Granulomatous sarcoid nephropathy

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Summary

When sarcoidosis of the renal parenchyma occurs it is rarely of major clinical significance. The clinical features and post-mortem findings of a case of subacute uraemic syndrome due to severe granulomatous involvement of the kidney by sarcoidosis are described. The case is unusual in that granulomatous sarcoid nephropathy resulted in death from renal failure within 4 months of onset of symptoms in the absence of clinically apparent sarcoidosis.

Introduction

Impairment of renal function in sarcoidosis is usually the result of nephrocalcinosis and nephrolithiasis following prolonged hypercalciuria or hypercalcaemia. When sarcoidosis of the renal parenchyma occurs it is rarely of major clinical significance. The clinical features and post-mortem findings are reported of a case of subacute uraemic syndrome due to severe granulomatous involvement of the kidney by sarcoidosis. The case is unusual in that the main clinical and pathological effects were due to granulomatous sarcoid nephropathy which in the absence of clinically apparent sarcoidosis rapidly resulted in death from renal failure.

Case history

A 60-year-old printer was admitted with a 3-month history of weight loss, nausea, anorexia, and subsequently vomiting with abdominal discomfort. There was no significant past medical history. He had frequency of micturition, and nocturia three times each night.

Clinical examination was normal apart from mild pallor and cachexia. The blood pressure was 130/80 mmHg and urinalysis showed a trace of protein with normal microscopy. Laboratory investigations: Haemoglobin 10.3 g/dl, MCV 88, white cell count $6.1 \times 10^9/l$. Blood urea 58 mmol/l, serum sodium 125 mmol/l, serum potassium 5.5 mmol/l and serum bicarbonate 13 mmol/l. Total protein 77 g/l, albumin 29%, Serum calcium 1.65 mmol/l, serum phosphate 2.52 mmol/l and alkaline phosphatase 188 u./l.

Arterial blood gas analysis confirmed a metabolic acidosis with a hydrogen ion concentration of 68 mmol/l, P_{O_2} 12 kPa and P_{CO_2} 4.1 kPa. Plasma creatinine was elevated at 1.4 mmol/l with a creatinine clearance of 0.03 ml/sec. Twenty-four hour urinary protein excretion was mildly elevated and ranged between 420 and 1230 mg. The chest X-ray revealed pulmonary venous congestion with bilateral pleural effusions and an increased cardiothoracic ratio without evidence of hilar lymphadenopathy. Radio-hippuran renography showed poor but symmetrical renal function without evidence of out-flow tract obstruction.

The patient was transferred to a Renal Dialysis Unit and peritoneal dialysis commenced. Unfortunately, following the second peritoneal dialysis he developed severe cardiogenic shock, pulmonary oedema and subsequently died.

At post-mortem both kidneys were pale and swollen, the right weighing 160 g and the left 170 g. Para-aortic lymph node enlargement was noted. The spleen was not enlarged. Large bilateral pleural effusions were present and the lungs were oedematous. The heart weighed 350 g and appeared normal. Histological examination revealed numerous typical sarcoid follicles with epithelioid cells and multi-nucleated giant cells throughout the lymph nodes and spleen. In the lung parenchyma multi-nucleated cells with Schaumann bodies were found: in the liver, fibrotic changes were noted around the portal tracts with sarcoid follicles present in liver parenchyma. Sarcoid follicles were also seen in the mucosa of the body and antrum of the stomach and in the pancreas. No evidence of sarcoidosis could be found on microscopy of heart muscle, brain, thyroid gland or small intestine.

Both kidneys showed extensive granulomatous sarcoid change without evidence of nephrocalcinosis or nephrolithiasis. The disease affected both the renal cortex and medulla and to some extent the calyces (Figs 1 and 2). Some of the sarcoid lesions were typical epithelioid and giant cell granulomata, others consisted of spindle cells interspersed with lymphocytes, and some were composed of discrete

FIG. 1. Sarcoidosis of renal cortex, HE $\times$ 200.

FIG. 2. Sarcoid giant cells near apex of renal papilla, HE $\times$ 200.

multi-nucleated cells with Schaumann bodies or calcispherules. Sarcoid follicles were also seen in the adventitia and intima of a few small arteries and veins. Though some of the glomeruli appeared normal others were severely damaged by sarcoid lesions in the adjoining stroma and a number were fibrotic. Casts were present in groups of collecting tubules and there was extensive degeneration of the convoluted tubular epithelium.

Discussion

Serious involvement of the genito-urinary tract in sarcoidosis is uncommon. Branson and Park (1954) reviewed over 650 cases of sarcoidosis including 138 post-mortems and found only 7% with sarcoid-associated lesions within the genito-urinary tract. When renal impairment becomes clinically apparent, hypercalciuria and hypercalcaemia are the commonest aetiological factors (Falls, Randall and Summers, 1972; Heptinstall, 1974).

Whilst granulomatous sarcoid nephropathy is now being recognized more frequently it rarely produces overt renal impairment and the clinical features of generalized sarcoidosis have usually become apparent well before the development of renal involvement. The clinical features often comprise only mild proteinuria, normal or slight enlargement of renal size radiologically and the absence of hypertension. Although granulomatous sarcoid nephropathy may be steroid responsive, relapses are frequent and death may result within 5 years from onset of clinically apparent renal involvement (Coburn, Hobbs and Johnston, 1967; Lebacqz, Desmet and Verhaegen, 1970).

The case just described is unusual in that granulomatous sarcoid nephropathy resulted in death from renal failure within 4 months of onset of symptoms in the absence of clinically apparent sarcoidosis.

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