Scleromyxedema In The Setting Of Inpatient Rehabilitation: A Case Report

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Introduction

Scleromyxedema is a rare skin disorder characterized by abnormal accumulation of mucin in the skin, resulting in papular and sclerodermoid bumps and eventual fibrosis and sclerosis. Currently, there is no standardized treatment and the exact cause of the disease is not known. Usually, this disorder affects those between the ages of thirty to fifty. Symptoms can include dysphagia, hoarseness, polyarthritis, proximal myopathy, cardiac abnormalities, eye abnormalities, difficulty breathing, and neurological dysfunction. The following is a case report of a fifty three year old male with scleromyxedema that was admitted to inpatient rehabilitation for higher level of care and facilitation of rehab needs with the ultimate goal to transition to home with 24/7 caregiver support from his family.

Case Description

The patient had a past medical history of non-toxic unimodular goiter, left hip osteoarthritis, and was a former Iron Man athlete. Prior to admission, he presented to his primary care physician with symptoms of chest and facial skin tightness and decreased appetite. He was admitted to the hospital, where he became progressively fatigued with more diffuse skin tightening, dysphagia, weight loss, lower extremity swelling, and muscle weakness. Eventually he underwent gastric tube placement for feeding due to significant dysphagia and weakness. Initially he was diagnosed with scleroderma, however he had a skin biopsy which further revealed scleromyxedema.

Further lab workup also revealed low level of Immuglobulin G-kappa monoclonal protein, suggestive of an associated monoclonal gammopathy. Thyroid levels at that time of diagnosis were within normal limits. He was also noted to have persistent tachycardia, low albumin levels, elevated creatine kinase, platelet, and c-reactive protein, although it is unclear if this was related to his disease process or an additional inflammatory process. Echocardiogram revealed ejection fraction of 55-60% with moderate mitral regurgitation and left moderate pleural effusion, but normal diastolic and systolic function. Doppler of the lower extremities was negative for a deep vein thrombosis. He was followed by rheumatology, dermatology, and hematology/oncology for his scleromyxedema, receiving scheduled chemotherapy with intravenous immunoglobulin (IVIG) and bortezomib, cyclophosphamide, and dexamethasone combination treatment. He was eventually stabilized and due to his significant proximal muscle weakness and decrease with independence of activities of daily living, patient was admitted to the inpatient rehabilitation team.

Inpatient rehabilitation team based approach can potentially **increase quality of life** in patients with **scleromyxedema**.

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Assessment/Results

He spent two weeks in the inpatient rehabilitation unit, where he was receiving multi-disciplinary rehab team approach. The rehabilitation team included physiatry, psychology, nutritionist, physical, occupational, recreational, and speech therapy. Over the course of two weeks he had made mostly minimal gains with his Functional Independence Measure (FIM) scores. However, despite this, the patient had reported that his quality of life improved. One aspect of improving his quality of life came from the nutritionist and dietitians that helped to individualize his diet, while meeting his caloric needs, and helping him be more independent with feeding to the point where through multiple therapy sessions he was able to how to administer his bolus tube feeds and flushing his gastric tube on his own. Having his family involved was crucial as well, and added to his motivation to participate with therapies so he could reach his disposition goal of living with his family. In addition, utilizing the appropriate assistive devices, such as a platform walker, provided comfort, ease of care and mobility. He was able to ambulate with one to two person assist for safety. Due to plateau of his FIM scores after two weeks of inpatient rehabilitation, he was eventually discharged to a community living center with the goal to transition to home with family.

Discussions/Conclusions

Scleromyxedema is a rare progressive skin disorder that can have extra-cutaneous involvement, making those affected severely debilitated, including: neurologic complications such as encephalopathy and seizures, gastrointestinal dysmotility and malabsorption, rheumatological manifestations such as joint contractures and muscle weakness, and cardiovascular and pulmonary complications like pulmonary hypertension and restrictive or obstructive lung disease. There is often time a high mortality rate due to respiratory complications. While there is no standard treatment, according to the literature, intravenous immunoglobulin has usually been used as initial treatment. Plasmapheresis can be a short-term treatment option. Other treatments have included thalidomide, systemic glucocorticoids, and in those without an adequate response, autologous bone marrow transplantation, melphalan, and bortezomib with dexamethasone have been used. There are case reports suggesting clinical improvement with chemotherapeutic agents such as cyclophosphamide.

Scleromyxedema can present with multiple challenges in the rehab settings that can make functional progress difficult. Ultimately, this patient did not improve in the inpatient rehabilitation unit based on his FIM scores. However, he reported that his quality of life improved with combination of medical treatment for his scleromyxedema and its complications and extra-cutaneous manifestations, and the multi-disciplinary approach from the inpatient rehabilitation team. While there is limited data for effectiveness of the current treatments due to the rarity of this disease, quality of life can still potentially be improved by using this team based approach.

There are no disclosures with this case report.

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