

Location: Outpatient clinic

Vitals: Temperature of 36.3C; H.R: 95/min; R.R: 22/min; Blood pressure is 85/50 lying down and 77/46 mm Hg standing.

C.C: Swelling

HPI:

The patient is a three-year-old white male who presents with his mother for evaluation of facial and scrotal swelling of ten days duration. Mother reports that the child had been well until one day prior to admission when she noticed the onset of swelling in his face. She also noted that he had scrotal swelling because he is almost potty trained. She notes some decrease in his urine output as well, although no change in color of the urine, other than becoming somewhat more concentrated. He has had no preceding diarrheal illness, sore throat, abdominal complaints, fevers, and rashes. Mother reports that the child has not complained of any pain syndrome. He does seem to be a little bit more tired than usual. Birth history is unremarkable. All of his immunizations are up to date. SH: He lives with his parents; two older siblings who are healthy, and have had no hospitalizations. There are no pets in the home. There are no smokers in the home. Risk for tuberculosis is low. Development has been normal.

How to approach this case?

Determine the nature and etiology of the "swelling", including whether it's edema or something like hives. Examine the patient to decide whether he needs inpatient or outpatient management.

Physical exam:

General appearance

HEENT/Neck

Heart

Lung

Abdomen

Genitourinary

Extremities

Skin

CNS

Results:

General: well-developed, well-nourished white male in no acute distress. HEENT: remarkable for periorbital edema. Mucous membranes are slightly dry. Neck is supple without lymphadenopathy or thyromegaly. Pupils normal. Cardiovascular: Regular rate and rhythm without murmurs, rubs or gallops. Lungs: very faint rales at the bases and otherwise clear. Abdomen is soft, nontender, nondistended. There are normoactive bowel sounds. There is 1+ sacral edema. + fluid wave. GU: There is scrotal edema present. There is no tenderness to palpation and the cremasteric reflex is intact bilaterally. Extremities: Pulses are 2/4 in the radial, femoral, and dorsalis pedis areas. Hands and feet show 2+ pitting edema and are otherwise unremarkable. Neurologic is nonfocal and appropriate.

Discussion:

This is a three-year-old patient who is manifesting signs of generalized edema, most prominent in his face hands and scrotum. The leading diagnosis in this age frame for such marked edema is nephrotic syndrome secondary to minimal change disease.

Order:

Urinalysis, stat

Basic metabolic panel, stat

CBC with differential, stat

LFTs

Lipid panel
PT/INR, PTT
Complement 3 and 4 levels

Results:

Urinalysis shows 4+ protein, no blood, no RBCs, specific gravity is 1.030, CBC shows a white count of 7, hemoglobin 12.6, hematocrit 36, and platelets 240. Complete metabolic panel (LFTs + BMP) reveals an albumin of 1.5, normal liver function tests, sodium 130, potassium 4.0, chloride 96, bicarbonate 20, BUN 10, creatinine 0.7, glucose 78, calcium 9.4, cholesterol level is 320 mg/dl. Serum albumin is 1.5 gm/dl. Serum protein is 3.7. PT, PTT are normal. Complement levels within normal. Patient has orthostatic hypotension and mild dehydration.

Order:

Admit to floor
Inputs/outputs
Vital signs q4; Continuous cardiorespiratory monitoring
Nephrology consult
Albumin 25% solution IV, 1 gr/kg body weight, infused over 8 hours
Lasix (Furosemide), 1 mg/kg, administered halfway through the albumin infusion
Complete metabolic panel q AM
No salt added, high protein diet.

Results:

Patient responds with good diuresis to albumin and lasix therapy over 24 hours. Vital signs remain stable. Orthostasis resolves. Electrolytes and renal function remain stable.

Order:

Prednisone 2 mg/kg per day, may give in divided dose, po
Vital signs q 12 hours
Repeat albumin and lasix therapy.

Results:

Patient tolerates prednisone.
Remains clinically stable.

Order review:

Discharge to home.
Prednisone for 4-6 weeks.
Follow up in 3-5 days.

Discussion:

The most likely cause of this patient's clinical syndrome is a nephrotic syndrome. The patient is a three year old with generalized edema and screening laboratory tests indicate low protein as well as proteinuria. He therefore meets the diagnostic criteria for nephrotic syndrome which are:

1. Generalized edema.
2. Hypoproteinemia, usually less 2 gm/dl, with a disproportionately low albumin level in relation to the globulin level.
3. Urine protein to urine creatinine ratio in excess of 2 on first morning void or a 24-hour urine protein that exceeds 50 mg/kg of body weight.
4. Hypercholesterolemia greater than 200 mg/dl.

In the vast majority of children in this age range the nephrotic syndrome is due to minimal change disease. This is the cause in approximately 76 percent of children in the one to 12 year age range who present with nephrotic syndrome in childhood. Other etiologies are focal segmental glomerulonephropathy, membranoproliferative glomerulonephropathy, membranous nephropathy and then other causes. For most children, clinical diagnosis is sufficient and

renal biopsy is not warranted unless the child is not responsive to steroids as noted below. CLINICAL FINDINGS in nephrotic syndrome include facial edema frequently periorbital, pretibial edema as well as swelling of the scrotum or labia which may be prominent. These children also have reduced perfusion of their splanchnic capillary bed and may have abdominal pain. As a consequence of their low intravascular oncotic pressure, they may have hypotension as well as pleural effusions with tachypnea and chest pain. As noted in the diagnostic criteria, they do have hypercholesterolemia with increases in their VLDL and LDL because of changes in hepatic catabolism and enhanced synthesis, respectively. However, the disturbances in lipid metabolism usually do not cause any other clinical findings. By contrast, changes in the protein levels of their coagulation cascade does put them at increased risk for thrombosis. Their tendency to form thrombus is due to a combination of factors including hyperaggregatable platelets, increased fibrinogen concentrations, loss of antithrombin III, increased blood viscosity and decreased blood flow. Venous thrombosis can occur in the deep veins of the extremities and the cerebrocortical system and renal veins and the pulmonary venous system and it is a source of increased morbidity for these children. As a consequence, one should obtain initial coagulation studies upon admission.

Laboratory analysis also shows reduced immunoglobulins, particularly IgG. This low level of IgG in combination with the steroids which are the primary treatment for nephrotic syndrome puts the children at increased risk for development of infections.

Peritonitis is one of the more serious complications of nephrotic syndrome. The causative organisms in children are streptococcus pneumoniae and Escherichia coli. One should consider peritonitis in any child who presents with significant abdominal complaints and paracentesis should be performed to identify the organism and confirm the diagnosis. For children in whom thrombosis is a serious consideration, one can start heparin therapy at a dose of 50 units/kg intravenously and 100 units/kg every four hours IV for maintenance therapy. Human nephrotic syndrome is supportive and aimed at increasing the intravascular oncotic pressure, decreasing third spaced fluid maintaining fluid balance, monitoring nutrition and treating any concomitant infection. Usually the typical approach is to give the patient intravenous albumin 1 gm/kg of a 25% solution for example and infuse it continuously over 8 to 12 hours under close supervision for the development of heart failure. Loop diuretics like Lasix can then be administered halfway through the albumin infusion or after it at a dose of 1-2 mg/kg IV and this effectively reduces third spaced fluids and pulls interstitial fluid into the vascular space. Prednisone is begun on patients at a dose of 60 mg/M² or 2 mg/kg and the daily dose is maintained for four to six weeks. It is advisable to obtain a tuberculosis screen test prior to initiating the steroid therapy. Most children will begin to show a decrease in their urine protein excretion after about seven to ten days following the initiation of steroids. If the child is stable with just mild to moderate edema, no pulmonary edema and has good diuretic response, then they do not need to stay in the hospital until their protein excretion is reduced and instead most of them can be discharged within two to three days. Fluid balance in the hospital setting is important and should be monitored closely during the initial diuresis. Patient should be given a high quality, high protein diet to improve their growth and because they have an increased protein need to regenerate their albumin. They should have a no salt added diet to discourage further edema formation. About 85-90% of children treated in fashion will have a satisfactory response and obtain remission. The mortality in minimal change nephrotic syndrome is approximately 2% with the majority of deaths due to peritonitis or thrombus formation and these complications can occur even under ideal treatment circumstances. The other 98% of children who develop this syndrome are likely to have a good response to steroids and return to a normal state of health. About two-thirds of them experience at least a single relapse and another third go on to have a series of relapses over the course of many years.

For children who remain symptom free for over two years without any medications, their prognosis is the best and they are considered recovered.

Primary diagnosis:
NEPHROTIC SYNDROME

