A rare case of paraneoplastic dermatomyositis secondary to smouldering multiple myeloma referred for proteinuria

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Background. Dermatomyositis (DM) is often associated with malignancies but less commonly with plasma cell dyscrasias. Myoglobin nephropathy is an uncommon complication of inflammatory myopathies. This case describes a patient who presented with proteinuria because of myoglobin nephropathy caused by dermatomyositis. The dermatomyositis was paraneoplastic with an underlying smouldering multiple myeloma (SMM) diagnosed on investigation.

Case report. A 58-year-old mixed-race gentleman was referred for sub-nephrotic proteinuria. Further history taking revealed a 1-month history of proximal muscle weakness and a 5-month history of skin changes suggestive of dermatomyositis. Additional testing showed no associated albuminuria. There was haematuria on the urine microscopy which, with the accompanying raised creatine kinase levels, suggested myoglobinuria. Kidney biopsy confirmed myoglobin nephropathy. Further testing was done to exclude paraneoplastic DM. Investigations for solid organ malignancies were negative. Plasma cell dyscrasia screening revealed an IgA kappa monoclonal band, which was confirmed with serum free light chain testing. A bone marrow biopsy confirmed the diagnosis of SMM based on the revised working group diagnostic criteria for multiple myeloma.

Conclusion. This case serves as a reminder of the importance of clinical history and examination. Investigation in this case revealed an extrarenal cause of the proteinuria, and recognition of the presentation of DM led to further investigation for secondary causes.
Routine screening for plasma cell dyscrasias is recommended as part of DM work-up to allow the diagnosis to be made early, prior to organ damage.

Keywords: dermatomyositis, smouldering multiple myeloma, myoglobin nephropathy

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Dermatomyositis (DM) is an inflammatory myopathy that may form part of a paraneoplastic presentation secondary to various malignancies. An uncommon association is with plasma cell dyscrasias.

Myoglobin nephropathy is an uncommon complication of inflammatory myopathies, which include polymyositis (PM) and dermatomyositis (DM). The mechanism of injury is rhabdomyolysis, which causes muscle destruction leading to myoglobinuria that precipitates in kidney tubules causing acute tubular necrosis (ATN).[1] This case describes such a presentation where the reason for referral was proteinuria.

Case description

A 58-year-old mixed-race gentleman presented as an outpatient with the primary reason for referral being sub-nephrotic range proteinuria on a 24-hour urine specimen. On consultation the patient reported a 5-month history of a erythematous, non-pruritic rash around the eyes, elbows and the upper chest. He also reported a 1-month history of bilateral proximal arm weakness. There was no history of peripheral oedema, macroscopic haematuria or frothy urine.

On examination the patient had a typical heliotrope rash, an erythematous rash on the extensor aspect of both elbows, and a V-sign on the upper chest. Typical Gottron's papules (Fig. 1) were observed on his metacarpophalangeal joints and periungal telangiectasia (Fig. 2) on each nail bed. The power in both arms was 3/5 on the Oxford scale. These findings were suggestive of dermatomyositis.

The only comorbidity was hypertension for which he was taking a combination tablet consisting of 10 mg amlodipine, 160 mg valsartan and 12.5 mg hydrochlorothiazide. There was no history of non-steroidal anti-inflammatory or illicit drug use.

The patient was admitted to hospital for further work-up. There was no significant albuminuria, and this pointed to another protein in the urine besides albumin. The creatine kinase was significantly raised (8 210 U/L) and there was microscopic haematuria, which suggested that the cause of the proteinuria may be myoglobinuria (Table 1). Urine dipstick detects peroxidase activity of heme, and this is present both in red blood cells and myoglobin. A kidney biopsy was performed to elicit the cause of the proteinuria

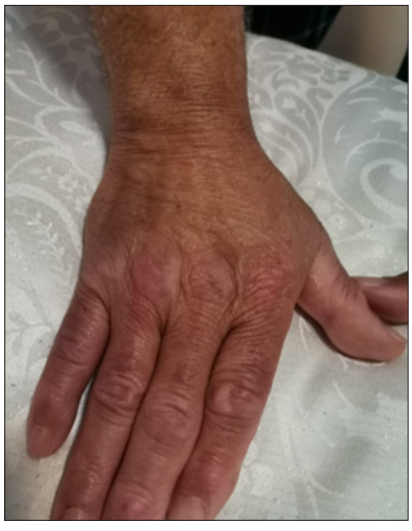

Fig. 1. Gottron's papules.

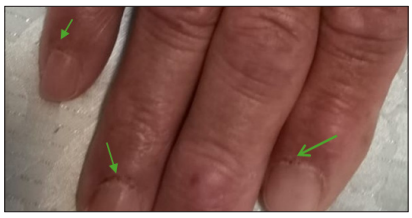

Fig. 2. Periungal telangiectasia.

Table 1. Biochemistry results on presentation and after six cycles of chemotherapy

Blood results	On presentation	Reference range	After 6 cycles of chemotherapy
Na (mmol/L)	136	136 - 145	142
K (mmol/L)	4.2	3.5 - 5.1	3.8
Cl (mmol/L)	97	98 - 107	104
CO_2(mmol/L)	33	22 - 29	29
Urea(mmol/L)	9.1	<8.4	6.8
Creatinine (µmol/L)	105	64 - 104	93
eGFR (CKD-EPI)	67	>90	78
Urine dipstick	1+ protein, 2+ blood	-	-
Urine MCS	1 - 5 red blood cells. No pus cells, crystals or casts.	<5/µL	-
24-h urine creatinine clearance (mg/24 h)	1786	<150	-
Albumin : creatinine ratio (mg/mmol)	4.6	<3.0	-
ANA	Positive >1:2 560	<1:80	-
p-ANCA	Negative	-	-
c-ANCA	Negative	-	-
CK U/L	8210	39 - 308	-
Hb (g/dl)	13.4	13.6 - 17.6	12.9
White cell count (g/L)	6.48	4.31 - 12.39	6.92
Platelets (g/L)	272	175 - 375	203
AST (U/L)	323	<38	-
ALT (U/L)	239	<50	-
DsDNA	Negative	-	-
HepC Ab	Negative	-	-
ESR (mm/h)	29	-	-
Ca(mmol/L)	2.43	2.15 - 2.50	2.32
Total protein (g/L)	62	60 - 82	58
Albumin (g/L)	37	38 - 51	41
SFLC kappa (mg/L)	253.0	6.7 - 22.4	49.6
SFLC lambda (mg/L)	15.8	8.3 - 27.0	10.7
S-kappa/lambda	16.01	0.31 - 1.56	4.64
Immunosubtraction	IgA kappa	-	-
SPEP	M protein-IgA kappa	-	-
Bence Jones protein	Positive	-	-
Mi-2 alpha myositis Ab	Mid positive	-	-
Mi-2 beta myositis Ab	High positive	-	-
TIF-1ɤ myositis profile	Negative	-	-
NXP2 myositis profile	Negative	-	-
SAE1 myositis profile	Negative	-	-

Ab = antibody; ALT = alanine aminotransferase; ANA = antinuclear antibody; AST = aspartate aminotransferase; cANCA = cytoplasmic antineutrophil cytoplasmic antibody; CK = creatine kinase; CKD-EPI = Chronic Kidney Disease Epidemiology Collaboration formula; DsDNA = double-stranded DNA antibody; eGFR = estimated glomerular filtration rate; ESR = erythrocyte sedimentation rate; HepC = hepatitis C; MCS = microscopy; pANCA: = perinuclear antineutrophil cytoplasmic antibody; SFLC = serum free light chain; SPEP = serum protein electrophoresis.

(Fig. 3). The light microscopy showed features of ATN with simplification of the tubular epithelium with loss of the brush border (Figs 3A - 3D). The immunohistochemical myoglobin stain was positive (Fig. 3E and 3F). Congo red stain was negative for amyloid. Immunohistochemistry was negative for kappa and lambda light chains (Fig 3G and 3H).

Laboratory results are shown in Table 1. The clinical, histological and biochemical evidence led to a diagnosis of DM complicated by myoglobin nephropathy.

Further tests were performed to exclude underlying malignancy in the presence of DM in a 58-year-old gentleman. The chest and abdomen computed tomography scans, gastroscopy and colonoscopy did not show any malignancy.

Serum electrophoresis and serum free light chains were performed and showed IgA kappa predominance. The urine was also positive for Bence Jones protein, which are free immunoglobulin light chains filtered in the urine. This also contributed to the proteinuria together with the myoglobinuria. The subsequent bone marrow aspirate and biopsy (Fig. 4A - E) showed an increased number of plasma cells, estimated at 15 - 20% (Fig. 3B, 3E). The majority of the plasma cells were positive for kappa light chain (Fig. 3G).

The patient was diagnosed with IgA kappa smouldering multiple myeloma (SMM) because of a lack of CRAB (hypercalcaemia, renal failure, anaemia and bone lytic lesions) criteria.

Discussion

DM forms part of a group of diseases classified as idiopathic inflammatory myopathies (IIM) and includes polymyositis (PM). DM is characterised by the presence of characteristic cutaneous lesions (heliotrope rash, Gottron's papules, periungal telangiectasia, shawl sign and V sign) with or without proximal muscle weakness. DM is

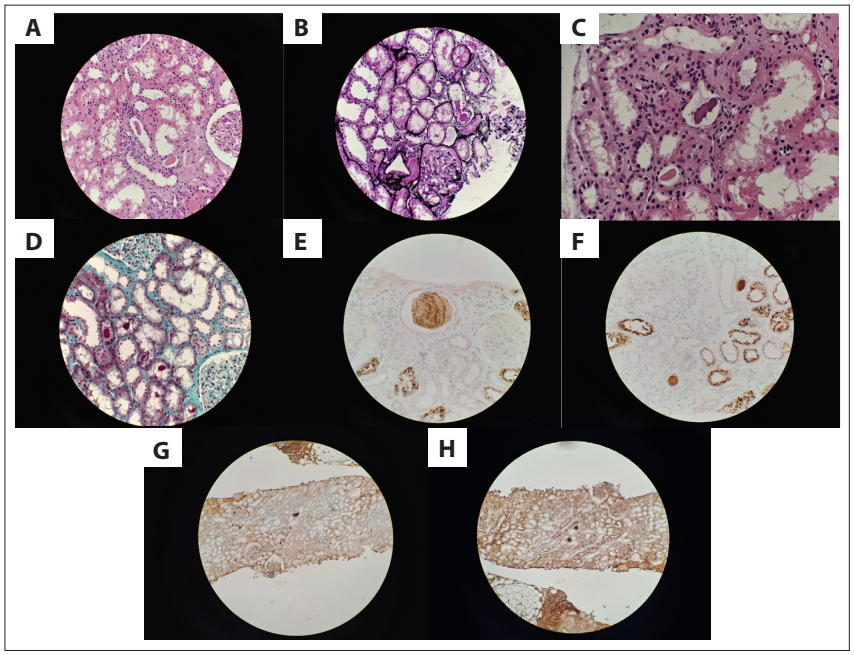

Fig. 3. Kidney biopsy. (A) Light microscopy – haematoxylin and eosin (H&E) stain: ATN with central casts; (B) Jones silver stain: ATN, simplification of tubular epithelium; (C) H&E stain – globular casts; (D) globular casts; (E) myoglobulin staining in tubular casts; (F) myoglobulin resorption droplets in cytoplasm; (G) negative lambda stain; (H) negative kappa stain.

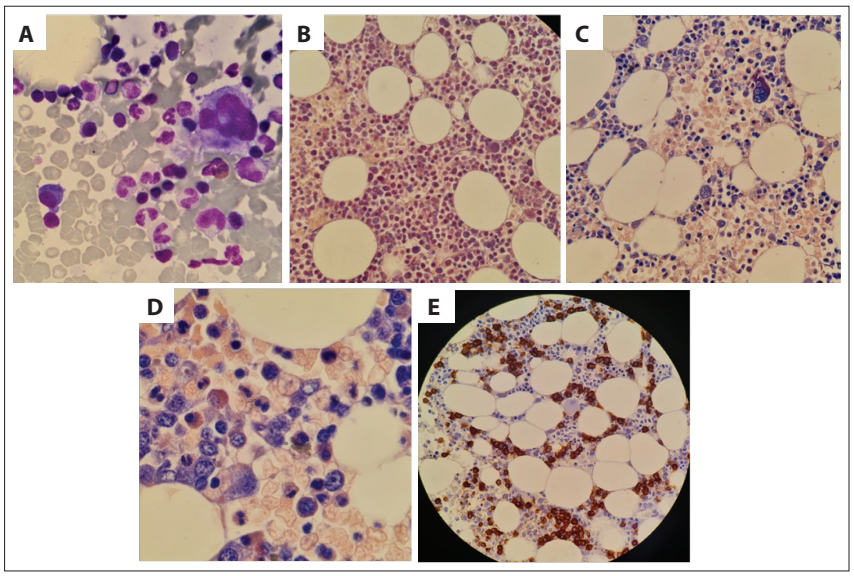

Fig. 4. Bone marrow aspirate and biopsy. (A) Aspirate showing 2 - 4% plasma cells; (B) trephine: Mayer's stain; (C) and (D) trephine: May-Grünwald Giemsa (MGG) stain; (E) immunohistochemistry – CD138 showing increased plasma cells in the interstitium and forming small groups in some areas.

paraneoplastic in up to 25% of cases.[2] The risk of malignancy is similar in myopathic and amyopathic cases.[3] Association between myositis-related antibodies and paraneoplastic dermatomyositis is lower compared with antitranscriptional intermediary factor 1 gamma (TIF-1ɤ) and is categorised as moderate risk for malignancy.[4,5] The current opinion is to treat all adult patients with DM, PM and immune complex-mediated necrotising myositis with the same index of suspicion for malignancy screening as patients with more strongly associated antibody positivity.[5] There is a strong association between TIF-1ɤ, antinuclear matrix protein 2 (NXP-2) and anti-small ubiquitin-like modifier activating enzyme (SAE) antibodies and malignancy-associated myositis.[2,3]

The most associated malignancies are solid-organ tumours, most frequently breast, lung, ovarian and urothelial cancers.[2,3] Haematological malignancies are less common, with only a handful of case reports in the literature. The more commonly associated haematological malignancies are lymphomas.[6] From a large retrospective cohort involving 4 641 patients with MM or monoclonal gammopathy of unknown significance (MGUS), there was a 2.29 times relative risk of developing plasma cell dyscrasia in patients who presented with IIM.[7]

Cases of DM associated with multiple myeloma are rare, with only a few documented case reports. The timeline of these cases is also variable, with the diagnosis of multiple myeloma either preceding or following the diagnosis of DM.[8-10] Multiple myeloma makes up 1.8% of all malignancies; the median age at diagnosis is ~65 years and it is almost two times more common in the black population.[7] This case is the first case in literature documenting paraneoplastic DM as the form of presentation of SMM and it allowed for early detection and treatment to prevent organ damage.

Inflammatory myopathies are associated with malignancies, but DM is significantly more associated than PM. In a cohort study, the prevalence of malignancies was 32% in patients with DM and 15% in patients with PM.[8] Chronic immune stimulation is considered the underlying mechanism that leads to T-cell and B-cell activation, leading to the development of haematological malignancies like multiple myeloma.[5] Crossover immunity between myofibroblasts and tumour cells may reflect the simultaneous presentation of the two diseases.[8]

In this case, the presentation of DM occurred concurrently at the time of diagnosis of SMM. What makes this case interesting is the lack of biochemical or radiological features to suggest plasma cell dyscrasia, and had there not been presentation with DM, the diagnosis of SMM may have been missed. It also emphasises that although haematological malignancies are less commonly associated with paraneoplastic DM, screening for haematological malignancies should be routine, together with other investigations. The revised international working group diagnostic criteria for multiple myeloma facilitates the early diagnosis of plasma cell disorders to allow for early treatment to avoid progression to end-organ damage. [11] This case serves as a clear example of its utility in early diagnosis. This patient was referred to a haemato-oncologist and received six cycles of bortezomib/leflunomide and dexamethasone. His most recent blood results are shown in Table 1.

A key point of this case study is that the reason for the referral was proteinuria, and this serves as a reminder of the core principles of history taking and clinical

examination. This prompted further investigation into the type of protein in the urine. This case was referred to a nephrologist and highlights how urine analysis can be the harbinger of diagnosis of a systemic illness and must be performed routinely in the form of a simple bedside urine dipstick test – its value should not be underestimated.

Declaration. Patient anonymity was maintained, and written informed consent was obtained from the patient.

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Conflict of interest. None.

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