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Case Report

Case report of Sleromyxedema with sensory motor axonal polyneuropathy treated with immunoglobulin therapy

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ABSTRACT

Sleromyxedema is a rare disease characterised by extensive mucin deposition with fibrosis in dermis. It is often associated with monoclonal gammopathy. Its true prevalence and/or cause is unknown due to its rarity. Various extra-cutaneous manifestations can be associated with sleromyxedema such as dermoneuro syndrome, neuropathy, rheumatological problems or various malignancies. This case report showed that a patient with sleromyxedema and neuropathy had positive outcomes when treated with intravenous immunoglobulin (IVIg) and steroids pulses. The underlying cause of the patient's condition was unknown.

1. Introduction

Sleromyxedema is a rare condition with extensive mucin deposition along with fibrosis in dermis. It is often associated with monoclonal gammopathy. Its exact prevalence and definitive treatment is not known and available literature is only in the forms of case reports. Hereby we are discussing a case report of Sleromyxedema with sensory motor axonal polyneuropathy which was successfully treated with immunoglobulin therapy and steroids.

2. Case Report

A 52-year-old male, who was businessman, chronic smoker, and resident of New Delhi, presented with complaints of burning and needle like sensations all over the body for the last 6 years and generalised skin tightening for the last 5 years. The patient was completely asymptomatic 6 years prior to experiencing burning and needle like sensations over the skin of right ante-cubital region, which was initially mild and progressed and spread to right upper limb below elbow the over a period of 7 days. The left upper limb became involved in similar pattern to the right upper limb as well as lower abdomen around umbilicus over the next 2-4 weeks. The lower extremities became involved over next 2 months. Then over next 2-3 months the patient noticed the abnormal sensations all over the body. The symptoms gradually progressed from bilateral upper limbs to lower limbs in a proximal to distal manner. The patient started feeling numbness in his both 4^{th} and 5^{th} digits2-3 times daily which resolved by the patient shaking hands. [1–6]

The patient started experiencing swelling with redness over the skin involving both upper limb and lower limb with shinning of skin and stiffening of skin after one year of being diagnosed with the condition. It becomes difficult for the patient to bend his trunk and joints. The patient also experienced difficulty with walking and weakness of hand grip due to tightness. There was no history of Raynaud's disease, digital ulcers, arthritis, sicca symptoms, skin papules, dysphagia,

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dyspepsia, hair loss, epistaxis, haematuria, claudication, foot /wrist drop, proximal muscle weakness, weight loss, night sweats, seizures, LOC, nor any history suggestive of cranial nerve involvement. Before coming to us, the patient had received treatment at multiple places, both allopathic (steroids, azathioprine, methotrexate and pregabalin, methylcobalamin, gabapentin and duloxetine) and alternative medicine without any improvement. There were no history of diabetes, hypertension, coronary artery disease (CAD), cerebrovascular accident (CVA), or tuberculosis (TB). There were no similar complaints in family or significant family medical history. The patient is chronic smoker who smoked 2-3 packs of bidi daily, and stopped alcohol drinking 6 years before. There was no illicit drug abuse or high-risk behaviour. The patient's appetite, bowel, bladder and sleep were normal.

On examination the patient's vitals were normal, all peripheral pulses palpable, and no postural variation drop in blood pressure, no pallor, icterus, lymphadenopathy, clubbing, nor cyanosis. Shiny skin was present in hands, feet, legs and arms with puffiness without any pitting edema figure 1), Biopsy scar present over lateral aspect of left elbow. His respiratory, cardiovascular, abdominal, and musculoskeletal examination was normal, except for disturbed gait and decreased range of joint moments due to skin tightness. On neurological examination, the patient was conscious, oriented and cooperative. Muscle strength was 4/5 in distal groups of muscles both in arms and legs. Tone and reflexes were normal. The patient reported positive sensory symptoms (needle like and burning sensation), all over the body below neck without any area of sensory loss. There were no cerebellar signs and abnormal movements.[7–10]

All routine investigations were normal. Autoimmune work up including ANA, ENA, ds-DNA, SCL-70, RO/LA, RF, ANCA, Anti-CCP, were all negative, along with TSH, cryoglobulin levels, CPK, LDH, ESR, CRP, vitamin B12, Folic acid all were in normal range. Viral profiles HBsAg, Anti-HCV, HIV were negative. Imaging including triple phase bone scan, positron emission tomography (PET) Scan, Contrast enhanced computed tomography (CECT) chest and abdomen, MRI brain and spine,2D echo, x-ray hand all were negative, prostate specific antigen (PSA) negative, serum protein and urine protein electrophoresis were negative, Bence Jones protein(BJP) negative. Nerve conduction velocity (NCV) initially showed abnormal sudomotor response, after few years shows sensory motor axonal neuropathy. Electromyography (EMG) was normal. Abdominal fat pad biopsy was normal, sural nerve biopsy shows mild patchy demyelination without any evidence of vasculitis or amyloidosis. Skin biopsy showed cutaneous mucinosis with panniculitis compatible with Sleromyxedema. Based on histopathology and clinical features, the patient was diagnosed with Sleromyxedema with sensory motor axonal polyneuropathy and therapy with intravenous immunoglobulin (IVIg) was initiated with minimal improvement after first infusion.

After 3 infusions there was around 20-30 percent resolution of symptoms. The patient was treated with immunoglobulin (IVIg) for total 6 months and did not experience symptoms relapse during his 6 months follow up. Repeated NCV testing 3 months and 6 months showed significant improvement with persistence of very mild peripheral neuropathy.

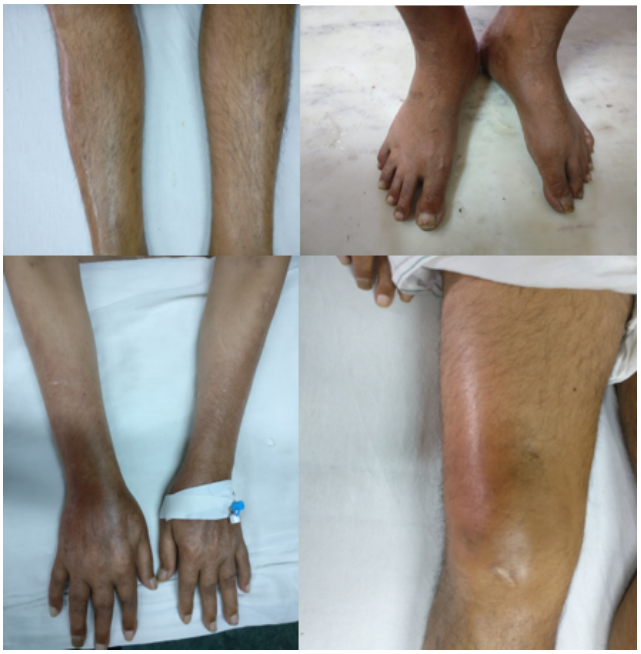

Fig. 1: Shiny skin of hands, feet, legs and arms with puffiness without any pitting oedema.

3. Discussion

Sleromyxedema is a rare disease characterised by extensive mucin deposition with fibrosis in dermis. Mostly it is associated with monoclonal gammopathy but not always necessary. Due to its rarity its true prevalence is not known. Various extra-cutaneous manifestations can be associated with it like dermoneuro syndrome, neuropathy,[1] rheumatological problems or various malignancies.

It is considered as a one of the rare manifestations of para-neoplastic disorder, mostly haematological origin. This can precede or coexist with the primary malignant disorder and it is a, chronic, progressive manifestation of the Lichen myxedematous (LM) disorder groups. This case report showed that a patient with sleromyxedema and neuropathy had positive outcomes when treated with intravenous immunoglobulin (IVIg) and steroids pulses.[2–4] According to this case report, IVIg should be considered for treatment of sleromyxedema with neurological involvement.

There are very few case reports of use of IVIg in treatment of Sleromyxedema[5–10] and neurological manifestation are rarely reported.[11–14]

4. Conclusion

Intravenous immunoglobulin (IVIg) and steroids can be tried in the difficult to treat sleromyxedema cases.

5. Source of Funding

None.

6. Conflict of Interest

None.

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