Case report

Morphoea (localised scleroderma) in a patient with mixed connective tissue disease

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SUMMARY An 18 year old girl concurrently developed skin lesions of morphoea (localised scleroderma) and pain and swelling of the hands and fingers. There were no dermatological or systemic signs of systemic sclerosis. The immunological features (high titred speckled antinuclear antibody, negative DNA binding, high titred positive anti-RNP and negative anti-Sm antibodies, speckled nuclear IgG fluorescence in the epidermis of the skin lesions) were consistent with mixed connective tissue disease, and it is suggested that the morphoea represented a component of this condition.

Key words: arthralgia, anti-RNP.

Since the syndrome of mixed connective tissue disease (MCTD) was first proposed in 1972 there has been argument between rheumatologists, immunologists, and others as to whether the condition is a true entity, or whether it merely represents an onset variant of one of the defined connective tissue diseases, such as systemic lupus or systemic sclerosis. MCTD is usually characterised by the coincidence of two or more connective tissue disorders and an immunological picture of high titred, speckled antinuclear antigen on autoantibody screen, direct immunofluorescence of the skin biopsy showing speckled nuclear IgG fluorescence in the epidermis, negative DNA binding (anti double stranded DNA antibodies), and high titred antibodies to nuclear ribonucleoprotein (anti-RNP). The argument against MCTD as an entity arises from the facts that (a) if observed for long enough many cases are now known to 'polaris' towards one or other connective tissue disorder, developing characteristic features and course; and (b) anti-RNP antibodies, formerly thought to be specific for MCTD, have been found in patients with a 'classic' connective tissue disorder in the absence of any of the features of MCTD. Nevertheless, a body of workers continues to believe in MCTD as a distinct entity. Recently there have been reports of a subvariety of MCTD, the 'undiifferentiated syndrome' of Calderon et al., who found that among 68 patients with anti-RNP antibodies 41 had a 'pure' systemic connective tissue disorder (usually systemic lupus). The remaining 27 (40%) had a syndrome consisting of Raynaud's phenomenon, swollen fingers, and arthralgia of the hands, normal serum complement, absent immune complexes, positive antinuclear antibodies (ANA), negative DNA binding, positive anti-RNP, and negative anti-Sm antibodies. One third had positive rheumatoid factor and only one patient evolved into a 'pure' collagen disease.

MCTD is a clustering of symptoms usually seen separately in various connective tissue disorders. It has a common bond of anti-RNP antibodies producing a statistically defined entity with overlapping features occurring together more frequently than would be expected by chance. Skin involvement, however, occurs in nearly every case—either by systemic lupus erythematosus (SLE), systemic sclerosis, or dermatomyositis lesions, usually starting with Raynaud's phenomenon and puffy hands. Localised forms of scleroderma such as morphoea have not, however, been reported in MCTD, and it is suggested that the following case represents the first example of this association.
An 18 year old girl discovered a 'lump' on the left lower chest wall which consisted of indurated, sclerotic, morphoeic plaques with surrounding pigmentation in a zosteriform distribution. Further lesions of morphoea then developed in the front of the left thigh and trunk. The fingers tended to go white in cold weather, but there was no definite Raynaud's phenomenon. There was no sign of acrosclerosis of the fingers or toes or other evidence of systemic sclerosis. A few weeks later she developed painful swellings of the finger joints and pain in the hands and feet resembling rheumatoid arthritis, involving the wrists and proximal interphalangeal joints of the fingers. Systemic examination showed no other abnormality.

Investigations: Hb 13.7 g/dl (137 g/l), white blood cell count (WBC) 6800/mm$^3$ (6.8×10$^9$/l) (differential WBC normal), erythrocyte sedimentation rate 31 mm/1st h, Rose-Waaler test positive 1/320, ANA test positive 1/1280 (speckled pattern), DNA binding negative, anti-RNP antibodies positive greater than 1/32 (strongly positive, curved immunoelectrophoresis technique with specificity determined by enzyme and other treatments of the antigen used and cross reactions between patient's serum and certified sera), anti-Sm and anti-SSB negative, serum immunoglobulins IgG 168 µg/l (normal <55), IgM 82 µg/l (normal <23), IgA 4 µg/l (normal <16), serum complement C3 and C4 normal, Clq 159 µg/l (normal <28), immune complexes absent from the blood.

Radiographs of the hands showed moderate osteoporosis of carpal and wrist bones with some cystic changes, consistent with early, non-erosive rheumatoid arthritis. A chest x ray and barium swallow were normal.

Skin biopsies of the lesions on the trunk and left thigh were typical of morphoea, there being a thin epidermis, striking increase in dermal collagen, loss of dermal appendages, and a mild lymphocytic perivascular infiltrate around the dermal vessels. Immunofluorescent staining showed a few granular C3 deposits in the blood vessels and strong speckled staining for IgG in the epidermis.

Course: the joint features responded to prednisolone 5–10 mg daily. She has remained well up to the latest follow up, two years from the onset.

Discussion

The patient had morphoea, arthralgia of the fingers, hands, and feet, positive rheumatoid factor, strongly positive speckled antinuclear antibody, speckled nuclear IgG fluroescence in the epidermis of the
skin lesions, negative DNA binding, strongly positive anti-RNP, and negative anti-Smith antibodies. Other than the skin lesions, these are the features of MCTD and in particular they call to mind the specific group described as the 'undifferentiated syndrome'.

In MCTD the epidermal nuclei of skin lesions often show a striking speckled pattern of nuclear IgG fluorescence. The deposition of immunoglobulins and complement components at the dermo-epidermal junction of non-lesional skin (the lupus band test), formerly thought to be specific for SLE, has been reported positive in MCTD, though it has been suggested that the reaction could represent an artefact associated with very high levels of anti-RNP.

Morphoea must be clearly distinguished from systemic sclerosis, there being complete absence of the systemic lesions so common in the latter. The skin lesions are localised or generalised, and it is rare for either variety to develop into systemic sclerosis (Fig. 1). Usually the lesions gradually soften, leaving areas of atrophy and abnormal pigmentation. The histology of affected skin is different in the two conditions: in morphoea the epidermis is thin and the dermis hypertrophic with excessive abnormal collagen; in systemic sclerosis the epidermis is normal and the dermis rather atrophic. The lesions of morphoea feel indurated, with ivory patches having purple or lilac edges. The skin and subcutaneous tissue are often bound to underlying muscle, which (as in systemic sclerosis) often shows electromyographic abnormalities of myopathy, though serum muscle enzymes are normal. Abnormal arterioles, often seen in systemic sclerosis, are not found in morphoea. Various immunological abnormalities have been described in patients with morphoea—antinuclear antibodies in up to 73% and increased DNA binding, though in low titres, in some. Anti-RNP antibodies have not so far been reported in this condition.

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References

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