To know more....

Sarcoidosis is an inflammatory disease of unknown etiology characterized by non-caseating granulomas and by variable clinical course which can affect every organ and system. Although the diagnosis of sarcoidosis can be made on clinical grounds when several specific clinical findings are present, in the majority of cases it is not so easy, especially when the affected organs are the ones less commonly involved, such as heart and kidney . Cardiac and renal sarcoidosis are rarely diagnosed. Of note the involvement of heart and kidney is not so rare in the course of the disease since according to autopsy studies its prevalence is at least 20 to 30% nonetheless, less than 5% of the patients suffer from clinical cardiac or kidney sarcoidosis.

Cardiac sarcoidosis may be completely silent or induce several cardiac changes which may vary from conduction abnormalities to sudden death . Despite the highly variable presentation, the prognosis of cardiac sarcoidosis is severe; thus it is imperative to make a diagnosis and start a treatment as soon as possible. The diagnosis of cardiac involvement in sarcoidosis is extremely challenging because of a lack of a gold standard test. Even the myocardial biopsy, which was also performed in our patient, may not be diagnostic due to patchy distribution of the disease . Algorithm and diagnostic criteria have been developed and considered as established criteria till recently but they have proved to be insensitive for detecting cardiac involvement . These studies however have shown that CMR and PET have a central role in the diagnosis and the management of cardiac sarcoidosis. In the present study CMR and endomyocardial biopsy were performed to determine the cause of the dilated cardiomyopathy but did not yield any results. A heart biopsy may result negative in case of sarcoidosis because sarcoid granulomas are less commonly located in the right ventricle where biopsies are commonly taken. 18F-FDG PET, which enables visualization of sarcoid lesions in various organs including the heart, was not performed in our patient and a diagnosis of dilated cardiomyopathy was made even though the majority of the conditions causing a cardiomyopathy could be ruled out based on medical and familial history and demographic data. No attempt was made to administer steroid as an ex adiuvantibus treatment probably because the possibility of a case of cardiac sarcoidosis never occurred to the cardiologists.

When the patient was admitted to the Nephrology Unit the presence of unexplained hypercalcemia, the reduction of GFR and the recent episode of HF led to further investigations and to perform the kidney biopsy even though there were no strong indications but only a mild increase of serum creatinine and of proteinuria . All the known causes of hypercalemia such as primary hyperparathyroidism, vitamin D intoxication, malignancies, thiazides, familial hypocalciuric hypercalcemia, milk alkali syndrome, were ruled out . Physical examination, serum angiotensin converting enzyme (sACE) levelsand chest x ray, considered central in the work-up for the diagnosis of sarcoidosis were negative or normal. The presence of non caseating granulomas with epithelioid-type multinucleated giant cells and associated interstitial nephritis and calcification at the kidney biopsy allowed the diagnosis in our patient . The rapid improvement of renal function and the progressive amelioration of cardiac ejection fraction with steroid treatment confirmed our diagnosis of cardiac and renal sarcoidosis ruling out other granulomatosis diseases .