Late diagnosis and management challenges of systemic sarcoidosis with renal involvement

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Systemic Sarcoidosis (SS) is a multisystem granulomatous disease rarely affecting the kidney. The noncaseating granulomatous interstitial nephritis is the typical histological finding; other forms such as calcium metabolism disorders, glomerular disease, tubular dysfunction, obstructive and vascular uropathy have been reported

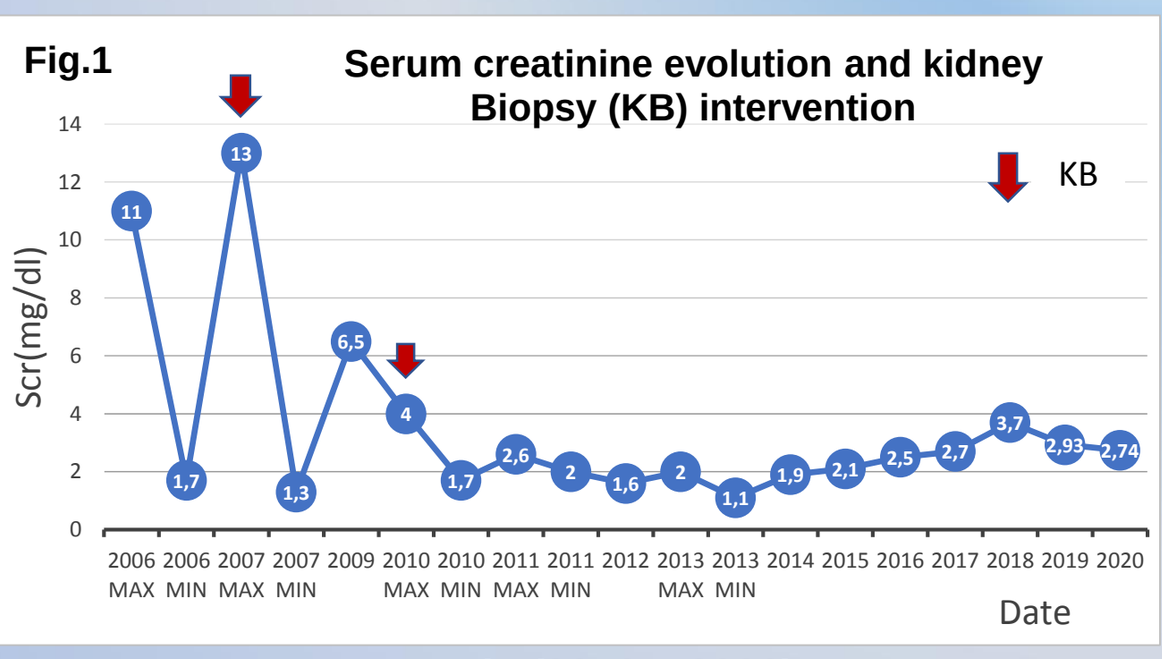
Case report

Algeria (2006 - 2010)

- **2006** (14 years old): 1st admission for iridocyclitis, hepatitis and acute kidney injury [AKI]. Suspicion of tubulointerstitial nephritis and uveitis (TINU) syndrome, → corticosteroids (methylprednisolone 1g /1.73 m^2) followed by oral intake led to a significant but transient improvement of kidney function (Fig. 1)
- **2007**: recurrent iridocyclitis and AKI episodes associated with severe hypokalaemia, hyponatremia and metabolic alkalosis suggested an acquired Bartter-like syndrome.(fig.3) Indomethacin treatment and kalium supplements were initiated. Intravenous corticosteroids improved kidney function (Fig.1)

  1st kidney biopsy: noncaseating granulomas (Fig. 2A), mononuclear cells and epithelioid histiocytes (Fig,2B)
- **2009**: diagnosis of renal sarcoidosis suggested by an increased serum level of angiotensin converting enzyme (ACE) (193 U, N: 12-68) in a French laboratory

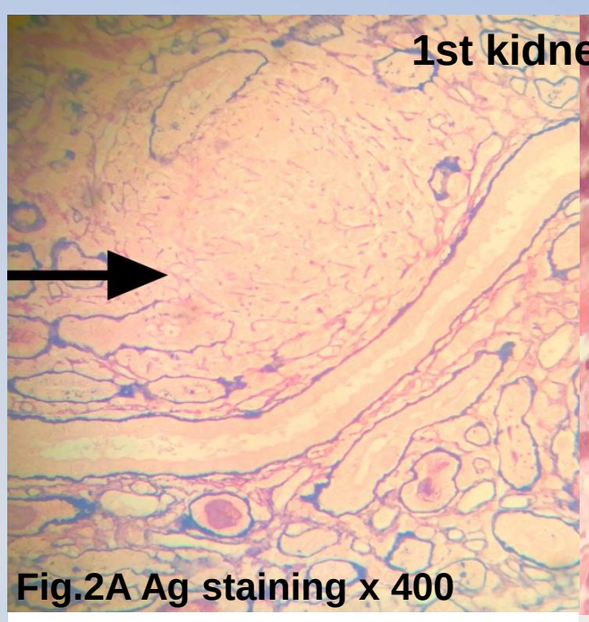
Fig.1 Serum creatinine evolution and kidney Biopsy (KB) intervention

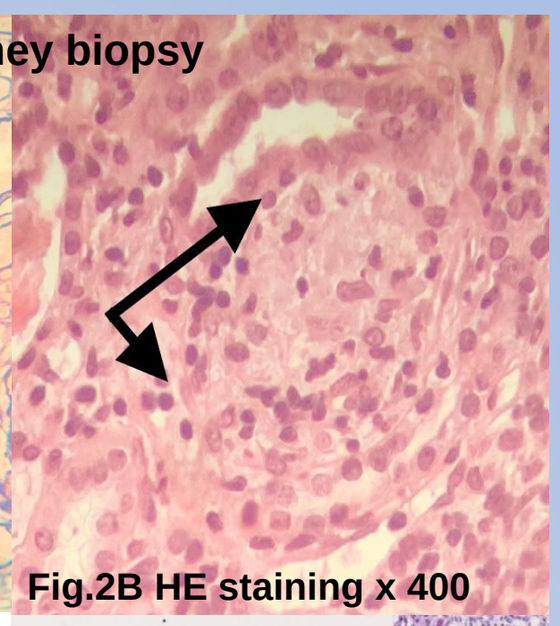


Brussels (2010 – 2020)

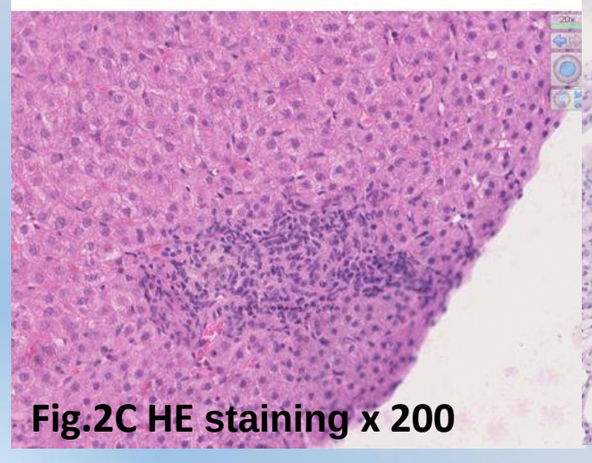
- **2010**: Indomethacin stopped and oral corticosteroids maintained (prednisolone 30 mg/day). A **liver biopsy** showed unilobular granuloma confirming the diagnosis of SS (Fig. 2C). A **2nd kidney biopsy** revealed extensive lesions of tubulointerstitial nephritis (Fig. 2D)
- Two attempts of corticosteroids tapering due to severe osteoporosis (Lumbar spine [LS] T-score 4.9 and femoral neck [FN] T-score =2.9) faild in 2011 and 2016
- **2017**: 3rd AKI episode concomitant to hypercalcemia (2.93 mmol/l) and very high level of 1.25 dihydroxy-vitamin D (268 N :29-84 ng/l) was attributed to sarcoidosis relapse
- **2020**: pejorative evolution with progression to chronic kidney disease stage 4

1st kidney biopsy





Liver biopsy



2nd kidney biopsy

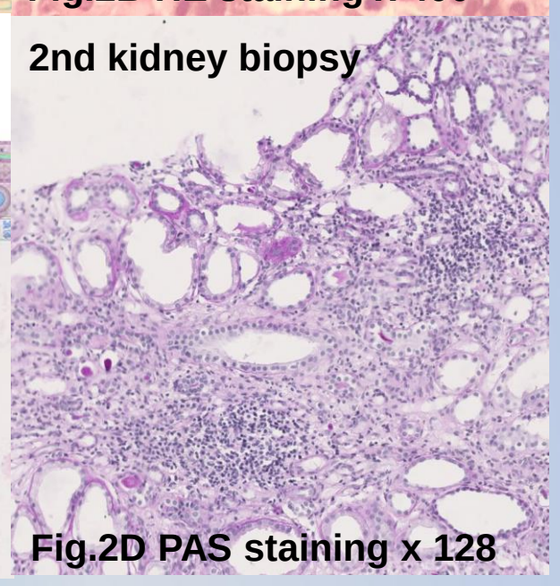
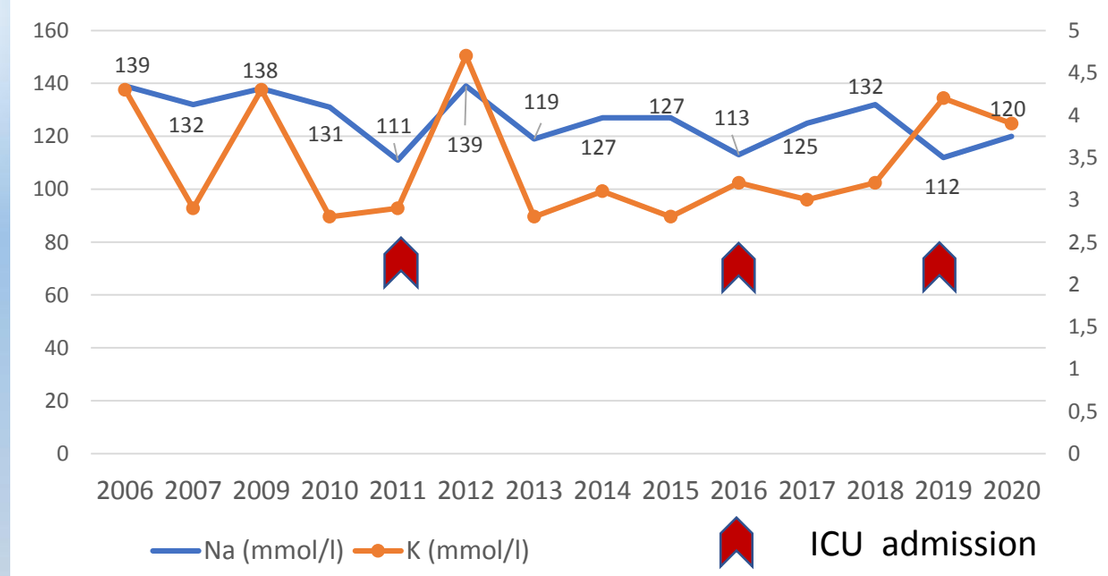


Fig.3 Na / K evolution and ICU related admissions



Discussion

- During the last 10 years, six ICU admissions in particular for severe hyponatremia with life-threatening neurologic complications such as seizures, confusion and behaviour disorders (Fig. 3)
- Worsening of osteoporosis: LS T-Score = 6.2 and FN T-Score = 3.2
- Bad side effects of long-term corticosteroids: Cushing syndrome with spinal deformations, amyotrophy and chronic gastropathy

Conclusions

- The diagnosis of SS with renal involvement is often delayed and the clinical management is very difficult
- **Sarcoidosis associated with salt loosing nephropathy (acquired Bartter-like syndrome)** is a real life-threatening condition. To our knowledge, this is the 2nd reported case.