTIGATIONS

Our patient was initially investigated at his local hospital with an ECG which showed evidence of LVH. Echocardiography confirmed LVH but found no cardiac

cause for the hypertension. Other preliminary investigations including renal function, renal ultrasound scan (USS) with dopplers and urinary catecholamine screen were normal.

Our patient was started on treatment, but persistent hypertension led to referral to our paediatric nephrology centre. Antihypertensive treatment was optimised to two agents and subsequent ambulatory blood pressure monitoring (ABPM) demonstrated a good response with a mean systolic blood pressure of 120–130 mm Hg (on the 75th centile for height). We then arranged a dimercaptosuccinic acid (DMSA) scan which demonstrated split right:left function of 63%:37%. The left kidney was slender and had cortical loss at the upper pole. However, our patient's hypertension was felt to be disproportionately severe to be attributed to these findings alone. Furthermore, there was no antecedent history of UTI. This raised the suspicion of fibromuscular disease (FMD) and MR angiography (MRA) was arranged. MRA revealed a 1.5 cm saccular aneurysm at the arterial trifurcation in the right kidney, with associated scarring on that side and normal appearances of the left kidney. Notably, there was no FMD in the main arteries and no renal artery stenosis (RAS). Six months after presentation, formal angiography was arranged to look for further evidence of FMD in the left kidney to try and explain the DMSA findings and to delineate the right lower pole aneurysm further. Angiography demonstrated normal left renal vessels and confirmed a 1.88 cm right-sided aneurysm from which four significant vessels arose (figure 1).

TREATMENT

Our patient had been started on an ACE inhibitor (ACEi) at his local hospital. On initial review by our department, the ACEi was stopped in light of the risk of RAS. The use of beta blockers was precluded by the history of asthma,

Figure 1 Angiography image showing right renal artery aneurysms.

so antihypertensive treatment was gradually altered to amlodipine 10 mg once daily and doxazosin 2 mg once daily. Although he responded well to conservative treatment with antihypertensives, our patient was a contact sports enthusiast and keen to pursue more definitive treatment. Formal angiography was undertaken, as not only did it allow for more accurate imaging of the renal vessels but also consideration for endovascular treatment of the right-sided aneurysm. During the procedure, stent branch occlusion of the aneurysm was abandoned, because an estimated 50% of the right kidney, the dominant side according to DMSA, was supplied by branches of the aneurysm. After much deliberation, a surgical vascular opinion was sought. The patient was counselled at length regarding the risks of surgical repair of the aneurysm and the possibility of persisting hypertension after the operation. Nine months after presentation, our patient underwent a right nephrectomy, ex vivo repair of the aneurysm and autotransplantation into the right iliac fossa. Histological examination of the aneurysm confirmed FMD. Postoperative complications included a wound infection and minor renal pelvis dilatation, which resolved spontaneously.

OUTCOME AND FOLLOW-UP

Twelve months after presentation, our patient's symptoms had resolved. Repeat ABPM and echocardiography demonstrated a mean systolic blood pressure of 107 mm Hg on no antihypertensive treatment and resolution of LVH.

DISCUSSION

The prevalence of primary hypertension is increasing in paediatrics, and the rise in childhood obesity has been hypothesised as a cause for this.² However, it remains essential to identify secondary treatable causes of raised blood pressure like RVD in this age group. RAAs are an uncommon cause of disease of the renal vessels with reports estimating a prevalence of 0.01–1% in the general population though their true prevalence is uncertain.^{5 6} Rarely, RAAs are idiopathic but, more often, an underlying cause can be identified. In adults, atherosclerosis and

FMD are proposed causes of RAAs.⁵ FMD is a non-inflammatory disease of the vasculature, mainly affecting women aged 20–60 years and commonly affecting the carotid and renal vessels.⁷ FMD can occur in childhood and, indeed, there are a few reports linking FMD to RAAs and hypertension in paediatric patients.^{8–12} More commonly, RAAs in childhood are caused by intrinsic collagen disorders (Ehlers Danlos syndrome and Marfan's syndrome), neurofibromatosis, tuberous sclerosis and blunt abdominal trauma.⁵

The underlying mechanism of how RAAs cause hypertension is not well understood, particularly, in the absence of RAS as with our patient. It may relate to thromboemboli from the aneurysmal sac causing areas of renal ischaemia, or external mass effect causing intrarenal kinking of the renal arteries.⁵ Irrespective of what factors drive hypertension in patients with RAAs, they are an important pathology to identify as they are potentially amenable to treatment.

The literature reports that up to 70% of children with RVD are asymptomatic and incidentally found to have hypertension.⁴ Infrequent and inaccurate measurement of blood pressure in children can delay the diagnosis of hypertension being made. Tools like ABPM can be useful not only in demonstrating true hypertension but also delineating its severity.² Inferences on the severity and duration of hypertension can also be made by demonstrating evidence of end organ damage using echocardiography and fundoscopy.

Once the diagnosis of hypertension is established, further investigations are guided by the clinical picture. Suspicion of RVD necessitates the use of imaging modalities. USS with dopplers, DMSA and ^{99m}Tc-mercaptoacetyltriglycine (MAG3) are low risk and relatively non-invasive imaging techniques but a recent review would suggest that these investigations have limited sensitivity in RVD.¹³ CT angiography (CTA) and, particularly, MRA are good tools for delineating RAAs, formal angiography only being necessary when anatomy is complex or endovascular treatment is being considered.⁵ However, the sensitivity of both these modalities in RAS, the more likely pathology when searching for a renovascular cause for hypertension, is still felt to be lacking.¹³ Digital subtraction angiography (DSA) is suggested as the gold standard for investigating RVD.¹³ DSA usually requires an anaesthetic and is invasive, so its use in children and younger patients requires justification. Criteria that increase the likelihood of RVD, necessitating DSA, have been proposed. These include very high blood pressure, symptomatic hypertension, hypertension not controlled on two antihypertensives, raised serum renin or a syndrome with increased risk of vascular disease.¹³

Conservative treatment of RVD can often be unfavourable owing to the serious long-term ramifications of sub-optimal control of hypertension. Other options include using endovascular intervention which has been used successfully to treat adult patients with RVD and is increasingly being used in the paediatric population.^{4 6 13} There is also a large cohort of paediatric patients with RAS, described in the literature, who have undergone numerous different surgical procedures including resection and reimplantation, bypass using veins or arteries as grafts or primary nephrectomy.¹⁴ The surgical management of RAAs in younger patients is less well documented.

In adult practice, expectant treatment of RAAs with surveillance imaging is used with indications for treatment including aneurysms >2 cm in size, dissecting RAA, pregnancy or potential pregnancy.⁵ Observation may not be an appropriate approach to management in younger patients for two reasons. As already stated, these patients are at greater risk of the long-term effects of sustained hypertension, and there can be a significant impact on lifestyle choices because of the risk of aneurysmal rupture. Endovascular treatment can be used to treat RAAs but as with our patient, it can be contraindicated because of the risk to renal parenchyma. In such situations, we propose that invasive surgical intervention with nephrectomy, ex vivo aneurysmal repair and autotransplantation is a viable approach to management. There are reports in the literature supporting this mode of treatment for complex RAAs in adults but very few in children and adolescents.^{15 10} Our report adds to existing literature the case of a 15-year-old man with symptomatic hypertension and end organ damage, caused by a RAA without RAS. Complex anatomy posed diagnostic challenges and necessitated surgical intervention, which ultimately led to complete resolution of his hypertension.

Learning points

- ▶ Although primary hypertension is becoming more prevalent in young patients, investigation to look for underlying causes is always required.
- ▶ Renovascular disease (RVD) is a recognised cause of hypertension in young patients and is important to identify as it is amenable to treatment.
- ▶ In certain patients who have hypertension secondary to RVD, invasive surgical intervention may be considered as a more absolute treatment modality.

Competing interests None.

Patient consent Obtained.

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