QUIZ SECTION

An Elderly Man with Facial Oedema and a Photosensitive Rash: A Quiz

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An 81-year-old male patient was diagnosed with adenocarcinoma of the prostate in 2020, characterized by focal neuroendocrine differentiation. Hormonal therapy with leuprorelin was initiated for bone metastases. Due to disease progression, the patient was subsequently treated with docetaxel and zoledronic acid. In February 2022, he commenced second-line therapy with enzalutamide. In April 2022, the patient developed muscle weakness and a localized erythematous rash on the face and neck, in addition to a violet rash on the dorsal surface of he hands. A skin biopsy revealed interface dermatitis consistent with lupus erythematosus. Screening for antinuclear antibodies (ANA) was positive, while serum creatine kinase (CK) levels remained within normal limits (Table I). Due to a suspicion of drug-induced cutaneous lupus, the oncology department discontinued enzalutamide. Palliative radiotherapy was initiated due to tumour infiltration into the bladder. A month later he was referred to a rheumatologist due to generalized muscle weakness, slurred speech, and swallowing difficulties necessitating a feeding tube. Clinical examination revealed heliotrope discoloration, Gottron's papules, Shawl sign, and significant swelling of the hands and fingers (Fig. 1A and B). Laboratory tests indicated elevated serum CK levels of 583 IU/l (Table I). The case was discussed with the rheumatology department at a university hospital, where a general sarcopenia was suspected, and no further diagnostic tests were suggested. By January 2023, the patient was classified as terminally ill, with recommendations for continued follow-up by his general practitioner. Due to persistent facial and extremity oedema, the general practitioner suspected angioedema, and referred the patient to teledermatology by the local dermatologist, who was provided the patients' medical history and photographs (Fig. 1C and D).

Table I. Laboratory investigations with autoantibodies and creatine kinase

Laboratory investigations	July 2022	October 2022	November 2022	February 2023
Creatine kinase (IU/l)	133		583↑	133
U1 snRNP A-IgG		1.1↑	0.8	0.5
Anti-Ro/SSA-52 kDa Ro protein IgG			531↑	> 1,685↑
Anti-Ro/SSA-60 kDa Ro protein IgG	0.3	< 0.2		
DNA topoisomerase 1-antibody (IgG)		> 8.0↑	1.4↑	5.7↑
ENA7-screening				Positive
ANA-screening	Positive			
TIF1-gamma-IgG			Positive	Positive
Rheumatoid factor				85
E3 ubiquitin-protein ligase TRIM21-antibody (IgG)				>8

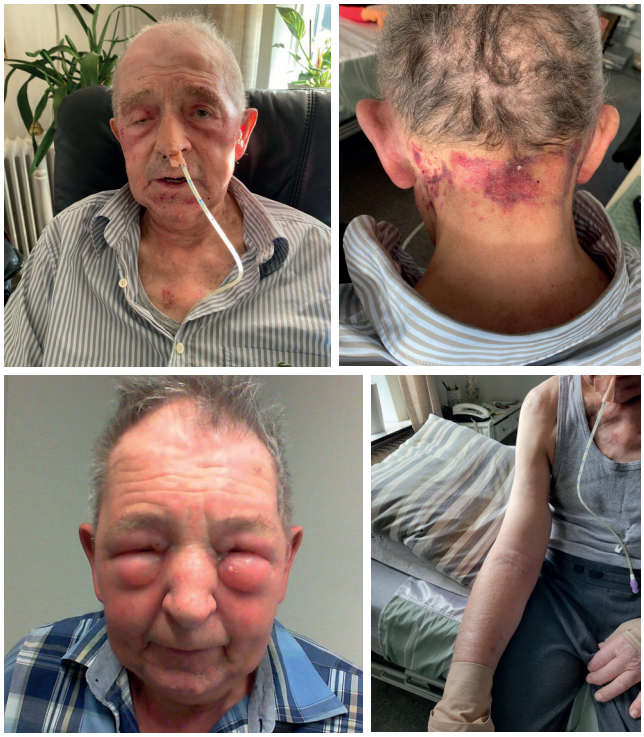

Fig. 1. (A) Heliotrope rash and muscle weakness with difficulty swallowing necessitating a feeding tube. (B) Shawl sign on the neck. (C) Periorbital oedema and heliotrope rash. (D) Oedema of the upper extremity. The patient and his next-of-kin both provided written permission to publish the photos.

What is your diagnosis?

Differential diagnosis 1: Systemic lupus erythematosus

Differential diagnosis 2: Paraneoplastic dermatomyositis

Differential diagnosis 3: CREST syndrome

Differential diagnosis 4: Polymyositis

See next page for answer.

ANSWERS TO QUIZ

An Elderly Man with Facial Oedema and a Photosensitive Rash: A Commentary

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Diagnosis: **Paraneoplastic dermatomyositis**

Upon evaluation, the dermatologist suspected cancer-associated dermatomyositis, which was corroborated by a strongly positive TIF1-γ-IgG antibody test. Following the teledermatological consultation, the patient was started on oral prednisolone 37.5 mg OD, resulting in notable improvements in oedema, muscle strength, and overall condition. Given that the patient had remained professionally active until the previous summer and exhibited significant vitality, he was referred to a specialized department with expertise in dermatomyositis. A subsequent muscle biopsy confirmed the diagnosis; however, the patient regrettably passed away before further treatment could be initiated.

Dermatomyositis is an inflammatory myopathy characterized by muscle weakness and distinctive skin manifestations, including heliotrope rash, Gottron's papules, and Shawl sign. Although rare, both facial and generalized subcutaneous oedema have been reported in the literature, in some cases as the presenting symptom (2, 3). Dermatomyositis can occur as a primary idiopathic disorder or as a paraneoplastic syndrome associated with underlying malignancies (1). Treatment for dermatomyositis typically involves corticosteroids as first-line therapy, which can be effective in reducing muscle inflammation and improving strength. In cases associated with malignancy, the management may also necessitate addressing the underlying cancer, as successful treatment of the malignancy can lead to improvement in dermatomyositis symptoms. Immunosuppressive agents, such as azathioprine, methotrexate, and mycophenolate mofetil, are second-line therapies (4).

In this case report, we observed a unique presentation of dermatomyositis associated with prostate cancer, which is rarely seen (5). Furthermore, the case highlights the complexities of diagnosing paraneoplastic dermatomyositis and the challenges by overlapping clinical and histological features of cutaneous lupus erythematosus (1). Paraneoplastic dermatomyositis is particularly noteworthy as it can serve as an important clinical marker for the presence of malignancy (1). In this patient, the occurrence of dermatomyositis symptoms coincided with the diagnosis of prostate cancer and the initiation of treatment with enzalutamide. While the initial interpretation of the skin manifestations suggested drug-induced lupus erythematosus, the subsequent identification of TIF1-γ-IgG antibodies pointed towards a diagnosis of paraneoplastic dermatomyositis (6). This underscores the crucial need for clinicians to maintain a high index of suspicion for dermatomyositis in patients with malignancies presenting with skin changes and muscle weakness. The relationship between dermatomyositis and cancer is complex and multifaceted (1, 6). Certain autoantibodies, such as TIF1-γ-IgG, have been associated with specific malignancies, including breast, ovarian, and haematological cancers, but their association with prostate cancer is less well established (6, 7). This case contributes to the growing body of literature suggesting that dermatomyositis, while rare, can occur in association with prostate cancer, particularly in older patients. The clinical presentation in this patient, featuring classic dermatomyositis signs following sun exposure and exacerbation after radiotherapy, aligns with previously reported cases where tumour antigen release post-treatment may trigger autoimmune responses (8).

The patient's journey included 13 different specialties, illustrating the challenges faced by an elderly patient. The "pinball effect", where patients are bounced between multiple specialties without a coherent care plan, can lead to delays in diagnosis and treatment. In this case, the patient's extensive medical history and the involvement of numerous specialists complicated the timely identification of dermatomyositis. It highlights the importance of effective communication and collaboration among healthcare providers to ensure that patients receive a comprehensive and coordinated approach to their care (9). The integration of telemedicine, particularly teledermatology, emerged as a pivotal tool in this case (10).

Dermatology is a unique medical specialty characterized by its visual nature, which relies heavily on the observation of cutaneous signs and symptoms to guide diagnosis and treatment. Lengthy waiting times associated with in-person consultations have made the field particularly suitable for the integration of telemedicine solutions. Initially, teledermatology was primarily utilized for the management of minor dermatological conditions, such as urticaria, acne, eczema, psoriasis**,** and fungal infections. However, the potential of teledermatology extends far beyond the management of common skin issues, as it can also play a critical role in the diagnosis and management of rare diseases, as evidenced by this report. This case exemplifies how telemedicine can bridge gaps in care, especially for patients with complex medical histories who may struggle to navigate the healthcare system. In conclusion, this case report underscores the significance of recognizing dermatomyositis as a potential paraneoplastic syndrome in patients with malignancies, including prostate cancer. The overlapping clinical features with other conditions, such as drug-induced lupus, necessitate a thorough and nuanced approach to diagnosis. Furthermore, the challenges faced by elderly patients in accessing care highlight the need for improved communication and collaboration among specialists. The utilization of telemedicine presents a promising avenue for enhancing patient care and ensuring that complex cases are managed effectively. Future research should continue to explore the relationship between dermatomyositis and various malignancies, as well as the role of telemedicine in optimizing patient outcomes in dermatological practice.

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