The mystery of hypoproteinemia and shock in a case of steatocystoma multiplex scrotum and subsequent multiorgan dysfunction syndrome.

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Abstract -- Thyroid is one of the vital hormones of the body. Myxedema is a rare and fatal condition associated with severely decompensated thyroid hormone. Thyroid hormone effects all the organs of the body adversely. Diastolic hypertension is one the cardiac compensatory mechanisms in hypothyroidism along with torsade's de pointes, hyperventilation and increased risk of bleeding are also manifestations. This case is a complex interplay between various organ systems as a result of thyroid hormone malfunction. The uncorrected hypoproteinemia and steatocystoma further complicating the situation leading to multiorgan dysfunction syndrome.

Key words -- Thyroid, steatocystoma multiplex, multi organ dysfunction syndrome, hypoproteinemia

Introduction:
Steatocystoma multiplex is a rare genetic infectious disease in which pilosebacious ducts get infected. The symptoms with which patient presented is quite typical and the progression was uncertain revealing the malfunction of organ systems one by one at every stage of investigation. The cause of hypoproteinemia remain uncovered even after supplementation and correction of protein levels due to kidney injury making the treatment even worse. Hypoproteinemia remains the marker for severity of sepsis in many researches. In this case patient has loss of protein in urine and probably suspected nutritional deficiency of albumin which was corrected by giving bolus and infusion of albumin to which patient’s response was not satisfactory. When the level of albumin reached to the extent that its further administration would lead to volume overload and pulmonary edema its further administration was stopped. Severe hypothyroidism led to deranged mental status hypothermia and slowing down of multiple organs in the body leading to multiorgan dysfunction syndrome.

Case presentation:
A 45-year-old male patient who is a farmer by occupation belonging to low socioeconomic class came to the clinic with a chief complaint of chest pain since 2 days which is of stabbing type radiating to left arm aggravated on physical activity and relieved on taking proton pump inhibitors. An electrocardiogram was taken which showed ST segment elevation and so the patient was referred to higher Centre to which patient presented with complaints of pedal edema which was of non-pitting type since 4 months on and off aggravated since 5 days, burning micturition since 4 days not associated with increased or decreased frequency of micturition and vomiting since two days which is non bilious non projectile non blood stained and content is food particles 4 to 5 episodes per day. Patient has no complaints of cold, cough, fever, shortness of breath, palpitations, abdomen pain, distension, loose stools or bleeding manifestations. Patient has a history of septic shock with metabolic acidosis 4 months back and had a history of blood transfusion and feroxuscarboxymaltose transfusion to which patient developed itching all over the body. Patient was a known case of diabetes since 5 years and is not a known case of hypertension, asthma, epilepsy, coronary artery disease or cerebro vascular accident and tuberculosis. Patient underwent septal drainage few months back. Patient takes mixed diet, normal sleep cycles and decreased appetite since one year. Normal bladder habits. He is an alcoholic since 15 years and chewed tobacco for one year and stopped this habit 4 months back. He is Allergic to iron apart from which no known drug or food allergies were present. On general examination patient was conscious, coherent well oriented to time place and person. signs of pallor were seen with red beefy and bald tongue. Edema of pitting type was present. No signs of cyanosis, clubbing and lymphadenopathy. Vitals were within normal limits apart from grbs which was recorded as 186 mg/dl. Hyper pigmented scaly skin was noted over bilateral lower limbs. Cardiovascular, respiratory and central nervous system examination along with gastrointestinal examination was normal. Further investigations were based on the examination findings and history. To rule out anemia serum iron ferritin and TIBC was done along with a haemogram.Elevated serum ferritin with normal TIBC was found implying inflammatory causes which lead to its raise in blood. Haemogram showed normocytic normochromic anemia with reticulocytosis and elevated esr and normal platelet count. Viral markers were as a part of routine examination and were found to be non-reactive. Serum calcium bicarbonate was deceased while uric acid and phosphorous were found to be elevated.INR was on the higher side while PT and APTT were normal. Urine examination showed proteinuria with pus cells and macrocytic hematuria which made us raise a doubt of urinary tract infection. Hbaic was high suggesting of diabetes. serum creatinine and chloride levels were on the higher side spot urine protein creatinine ratio peaked to 1.29. Thyroid stimulating hormone was raised with deceased serum T3,T4 levels showing hypothyroidism liver function tests showed hypoalbuminemia with elevated globulin levels. Ecg and symptomatology of chest pain suggested of coronary syndrome. Patient had features of abnormal sensations in hands and feet suggesting diabetic neuropathy. Red cell casts in urine suggested acute kidney injury and altered renal function tests confirmed it. Fundal examination was done which revealed the presence of hard exudates, cotton wool spots and hemorrhages with decreased cdr ratio. Patient was admitted into the intensive care
Patient complained of multiple itchy lesions over the scrotum and testicular swelling associated with pain since 20 days a dermatology referral was done which revealed hypo pigmented nodules on the scrotum and was diagnosed with steatocystoma multiplex scrotum. A 2d echo was done which revealed changes of later wall myocardial infarction. Ultrasound revealed bilateral grade 2 renal parenchymal changes along with cystitis changes and minimal ascites. Patient landed in shock secondary to myxedema and was shifted to cardiac intensive care unit. Treatment regimen began with antibiotics, antifibrinolytics and anticoagulants along with statins and intravenous bolus of albumin was given. Medications included inj ceftriaxone twice daily one gram intravenously, inj pantoprazole, inj odoncetron, inj heparin, tablet ecosprin, tablet clopidogrel, tablet atorvastatin, tablet orofer, tablet thyronorm and injection optinuerin was given. 24 hours after admition patients blood pressure and sat trembled and was put on nor adrenaline to which patient responded well. Patient developed rash to heparin and potassium was given. Magnesium sulfate dressing was given. Next day patients saturation became unstable and was put on nor adrenaline again but inspite of efforts patient’s condition deteriorated and he was declared dead due to severe myocardial infarction.

Discussion:

Myxedema is difficult to recognise, but early treatment is imperative. Management is three-fold and consists of rapid thyroid hormone replacement, supportive measures and the treatment of coexisting problems, most importantly infection. This is most effectively done in a critical care environment, at least in the acute phase. If stress factors are identified in such patients, the diagnosis of myxedema coma in anyone with shock and multiple organ failure should be considered. Prompt treatment with thyroid hormone replacement and diligent supportive therapy following the diagnosis of myxedema coma can reduce mortality and morbidity.

Acknowledgment:

No sponsors

References:

5. Davey MA, Burkhart CN, Morrell DS. Steatocystoma multiplex: Treatment and medication. Available from: