Pulmonary hydatid disease associated with nephrotic syndrome in a paediatric patient

Sir,

Renal diseases associated with infectious and parasitic diseases have been well documented [1,2]. The nephrotic syndrome of quartan malaria and glomerulonephritis associated with the hepatosplenic form of schistosomiasis are examples of such associations. Infectious agents may serve as triggers for immunological kidney diseases. In turn these antigen–antibody complexes may form depositions in the glomerular basement membrane [1]. Sanchez et al. [3] have described Echinococcus granulosus antigen in a renal biopsy performed on a 67-year-old woman suffering from nephrotic syndrome due to membranous glomerulonephritis and hydatid disease of the liver. We report a paediatric patient with pulmonary hydatic cysts and associated nephrotic syndrome.

Case. A 3-year-old boy presented with a history of swelling of the face and was diagnosed as having minimal change nephrotic syndrome. On admission, physical examination showed periorbital and pretibial oedema. Laboratory tests revealed a complete blood count showing haemocoagulation and normal serum electrolytes and renal function. However, heavy proteinuria with a protein/creatinine ratio of 30 and low serum albumin (1.2 g/dl) and hypercholesterolaemia (653 mg/dl) were found. Corticosteroid treatment was started at a conventional dose of 2 mg/kg/day. He presented a steroid-dependent course of treatment following each completion of steroid therapy, the symptoms and laboratory findings showed a relapse of nephrotic syndrome, a situation which occurred more than six times over one and a half years. The family refused renal biopsy.

In his fourth year he was referred to the paediatric pulmonology department because of his cough, fever and nodular lesions seen on the postero-anterior chest X-ray. He was still on 10 mg oral corticosteroid every other day when a physical examination revealed a fever of 39.4°C, rales with rare wheezes in the middle and lower zones. Chest X-ray showed two lesions. Thoracal computerized tomography showed two homogenous cysts; one, in the posterior of the right lower lobe with a 35 mm diameter and the other in the basal medioposterior region of the left lung with a 33 mm diameter. The densities of the lesions were similar to fluid densities compatible with a hydatic cyst. There were no other lesions in the lung parenchyma. Ultrasonographically there were no cysts in the liver. The diagnosis of hydatid disease was confirmed serologically by positive Echinococcus passive haemagglutination test in titer of 1/1200 (ELISA). The patient was put on albendazole treatment. Steroid treatment was tapered and discontinued 8 weeks after the commencement of albendazole. Albendazole was continued for 6 months during which the size of the lesions decreased almost totally. He remained on full remission for the nephrotic syndrome for 5 years after cessation of albendazole therapy, during which time he required no further steroid therapy.

Comment. This is the first report of successful therapy with albendazole in a childhood nephrotic syndrome with pulmonary cyst hydatid. The relationship of hydatid disease and nephrotic syndrome has been previously suggested in the case of an elderly woman where renal hydatid antigens were demonstrated by immunoffluorescent staining of a renal biopsy using antihydatid serum [3]. Hydatid disease has also been associated with a vasculitic disease, namely, polyarteritis nodosa [4]. Covic et al. [5] reported a 67-year-old patient who had developed mesangiocapillary glomerulonephritis secondary to hepatic hydatid disease who responded to the removal of the echinococcal cysts. Gelman et al. [6] have also described a 63-year-old man with minimal change glomerulonephritis (MCGN) associated with hepatic hydatid disease. Miatello et al. [7] have described a patient in whom nephrotic syndrome disappeared following the excision of a pulmonary hydatic cyst.

This is the first report of paediatric nephrotic syndrome associated with pulmonary hydatid lesions. An interesting feature in our patient was that the nephrotic syndrome was diagnosed earlier than the hydatic lesions in the lung. He was treated as minimal change nephrotic syndrome before the hydatic lesions in the lung were diagnosed. The occurrence of hydatid disease in this nephrotic patient may well have been considered as being secondary to the relative immune deficient state of corticosteroid therapy. However, prednisolone therapy failed in this patient and no complete remission could be obtained until albendazole treatment was commenced. Again a complete remission for 5 years was achieved only after the treatment of hydatid disease. There are some studies suggesting that the clinical and biochemical manifestations of the nephrotic syndrome of childhood may be related to hypersensitivity [8]. A T cell defect, characterized by a decreased number of Th1 cells and increased number of Th2 cells, has been reported in children with MCGN [9]. In allergic reactions there are Th2 helper lymphocytes producing the cytokines interleukin-4 (IL-4), IL-5 and IL-6, and long-lasting antibody response with production of IgE and IgG4 antibodies. This is true also for the defence against helminth parasites. Continual antigen presentation as an E. granulosus antigen
may cause long-lasting antibody response that causes antigen-antibody deposition in the glomerular basement membrane [3,10].

Although we were unable to demonstrate the E. granulosus antigens in the basement membrane of our patient, the aforementioned clinical course favours an association between this infectious agent and the nephrotic syndrome of this patient. We suggest that Echinococcus should be included in the aetiologic classification of secondary nephrotic syndrome especially in the country where hydatid disease is endemic. We draw attention to the origins of recent literature on this subject around the Mediterranean area.

Hacettepe University Medical Faculty Cansin Saçkesen
Ihsan Doğramacı Children’s Hospital Halil Atasoy
Ankara, Turkey Nural Kiper
Email: sozen@gen.hun.edu.tr Seza Ozen