Myxedema Ascites: A Rare Case Report
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Abstract
Congenital hypothyroidism is a very common condition in developing world, but myxedema ascites is very rare in pediatric population. We report a case of 13 years old girl with congenital hypothyroidism on irregular thyroxin started on second half of infancy, presented with recurrent ascites excluding all other causes and discuss different aspects of the condition on the basis of literature review.

Key words: Hypothyroidism, Myxedema, Ascites

Introduction
Myxedema is a term used synonymously with severe hypothyroidism. It refers to deposition of mucopolysaccharides in the dermis, which results in swelling of the affected area. One manifestation of myxedema occurring in the lower limb is pretibial myxedema, although it is most often confined to the pretibial area, it may occur anywhere on the skin, especially the ankle, dorsum of the foot, knees, elbows, upper back.¹ Myxedema ascites was first described in 1886 and occurs in 4% of total and less than 1% of newly diagnosed cases in adults.² Exact incidence in pediatric patients is not clear. Severe, uncontrolled hypothyroidism can lead to accumulation of fluid in body cavities, presenting as pericardial effusion, pleural effusion and ascites. Hypothyroidism is also an uncommon cause of ascites and is seen in only one to five percent of patients with ascitis.³ Treatment of underlying hypothyroidism leads to quick clinical improvement.⁴

Case Report
A 13-year-old girl known case of congenital hypothyroidism on irregular thyroxin started on second half of infancy presented with swelling of both legs and abdomen for 1 month and fever for 3 days. She had similar history 4 times before within last 2 years. Her urine output was normal every time she had swelling. There was no history of abdominal pain, jaundice, abnormal stool color, breathing difficulty, recurrent infection, rash, joint pain or swelling. She attained the developmental milestones quite delayed with below average IQ. She has not yet experienced menarche with no secondary sexual characteristics. Other 2 siblings are healthy with no such family history. On examination, she had dull appearance with frontal bossing, depressed nasal bridge and dry, rough wrinkled skin. She had bipedal edema with ascites but no organomegaly and vital signs were within normal limit. There was distant heart sounds and delayed relaxation of ankle jerks. Her weight and height were below 3rd centile.

Investigations revealed Hb 8.3 gm/dl with neutrophilic leukocytosis, Liver function tests were normal except Serum albumin, which was 1.9 gm/dl. Urine R/M/E & C/S and 24 hours urinary total protein were normal. Serum cholesterol, renal function test and chest X-ray were normal. Serum FT4 was low (0.28ng/dl) along with high TSH (>100 µIU/ml). Serum Ascitic Albumin Gradient (SAAG) was <1.1. ECG revealed low voltage reading with mild pericardial effusion on echocardiography. USG of thyroid showed thyroid

Fig.-1: Patient with myxedema ascites
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Agenesis and bone age was delayed. The abdominal ultrasound showed normal liver parenchyma and the CT scan did not reveal any abdominal masses. Anti-Nuclear Antibody (ANA) was negative. So, the patient was diagnosed as myxedema ascites. She was given diuretic and thyroxin (2µg/kg/day) along with supportive treatment. After 4 weeks, edema and ascites subsided completely.

Discussion

Ascites is an uncommon manifestation of hypothyroidism and rarely the primary presenting symptom. The pathophysiology of hypothyroidism-induced ascites is still not fully known. The ascites in patients with myxedema has multiple causes, including increased capillary permeability, impaired lymphatic flow, as well as accumulation of hyaluronic acid-albumin complexes that prevent lymphatic drainage of extravasated albumin forming hyaluronic acid-albumin complexes that release substances that increase capillary permeability. An increase in VEGF also leads to increased extravasation of plasma proteins because of abnormal capillary permeability. Levels of VEGF have been reported to be high in hypothyroid patients with a decline in level to normal values with thyroid replacement therapy.

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Ascitic fluid analysis is the key in guiding the course of investigations needed to establish the diagnosis. A high serum-ascites albumin gradient (SAAG; >1.1 g/dl) suggests that the ascitic fluid is caused by portal hypertension with a 97% diagnostic accuracy. A low SAAG (<1.1 g/dl) suggests that the ascitic fluid is not due to portal hypertension.

Ascitic fluid in case of hypothyroidism reveals high protein (>2.5 g/dl) and SAAG value is usually low although it may vary. Review of literature revealed 52 documented cases of myxedema ascites in adults. A very consistent finding was high total protein level (>2.5 g/dl) in almost all cases with mean SAAG of 1.5 g/dl with a range of 0.8-2.3 g/dl. In another study, SAAG may exceed 1.1 in patients with myxedema ascites, based on a review of eight patients. Because so few cases have been studied about ascites in patients with myxedema, we cannot conclude that a high SAAG is a typical feature in this disease. Moreover, the patient reported here showed low SAAG. Multiple etiologies to our patient’s anasarca were considered. She did not have a history or clinical findings suggestive of nephrotic syndrome or liver disease, tuberculosis or pancreatitis. No significant proteinuria was present to suggest a renal contribution to her presenting complaints. The abdominal ultrasound showed normal liver parenchyma and the CT scan did not reveal any abdominal masses. Hypoalbuminemia was noted to have preceded clinical presentation but malnutrition or synthetic liver dysfunction was a less likely contributor. Ascitic fluid cultures were negative for bacterial or tuberculous infections and also showed negative cytology. In the absence of other causes of ascites and in the presence of long duration of inadequately treated hypothyroidism, myxedema ascites was established as the cause of the patient’s ascites as complete symptom resolution occurred after 4 weeks on levothyroxine.
Conclusion
Hypothyroidism rarely presents with ascites but can be considered after common causes have been ruled out. Diagnostic workup should include a paracentesis with determination of the SAAG. Treatment with thyroid hormone remains an effective therapy with excellent prognosis.

References