CLINICAL QUIZ (p214-p215) ANSWER

Case 1

Question 1

Renal arteriogram were showed in Figures 1 and 2. Typical "string of beads" appearance of distal portion of left renal artery and right segmental renal arteries with probably involvement of distal right main renal artery is noted. String of beads mean alternating areas of stenoses and aneurysm.

Question 2

Renal artery stenosis (RAS) involved bilateral main renal arteries and intrarenal arteries. The most likely pathological diagnosis is fibromuscular dysplasia (FMD). Renal artery stenosis due to FMD usually involves the distal two third of the renal artery and its branches. FMD are characterised by focal thickening of the vascular wall and aneurysm formation.[1,2] Therefore the string of beads appearance in renal arteriogram is pathognomonic of renal artery stenosis due to FMD. In patients with renovascular hypertension, the plasma rennin level may be normal or low if the hypertension is sustained, termed as "reverse tachyphylaxis".[1] FMD is the most common cause of renovascular hypertension under forty years old.

Question 3

Control of hypertension is usually difficult and requires multiple anti-hypertensive agents. Hypertension resulted from RAS usually responds well to angiotensin-converting enzyme inhibitor but there is a risk of impaired renal failure. Refractory hypertension may respond to percutaneous balloon angioplasty. The immediate success rate after balloon angioplasty is about 95%,[3,4] which remained cured or improved in 74% at 12 months of follow up.[4] The re-stenosis rate is about 10-25%.[3] However angioplasty seems ineffective in children with multiple stenoses.[5] A recent new technique of using cutting balloon catheter to dilate resistant renal artery stenosis was reported but its effect on long- term outcome is not available.[6] The prognosis of RAS from FMD is relatively better as compared with those from artherosclerosis as the risk of ischaemic nephropathy and excretory dysfunction is low.[1,2] However, data on long-term outcome of renal function and impact of sustained hypertension is lacking in children with FMD.

References

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4. Birrer M, Do DD, Mahler F, Triller J, Baumgartner I. Treatment of renal artery fibromuscular dysplasia with balloon angioplasty: a prospective follow-up study. Eur J Vasc Endovasc Surg 2002;23:146-52.
5. Courtel JV, Soto B, Niaudet P, et al. Percutaneous transluminal angioplasty of renal artery stenosis in children. Pediatr Radiol 1998;28:59-63.
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Case 2

Answer: Congenital Syphilis

Congenital syphilis is transmitted vertically from infected mother to her baby during pregnancy via placental circulation. This unmarried mother was non-reactive to VDRL test in first trimester, and did not aware that she was infected in early third trimester. The diagnosis was confirmed by checking mother and baby blood for VDRL and FTA-ABS titres. (Baby VDRL: 1: 1024, Mother VDRL: 1:64), and FTA-ABS were strongly positive in them.

Infected baby may be asymptomatic after birth and signs then develop afterward. The first antenatal blood test for VDRL is not helpful if pregnant women acquire the infection late in pregnancy and infected women may be asymptomatic.

Other tests for evaluation for probable and confirmed congenital syphilis include:

1) Placenta histology, and fresh body tissues/fluid for Dark field examination for spirochetes; Complete blood count, Liver function test; Lumbar puncture for CNS involvement and Radiology of long bones to look for periositis and metaphysitis, and Chest X-rays for pneumonitis.
2) Mother should be screened for HIV and other sexual transmitted diseases.

The recommended treatment is aqueous crystalline penicillin G (100 000 to 150 000 Unit/kg/day given every 8 to 12 hours) for 10-14 days. Treatment should last for 21 days for CNS involvement. Serial evaluation for physical signs and VDRL titres are mandatory for monitoring treatment success.