The Interesting Case

Phaeochromocytoma associated with reversible renal artery stenosis

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Introduction

Phaeochromocytoma is a catecholamine-secreting tumour. It is very rare in children. The importance of these tumours is out of proportion to their incidence because they may be associated with catastrophic increases in blood pressure, they are difficult to diagnose and treat, but they have potential for complete cure by surgical removal [1]. The cardinal symptom is hypertension, as a result of excessive catecholamine secretion. Occasionally, renovascular hypertension may also occur due to external compression of a renal artery by the tumour mass [2].

We here report a child presenting with sustained hypertension and renal artery stenosis due to external compression by a phaeochromocytoma.

Case

A previously fit 12-year-old boy was admitted with seizures. His past history revealed that he had been complaining of headache, nausea and vomiting for a month. No family history of seizures, hypertension or nephropathy was recorded. His body weight was 30 kg, temperature 36.4°C, and pulse rate 50 beats/min. Blood pressure was 210/180 mmHg, as measured on all four extremities. By auscultation, there were no pathological murmurs over the heart, abdomen or peripheral sites. He was treated with sublingual nifedipine initially and propanolol thereafter. The blood pressure rapidly fell to an average of 160/105 mmHg.

Initial laboratory data were: haematocrit 0.35, white blood cell count 8.2×10⁹/l, sodium 138 mmol/l, potassium 4.1 mmol/l, calcium 2.4 mmol/l, phosphorus 1.3 mmol/l, glucose 7 mmol/l, blood urea 4.5 mmol/l, creatinine 25 μmol/l, SR 70/30, bilirubin 18 mmol/l (direct fraction 10 mmol/l), SGOT 48 U/l, SGPT 36 U/l, C3 1.2 mg/l, and C4 0.7 mg/l. Urinalysis failed to reveal proteinuria, haematuria, leukocyturia and casts. Diuresis was 1200 ml/24 h. Urine culture was negative. Electroencephalography showed generalized teta-delta dysrhythmia. Brain computerized tomography (CT) was normal. Echocardiography excluded aortic coarctation.

On abdominal ultrasonography the kidneys and other visceral organs appeared normal. PRA was not determined. Urinary excretion of 17-hydroxycorticosteroids was 31.6 μmol/24 h and of 17-ketosteroids 11.4 μmol/24 h (normal). Urinary excretion of vanilmandelic acid was 39 nmol/24 h (normal <38 nmol/24 h) and that of catecholamines was 1170 μg/24 h at a first examination and 90 μg/24 h at a second sampling (normal <105 μg/24 h).

131I-metaiodobenzylguanadine (MIBG) scan did not show radionuclide accumulation in the suprarenal region, or in neighbouring paraortal ganglia, abdomen and thoracic cavity. A 99m-technetium dimercapto-succinic acid (DMSA) scan showed a small left kidney, accounting for approximately 27% of total uptake. This finding is usually associated with vesico-ureteric reflux and reflux nephropathy in children, however, voiding urothrocystography (VCUG) was normal and intravenous pyelography revealed a slight reduction in contrast media concentration in the left kidney, with a normal aspect of the excretory tract.

Renal arteriography revealed two arteries on the left side. The superior artery showed a 90% stenosis, 1 cm in length, and a microaneurysmatic poststenotic dilatation (Figure 1). Taking into account the high diagnostic value of extremely elevated urinary catecholamines, the possibility of a catecholamine-secreting tumour causing renal artery compression was given strong consideration. Repeated abdominal Doppler ultrasound detected a left adrenal tumour mass and CT of the abdomen showed a 5×4×3 cm mass thought to arise from the left adrenal gland (Figure 2).

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During surgery, the left superior branch of the renal artery was found to be draped around a renal hilar mass arising from the adrenal gland. The tumour and the adrenal gland were resected, and light microscopy examination confirmed phaeochromocytoma. Postoperatively, the patient had the clinical diagnosis of mild hypertension, which resolved within a few days.

A control DMSA scan performed 6 months later showed a right kidney function of 57% and a left kidney function of 43%. The child has been followed subsequently for 6 years, with no recurrence at present of hypertension or tumour.

Discussion

Severe sustained hypertension occurs in only 0.1% of the paediatric population and only about 2% of these patients will have an underlying endocrine cause [2]. Phaeochromocytoma is a catecholamine-secreting tumour, which is exceedingly rare in children. Extra-adrenal tumours or paragangliomas are more commonly seen in children than in adults, and the renal hilum is the most common ectopic location [2].

The coexistence of phaeochromocytoma and renal artery stenosis is a very rare occurrence. In total, 87 cases have been reported in the literature of whom 48 (52.2%) had been diagnosed preoperatively [3]. Among the 87 reported cases, phaeochromocytomas were of extra-adrenal location in 40 (46%), of which 31 were in the renal hilum, as illustrated in our patient. In most cases of renal artery stenosis caused by phaeochromocytoma, the tumour was benign. This was also the case in the present patient. Most of the patients with renal artery compression were adults, with only about 14% occurring in children aged 5 years or more [3,4].

When renal artery stenosis is associated with phaeochromocytoma, the tumour may compress the renal artery; however, in other instances, it may only be near to, or in contact with, a stenosed artery without compressing it. How a phaeochromocytoma adjacent to a renal artery, without compressing it, can cause stenosis is not clear. This association suggests a local effect of the tumour. Chronic spasm may arise from local diffusion of catecholamines [5], and eventually this can lead to fibrosis. Another possibility is that fibrous bands emanating from the tumour may narrow the renal artery, thereby causing hypertension [6].

Correct preoperative diagnosis is highly desirable to minimize the risk of surgery and anaesthesia. The investigative protocol for identifying phaeochromocytoma and coexistent renovascular disease as a cause of hypertension in a child should follow the general strategy of evaluating secondary causes of hypertension [2]. The evaluation is centre dependent because of the varying availability of diagnostic resources and variable physician preferences. Measurement of plasma levels of renin and 24-h excretion rates of urinary catecholamines or their metabolites should be considered on a case-by-case basis [2]. Renal imaging studies are designed to delineate renal parenchymal disease and/or renovascular disease. Such studies include ultrasound, radionuclide scintigraphy, Doppler flow study, CT scan, magnetic resonance imaging (MRI), and/or arteriography. Other imaging or isotopic studies are directed toward endocrine evaluation including adrenal ultrasonography, [131I]- or [123I]MIBG scans, and CT or MRI scans. The diagnostic value of various radiological/radionuclide procedures in identifying phaeochromocytoma and renal artery stenosis respectively is high, although not 100% [3]. However, in clinical practice, an unpredictable coincidence of two apparently different diseases is always possible. To avoid potential diagnostic and therapeutic pitfalls, a careful differential diagnosis is mandatory in each individual case. A high degree of suspicion is required. Thus, the underlying phaeochromocytoma in our case might have been missed based on the preoperative angiography alone [7].
In the present case, there was a certain diagnostic confusion at the beginning, arising from the false negative findings of the initial ultrasound and MIBG scan. However, other imaging studies performed thereafter (DMSA scan showing a small hypofunctioning left kidney and i.v. pyelography showing a diminished concentration phase and normal pelvicaliceal system) suggested that renal blood flow might be impaired. Selective renal arteriography justified this consideration, detecting renal artery stenosis. Fibromuscular hyperplasia is the most common isolated cause of renal artery stenosis in children and it is usually treated with luminal angioplasty. In fact, we did plan angioplasty and aborted this idea in the very last moment when the suspicion of a concomitant tumour compressing the renal artery arose and CT could confirm this. Had the pheochromocytoma remained unsuspected preoperatively, the unprepared patient would have been subjected to the well-known hazards of uncontrolled intraoperative catecholamine release. The risk of major cardiac complications and perioperative mortality in such cases is high [3].

Opinions differ as to whether a coexistent renal artery stenosis should be corrected [3,8,9], as a number of cases of spontaneous remission of such a stenosis have been documented after removal of the pheochromocytoma [10,11]. The surgical removal of the tumour in our patient resulted in rapid normalization of blood pressure and in the subsequent recovery of left kidney function, as shown by DMSA scan (from 27% to 43%), reflecting restitution of normal renal blood flow. Renal vein renin levels may help to decide if renal artery stenosis should be corrected [12]. However, it is up to the surgeon’s expertise during the operation to choose the best approach in each individual case. When the degree of renal artery stenosis is uncertain, intraoperative Doppler flow measurement may be helpful [3]. Missing a functionally significant lesion may lead to persistent hypertension postoperatively [3].

In summary, we report a rare case of pheochromocytoma with coexisting renal artery stenosis. A high degree of suspicion and awareness of the clinical spectrum of pheochromocytoma are necessary for both entities to be diagnosed preoperatively. When an adrenal tumour mass is detected by imaging techniques, the possibility of a coexistent renal artery stenosis should be considered and, vice versa, when renal artery stenosis is diagnosed first by arteriography, external compression should be considered. Spontaneous remission of the stenosis after removal of the pheochromocytoma is possible, without the need for renal artery surgery.

References

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