This case report presents a neglected syndrome of precordial pain. JAMA 1955; 159: 1364-5.

**Summary**

An 11-year-old girl presented with acute renal failure and severe hypertension. The blood film showed thrombocytopenia, numerous fragmented red blood cells, and a reticulocyte count of 10%. An intravenous pyelogram showed a small contracted left kidney, and plasma renin activity was increased in the left renal vein. Treatment with minoxidil and propranolol controlled the hypertension. After nephrectomy the hypertension resolved. Light microscopic examination of the left kidney showed a segmental renal hypoplasia. Malignant arterial hypertension can provoke a syndrome of haemolysis and uraemia in children. Aggressive lowering of blood pressure leads to an improvement in renal function.

**Severe hypertension in haemolytic uraemic syndrome**

Severe hypertension in haemolytic uraemic syndrome is common, and is often a complication of the renal disease. However, in adults, haemolysis and uraemia are present in some cases of malignant hypertension. This case report shows that this can be so in children too.

**Case report**

An 11-year-old girl presented with a history of polyuria, headaches, nausea, and vomiting for one year. Blood pressure (BP) had not been measured. Gross haematuria, pallor, and purpura had developed 3 days before admission. The day she was admitted BP was 250/130 mmHg. The liver was enlarged and there was bilateral oedema on the legs. Haematocrit was 23%, haemoglobin 5 g/dl, white blood count $12.9 \times 10^9$/l, platelets $10.0 \times 10^9$/l. The blood film showed numerous fragmented red blood cells, and the reticulocyte count was 10%. Urine analysis showed protein (+ + +) and numerous red blood cells. Other values were as follows: blood urea 60 mg/100 ml (9.9 mmol/l) on admission rising to 185 mg/100 ml (31 mmol/l). Plasma creatinine level was 3 mg/100 ml (265 µmol/l), fibrinogen degradation products were between 25 and 40 mg/ml. The C3 fraction of complement was 135 mg/100 ml and the C4 was 49 mg/100 ml. Plasma renin activity, measured after the administration of 40 mg furosemide and 20 mg hydralazine, was found to be 708 ng/ml per hour. The normal value at this age, in the supine position and on a normal diet, is 3 ± 2 ng/ml per hour in our laboratory. During the next 3 weeks the child received 80 mg hydralazine (4 mg/kg) and 200 mg acetobutolone (10 mg/kg) each day, and intravenous heparin at a dose of 5 mg/kg a day, for 7 days only. After this treatment BP, blood urea, reticulocyte count, and platelet count returned to normal. However, after a few days the blood pressure again rose to 160/130 mmHg, despite continuous antihypertensive treatment. One intravenous injection of 200 mg diazoxide was given every day for 8 days. Treatment with minoxidil was started at a dose of 1 mg/kg a day with propranolol (40 mg daily) and furosemide (40 mg daily). After 3 days the BP had fallen to a normal level. The treatment was maintained at the same dose for 3 months. An intravenous pyelogram showed a small, contracted left kidney. The cystourethrogram was normal, without evidence of reflux. Left renal arteriography showed no evidence of stenosis, but...
there was poor filling of vessels in the lower pole of
the kidney. During treatment with hydralazine and
acetobutolone, venous samples from right and left
renal veins and from the inferior vena cava, below
and above the renal veins, were taken. The plasma
renin activity of venous blood from the inferior vena
cava, below and above the renal veins was 4·8 and
7·4 ng/ml per hour. In the left renal vein plasma
renin activity was 14·6 ng/ml per hour, while it
was 4·6 ng/ml per hour in the right. A right kidney
biopsy was performed. On light microscopical
examination focal and segmental glomerulosclerosis
without pronounced vessel lesions could be seen.
Because of the high level of plasma renin activity in
the left renal vein, a left nephrectomy was performed.
A small contracted kidney weighing 20 g and
measuring $6 \times 2 \times 1$ cm was removed. Light
microscopical examination showed segmental areas
of severe cortical and medullary abnormality. The
affected segments contained a few sclerosed glom-
eruli, many atrophic and dilated tubules, and
thickened blood vessels (Figure). After nephrectomy
the hypertension resolved and the plasma renin level
fell to 1 ng/ml per hour.

Discussion

The association between segmental renal hypoplasia
(Ask-Upmark kidney) and hypertension has been
known for many years. The pathogenesis of the
hypertension in this disease is not perfectly under-
stood. An abnormal renin secretion has been
suggested in the course of this disease. Although
the measurements were made after treatment had
started in our patient, the renin level was three times
the normal rate on the side of the diseased kidney.

After nephrectomy the hypertension resolved and
plasma renin activity fell to below the normal value.
However, as is usual in the hypoplastic segment,
no juxtaglomerular cells could be seen, thus raising
the question of the origin of the secreted renin.

Malignant arterial hypertension can provoke a
syndrome of haemolysis and uraemia in adults, but
this has not been reported in children. It has been
suggested that an increased BP would result in
accumulation of fibrinogen and in the initiation of
coaulation in the wall of the arteries. The fibrinoid
vascular lesion might induce mechanical damage of
the red blood cells and provoke a microangiopathic
haemolytic anaemia. Furthermore, reversible acute
renal failure can be the consequence of malignant
hypertension. Aggressive lowering of BP commonly
leads to an improvement in renal function.

Minoxidil is a new vasodilator, and studies show that
it is more effective than hydralazine. In this patient
minoxidil combined with propanolol controlled
hypertension. There was sodium and water retention
and severe hypertrichosis. In a child with unilateral
kidney disease, nephrectomy is undoubtedly the
treatment of choice.

Conclusion

Renal segmental hypoplasia may be the cause of
increased hypertension associated with haemolytic
uraemic syndrome. Although treatment with
minoxidil was effective for this type of hypertension,
nephrectomy was performed because of side effects.

References

1. Linton A L, Gavras H, Gleade R I. Microangiopathic
haemolytic anaemia and the pathogenesis of malignant
2. Godard G, Valloton M B, Broyer M. Plasma renin
activity in segmental hypoplasia of the kidneys with
3. Chester E M, Agamanolis D P, Banker B Q, Victor M.
Hypertensive encephalopathy. A clinicopathologic study
5. Eknoyan G, Siegel M B. Recovery from anuria due to
6. Mandani B H, Lim V S, Mahurkar S D. Recovery from
prolonged renal failure in patients with accelerated
Minoxidil treatment of malignant hypertension. Recovery

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