HYPERTENSION AND UNILATERAL RENAL ISCHAEMIA CAUSED BY COARCTATION OF THE ABDOMINAL AORTA

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In 1934 Goldblatt and his colleagues established that reduction of the renal blood flow in the experimental animal could produce reversible arterial hypertension. Recently some cases of this condition in man have been shown to be due to a lesion of the renal artery, and a normal blood pressure has been restored by treatment of this lesion.[1-3]

It is therefore important that a diligent search should be made for causes of hypertension that can be cured by surgery. Such secondary causes include:

1. Occlusive diseases of the renal artery.
2. Coarctation of the aorta.
3. Tumours of the adrenal medulla.
4. Tumours of the adrenal cortex.
5. Unilateral disease of the renal parenchyma (e.g. atrophic pyelonephritis, hydronephrosis, tuberculosis, polycystic disease).

PATHOLOGY OF VARIOUS OCCLUSIVE DISEASES OF THE RENAL ARTERY

Atheroma. The most common type of lesion affecting the renal artery is an atheromatous plaque, usually close to the origin of the vessel, producing stenosis or occlusion. This is generally a manifestation of widespread atheromatous disease elsewhere in the body.

Embolism. When this produces partial infarction, hypertension may result. An originating focus is usually suspected, rheumatic heart disease being the commonest.

Fibromuscular medial hyperplasia. The cause of this lesion is obscure; it is primarily a disease of young adults and may be congenital. Angiography shows 'beading' of the artery. It is a recently described condition, characterized by irregular areas of fibromuscular hyperplasia of the arterial wall.

Aneurysms. These may occur rarely as primary lesions in the renal artery and their partial thrombosis may cause renal ischaemia; they may also develop secondarily to atheromatous stenosis. Calcification may occur.

Stenosis. This may be due to congenital malformations or other rare causes. Post-stenotic dilatation may occur. Most cases probably represent localized areas of the fibromuscular hyperplasia described above.

Pathological Changes in Relation to Choice of Treatment

The kidney on the affected side may be protected from hypertensive damage because the pressure distal to the stenosis is low. It will eventually undergo secondary fibrosis and atrophy itself when the renal ischaemia is extreme. In fact the affected kidney may even be the better of the two, the contralateral kidney taking the full brunt of the hypertensive damage. Careful appraisal will therefore dictate the best form of surgery, either vascular with conservation of the affected kidney, or nephrectomy.

A number of operative procedures are available, the type of operation depending on the local condition and the experience of the surgeon. Direct surgery to replace or bypass the obstructed renal artery is the procedure of choice, since it conserves a kidney which may be the better one when normal blood flow is restored. Operative procedures that have been successfully performed include: endarterectomy of the renal artery, end-to-end reconstruction, spleno-renal anastomosis, reimplantation of renal artery to aorta, and arterial homografts.

Nephrectomy is the procedure of choice in patients whose general condition is poor, or if a severely damaged or atrophied kidney exists. To decide on nephrectomy when renal tissue is still contributing some excretory function, must be considered bad practice. Nephrectomy is also indicated when an attempted conservative operation has failed.

CRITERIA FOR SUSPECTING RENAL-ARTERY LESIONS IN HYPERTENSIVE SUBJECTS

Patients with renal-artery lesions may present with any of the clinical manifestations of arterial hypertension, and there may be no distinguishing features in the history to suggest renal ischaemia. Helpful clues, however, in selecting patients for investigation, are:

1. The usual absence of a family history, such as is commonly present in essential hypertension.
2. Hypertension beginning under the age of 20 or over the age of 50.
3. Sudden onset of severe hypertension.
4. Sudden increase in severity of existing hypertension.
5. Symptoms or signs of other vascular occlusions (coronary, cerebral or peripheral).
6. An attack of flank pain followed by hypertension (renal infarct).

Patients over 30 but under 65 years with mild hypertension and minimal elevation in the diastolic blood pressure should probably not be investigated.[8] In this group we must use the previously mentioned clues together

with clinical judgement, in deciding whether to launch out on the somewhat costly investigations and hazardous surgical intervention.

PHYSICAL EXAMINATION

In addition to the normal examination, auscultation of the abdomen should be performed. This may reveal a systolic murmur audible along the course of the renal artery. It is best heard at a point one-and-a-half inches above and to the right or left of the umbilicus, with the stethoscope pressed firmly against the abdominal wall.[4] A systolic bruit over the renal areas may be present independently.

SPECIAL INVESTIGATIONS

Intravenous Pyelogram

All patients with hypertension should have an intravenous pyelogram.[5] A careful study of the films is important, since this may show a *difference in size* between the kidneys — even 1 cm. suggesting the possibility of diminished blood supply on the smaller side.

Renal function, as judged by urography, may be reduced or even absent. Certain patients may hyperconcentrate urine on the affected side, with *increased* concentration of dye, giving an erroneous impression of an impaired kidney on the side that is actually normal.

Any other unilateral lesion may be discovered.

Divided Renal-function Tests

Evaluating any disparity may point to the poorer functioning of 2 kidneys, or to the involved kidney in unilateral renal disease. In essential hypertension, impairment of function tends to be equal and parallel in the 2 kidneys. A sensitive index of reduced renal blood flow is the depression of the sodium *and* water excretion by the affected ischaemic kidney. This is the basis of the Howard test,[6] in which simultaneous measurements are made of ureteric catheter specimens from 2 kidneys. It has been postulated that these changes are due to the depression of the glomerular filtration rate with maintenance of relatively normal tubular function, so that a greater proportion of the glomerular filtrate is reabsorbed. Reduction of the urine volume should be 50% or more, and the reduction of the sodium concentration at least 15% on the affected side.

In unilateral *parenchymal* disease, however, the urinary volume is decreased while the level of sodium remains the same or shows a slight elevation on the affected side.

One technical difficulty invalidating the Howard test is the leakage of urine around the ureteric catheters. Rapoport[9] found that on the affected side the creatinine concentration of the urine is higher than on the normal side. Thus the urine-to-plasma ratio of creatinine is higher, and that of sodium lower, on the affected side; and the difference between the 2 sides can be emphasized by dividing the one ratio by the other. The tubular-rejection-fraction ratio (TRFR) thus obtained is expressed as

$$\text{TRFR} = \frac{\text{Urinary sodium concentration (left)}}{\text{Urinary creatinine concentration (left)}} \times \frac{\text{Urinary creatinine concentration (right)}}{\text{Urinary sodium concentration (right)}}$$

and its calculation does not involve estimations of plasma concentration or an accurate knowledge of the urinary volumes. The range for the TRFR in hypertension from causes other than unilateral partial renal artery obstruction is 0·62 - 1·62. With obstruction on the left side, the TRFR is below the lower limit; with obstruction on the right side the TRFR is above the upper limit. Unilateral pyelonephritis falls within the normal range. The TRFR does not reveal obstruction to a branch of the renal artery even if this is causing hypertension, nor does it reveal bilateral and equal occlusion.

The TRFR is thus a simpler technical investigation and actually forecasts the side of the renal-artery occlusion, so that aortography should only be undertaken when the TRFR is abnormal. Since renal-artery disease is responsible for 5 - 10% of all cases of severe hypertension, it may be necessary to determine the TRFR in every case where the cause is obscure.[10]

Arteriography

One is naturally reluctant to submit large numbers of patients with essential hypertension to such a major investigation, but greater use of arterial auscultation and the selective criteria may prove rewarding. It should certainly be done in a patient with any suggestive feature in the history or preliminary investigations. It may have to be done as a final measure in suspicious cases in order to visualize the arterial lesion.

A translumbar or percutaneous femoral approach may be employed to demonstrate arterial lesions radiologically. The nephrogram obtained may confirm the difference in size between the kidneys, or areas of infarction may be seen. Radiographic findings should be correlated with the functional tests. Aortography is capable of producing severe renal arteriolar spasm[7] with overall depression of renal function and a high filtration fraction. These findings may persist for as long as 3 days. Therefore differential renal clearance studies should be performed *before* aortography. Operation should not be undertaken following a radiological indication alone.[4]

REPORT OF A CASE

The following case is reported and bears reference to many of the features commented on above.

S.S., a female aged 25, was first seen on 12 November 1960, complaining of headaches for the past 5 years. These had been fairly severe, generalized and throbbing in nature. About 1 year before this she had had an attack of left loin pain requiring 3 or 4 injections for relief.

During a routine physical examination by her doctor 4 weeks before being seen, her blood pressure was found to be 200/140 mm.Hg. On other occasions it was 220/160 and 160/100 mm.Hg.

Intravenous pyelography at that time showed a non-functioning left kidney. The right kidney functioned normally and appeared hypertrophied. (Fig. 1).

Chest X-ray showed left ventricular hypertrophy, but clear lung fields.

Blood investigations: Blood urea — 22 mg. per 100 ml., blood cholesterol — 187 mg. per 100 ml., haemoglobin — 85%, with a mild iron-deficiency anaemia.

Physical examination. Apart from a blood pressure of 180/100 mm.Hg, no significant abnormalities were encountered. Fundoscopy was not performed. All peripheral pulses were present and normal.

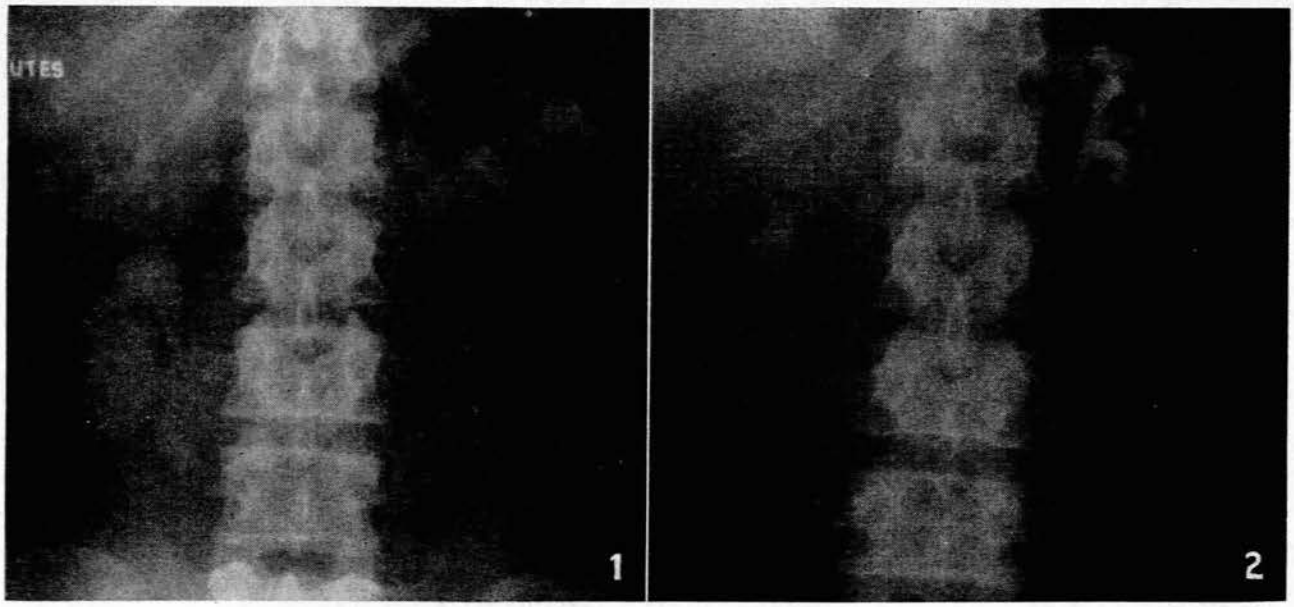

Fig. 1. Intravenous pyelogram showing non-function on the left side, with normal function on the right side and parenchymal hypertrophy of the right kidney.
Fig. 2. Retrograde pyelogram showing diminutive, atrophic renal architecture on the left side with reduced renal outline.

Cystoscopy and Retrograde Pyelogram (14 November)

Cystoscopy. The bladder was normal. There was a clear efflux from the right ureteric orifice, but none was observed from a normal left ureteric orifice. Catheters passed freely to both renal pelves. Urine dripped from the right side, but there was virtually no excretion on the left.

Pyelography showed a normal right upper urinary tract. The left renal outline was reduced in size. The left renal architecture was diminutive and mildly overdistended with dye (Fig. 2), indicating a markedly atrophic kidney.

Urinalysis and Divided Renal-function Test

Bladder urine. Moderate cloud of albumin. Many squamous and epithelial cells, finely granular and hyaline casts; no growth on culture.

Right kidney urine. Insufficient for albumin estimation. Isolated pus and red cells. Casts not observed.

Left kidney urine. Since no urinary excretion occurred from this side, the Howard test could not be performed.

Arteriography (17 November)

The examination was done through a Seldinger catheter

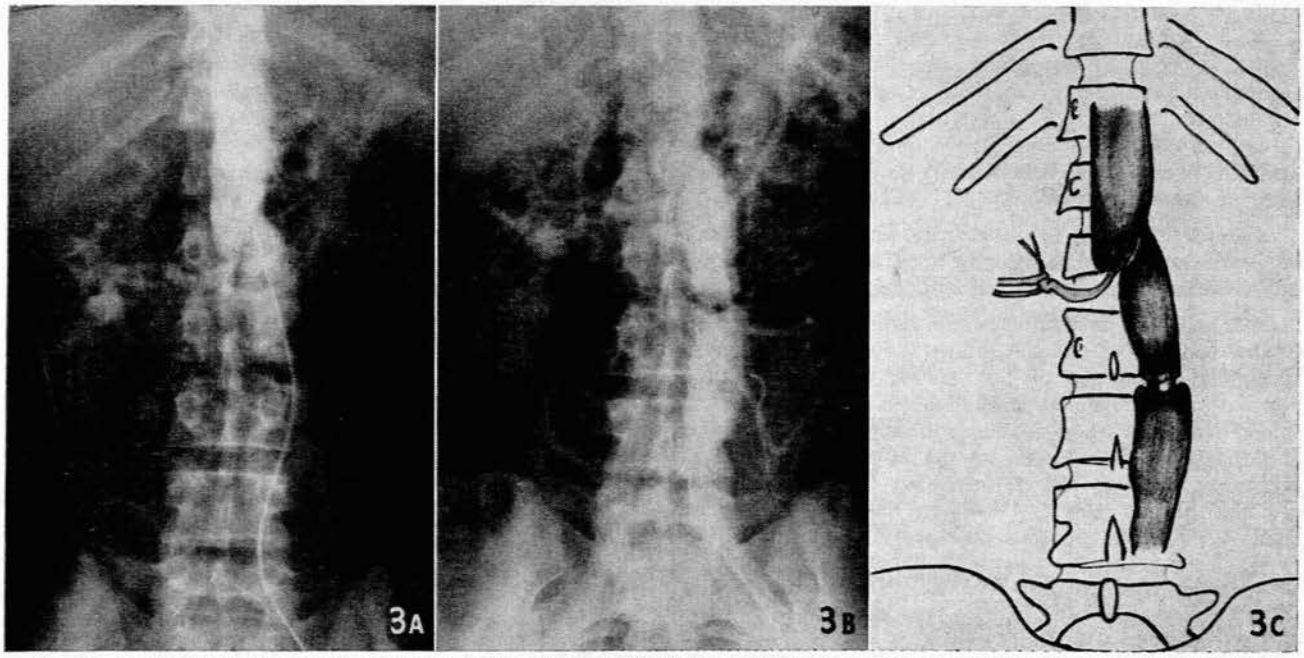

Fig. 3A. Arteriogram showing primary coarctation of the abdominal aorta at the level of the renal arteries, sloping obliquely down from left to right. The left renal artery is occluded at its origin and fails to fill with dye. The right renal artery has a normal take-off from the aorta just proximal to the lowest level of the stenosis.
Fig. 3B. Arteriogram showing a second further acute narrowing of the abdominal aorta at the L2/3 disc-space level.
Fig. 3C. A composite diagrammatic representation of 3A and 3B.

inserted into the left femoral artery. This showed absence of filling of the left renal artery and non-function of the left kidney.

There was a double coarctation, with marked twisting, involving the abdominal aorta. The primary coarctation was oblique and at the level of the renal arteries, sloping down from left to right. The right renal artery had a normal take-off from the aorta just proximal to the lowest level of the stenosis, but the left renal artery appeared to be incorporated in the uppermost level of this oblique stenosis (Fig. 3A).

Apart from the first-mentioned site of coarctation, there appeared to be a further acute narrowing of the aorta at the L2/3 disc space (Fig. 3B).

The right renal artery and its ramifications were normal.

Choice of Treatment

Since the left kidney was obviously non-functioning and atrophied, it was decided to perform a nephrectomy.

Operation (28 November)

A left loin incision was made. The kidney was found to be very small and atrophic. The renal pedicle consisted of small string-like vessels, thin-walled and atrophic. Nephrectomy was performed. Palpation of the abdominal aorta thereafter clearly revealed a constriction at the level of the renal vessels, with a second constriction about 2½ inches distally, confirming the aortography findings. The aorta between and below these constrictions was not aneurysmal.

Histology of the Left Kidney

Small kidney, weight 25 G., with normal configuration. The capsule stripped easily, leaving a slightly granular surface. 'Sections of the kidney show marked fibrosis of 90% of the glomeruli. Surviving glomeruli are normal. There is minimal fibrinoid necrosis in the fibrosed glomeruli. Most of the tubules are atrophied, remaining tubules are dilated. The arteries and arterioles show marked intimal proliferation and, in places, reduplication of the internal elastic lamella. There is mild stromal round-cell infiltration. Picture of advanced nephrosclerosis' (Fig. 4).

Progress

The patient made an uninterrupted recovery and was discharged 10 days later. From the day of her operation her blood pressure reverted to normal and has remained so one year later (120/88 mm.Hg). There is no clinical impairment of the circulation below the level of the coarctation.

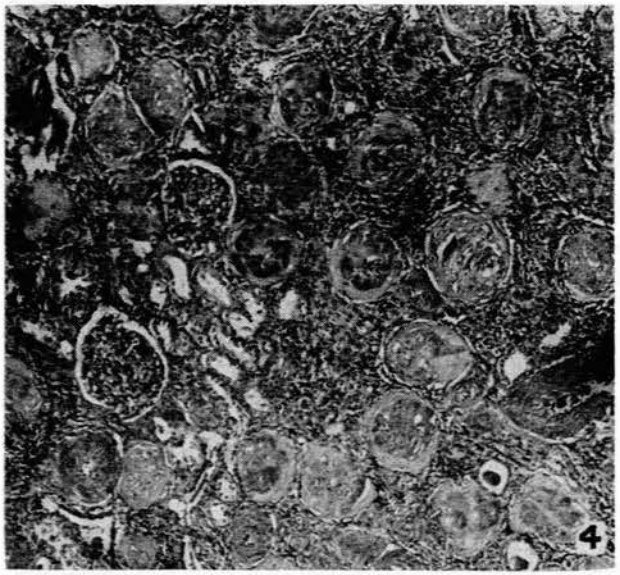

Fig. 4. Section of kidney showing glomerular fibrosis and 2 normal surviving glomeruli. The arterioles show intimal thickening with narrowing of the lumen. Tubular atrophy is apparent; a few surviving tubules show hypertrophy (× 70).

SUMMARY

Criteria for suspecting renal-artery lesions as a cause of secondary hypertension are presented. Since these lesions may be corrected by appropriate surgery and the hypertension abolished, investigations should be conducted in selected cases, and these are outlined.

The types of corrective procedures available are briefly referred to, with emphasis on conservation of renal tissue when possible.

A case of hypertension and unilateral renal ischaemia is described, caused by a double coarctation of the abdominal aorta, the primary coarctation occluding the origin of the left renal artery. Nephrectomy was carried out and normotension was still present one year later.

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