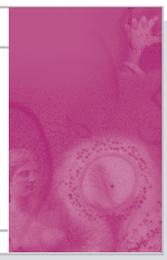
SCIENTIFIC LETTER

Renal cortical necrosis and HELLP syndrome: A case presentation



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To the Editor: Renal cortical necrosis (RCN) is a rare cause of acute renal failure (ARF), accounting for 3% of all ARF cases in adults in developing countries, but up to 15% in pregnancy-related ARF (PR-ARF).[1] It is caused by a significant reduction in renal perfusion due to vasospasm, microvascular injury, intravascular coagulation or a combination of these factors. Histology shows a diffuse or patchy necrosis of the renal cortex; the medulla and a thin rim of subcapsular cortex are usually spared.[2] HELLP syndrome (haemolysis, elevated liver enzymes and low platelets) is a variant of severe pre-eclampsia which often complicates with renal failure, most of the time because of acute tubular necrosis (ATN) but occasionally because of RCN, through a thrombotic micro-angiopathic process.[3] We report on a case of HELLP syndrome where the patient developed ARF as a result of RCN.

Case report

A 25-year-old pregnant woman, gravida 2, para 0, abortion 1, presented at 34 weeks of pregnancy with vomiting, haematemesis and vaginal bleeding. During a consultation 3 weeks before admission, she had been found to be hypertensive (140/100 mmHg) and proteinuric (1+). On examination at her district hospital she was pale and oedematous, with a blood pressure of 190/111 mmHg; the fetal heart rate was 156/min, and gastric aspiration confirmed upper digestive bleeding. Her condition deteriorated, and she became anuric and confused. On arrival at Polokwane Hospital she was very ill, mildly jaundiced and hypertensive, and the fetus was dead. Fundoscopy showed bilateral papilloedema and flame haemorrhages. She delivered a fresh stillborn fetus of 1 400 g after induction of labour. Further findings were: haemoglobin 6.3 g/dl; white cell count 21.5×10^9/l; platelets 23×10^9/l; mean cell volume 82 fl; peripheral smear showed anisocytosis, target cells and schistocytes; lactate dehydrogenase 612 U/l; alanine transaminase 446 U/l; aspartate transaminase 191 U/l; total bilirubin 34 mmol/l; direct bilirubin 12 mmol/l; total protein 46 g/l; albumin 20 g/dl; international normalised ratio 2.5; partial thromboplastin time 53.1 seconds (control 33 seconds); fibrinogen 0.6 g/l; D-dimer 1.4 (normal <0.25); creatinine 406 μg/dl; urea 9.3 mmol/l; uric acid 0.48 mmol/l; HIV non-reactive; hepatitis B and C negative; and antinuclear, antineutrophil cytoplasmic and antiphospholipid antibodies all negative. An abdominal ultrasonogram showed both kidneys to be of normal size but with increased echogenicity. She remained anuric and went into pulmonary oedema, needing mechanical ventilation and haemodialysis. Renal function did not recover after 4 weeks, and a kidney biopsy then showed necrosis of most glomeruli (Fig. 1) and widespread thrombi within glomerular capillaries (Fig. 2). An abdominal radiograph after 12 weeks showed calcified kidneys. The patient regained consciousness but remained anuric, and developed nosocomial pneumonia and severe hypoalbuminemia. She died after 4 months in the ICU as a result of a ventilator-related nosocomial pneumonia caused by extensively resistant *Klebsiella pneumoniae*.

Discussion

RCN is a very rare event that occurs in less than 1 in 80 000 pregnancies, accounting for just 3% of all cases of ARF in adults in developing countries and 15% of pregnancy-related ARF cases.[1] Various conditions, both obstetric and non-obstetric, can cause RCN; among the former are septic abortion, pre-eclampsia, eclampsia, HELLP syndrome, abruptio placentae and prolonged intrauterine fetal death; among the latter are snakebite, renal allograft rejection, uraemic-haemolytic syndrome, enterocolitis, pancreatitis, sepsis, etc. More than 50% of cases are pregnancy-associated, however, and most

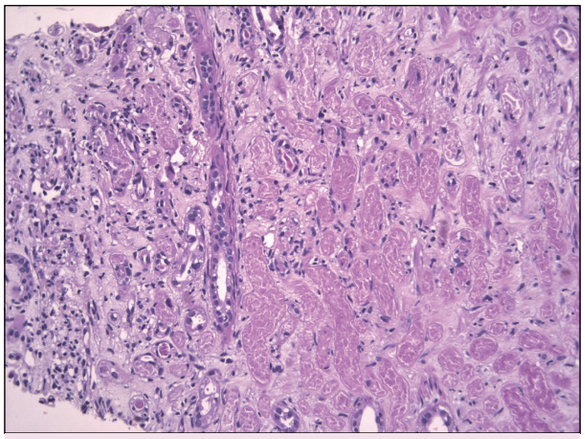

Fig. 1. Haematoxylin and eosin stain x200. Junction between necrotic cortex on the right-hand side and residual viable parenchyma to the left.

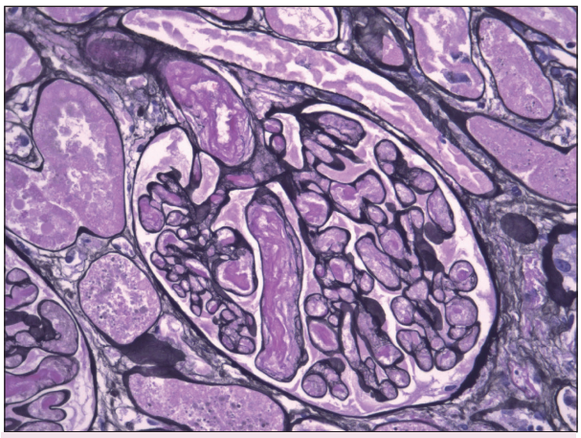

Fig. 2. Silver-methenamine stain x400. An area of cortical necrosis, depicting ghost outlines of tubules and a glomerulus. Fibrin-platelet thrombus material is visible within the glomerular capillary lumina.

of them occur during the third trimester, mainly as a consequence of abruptio placentae or HELLP syndrome.[4] We ascribe our case to the latter since, although the patient presented with vaginal bleeding and had a rapid fetal death suggestive of abruptio placentae, she never went into shock, and examination of the placenta did not reveal any retroplacental clot.

Maternal prognosis in PR-ARF is generally good, but poor if associated with RCN or disseminated intravascular coagulation;[5] the latter was reported by Sibai *et al*. in more than 80% of their patients, with a maternal mortality of 13%. In Prakash's study, 15% of the PR-ARF cases were caused by RCN; of this 15%, a third never recovered renal function and went to end-stage renal disease.[1-5] Our patient suffered from both diseases, as evidenced by histological analyses: cortical necrosis and signs of thrombotic microangiopathy (fibrin-platelet thrombus material within the glomerular capillary lumina) and abnormal clotting parameters with low fibrinogen, high D-dimer levels and low platelet counts. We can suspect the condition when a patient suffering from HELLP syndrome or abruptio placentae develops anuric ARF and fails to produce any urinary output after 3 or 4 weeks on dialysis – enough time for acute tubular necrosis to show any sign of recovery. Calcification of renal parenchyma has also been mentioned as a very specific characteristic of RCN but has been found to be insensitive. We persevered in looking for it almost weekly, and found it only after 12 weeks – too late to be useful. RCN diagnosis can also be made by renal arteriography or contrast-enhanced computed tomographic scan;[4] both of these, although less invasive (especially the latter), are difficult to perform on a critically ill patient on mechanical ventilation confined to an ICU bed. A kidney tissue sample can always be obtained, even in the critically ill, provided that normal clotting function and blood pressure control have been achieved; but carries the risk of missing the diagnosis in the patchy, rather than diffuse, type of RCN that has been reported in up to 30% of cases.[4]

Our case highlights the importance of kidney biopsy in ARF to clarify the diagnosis when the expected recovery does not occur, and also of early identification of pre-eclampsia for proper management, so as to avoid these and other fatal complications.

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2. Madias NE, Donohoe JF, Harrinton JT. Post ischemic acute renal failure. In: Brenner BM, Kazarus JM, eds. *Acute Renal Failure*. 2nd ed. New York: Churchill Livingstone, 1998: 597-690.
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