ABSTRACT
A 36-year-old female with diffuse cutaneous systemic sclerosis (dcSSc) developed macrohematuria due to vesical telangiectasias that was responsive to diathermocoagulation of the vasal lesions. This is the first report of a patient with dcSSc and vesical telangiectasias leading to severe macrohematuria that was successfully treated with diathermocoagulation.

Introduction
Systemic sclerosis (SSc) is a multi-system disorder characterised by vascular damage and the overproduction of collagen and other matrix constituents in the skin, gastrointestinal tract, lungs, heart, kidneys, joints and muscles (1-3). Clinically evident vascular manifestations mainly consist of Raynaud’s phenomenon and telangiectasias (4). Telangiectasias involve the skin, but have been also detected at the mucosal membranes, i.e. lips, tongue, nose, bladder, oesophagus, stomach, small intestine and colon-rectum (5-7), and are more prominent in patients with limited SSc (the so-called CREST syndrome) (8).

Here we report a patient with dcSSc who developed macroscopic haematuria caused by widespread vesical telangiectasias, in whom blood loss was stopped by diathermocoagulation of the vesal lesions.

Case report
A 35-year-old woman was admitted in September 1996 to our clinic because of Raynaud’s phenomenon with digital ulcers, anti-Scl70 positivity and skin thickening involving the arms, face and trunk (Table I).

The patient was administered D-penicillamine, calcium channel blockers and cisapride. In February 1997 a routine urinalysis revealed the presence of proteins (++). D-penicillamine was discontinued and cyclophosphamide 50 mg/die was introduced. In July 1997 the patient developed microhematuria (5-10 red blood cells/high power field) and in December 1997 macrohematuria. She was treated with tranexamic acid and N-acetylcisteine. Cyclophosphamide was stopped and never subsequently administered. The hematuria disappeared.

In April 1998 another episode of macrohematuria occurred. A renal ultrasonography was normal, but intravenous pyelogram showed left ureteral kneeling and cystoscopy showed mucosal inflammation of the bladder wall with several telangiectasias. In July 1998 the results of a routine urinalysis showed: no proteins, 7-15 red blood cells/high power field, no pyuria, no bacteriuria, and no cellular casts. A routine urine culture was negative.

In August 1998 the patient manifested marked macrohematuria poorly responsive to anti-fibrinolytic drugs. This lasted 3 weeks, causing severe anaemia and requiring a transfusion of 8 blood units. Pelvic echography showed bladder wall thickening; cystoscopy revealed confluent telangectatic lesions; and bladder wall biopsy showed telangiectasias, small vascular thrombosis in the right side wall, and glandular cystitis in the left side wall (Figs. 1, 2). In September 1998 the patient underwent further biopsies and diathermocoagulation of the vesal lesions. No bleeding occurred during bladder emptying. Bimanual bladder examination was negative. At present the macrohaematuria persists.

Discussion
Mucosal telangiectasias in SSc have been reported to cause bleeding ranging from occult blood loss to acute haemorrhages (9, 15). Bladder involvement in SSc has been recently reviewed by La Civita et al. (16), who described...
CASE REPORT

a case with bladder fibrosis. Vesical telangiectasias have been so far reported in 4 SSC patients, all with the limited subtype of SSC (lcSSc) (12-15), associated in 2 cases with microhematuria and in 2 with macrohematuria. In all of these cases the cause of the blood loss, the need for blood transfusions and the local therapy used, if any, were not mentioned.

Macrohematuria in SSC could arise from a number of causes other than vesical telangiectasias. Cystitis, kidney stones, nephritis, bladder carcinoma, and sickle cell anaemia were all ruled out in our patient, however. Cyclophosphamide also could have been the causative factor. We initially ascribed our patient’s macrohematuria to a toxic effect of the drug. However, the recurrence of hematuria after drug withdrawal and the absence of findings consistent with hemorrhagic cystitis at biopsy allowed us to attribute the hematuria to the disease itself.

The patient described here shows some peculiarities compared to those reported thus far in the literature. (1) This is the first patient with dcSSc and vesical telangiectasias, showing that mucosal as well as cutaneous telangiectasias are by no means confined to patients with lcSSc. (2) The blood loss was so great that the transfusion of 8 blood units was required. Indeed serious anaemia had developed (Hb 6 gr/dl). (3) This is the first report of a patient in whom diathermocoagulation, i.e. a topical therapy, has been showed to stop significant macrohematuria by vesical telangiectasias, and confirms the already reported effectiveness in SSC of diathermocoagulation on bleeding telangiectasias in other regions such as the gastrointestinal tract (10).

References