Corticosteroid induced remission of oesophageal involvement in mixed connective tissue disease

AMOS PINES
M.D.

NAOMI KAPLINSKY
M.D.

EHUD GOLDHAMMER
M.D.

JOSEPH BREGMAN
M.D.

OTTO FRANKL
M.D.

The Department of Medicine F, The Sheba Medical Center, Tel-Hashomer, Israel

Summary

Oesophageal involvement is known to be one of the most severe and resistant manifestations of connective tissue diseases, mainly progressive systemic sclerosis (PSS). A patient who had manifestations of systemic lupus erythematosus, PSS and polymyositis is described. Since some features of mixed connective tissue disease (MCTD) respond to corticosteroid treatment, fluocortolone was administered and both clinical and radiological remission of the oesophageal involvement were observed. At the same time, there was also a marked improvement of the cutaneous lesions. In view of the described observation, it seems advisable to try this treatment for oesophageal manifestations of MCTD.

Introduction

Oesophageal involvement is a common feature of progressive systemic sclerosis (PSS), known to be resistant to therapy. We report a patient who developed overlapping features of systemic lupus erythematosus (SLE), PSS and polymyositis, with severe oesophageal involvement which responded well to corticosteroid therapy.

Case report

A 45-year-old female patient was hospitalized because of vomiting and dysphagia following every meal, appearing 2 weeks before her admission.

Her past history revealed rheumatic fever at the age of 18 years, leading to mitral stenosis and chronic atrial fibrillation. At the age of 42 years, thrombocytopenic purpura and anaemia associated with hypertension and positive antinuclear factor (ANF) were found, and SLE was suspected. A year later,

mild renal failure was diagnosed, the patient refused a kidney biopsy, and responded well to low doses of corticosteroids. At the age of 44 years, sudden hemiplegia and aphasia appeared. Cerebral haemorrhage or embolus were suspected but since lupus encephalitis could not be excluded, a short trial with high dose of corticosteroids was initiated, with no improvement. At that time, the patient could swallow without difficulty.

At the age of 45 years, and 3 weeks before the present admission, difficulties in swallowing gradually developed and vomiting appeared following every meal, until she stopped eating and was hospitalized for evaluation and treatment.

On admission the physical examination revealed blood pressure of 130/90 mmHg, irregular pulse 80 per min, right hemiplegia and aphasia, muscle weakness of the left upper and lower extremities, decreased air entry to the base of the right lung, the first heart sound was accentuated, the abdomen was soft, the liver and the spleen were not palpated. The skin over the abdomen, arms and thighs appeared thickened, in great contrast to the texture of the skin on previous admissions.

The significant laboratory findings were: Hb 10.5 g/dl, WBC $5 \times 10^9/l$, platelet count $126 \times 10^9/l$, urea 16.6 mmol/l, creatinine 177 $\mu\text{mol/l}$, glutamic oxalic transaminase (SGOT) up to 120 u./l (normal < 16 u./l), positive ANF, LE cells were found, total haemolytic complement 24%. X-ray examinations revealed a small right pleural effusion and severe dilatation of the oesophagus, loss of peristalsis and reflux. The stomach and the small intestine appeared normal.

The patient continued to vomit food and fluids. A soft gastric tube was introduced in order to enable

safe feeding. The pleural effusion was aspirated and found to be an exudate. Skin biopsy showed thickened collagen fibres and mild atrophy of the sweat glands, compatible with early scleroderma.

Assuming that the patient had mixed connective tissue disease (MCTD) with features of SLE, PSS and myositis (as suggested by muscle weakness and muscle enzyme elevation), a trial with 20 mg of fluocortolone daily was given.

After 2 weeks there was an obvious improvement in the texture of the skin. The patient was discharged in May 1980, taking alternate day treatment of 20 mg of fluocortolone. In July 1980, she was able to eat without the gastric tube. In November 1980, the patient was hospitalized for re-evaluation. This time oesophageal radiograms showed that it was less widened, peristaltic waves were clearly seen and no reflux was demonstrated. There was also further improvement in the skin over the abdomen, while the skin over the thighs and arms seemed normal. In March 1981, the patient was still in our follow-up, taking fluocortolone, and stable clinically.

Discussion

Since the original description (Sharp *et al.*, 1972), it has become apparent that the definition of MCTD is not distinct. Two factors were basically needed for the diagnosis of MCTD: overlapping clinical features of more than one rheumatic disease and high titres of antibody to ribonucleoproteins (RNP) (Sharp, 1979). The boundaries of the above definition have since widened. Cases with overlapping features without antibodies to RNP (Clements *et al.*, 1978) and cases with high titres of antibody to RNP without overlapping clinical manifestations (Himelstein *et al.*, 1980) were described. At first, it seemed that all patients with MCTD have a better prognosis and respond well to corticosteroids (Sharp, 1979) but this assumption did not hold true (Himelstein *et al.*, 1980). Some investigators now think that it would be better to change the name of MCTD into the less committed name of undifferentiated connective tissue disease (LeRoy, Maricq and Kahaleh, 1980) in view of the fact that most of the patients eventually appear to have only one distinct rheumatic disorder, usually PSS (Himelstein *et al.*, 1980).

Although antibodies to RNP were not looked for in the presented case, it was assumed according to the current concepts that she had MCTD. Gastro-

intestinal involvement is common in PSS and could be expressed as oesophageal dilatation and atony, small bowel dilatation, pseudo-diverticula on the antimesenteric border of the colon and pneumatosis cystoides (Berk, 1974). The same seems to be true also for MCTD (Sharp, 1979; Himelstein, 1980; Norman and Fleishman, 1978; Sharp, 1975; Silver, Farber and Martel, 1976; O'Connell and Benett, 1977).

Corticosteroids have not been shown to have an effect on the natural course of PSS, but in the early inflammatory phase of the illness, transient improvement from corticosteroid therapy may be seen (Korn and LeRoy, 1979).

Since we assume that the oesophageal and skin involvement in this patient were present while still in the inflammatory stage, a trial with steroids was initiated and found to be effective. It seems advisable that patients with early gastrointestinal involvement of MCTD should be given the benefit of a trial with corticosteroid therapy.

References

- BERK, R.N. (1974) Scleroderma of the gastrointestinal tract. *American Journal of Gastroenterology*, **61**, 226.
- CLEMENTS, P. J., FARST, D.E., CAMPION, D.S., BOHAN, A., HARRIS, R., LEVY, J. & PAULUS, H.E. (1978) Muscle disease in progressive systemic sclerosis. *Arthritis and Rheumatism*, **21**, 62.
- HIMELSTEIN, S.H., BROODY, S., MCSHANE, D. & HOLMAN, H.R. (1980) Mixed connective tissue disease: A subsequent evaluation of the original 25 patients. *Medicine*, **59**, 239.
- KORN, J.H. & LEROY, E.C. (1979) Scleroderma. In: *The Science and Practice of Clinical Medicine Vol. 4*. (Ed by Cohen, A.S.), p. 258. Grune & Stratton Inc., New York.
- LEROY, E.C., MARICQ, H.R. & KAHALEH, M.B. (1980) Undifferentiated connective tissue syndromes. *Arthritis and Rheumatism*, **23**, 341.
- NORMAN, D.A. & FLEISCHMANN, R.M. (1978) Gastrointestinal systemic sclerosis in serologic mixed connective tissue disease. *Arthritis and Rheumatism*, **21**, 811.
- O'CONNELL, D.J. & BENNETT, R.M. (1977) MCTD—clinical and radiological aspects of 20 cases. *British Journal of Radiology*, **50**, 620.
- SHARP, G.C., IRWIN, W.S., TAN, E.M., GOULD, R.G. & HOMAN, H.R. (1972) Mixed connective tissue disease—an apparently distinct rheumatic disease syndrome associated with a specific antibody to an extractable nuclear antigen. *American Journal of Medicine*, **52**, 148.
- SHARP, G.C. (1975) Mixed connective tissue disease. *Bulletin on Rheumatic Diseases*, **25**, 828.
- SHARP, G.C. (1979) MCTD (Ed by MCCARTH, D.J.), p. 737. *Arthritis and Allied Condition* 9th edn. Lea and Febiger, Philadelphia.
- SILVER, T.M., FARBER, S.J. & MARTEL, W. (1976) Radiologic features of MCTD and scleroderma—SLE overlap. *Radiology*, **120**, 269.