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Case Report

Unexplained Coma with Linear Skin Papules: A Diagnostic Trap Lest We Forget! A Case Report

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ABSTRACT

Dermatoneuro syndrome presents with a sudden onset of coma of unknown origin, often in the setting of mucinosis. The linear array of skin-coloured papules is often the only clue to the diagnosis, and unless promptly, intravenous immunoglobulin at the dose of 2 g/kg divided over 5 days is started, most patients have a grim prognosis. The importance of recognition of this entity lies in the differential diagnosis of coma of unknown origin. Often, dermatologists are called after a significant number of days have lapsed since the patients have entered into coma, resulting in significant mortality. Sadly, all imaging studies and toxic screen/metabolic screen/autoantibody screen are normal. Herein, we report an unusual case of dermatoneuro syndrome secondary to scleromyxedema which went undiagnosed for a long duration owing to a lack of awareness of this entity amongst other specialities (such as neurology, where patients usually first present with coma).

Keywords: Dermatoneuro syndrome, Mucinosis, Scleromyxedema

INTRODUCTION

Dermatoneuro syndrome is a rare but life threatening manifestation of Scleromyxedema. Herein we report a case of a 50 year old man with unexplained coma which turned out to be dermatoneuro syndrome.

CASE REPORT

A 50-year-old man with no known comorbidities presented to the emergency department of a tertiary care centre with a sudden onset of deep coma and drowsiness of 2-day duration. The patient had a history of a similar episode 4 years ago, where he was in a coma for a duration of around 10 days; however, no definite diagnosis could be made at that time. The current episode had started with a history of fever with joint pains for 4 days. There was, however, no history of photosensitivity, oral ulcers or any drug intake before the current episode. There was no history of thyroid disorders, hypertension or type 2 diabetes mellitus. A meticulous history revealed that he had been experiencing skin tightening and development of skin-coloured dome-shaped papules gradually for the past 4 years; however, the patient did not seek medical attention for the skin lesions.

The Glasgow Coma Scale of the patient was E: Eye movement, V: Vocalisation, M: Muscle (E1V1M1). Metabolic workup for the coma was non-contributory. Imaging scan (computed tomography scan of the brain and magnetic resonance imaging) was normal. Polymerase chain reaction screening of the cerebrospinal fluid tap was negative for tubercular and herpetic aetiology.

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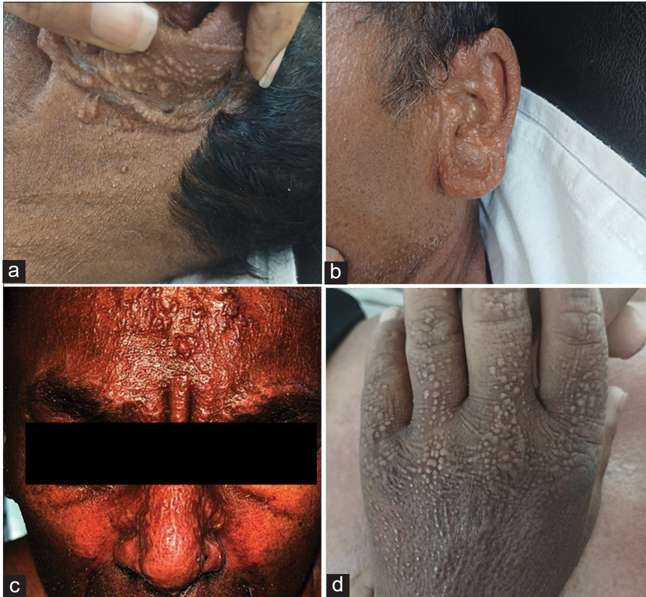

Figure 1: (a) Dome-shaped skin-coloured papules on the back of ear; (b) A few papules on the ear arranged linearly; (c) Linear deep forehead furrows suggestive of Shar pei sign - Also note a few sclerotic bands near the nose; (d) Linearly arranged papules on sclerotic skin on the dorsum of hand.

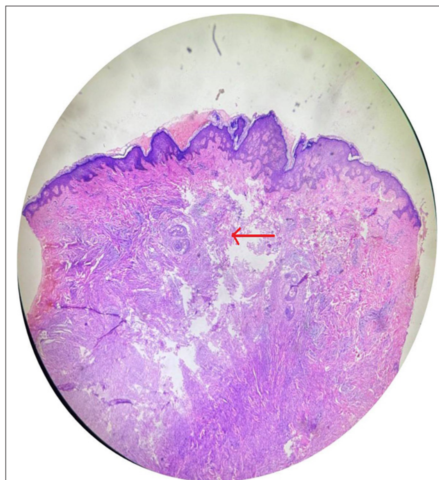

Figure 2: Histopathological section shows stringy basophilic deposits in the middle to deep dermis (×10 magnification, haematoxylin-eosin stain). Bluish stringy deposits of mucin in mid dermis (red arrow).

Viral encephalitis screen was also negative. An antinuclear antibody profile done by indirect immunofluorescence was also normal. Urine electrophoresis for abnormal light and heavy chains of Bence Jones protein was normal. Toxin screen of the serum was also normal. Thyroid screen was also normal. Skin examination revealed multiple dome-shaped skin-coloured papules arranged in linear arrays on the back of fingers, ears and trunk on the background of oedematous and doughy skin with mild background erythema. The skin of the forehead showed linear furrows. A skin biopsy showed fibroblastic proliferation with mucin deposition and some sclerosis.

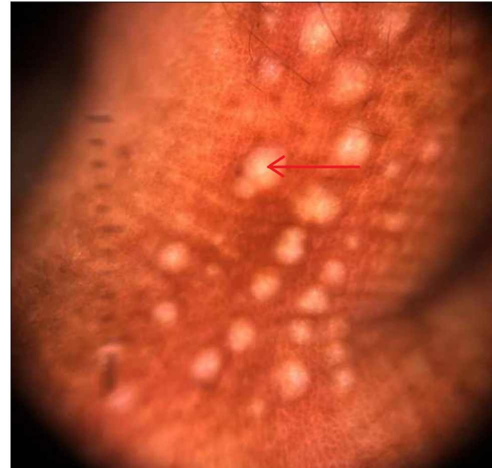

Figure 3: Non-contact polarised mode dermoscopy with DL-4 dermLite shows linear white structureless areas from one of the papules. White structure less areas seen on polarized dermoscope (red arrow).

Based on these findings [Figures 1-3], a diagnosis of dermatoneuro syndrome due to scleromyxedema was made and the patient started on intravenous immunoglobulins. He gradually recovered and could be extubated by 10th day of illness. A follow-up scheduled 1 month later revealed that he was well and off all medications, although the linear papules had persisted. He was scheduled for narrowband Ultraviolet B (UVB) for the skin lesions.

DISCUSSION

Dermatoneuro syndrome is an extremely rare yet a life-threatening manifestation of scleromyxedema.[1] This presents as a sudden onset coma with a short prodrome of fever and joint pain, often mistaken for viral infections. The only clue to the diagnosis is the presence of skin-coloured tiny papules, which are often grossly overlooked.[2] No metabolic or imaging workup can diagnose this disease, other than a routine skin biopsy from these papules. The aetiology of dermatoneuro syndrome is mysterious – often believed to be due to excess mucin deposition in the cerebral microcirculation, which interferes with axonal transport and thereby produces microvascular occlusion inside small vessels of the brain. Early institution of intravenous immunoglobulin at the rate of 2 g/kg for 5 days can be life-saving. Other therapies tried include cyclophosphamide and methotrexate but their slow onset of action makes

them rather ancillary in the management of dermatoneuro syndrome.[3] Unfortunately, the limited awareness of this extremely rare entity amongst internal medicine experts makes this a diagnosis nearly always a fatal condition!

CONSCLUSION

Dermatoneuro syndrome is a life threatening manifestation of systemic sclerosis which should be kept in the differential diagnosis of come of unknown origin

Ethical approval: Institutional Review Board approval is not required.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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