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Sjögren's Syndrome Revealed by Obstructive Renal Failure: A Case Report and Review of the Literature

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Abstract

Introduction: Primary Sjögren's syndrome (SS) is the most common connective tissue disease after rheumatoid arthritis and affects mostly women between 30 and 40 years of age with an estimated prevalence between 0.1% and 0.6%. This observation illustrates an incidental finding of a case of SS in a young female patient in a context of obstructive renal failure (ARF) due to uterine fibroids. **Observation:** This was a 31-year-old woman hospitalized for anuric AKI (Acute Kidney Injury) with a creatinine level of 1247 µmol/l. Her history included sickle cell disease A/C and an unoperated uterine fibroid diagnosed 3 years ago. Approximately 2 months before her admission, her symptomatology was made of dizziness, physical asthenia, vomiting, polyarthralgia, morning rash, pollakiuria and oral dryness. Abdominal examination showed a painless transverse mass in the pelvis. Biological examination showed a CRP (C-reactive protein) level of 488 mg/l. The cytobacteriological examination of the urine was normal and the proteinuria was 1.35 g/24 hours. The CT scan showed kidneys measuring 110 mm on the right and 113 mm on the left associated with bilateral pyelo-caliceal dilatation on a large polymyomatous uterus of interstitial and submucosal type. Immunologically, the anti-nuclear factor, the rheumatoid factor and the anti-SSA antibodies were positive. The resumption of the interrogation within the framework of the research of the subjective dry syndrome to find a notion of intermittent xerophthalmia 4 months ago. The Schirmer test was positive in the left eye. The initial management consisted of a polymyomectomy after 3 sessions of hemodialysis. Background

treatment combining prednisone 5 mg/day and methotrexate 20 mg/week was started in parallel with the use of artificial tears. The evolution after twelve (12) months of treatment was favorable with a complete disappearance of the signs dry syndrome and full recovery of renal function. **Conclusion:** SS can have an insidious evolution and remain stable for many years, hence its fortuitous discovery in this case of obstructive ARF on uterine fibroid. In this context we insist on the interest of the immunological assessment in a patient in period of genital activity with a significant proteinuria and non-specific extrarenal signs.

Keywords

Obstructive Renal Failure, Fibroid, Sjögren's Syndrome

1. Introduction

Sjögren's syndrome (SS) is a slowly progressive chronic autoimmune exocrinopathy characterized by lymphocytic infiltration of the exocrine glands [1]. The dry syndrome characteristic of Sjögren's syndrome may be frugal or even absent and the extra-glandular systemic manifestations may be the circumstance of discovery of the connective tissue disease [2].

By definition, Sjögren's syndrome is said to be primary or "sicca syndrom" when the manifestations are not associated with another systemic disease pulmonary fibrosis, (usually rheumatoid arthritis, but also systemic lupus erythematosus or primary biliary cirrhosis…). Sjögren's syndrome is classified as secondary when it is associated with other clearly defined systemic diseases [3].

Primary Sjögren's syndrome (SS) is the most common connective tissue disease after rheumatoid arthritis [1]. The disease predominantly affects women (90%) between the ages of 30 and 40 years, with an estimated prevalence of 0.1% - 0.6% according to US-European consensus criteria [4]. Secondary SS occurs in 30% of patients with rheumatoid arthritis, 10% of patients with systemic lupus erythematosus, and approximately 5% of patients with scleroderma [2]. The annual incidence is approximately 4 new cases per 100,000 population in Olmsted County and Slovenia [4] [5].

In the literature, the prevalence of renal involvement in SS varies considerably from 2% to 67%. The modes of manifestation are dominated by tubular involvement, with glomerular involvement being rarer and obstructive involvement exceptional [6].

2. Case Presentation

This is a 31-year-old woman hospitalized in the nephrology department of G point University Hospital for anuric AKI with 1247 µmol/l creatinine. Her history included sickle cell disease A/C and an unoperated uterine fibroid 3 years ago.

Approximately 2 months prior to admission, her symptomatology consisted of dizziness, physical asthenia, vomiting, polyarthralgias, morning twitching, pollakiuria and dry mouth.

Physical examination on admission revealed a temperature of 37.2˚C, blood pressure of 130/80 mmHg, and heart rate of 80 bpm. Cardiopulmonary auscultation was normal. Abdominal examination revealed a painless transverse mass in the pelvis.

On ultrasound and CT scan, the kidneys measured 110 × 77 × 47 mm on the right and 113 × 72 × 57 mm associated with bilateral pyelo-caliceal dilatation over a large polymyomatous interstitial (measuring 30 × 22 mm and 24 × 16 mm, respectively) and submucosal (measuring 24 × 14 mm and 18 × 14 mm, respectively) uterus (**Figure 1** and **Figure 2**). The frontal chest radiograph was normal.

The biological workup showed a biological inflammatory syndrome with a CRP of 488 mg/L. Urine cytobacteriological examination was normal and proteinuria was 1.35 g/24h.

Immunologically, anti-nuclear factor, anti-rheumatoid factor and anti-SSA antibodies were positive. Thyroid function was normal.

The resumption of the interrogation within the framework of the research of the subjective dry syndrome to find a notion of intermittent xerophthalmia 4 months ago. Schirmer's test was positive with a wetting of the blotter paper strip of less than 4 mm at five minutes in the left eye. A biopsy of the accessory gland was not performed.

The management was multidisciplinary with an initial treatment of the patient by gynecology which consisted in the realization of a polymyomectomy by laparotomy (**Figure 3**) at D5 of the hospitalization after 3 sessions of hemodialysis whose indication was a total anuria on IRA stage III of KDIGO. Follow-up was simple, with resumption of diuresis and normalization of creatinine level to 74 µmol/L on day 35.

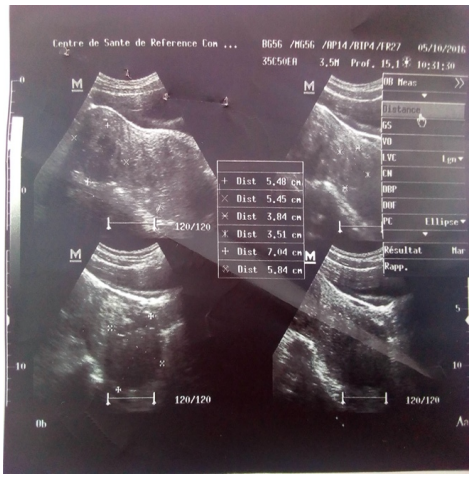

Figure 1. Echotomographic evidence of a globular uterus with the presence of four (4) subserosal and intramural myomas.

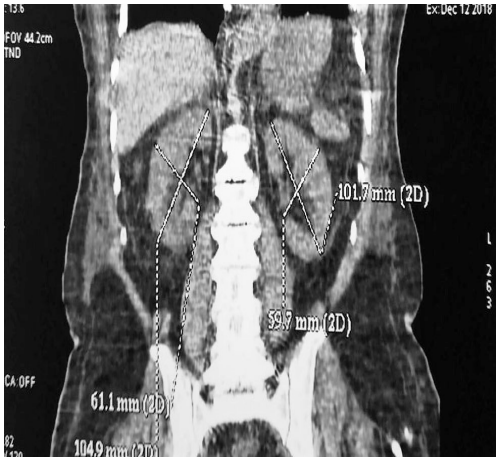

Figure 2. CT scan showing dilatation of the uretero-pyelo-caliceal cavities.

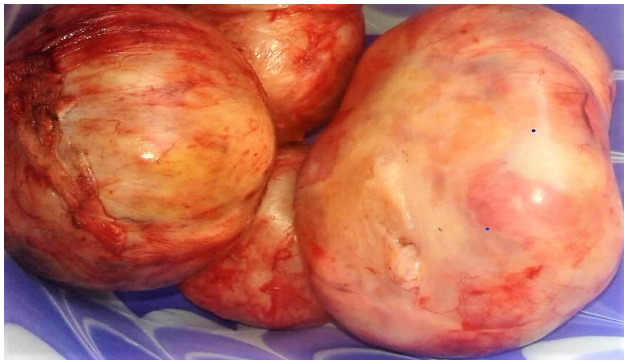

Figure 3. Surgical photo of the four myomas after excision.

The non-specific treatment involved artificial tears and other associated measures (smoking cessation and avoidance of certain drugs).

Basic treatment of SS syndrome was initiated with low-dose prednisone 5 mg/day and methotrexate 20 mg/week. Regular follow-up based on adapted clinical and biological examinations was performed at M1, M3, M6 and M12 respectively.

The evolution at M12 was favorable with complete disappearance of the signs of dry syndrome and complete recovery of renal function.

3. Discussion

SS can occur at any age with a clear female predominance: 9 females to 1 male [7] [8]. Renal involvement is in the form of tubulopathy in 20% of patients or more exceptionally glomerular nephropathy with or without cryoglobulin [6].

Our 31-year-old patient presented with obstructive AKI due to polymyomatous uterus.

The systemic manifestation of immuno-inflammatory nature reflects an infiltration of the exocrine glands, predominantly in the salivary and lacrimal glands, responsible for xerostomia and xerophthalmia. Other extrarenal signs may be found such as joint pain or pulmonary signs [2].

In our case, apart from dry mouth, no other signs of a subjective dry syndrome were reported by the patient at first sight. It was in the face of an immunological workup in favor of SS that the history was taken and the Schirmer test was positive.

The medications previously taken by the patient were analyzed and were not responsible for ocular and/or oral dry syndrome.

The dry syndrome may be symptomless or even absent during the first months of the disease, with sometimes even a paradoxical hyper-lacrimation or hypersalivation that may lead to a misdiagnosis [9]. Thus, the diagnosis is not always easy to make and is often delayed by several years due to the variability of the clinical presentation and the insidious progression of the disease. For a better homogeneous diagnostic approach, the European and American consensus diagnostic criteria, currently used in routine practice, take into account subjective and objective clinical parameters, as well as immunological and histological biological criteria (**Table 1**) [10]. New criteria were proposed in 2012 by the American College of Rheumatology (ACR), which consider only objective dry eye, lymphocytic salivary infiltrate, and the presence of immunologic abnormalities (**Table 2**) [11].

Table 1. AECG diagnostic criteria for SS [10].

1) Ocular symptoms	At least one of the following 3 criteria - Daily, persistent and bothersome sensation of dry eyes for more than 3 months - Frequent sensation of "sand in the eyes" - Use of artificial tears more than 3 times a day
2) Oral symptoms	At least one of the following 3 criteria - Daily sensation of dry mouth for more than 3 months - As an adult: recurrent or permanent episodes of parotid swelling - Frequent consumption of liquid to swallow dry food 2/Oral symptoms
3) Objective signs of ocular involvement	At least one of the 2 tests below positive - Schirmer's test ≤ 5 mm in one of the 2 eyes - Van Bijsterveld score ≥ 4
4) Objective signs of salivary involvement	At least one of the 3 tests below positive - Unstimulated salivary flow - Salivary scintigraphy - Parotid scintigraphy
5) Histological signs	Lymphocytic sialadenitis (focus score ≥ 1 on BGSA) or Chisholm grade 3 or 4
6) Presence of autoantibodies	Presence of anti-Ssa (Ro) or anti-SSb (La) antibodies

The diagnosis of primary SS is made when: four out of six items are present, with the mandatory presence of item 5 (histology) or item 6 (immunology); three of the four objective items (items 3, 4, 5 and 6) are present. The diagnosis of secondary GSS is made when item 1 or 2 is present and two of items 3, 4 and 5 are present.

Table 2. Criteria proposed by the ACR in 2012 [11].

Anti-SSA and/or anti-SSB antibodies or (anti-nuclear antibodies at a level ≥ 1/320 and rheumatoid factor positive)
Presence of lymphocytic infiltrate on accessory salivary gland biopsy with focus score ≥ 1
Ocular staining score after examination with Lissamine green and fluorescein ≥ 3 (Ocular Staining Score)

According to these criteria, the diagnosis of Sjögren's syndrome can be made when 2 of the 3 criteria above are present.

SS may be primary or associated with another systemic disease (rheumatoid arthritis, systemic lupus erythematosus, inflammatory myopathies or scleroderma) [12].

Our case redundantly met the primary diagnostic criteria of primary SS according to the criteria established by the American European Consensus Group (AECG).

Adnexal gland biopsy, ophthalmic staining with Lissamine green and fluorescein were not performed due to insufficient technical facilities.

The data in the literature concerning the impact of SS on the occurrence of obstetrical events are few, some studies find a higher rate of prematurity (birth < 37 weeks). Other studies have found a higher rate of low birth weight for gestational age and a higher rate of caesarean sections [13].

In our case it was a nulligravida patient and we can thus clearly say that the association of dry syndrome and obstructive renal failure by uterine fibroid would be the result of a coincidence. However, it is clear that the extra renal signs found in our patient were manifestations very often described in the literature.

4. Conclusions

SS requires an important diagnostic rigor in view of the non-specific character and the diversity of the clinical manifestations and the biological anomalies observed.

It can have an insidious evolution and remain stable for many years, hence its fortuitous discovery in this case of obstructive ARF on uterine fibroid. In this context we insist on the interest of the immunological assessment in a patient in period of genital activity with a significant proteinuria and non specific extra-renal signs.

Consent

Informed consent was obtained from patients.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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