Case Report

Reversible nephrotic syndrome due to mesangiocapillary glomerulonephritis secondary to hepatic hydatid disease

A. Covic1, I. Mititiuc1, L. Caruntu1 and D. J. A. Goldsmith2

1University Hospital of Lasi, Lasi, Romania, and 2Royal Sussex County Hospital, Brighton, UK

Key words: hydatid disease; mesangiocapillary glomerulonephritis; nephrotic syndrome

Introduction

Hydatid disease (echinococcosis) is a cyclozoonotic infection with the dog tapeworm Echinococcus granulosus, more commonly seen in men than in women. The worm is endemic to many parts of the world, including Europe, Asia and the Americas. The usual life-cycle is between dogs and sheep, with humans infrequently accidental hosts. Rarer primary hosts are foxes, horses, wolves and jackals. The life-cycle is shown in Figure 1.

There are no specific local or general signs and symptoms, while the distribution and manifestations of hydatidosis are ubiquitous and protean. Liver and lung are the two commonest organs involved (one or both in 90% of cases), but the kidney is very rarely affected primarily (2% in 1802 cases in the Australasian Hydatid Register [1]). Usually symptoms arise from the expanding presence of the cysts inside organs, or after cysts rupture. Systemic ill-effects are rare. Diagnosis is made by history of exposure, by radiological and ultrasonographic cyst detection, and by serology [1].

Secondary renal involvement as a response to the presence of Echinococcus in other parts of the body is exceptionally unusual, with sporadic reporting of glomerular lesions (IgA nephropathy [2], membranous glomerulopathy [3] and mesangiocapillary glomerulonephritis [4]). We report a unique case of a female who presented with nephrotic syndrome due mesangiocapillary glomerulonephritis (MG) with a large hepatic hydatid cyst. Proteinuria was abolished by surgical cyst removal, but later returned with a relapse of hydatid disease, a renal biopsy again confirming MG. Definitive treatment for hydatid disease abolished all proteinuria and restored the patient to sustained good health.

Case report

A 67-year-old woman complained of ankle swelling of recent onset in January 1995. This worsened despite initial therapy with digitalis and diuretics. She rapidly became severely hypertensive, and presented to hospital with hypertensive encephalopathy and gross oedema in February 1995.

On admission to hospital she had anasarca, though without significant pleural or pericardial effusions, or ascites. Blood pressure was 220/120 mmHg. Her liver was grossly enlarged, firm and tender. The spleen was not enlarged. She was afebrile.

Her urine was frothy, tested positive to blood and protein on dipsticking, and contained dysmorphic red cells and proteinaceous casts. She was excreting 24 g of urinary protein per 24 h (non-selective; albumin-globulin ratio of 1.32). Renal function was abnormal (urea 9 mmol/l; creatinine 180 μmol/l).

Haemoglobin was 9.6 g/l with a normochromic, normocytic blood film and no evidence of red cell fragmentation. Serum iron was normal. The white count was elevated at 28 000, with a neutrophil leukocytosis but no eosinophilia. Platelets were normal at 180 000. The ESR was 160 mm in the first hour. Liver function tests were normal. Autoantibody screening was negative (including ANA and ANCA), and serum

© 1996 European Renal Association–European Dialysis and Transplant Association
Immunoglobulins normal. C₃ was at the lower limit of normal for an adult at 0.7 g/l, while C₄ was normal at 0.2 g/l. Antistreptolysin O titres (ASOT) were not raised. Plasma albumin was 14 g/l. Plasma lipids were elevated significantly, in keeping with severe nephrotic syndrome.

The chest X-ray showed a raised right hemidiaphragm. Renal ultrasound showed two normal sized kidneys with a normal corticomedullary pattern. Liver ultrasound showed a large fluid-filled cavity in the posterior segment of the right lobe of the liver, with a circumferential distribution of smaller cysts. Hepatic scintigraphy confirmed the presence of a hepatic space-occupying lesion.

Clinical suspicion of hydatid disease was high, and the diagnosis was confirmed by a strongly positive serum ELISA for Echinococcus. A renal biopsy was performed which showed MG and hypertensive changes (Figure 2).

Treatment was started with a high protein intake, diuretics and high-dose captopril. BP fell to 150/80 mmHg, oedema improved but remained, and proteinuria fell to 8 g/24 h. Plasma albumin rose to 23 g/l. In May 1995, hepatic cyst resection was attempted. The clinical diagnosis was confirmed at laparotomy, and several cysts were removed from the liver without incident.

One month later the patient was well, with BP 130/90 mmHg, off anti hypertensives and diuretics, plasma albumin 36 g/l, 24-h urinary protein 1.3 g, and renal function now normal. The serum ELISA for Echinococcus was borderline positive. One month later, all proteinuria and microscopic haematuria had resolved. The ESR was now 16 mm in the first hour, serum urea 3 mmol/l and plasma creatinine 80 µmol/l. She was on no drug therapy.

Five months later (December 1995) the patient re-presented with severe oedema, and raised blood pressure. Plasma albumin was 20 g/l, 24-h urinary protein loss was 12 g. Dysmorphic red blood cells were once again found on urine microscopy. ESR was 120 mm in the first hour. Serum urea was 7.5 mmol/l and plasma creatinine 156 µmol/l. Liver ultrasound showed another hepatic cyst. The serum ELISA for Echinococcus was strongly positive. Another renal biopsy showed MG.

One month later, another laparotomy was performed at which a single large echinococcal cyst was removed from the left lobe of the liver without incident. Over the next 2 months all proteinuria and manifestations of the nephrotic syndrome resolved. The patient remains well, with normal blood pressure and renal function, with no evidence of residual hepatic echinococcal disease.

Discussion

Echinococcal disease is still significant in the developing world, and in some parts of the developed world (e.g. Wales, Tasmania). In north-east Romania, hydatid disease is a significant source of chronic ill-health; here, dogs and bats are vectors. Definitive treatment is surgical resection, though this is not without risk, especially if cysts rupture, spilling their contents. Relapse (presumably due to further growth of cysts that are small at the time of first surgery and are therefore missed or are inaccessible in situ) is not unusual.

Mesangiocapillary glomerulonephritis is described as four distinct histological variants [5]. Type I, characterized by mesangial cellular proliferation and an increase in mesangial matrix, is considered as a glomerular response to antigen–antibody complexes (there are histological similarities with immune-complex disease). Hypocomplementaemia is frequent, but less common in adults. A histologically identical appearance to type I MCGN is seen in shunt nephritis (mostly staphylococcal), postinfectious nephritis (mostly streptococcal), sickle-cell disease, loiasis, onchocerciasis, alpha-1-antitrypsin deficiency, and systemic lupus [5].

There are very scanty reports of hydatid-associated glomerular lesions in the literature, and none with such a clear relationship to clinical disease activity. Membranous glomerulonephritis and echinococcal disease was reported in an adult shown to have early membranous glomerulonephritis after renal biopsy during liver-cyst resection [3]. In that case no proteinuria was present and renal function was normal. Immunoperoxidase studies showed echinococcal antigen and corresponding antibody in the glomeruli. Other hydatid-associated glomerulonephritides have been reported, though cause and effect was not well established, including one case of ‘postinfectious glomerulonephritis’ [6] and another of mesangiocapillary disease [4]. Our case here presented is unique in demonstrating a relapsing—remitting course of mesangiocapillary glomerulonephritis-associated nephrotic syndrome matching pari passu the presence or absence
Fig. 3. Relationship between proteinuria, renal function, blood pressure, serum ELISA for Echinococcus, and hepatic resection of hydatid cysts.

of hepatic echinococcal cysts, and positive serum ELISA. This is shown in Figure 3.

References


Received for publication: 31.5.96
Accepted: 3.6.96