PHOTO QUIZ

A patient with acute renal failure

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CASE REPORT

A 78-year-old female was admitted with renal insufficiency. She presented with dyspnoea on exertion and complained of painful and stiff hands. Photographs of her hands *(figure 1)* and face (*figure 2)* are shown. Blood pressure was 150/95 mmHg.

Laboratory tests showed an ESR 29 mm/h, WBC count 13.8 G/l, haemoglobin 8.3 mmol/l (nadir 6.3), platelet count 177 G/l (nadir 96), sodium 141 mmol/l, urea 10.8 mmol/l, creatinine 144 µmol/l (zenith 299), potassium 3.5 mmol/l, lactate dehydrogenase 1162 U/l (zenith 1500) and plasma renin activity (PRA) 7270 fmol/l/sec (zenith 9950).

ANA was ≥1:640, anti-dsDNA and anti-centromere antibodies were negative but anti-Scl70 was positive. Urinalysis showed protein 1.1 g/l, leucocytes 0-5 (high power fields; HPF) and erythrocytes 40-50 (HPF).

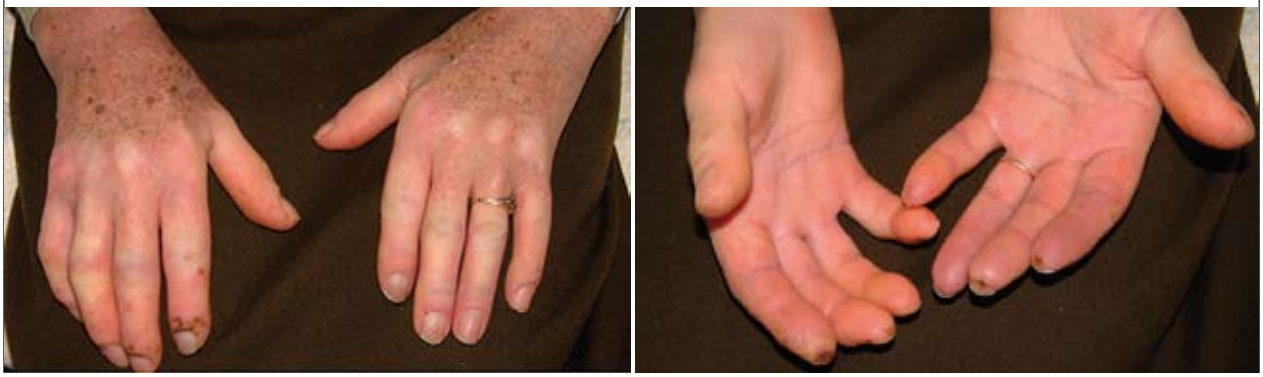
WHAT IS YOUR DIAGNOSIS?

See page 361 for the answer to this photo quiz.

Figure 2. *Patient's face*

The patient has consented to the use of her pictures.

Figure 1. *Patient's hands*



ANSWER TO PHOTO QUIZ (ON PAGE 360)
A PATIENT WITH ACUTE RENAL FAILURE

DIAGNOSIS

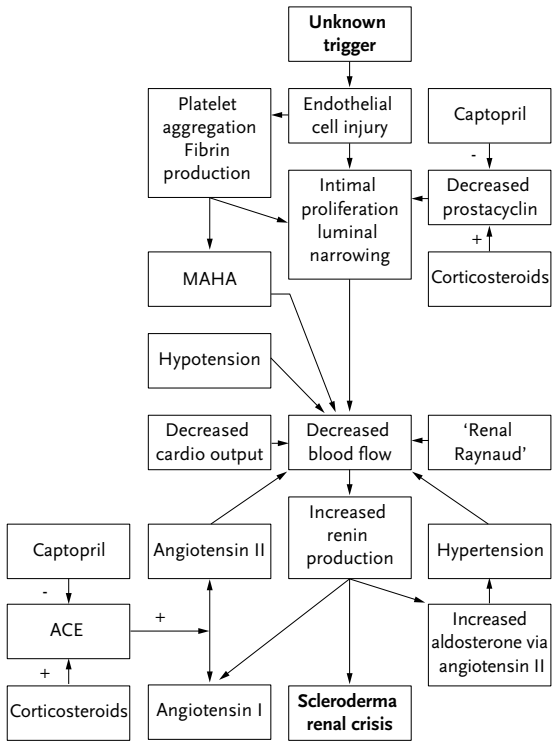
The patient was diagnosed with diffuse cutaneous systemic sclerosis three years ago. There was evidence of cardiac and pulmonary involvement. She was treated with prednisone 15 mg/day besides methotrexate, acenocoumarol, pantoprazole, folic acid, furosemide and carbasalate calcium. She now presented with a scleroderma crisis (SRC), with renal failure, thrombotic microangiopathy, hyper-reninaemia and hypertension. Her skin abnormalities are typical for scleroderma with sclerodactyly and acral ulceration (*figure 1*) and absence of wrinkling of her facial skin (*figure 2*).

Captopril 25 mg, three time a day was given. The blood pressure diminished to 140/90 mmHg and after adding labetolol to 120/70 mmHg. Prednisone was reduced to 5 mg/day and methotrexate was stopped. The platelet count and lactate dehydrogenase normalised. Renal function and plasma renin activity improved slowly. Symptoms of dyspnoea and acral ulcerations improved in the course of time. Two years after admission the patient was in a stable clinical condition with a creatinine of 146 µmol/l.

SRC is a syndrome that is characterised by accelerated hypertension, rapidly progressive renal failure, and hyper-reninaemia. Of note, hypertension blood pressure can be normal in SRC. The frequency of SRC in diffuse cutaneous systemic sclerosis (dcSSc) is 15 to 20%. The renin-angiotensin system plays an important role in its pathogenesis. Decreased blood flow, caused by structural vascular changes and possibly vasospasm (renal Raynaud's phenomenon),[1] lead to decreased renal perfusion. This causes excessive release of renin and subsequently formation of large amounts of angiotensin II, resulting in further vasoconstriction, raised blood pressure and renal ischaemia. The end result is a vicious cycle that is believed to be the cause of SRC.

Our patient had several characteristics indicating an increased risk for SRC: diffuse form of SSc, rapid progression of skin thickening and a duration of SSc less than four years.[2] Factors possibly provoking SRC in our patient were (1) daily intake of corticosteroids, (2) cold season and (3) cardiac dysfunction. (Ad 1) Corticosteroids increase ACE activity via inhibition of prostacyclin production and may thus contribute to the pathogenesis of SRC. Therefore, the dose of prednisone was reduced to 5 mg/day. (Ad 2) The patient developed SRC in the wintertime. SRC onset has been reported more often during winter, suggesting that renal Raynaud's phenomenon may be a contributing factor.[1] (Ad 3) The patient suffered from arrhythmias. Cardiac dysfunction precedes SRC in some patients.[2] All mentioned factors contribute to decreased renal perfusion, leading to excessive secretion of renin. The hyper-reninaemia maintains SRC via a vicious cycle (*figure 3*). The vicious cycle can be interrupted by ACE inhibitors. Since the introduction of ACE inhibitors one-year survival has increased from 10% to 76%.[2] ACE inhibitors inhibit the conversion of angiotensin I to angiotensin II, resulting in a prompt drop in blood pressure in many patients with SRC. In our patient, after treatment with captopril, the blood pressure decreased and renal function improved slowly.

Figure 3. *Pathophysiology of scleroderma renal crisis*



MAHA = microangiopathic haemolytic anaemia.

REFERENCES

1. Cannon PJ. The relationship of hypertension and renal failure in scleroderma (progressive systemic sclerosis) to structural and functional abnormalities of the renal cortical circulation. Medicine (Baltimore) 1974;53:1-46.
2. Steen VD, Medsger TA Jr, Osial TA Jr, et al. Factors predicting development of renal; involvement in progressive systemic sclerosis. Am J Med 1984;76:779-86.