Nephrology Dialysis Transplantation

Nephroquiz for the Beginner
(Section Editor: T. J. Rabelink)

An unusual complication of pregnancy

A 31-year-old primigravida, previously fit and well, was noted by her health visitor to be pale 6 weeks after a normal full-term pregnancy. A routine full blood count and film showed evidence of a microangiopathic haemolytic anaemia (Hb 9 g/dl) and thrombocytopenia (PLT 82 × 10⁹/l) but a normal coagulation screen. On further questioning, she admitted to generalized aching and lethargy 1 week after delivery and had noted increasing tiredness, breathlessness, proximal muscle weakness and swelling of her hands and feet 3 weeks later. Renal function was impaired (Cr 142), urinalysis showed only a trace of protein and +1 blood and microscopy showed an inactive urinary sediment. Blood pressure was elevated (160/110) but fundoscopy was normal. Autoimmune serology (antinuclear antibodies (ANA) and anticardiolipin antibodies) were negative and complement levels (C₃, C₄) were normal. A diagnosis of idiopathic post-partum haemolytic uraemic syndrome (HUS) was made and she was started on antihypertensives and aspirin. Because of persisting thrombocytopenia, haemolysis and deteriorating renal function, plasma infusion followed by plasma exchange with fresh frozen plasma (FFP) was commenced from day 9 of presentation and a renal biopsy (Figure 1) performed on day 22.

Despite daily plasma exchange with FFP and cryosupernatant, she became dialysis dependent by day 24. Her course was complicated by refractory hypertension, profound ischaemic retinopathy (Figure 2), continuing haemolysis and severe left ventricular failure, the latter requiring treatment with daily volume removal, digoxin and captopril. Somewhat unexpec-

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Fig. 1. (a and b) Histology at renal biopsy.

Fig. 2. Retinal photograph taken at day 43 showing the presence of ‘cotton wool’ exudates and occasional haemorrhages, in particular surrounding the optic disc. At this point, the patient had complained of ‘moth eaten’ vision. Six months later, these changes had resolved and her vision had returned to normal.

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Fig. 3. (a) Photograph of both hands taken 8 months after the onset of illness demonstrating the marked shiny skin thickening overlying the fingers. (b) Photograph of the left hand with the fingers fully extended to illustrate the development of flexion deformities of the fingers.

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...lysis has ceased but the skin changes in her hands have progressed (Figure 3a and b). Her resting ECG remains abnormal with widespread T wave inversion (Figure 4) but cardiac function is only mildly impaired. Pulmonary function tests performed on day 51 showed mild abnormalities: vital capacity (VC) 82% of predicted, total lung capacity (TLC) 88% of predicted and transfer factor (KCO) 80% of predicted. Subsequent tests have revealed detectable autoantibodies to RNA polymerase I and III, persisting from day 36 of her illness. Nine months from the start of her illness, she has recovered minimal renal function (CrCl 5 ml/min) and remains on CAPD.

Question

What is the diagnosis?

(Answer on next page)
Answer to quiz on preceding page

Post-partum HUS secondary to scleroderma

The biopsy shows glomeruli of normal cellularity and open capillary loops but the glomerular tufts are slightly shrunken (Figure 1a). The major abnormality is in the arteries where a striking concentric intimal proliferation can be seen (Figure 1b). These changes have been reported in idiopathic post-partum HUS but the arterial changes are more typical of other conditions such as malignant hypertension and scleroderma [1].

Idiopathic post-partum HUS is a rare complication of pregnancy which can lead to irreversible renal failure [2]. The aetiology of this condition is unknown but it may recur in subsequent pregnancies [3] or with the subsequent use of the oral contraceptive pill [2]. Several cases of idiopathic post-partum renal failure associated with congestive heart failure and haemolysis have also been described [4], but it seems likely that they represent the severe end of the disease spectrum in this condition rather than a separate disease entity.

The effect of pregnancy on scleroderma is unpredictable but post-partum renal failure in patients with pre-existing scleroderma has been reported [5]. Our patient was unusual in a number of respects; (i) post-partum HUS was the presenting feature of her scleroderma; (ii) she did not develop malignant hypertension but still had marked ischaemic retinopathy; (iii) she remains ANA negative. None of these features however exclude an atypical de novo presentation of scleroderma [6] and normotensive crises may account for 25% of scleroderma renal crises [1]. Less than 10% of all scleroderma patients are ANA negative [7]. However, the presence of autoantibodies to RNA polymerase is more prevalent in scleroderma patients with renal involvement [7].

A proportion of patients with scleroderma renal crises maintained on ACE inhibitors have been able to discontinue dialysis up to 15 months later [8]. Cases of idiopathic post-partum HUS with delayed recovery of renal function have also been reported [2].

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Suggested reading


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