RENAL VEIN THROMBOSIS IN INFANTS*

BY

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Renal vein thrombosis in children is an uncommon but not a rare condition that carries a grave prognosis unless recognized early and properly treated. Over 300 cases of various ages have been reported. About two-thirds of these occur in the paediatric age-group with a majority seen in the neonatal period. In young infants, renal vein thrombosis is often accompanied by haemorrhagic infarction of the involved kidney (Williams, 1958).

Renal vein thrombosis has been diagnosed in this clinic in 12 patients in the past six years. Half of the cases were unilateral and half bilateral; 10 of the 12 were under 3 months of age. Only one patient with unilateral thrombosis and one with bilateral involvement survived. In as much as these were the only patients surgically treated in each group it seemed worth while to record our experience in these two cases.

Pathogenesis

Though the pathogenesis of renal vein thrombosis in infants is still obscure, severe gastro-intestinal symptoms are often noted as initial or accompanying symptoms; less frequently, the condition develops suddenly in a previously healthy infant. In older children, renal vein thrombosis may develop in a clinical history dominated by severe illness with toxæmia and sepsis. In the latter variety, the diagnosis is often made only at necropsy with the pathologist finding signs of incipient renal vein thrombosis which is attributed to marasmus. These two distinct clinical entities led Sandblom (1948) to separate a primary 'idiopathic' from the more common secondary variety.

From the time of its original description in an adult patient 125 years ago until 1932 when Grüneberg reported a nephrectomy for an infarcted kidney in a 3-month-old infant, renal vein thrombosis remained a post-mortem diagnosis. The successful outcome of this case resulted in an increased clinical interest in a condition previously considered to be

hopeless. It soon became evident that the acute unilateral form of renal vein thrombosis lent itself to successful surgical management, and that early recognition was an important part of effective treatment (Campbell and Matthews, 1942). Several reports (Clatworthy, Dickens, and McClave, 1953; Kaplan, Straus, Grumbach, Dubois, Blondel, and Drapeau, 1958; Milburn, 1952; de la Torre, Villalpando, Esparza, and Olarte, 1958; Tveterås and Rudström, 1956) of children treated for renal vein thrombosis have been published, chiefly as scattered case reports. The youngest successful nephrectomy for unilateral renal vein thrombosis was in a 4-day-old infant (Miller and Benjamin, 1962).

Case Reports

Case 1. A white male infant weighing 2,150 g. was born prematurely on August 17, 1959 to an 18-year-old toxæmic primipara. Urine passed on the first day was brick red in colour, and tetany-like tremors of the upper extremities were observed. General condition was considered only fair. A large, smooth globular mass was felt in the right upper quadrant. Serum electrolytes, including calcium, were normal. Blood urea nitrogen (BUN) was 45 mg./100 ml., a moderate leucocytosis existed and urinalysis showed haematuria. Blood culture was negative. Intravenous urogram showed a normal functioning left kidney but no visualization of the right. Pre-operative diagnosis was polycystic kidney.

At operation on August 24, the right kidney was three times normal size, and red-brown in colour as was the right adrenal. The left kidney was of normal size and consistency. The right kidney and adrenal were removed, the right renal vein being filled with a thrombus that involved the entire inferior vena cava below the renal vein. The suprarenal segment of the inferior vena cava was free of thrombosis and dilated. No attempt was made to remove the thrombus from the vena cava and no ligation of the vena cava was carried out.

After operation the child was treated with heparin. Histological sections revealed renal vein thrombosis and infarction of the right kidney and autolysis of the right adrenal. Except for a minor wound infection, the child made a satisfactory recovery with no oedema of the lower extremities, and no haematuria. BUN came down to 21 mg./100 ml. He left hospital on September 17, 1959,

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FIG. 1a and b.—Inferior vena cavagram revealing obliteration of IVC, with collateral flow through the azygos veins.

and a follow-up five years later showed a normal child without evidence of abnormal renal function.

Case 2. A 2-week-old negro male infant, born October 15, 1963, to a 17-year-old primipara of a full-term pregnancy, weighed 3,750 g. He refused feedings and then began to vomit and to have loose stools on the day before admission. Except for malnutrition and moderate dehydration he was alert and responsive. An abdominal mass was felt in the right upper quadrant and 'sclerema' of the legs was noted. Haematuria was present and electrolytes showed evidence of marked acidosis with a low serum calcium. Haemoglobin was 15.4 g./100 ml., platelets 24,000, leucocytes 23,100/c.mm. Anuria developed on the day of admission and the child had a convulsion.

On October 31 hydration was somewhat improved, but the 'sclerema' had extended to the trunk. Bilateral abdominal masses were now palpated. BUN was 160 mg./100 ml. The electrolyte imbalance was corrected,

the fluid regimen was regulated for renal failure, and calcium and anticonvulsive therapy was started. On November 1 the abdominal masses seemed to be larger and until that time only a few ml. bloody urine had been produced. The diagnosis of bilateral renal vein thrombosis was made. An inferior vena cavagram was made via a saphenous vein (Fig. 1a and b), showing obliteration of the lower inferior vena cava with extensive collateral circulation connecting with the azygos system. Platelets rose to 64,000, BUN was still 150 mg./100 ml.

Operation was undertaken on November 1 with a tentative diagnosis of combined thrombosis of inferior vena cava and both renal veins. Both kidneys were found to be three to four times as large as normal, brown in colour, with diffuse haemorrhage and covered with dilated lymphatics that contained bloody lymph. The renal segment of the inferior vena cava was dilated and contained a thrombus that extended for 2 cm. proximally into the suprarenal segment of the vena cava and also for a short distance into both renal veins, occluding them

both. The entire infrarenal segment of the inferior vena cava was irregularly narrowed and obviously obliterated with advanced thrombosis.

The inferior vena cava was transected 1 cm. distal to the take-off of the renal vein and the distal part was ligated. Through the proximal stump, thrombus material was removed both from the inferior vena cava and from the central part of the renal veins by extraction and saline irrigation until bleeding occurred. No heparin was used in view of the persistent thrombocytopenia. Histology showed beginning of organization of the thrombus. After operation a diuresis occurred and urine output returned gradually to normal as the haematuria disappeared. Continued low calcium levels required supplementary treatment and anticonvulsive therapy was continued. BUN came down to 17 mg./100 ml.

Intravenous urogram just before discharge showed kidneys of normal size with good function on both sides. The convulsions had subsided and calcium therapy was discontinued. Follow-up 9 months later showed, except for a low serum calcium, no other evidence of abnormal renal function. The BUN was 11 mg. Platelet count and urinalysis were also normal. There was no evidence of hypertension, but the child seemed developmentally slow.

Discussion

Although one had unilateral and the other bilateral involvement, there is a striking similarity in the clinical picture of the two patients in the urinary symptoms, operative findings, and in the favourable outcome after surgery. On the basis of the clinical course both cases could probably be considered compatible with the primary type of renal vein thrombosis, according to Sandblom's classification.

Of the 8 infants who died, 6 were found to have a pre-existing congenital lesion, the importance of which may be questioned. They were noted to have toxoplasmosis, sickle cell disease, cyanotic heart disease, inclusion body disease, hydronephrotic kidney, and polycystic kidney. All these deaths occurred in patients with marked dehydration, shock, or sepsis without exhibiting obvious renal symptoms. In them the renal vein thrombosis was found only at necropsy and in the majority the diagnosis was based only on microscopic findings. Massive infarctions were not encountered in this group, and we are inclined to classify them in the second category of renal vein thrombosis in accordance with Sandblom's description.

In both surviving patients, the thrombosis also involved the lower segment of the inferior vena cava. This observation made in two living patients with renal vein thrombosis led us to consider angiography of the inferior vena cava as a promising method in the study of similar patients. As an addition to routine intravenous urography, inferior vena cava visualization can give valuable information, especi-

ally in patients presenting with renal tumours (Allen, Morse, Frye, and Clatworthy, 1964).

When both kidneys are involved in a thrombotic process, a fatal outcome is almost certain. According to Kaufmann (1958), in an analysis of the published reports, more than 45% of 91 childhood cases occurred bilaterally. Campbell (1951) also mentioned equal distribution in the incidence of unilateral and bilateral cases. Few reports of surgical cure of an adult patient with the proven diagnosis of bilateral renal vein thrombosis have appeared (Austen, 1961). Isolated reports claim recovery of bilateral renal vein thrombosis with conservative methods only, but in none of these was the diagnosis unequivocal (Fallon, 1949; Gaillard, Delphin, Cajfinger, and Brugiere, 1960; Michon, Aubertin, Jagerschmidt, and Valleteau de Moulliac, 1962).

Since the therapeutic effect of anticoagulants on established thrombosis is uncertain, we prefer to consider renal vein thrombosis in its acute stage as an urgent surgical problem. Though we prefer nephrectomy in strict unilateral renal vein thrombosis with a normal contralateral kidney, restoration of the renal venous return by removal of thrombus may also be effective. This is demonstrated in our second case in which we were forced to rely on this method because of bilateral involvement. Contrary to reports of cure with heparin alone (Fournier and Pauli, 1955; Gaillard *et al.*, 1960; Michon *et al.*, 1962), we are inclined to consider anticoagulant treatment mainly complementary to nephrectomy or thrombectomy. In situations where surgical intervention might be contraindicated, heparin therapy should be used. Reports suggesting renal vein thrombosis as the underlying cause of hypertension (Perry and Taylor, 1940; Schöenberg and Staemmler, 1960) or the nephrotic syndrome (Feinerman, Burke, and Bahn, 1957; Torres, 1962) in some patients seem to underline our views on the management of patients with renal vein thrombosis.

Summary

Two cases of renal vein thrombosis, one unilateral and one bilateral, are reported. Both patients survived following operation. The importance of early diagnosis, the use of the vena cavagram as a pre-operative study, and the rationale of treatment are discussed.

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