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Epidemiological, Clinical and Follow-Up Data in a Series of Thirteen Renal Insufficiencies Complicating Sarcoidosis

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Abstract

The sarcoidosis is a systemic granulomatosis affecting most frequently the lungs and the mediastinum. An acute renal failure reveals exceptionally this disease. It's a retrospective study implicating 13 cases of sarcoidosis complicated of acute renal failure. The aim of this study is to determine epidemiological, clinical, biological and histological profile in these cases and the interest of considering sarcoidosis diagnosis in case of unexplained renal failure. Extra-renal complications, therapeutic modalities and the outcome were determined in all patients. Our series involved 13 women with an average age of 41 years. Biological investigations showed an abnormal normocalcemia in 8 cases, a hypercalcemia in 5 cases, a hypercalciuria in 11 cases and polyclonal hypergammaglobulinemia in 7 cases. An acute renal failure was found in all patients with a median creatinin of 540 umol/L. The renal echography was normal in all patients. The kidney biopsy performed in all patients showed tubulo-interstitial nephritis. The extra-renal signs were: pulmonary interstitial syndrome in 5 cases, a sicca syndrome in 4 cases, mediastinal lymphadenopathy in 2 cases, a lymphocytic alveolitis in 3 cases, an anterior granulomatous uveitis in 3 cases and a polyarthritis in 6 cases. Six patients benefited from hemodialysis. The treatment consisted of corticosteroid in all cases. The follow up was marked by complete resolution of clinical and biological signs. The diagnosis of renal sarcoidosis should be made rapidly in order to avoid end stage renal failure.

Keywords

Sarcoidosis, Acute Renal Failure, Interstitial Nephritis, Granuloma

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1. Introduction

Sarcoidosis is a systemic granulomatous disease which preferentially involves the lung and mediastinum. A revealing acute renal failure is rare and a cause of positive diagnosis lateness [1]. It is due most commonly to a balance anomaly of calcium or a parenchymal involvement. Renal biopsy allows the diagnosis by finding specific impairment of sarcoidosis. The prognostic factor for renal survival in sarcoidosis is the early diagnosis and response to treatment [1]-[3].

2. Materials and Methods

This is a retrospective study which collected 13 cases of sarcoidosis complicated by acute renal failure hospitalized in Internal medicine A department of Charles Nicolle Hospital. The study consisted of identifying epidemiological data (age, gender), clinical symptoms (renal and extra-renal signs), biological signs (serum calcium level, urinary calcium, proteinuria/24h, serum creatinine level) and histological results (renal biopsy), which confirmed the diagnosis. A study of the therapeutic and evolving profile (clinical and biological signs) was performed in all cases.

The purpose of this study was to determine the epidemiological, clinical, biological and histological characteristics in 13 patients with acute renal failure complicating systemic sarcoidosis, and emphasize the interest to consider this diagnosis in case of unexplained renal failure.

3. Results

This study collected 13 women with an average age was 41 years, ranging from 19 years to 72 years. Clinical examination found a preserved general state, apyrexia and normal blood pressure in all patients. No cases of peripheral lymphadenopathy or cutaneous sarcoid were mentioned. Cardiac, respiratory and neurological examinations were normal. Tuberculin skin test was negative in 8 cases.

Laboratory tests showed an acute renal failure with a median creatinine 540 umol/L and creatinine values ranging from 138 to 1520 mmol/L, an inflammatory syndrome in 12 cases, an abnormal normocalcemia in 8 cases, a hypercalcemia in 5 cases with an average of 2.8 mmol/L, a hypercalciuria in 11 cases, a polyclonal hypergammaglobulinemia in 7 cases and a negative proteinuria in 8 cases. Proteinuria was low in 5 patients with an average of 0.3 g/24h. Hypokalemia and metabolic acidosis were mentioned in 7 patients. The angiotensin-converting enzyme was elevated in 8 cases.

Renal ultrasound was normal in all patients. Renal biopsy performed in all cases showed tubulo-interstitial nephritis in all patients, with the presence of giant cells in 2 cases and associated with giant cell granuloma in 6 cases.

Extra-renal signs were: pulmonary interstitial syndrome in 5 cases, sicca syndrome in 4 cases, mediastinal lymphadenopathy in 2 cases, lymphocytic alveolitis in 3 cases, anterior granulomatous uveitis in 3 cases and polyarthritis in 6 cases.

According to the clinical, biological and histological results, the diagnosis of sarcoidosis complicated of acute renal failure was made. Six patients benefited of hemodialysis to severe renal impairment.

All patients received steroids, at a dose of 1 mg/kg/day with a gradual degression, since they had active inflammatory kidney damage. The outcome was marked by improvement of serum creatinine levels and then normalization of renal function in all patients and serum calcium level in pathological cases. No new recurrence of renal sarcoidosis was observed in all our series with mean recoil of 5 years.

4. Discussion

Sarcoidosis is a granulomatous giant cell non-caseating and multisystemic disease of unknown etiology. Mediastinal and pulmonary localizations are the most common [1].

Renal involvement in sarcoidosis is rare but can be severe by progressing to irreducible and end stage renal failure. It is most often the result of disorders of calcium metabolism inducing calcium renal deposits [2].

The parenchymal involvement is frequently tubulointerstitial nephritis. Glomerular lesions remain exceptional including essentially membranous glomerulonephritis, rarely amyloidosis and exceptionally IgA nephropathy [1] [3].

Renal failure associatied to hypokalemia and metabolic acidosis can be biological indicative signs of tubulo-

interstitial nephropathy as in the case of 7 patients in our series.

Renal biopsy has a very significant interest to the diagnosis of sarcoidosis especially in the absence of other extra-renal signs.

At histology, the lesions associated interstitial cellular infiltrates and tubular inflammation. The presence of non-caseating granulomas, which is not constant, is highly suggestive of sarcoidosis diagnosis [1] [4].

In the series of Löffler U *et al.*, including 27 cases, non-granulomatous tubulo-interstitial nephritis was the most common histological entity (44%), followed by granulomatous interstitial nephritis (30%), IgA glomerulonephritis (26%) and nephrocalcinosis (11%) [1].

Granulomatous tubulo-interstitial nephritis in sarcoidosis manifests rarely as acute renal failure feature [5] [6]. In fact, these lesions are often asymptomatic, and the prevalence is higher in autopsy series [1] [5].

Some rare cases of isolated sarcoidosis interstitial nephritis without visceral involvement had been published [1] [7] [8].

Granulomatous tubulo-interstitial nephritis secondary to sarcoidosis is a formal indication for systemic therapy because it threatens the renal functional outcome [1] [5] [9].

Corticosteroids remain the first line treatment of kidney damage. The dose and duration is variable and not codified. Recovery is more rapid and complete if the treatment was initiated at the early inflammatory stage and prior to fibrosis process [1] [9] [10]. In case of cortico-resistance, an immunosuppressor should be indicated.

The corticosteroid treatment results usually in rapid improvement of hypercalcemia, renal function, and complete resolution of extra-renal signs as is the case of our patients.

The prognosis of sarcoidosis is usually good with spontaneous remission but the chronic aspect can be seen in one third of cases and may be associated then with one or more organ dysfunctions [1] [7] [9] [10]. A chronic kidney failure may be secondary to sarcoidosis especially in cases of diagnostic and therapeutic delay lateness.

5. Conclusion

Renal involvement in sarcoidosis is rare but serious and can lead to chronic kidney failure. Early diagnosis and adapted treatment allows preserving renal function.

Disclosure Statement

The authors have nothing to disclose.

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