chyluria in this girl rules out hypoalbuminemia due to filariasis per se. However, microfilaria could not be documented by renal biopsy. Hence, filariasis is the probable cause for nephrotic syndrome in this girl.

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Pseudomonas Septicemia in Selective IgM Deficiency

A 7½-month-old male baby presented with cough 10 days, fever 7 days, altered sensorium 3 days, discharge both ears 3 days, and reddish lesions on body 2 days. In the past he had 3 episodes of chest infections. He was the only child born out of non-consanguinous marriage between apparently healthy parents. There was no history of intake of immuno-suppressive drugs. He was found to be very sick with marked tachycardia, respiratory distress, bilateral seropurulent ear discharge, multiple erythematous indurated patches with necrotic center, generalized lymphadenopathy, hepatomegaly and crepitations in the chest. He was deeply comatose, had brisk deep tendon reflexes, extensor planters and meningeal signs. There was polymorphonuclear leukocytosis and thrombocytopenia. CSF was suggestive of partially treated pyogenic meningitis, however, CSF culture was sterile. Gram stain of CSF and skin lesions revealed no bacteria. Culture of fluid from skin lesions, blood culture and ear discharge grew Pseudomonas aeruginosa. Serum IgG level was 930 mg/dL, IgA was 102 mg/dL, and IgM was 12 mg/dL. Serum IgM level of father was 19.5 mg/dL. The patient was administered appropriate antibiotics, platelet transfusion, fresh frozen plasma, IV immunoglobulins along with other supportive measures. His condition progressively deteriorated and he succumbed to disseminated intravascular coagulation and multiorgan dysfunction.

Selective IgM deficiency severe enough to cause symptoms is rare as a primary disorder though it is a common consequence of immunosuppression(1). Patients have IgM concentration <20 mg/dL with normal levels of other immunoglobulins(2). Inheritance is multifactorial, may be autosomal recessive but is presumed to be partly influenced by X-chromosomal products. Among healthy
controls low IgM levels are more common in males(1). Fathers of patients tend to have low levels too(3). IgM bearing B cells are present in normal numbers. Some patients have decreased helper T cell activity. Some patients are capable of normal antibody response in other immunoglobulin classes following specific immunization, whereas others respond poorly. Cell mediated immunity appears to be intact. It is hypothesized that selective IgM deficiency results from either insufficient T helper cells or increased T suppressor cell functions interfere with IgM committed B cell differentiation.

The deficiency predisposes them to overwhelming infection with polysaccharide containing organisms e.g., pneumococci, *H. influenzae*, meningococcal and pseudomonas(1,4). They may also have atopic dermatitis, recurrent respiratory infections, recurrent sepsis, urinary tract infections. Association has been found with autoimmune diseases (SLE, hemolytic anemia) and gastrointestinal conditions (Crohn’s disease, chronic diarrhea, lymphoid nodular hyperplasia, Whipple’s disease, splenomegaly)(5). In the absence of sufficient data on appropriate therapy it would seem logical to treat these patients similar to post splenectomy patients(4). Immediate antibiotics (penicillin/ampicillin) should be instituted for all infections or chemoprophylaxis offered. Fresh frozen plasma should be infused for serious infections. If patients are unable to form antibody to specific antigens, gamma-globulin therapy should be given. However, commercially available intravenous immunoglobulins have only traces of IgM. The prognosis is poor and a vast majority succumb to rapid hematogenous spread of infection.

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