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Lupus Nephropathy: Observation of a Case: Clinical Aspect, Evolution and Management at the West Guyana General Pediatric Service

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Abstract

Renal damage is one of the most frequent and serious manifestations of systemic lupus erythematosus (SLE). Even if this disease is the prerogative of female adults, it can affect children especially during the peri-pubertal period. The aim of this work is to highlight the clinical-biological and progressive characteristics of lupus nephropathy in children.

Keywords

Nephropathy, Lupus, Child

1. Introduction

Lupus nephropathy (NL) is a frequent complication of systemic lupus erythematosus (SLE): 30% to 50% of patients immediately suffer from kidney damage, and monitoring shows that 60% of adults and 80% of children will present early or later such an attack. Black patients are at greater risk of developing NL [1]. A better understanding of immunological mechanisms and an identification of risk factors have opened the way to a better therapeutic approach and made it possible to transform the prognosis of patients: the five-year survival of patients with proliferative glomerulonephritis increased from 17% before 1970 to more than 80% currently [2].

Often, the first signs are isolated proteinuria (>1 g/24h) or nephrotic syndrome, activation of the urinary sediment with leukocyturia and/or erythrocyturia and a reduction in glomerular filtration. Nearly 20% to 50% of patients with

SLE present with hypertension, a proportion increased in cases of NL and especially proliferative glomerulonephritis.

The biological picture is common to patients with or without NL: anemia, leukopenia, anti-phospholipids in more than 40% of NL [3] anti-dsDNA in almost 90% of cases and lowering of complement. Anti-Sm is found in 30% of LEDs, this frequency is increased in NL.

Various markers have been associated with an increased risk of renal failure in patients with SLE: an initial serum creatinine > 100 µmol/l, a hematocrit level as well as proteinuria > 3.5 g/24h, a lowered C3, hypertension, prolonged duration of renal damage, lack of normalization of creatinine after 48 weeks of treatment and age > 30 years. But it is above all certain elements of renal histology which have the most value in indicating a poor prognosis: presence of cellular crescents, interstitial fibrosis, tubular atrophy, high activity or chronicity index [4] [5].

Many studies have attempted to specify which marker would predict recurrence of NL, but without success; the various markers essentially help us confirm a lupus flare. An increase in anti-DNA titer has often been associated with relapse of NL, [6] and retrospectives even show up to 89% relapse within ten weeks following an increase in anti-dsDNA titer, but this marker may also remain elevated regardless of any recurrence. Other markers have been associated with recurrence of NL: elevation of C1q-binding protein or anti-endothelial cell IgG, decrease in complement (C3, C4) or drop in hematocrit level.

2. Observation

The observation focused on a case of lupus nephropathy diagnosed after an etiological assessment in a 15-year-old girl of Haitian origin who was treated for severe renal failure of unknown origin without favorable outcome, who arrived at the emergency room in a picture of septicemia on left femoral catheter in place in the fistula with peri-orificial inflammatory signs plus pain, associated with swelling of the left leg to be dialyzed. During this observation, in addition to the clinical aspect, psychosocial care was also taken into account with multidisciplinary involvement. All the staff in the pediatric department participated in the success of this treatment. The patient's sister was called upon, as she was the only responsible person who could address certain departmental concerns. Certain difficulties have been reported on a social level, making a hospital stay very prolonged.

The onset of the disease dates back around ten months with joint and muscle pain, repeated vomiting for a year and under corticosteroid therapy with prednisolone.

Admission examination: Weight: 52 kg, Temperature: 38.6 degrees Celsius, Blood pressure: 121/70mmhg, Heart rate: 134 beats/min; 100% oxygen saturation in ambient air. She presented with extreme fatigue and a deterioration in general condition, clear consciousness, no other neurological disorder was re-

ported. No swallowing, respiratory, sphincter or dysautonomic problems. Hemodynamic parameters were stable and good fluid status, no purpura. Heart sounds were audible and regular without signs of heart failure.

The entry blood test

Hemoglobin level: 5.7 g/dl; leukocytes: 6877/mm^3; Neutrophils: 4910/mm^3; platelets: 420,000/mm^3; Reticulocytes: 106,348/mm^3; Sodium: 133 mmol/l; Potassium: 5.3 mmol/l; Alkaline reserve: 18 mmol/l; serum calcium: 1.98 mmol/l; Urea: 16 mmol/l; Creatinine: 998 micro mol/l; CRP: 137 mg/l; PCT: 24.5 ng/l; Albumin: 21 g/l; Proteins: 71 g/l; ASAT: 28 IU/l; ALT: 17 IU/l; GGT: 32 IU/l; PAL: 130 IU/l; CPK: 565 IU/l; LDH: 463,463 IU/l; TP: 76%; TCA: patient: 32; TCA: witnesses 31; Uric acid: 238 mg/dl; Ferritin: 1103; blood group: O positive; hemoglobin electrophoresis: AA; Combs test: positive; Search for BK: negative.

The blood cultures taken on 03/07/20: show *Staphylococcus caprae* and the culture taken at the point of catheter removal revealed *Staphylococcus haemolyticus*;

Malaria research: negative; Beta HCG: negative; HBsAg serology: negative; DENGUE: IgG positive; HIV serology: negative; protein electrophoresis: gamma globulins: 31.5% or 21.7% (N: 7.4 - 14); ECBU: Klebsiella infection: treated with broad-spectrum antibiotic (Meropenem). Faced with this table, we retained the following diagnostic hypotheses:

- ✓ *Staphylococcus caprae* and *Staphylococcus haemoliticus* septicemia on catheter;
- ✓ Hemolytic anemia currently being explored;
- ✓ End-stage renal failure on dialysis
- ✓ Secondary hypertension probably linked to his nephropathy.

2.1. Diagnostic Suspicion

1) Acute tubular necrosis secondary to hemolysis

2) Rapidly progressive glomerulonephritis as part of a systemic disease or vasculitis

- ➢ Ultrasound of the soft parts of the neck: ultrasound appearance of bilateral parotitis predominantly on the left associated with bilateral inflammatory lymphadenopathy also predominant on the left in connection with lymphadenitis. No abscessed collection visualized. Edematous infiltration of bilateral soft tissues.
- ➢ Chest X-ray: absence of focus.
- ➢ Spine X-ray: for low back pain without bony abnormalities but stercoral stasis.
- ➢ Renal ultrasound: shows isolated hypotony of the calyceal font, bilateral and symmetrical, without visible mechanical obstacle, peritoneal fluid layer consistent with renal insufficiency described. Uncomplicated gallbladder lithiasis.
- ➢ Transthoracic ultrasound: no infective endocarditis.

- Brain MRI shows T2 and FLAIR, periventricular and focal cortical hypersignals of paramedian frontal location predominant on the right, bilateral paramedian parietal, right posterior temporal, suggesting in first hypothesis neurolupus.

2.2. Treatment on Admission

- ✓ Removal of the femoral catheter;
- ✓ Kayexalate 1 measuring spoon per day;
- ✓ Paracetamol 15 mg/kg/6hours if pain and/or fever;
- ✓ Water restriction; 1 liter of water per day at the beginning then 750 ml/day;
- ✓ Amlor: 5 mg/day;
- ✓ Lasilix: 80 mg morning noon evening then 500 mg per 24 hours using an electric syringe;
- ✓ Blood transfusion: 2 CGR then 48 hours after 04 CGR;
- ✓ Two dialysis performed within 5 days;
- ✓ Vancomycin: 1 gram per dialysis session then 1.5 gram;
- ✓ Gentamycin: 160 mg per dialysis session;
- ✓ Calciparin: 12,500 IU per day.

2.3. Evolution

Conscious patient, wide awake, not painful, afebrile, blood pressure peaks at 165/112mmhg, puffy face, takes approximately 900 ml of water for a diuresis of approximately 200 ml per day since her admission. Then a regression of diuresis to 60 ml of urine per day, difficult to obtain despite continuous Lasilix. A fistula was made bringing little urine less than 100 ml per day.

- **On the respiratory level:** eupneic in ambient air saturation 100%, no sign of respiratory struggle, no rales, symmetrical vesicular murmurs.
- **On the infectious level:** constant apyrexia under vancomycin at the time of dialysis. Repeat blood cultures were negative without germs.
- **On the digestive and nutritional level:** we noted abdominal pain without defense or contracture, no hepatosplenomegaly, the lymph node areas are free, no ascites, frequent constipation resolved by laxative and salt-free diet.
- **On the skin:** notion of pruritus appearing during hospitalization without joint damage.
- **On the metabolic and endocrine level:** hypocalcemia treated by the intake of Vitamin D, 25(OH) D3: 21.2 micrograms/l (N > 30), parathyroid hormone insufficiency: 315 pg/ml (N < 39).
- **Neurologically:** generalized tonic-clonic convulsions without sensory-motor deficit and meningeal syndrome: treated with valium and Dilantin during seizure episodes, then the introduction of URBANYL: 10 mg/day for 03 days every evening and KEPPRA 500 mg every 12 hours.
- **On the hematological and hemostasis level:**

Hemoglobin level: 4.6 g/dl; Leukocytes (WBC): 4020/mm^3; Polynuclear neu-

trophil (PNN): 1853/mm^3; Platelets: 437,000/mm^3; Reticulocytes: 592,000/mm^3; C-reactive protein (CRP): 50 mg/l; Procalcitonin (PCT): 24.5 ng/l; Urea: 13.6 mmol/l; Creatinine: 776 micromol/l; NTproBNP: 42,188.

Autoimmune anemia, transfusion during dialysis sessions, treated with Calciparin since its entry, then the relay made by COUMADINE at 2 mg per day, the effectiveness of the treatment estimated by measuring the INR which was at 2.9.

- **On the immunological level**:

✓ C3: 0.60 g/l (N: 0.90 - 1.80); C4: 0.13 g/l (N: 0.10 - 0.40), CH50: 24.1 U/ml (N > 31.6);
✓ AAN: speckled appearance; titration > 1280;
✓ Auto AC IgG Anti-native DNA: 115 IU/ml (N: <10);
✓ Auto AC Anti soluble nuclear Ag screening: 8.7 (N: <1.1);
✓ Auto AC Anti soluble nuclear Ag titration; SSA/Ro < 2 U/ml (N: <7);
✓ SSB/La: <2 U/ml (N: <7); Sm > 481 U/ml (N: <7); U1RNP: 29 U/ml (N: <5);
✓ Scl 70; 2 U/ml (N: <5);
✓ Anti-ARS antibodies (anti-Jo-1): <2 U/ml (N: <7) ;
✓ ANCA and specific ANC: negative;
✓ Auto AC Anti-dermo-epidermal basement membrane: <10 (N: <20);
✓ Auto AC IgG anti-beta 2 glycoprotein 1:10 U/ml (N < 7);
✓ Lupus anticoagulant antibodies: Patient/control ratio: 5.37 (N: <1.2).

A few follow-up reports:

Dates	18/03	30/03	02/04	04/04	05/04
Proteinuria g/l	4.01	13.07	11.32	13.01	9.18
Créatinine mmol/l		14	11	9	10
Rapport Alb/Créât	154	934	1029	1446	918

Results in favor of end-stage renal failure of lupus origin, associated with:

✓ A vitamin D deficiency which would be linked to high proteinuria accompanied by muscle weakness and reduced immunity;
✓ Hypocalcemia;
✓ Staphylococcus Caprae and Staphylococcus Haemoliticus septicemia on dialysis catheter;
✓ Secondary arterial hypertension (HTA).

Support before transfer:

Patient who received three boluses of Solumedrol for 72 hours then relayed with Prednisolone at 60 mg per day then supplemented with Plaquenyl 200 mg twice per day then adjuvant treatment was associated:

✓ Inexium: 40 mg in the evening;
✓ Calcidose: 500 mg per day;
✓ Foldin: 5 mg per day;
✓ Paracetamol: 500 mg every 6 hours if fever and/or pain;
✓ Movicol: 1 sachet morning, noon and evening;

- ✓ Morphine: dilution 1 mg for 1 ml: flow rate 0.6 mg/hour if pain not relieved by paracetamol;
- ✓ Coumadin: 2 mg per day;
- ✓ Dialysis three sessions per week.

The transfer was considered to the Cayenne Andrée Rosemon Hospital Center (CHAR) for a Renal Puncture Biopsy (PBR) which should be carried out there and a possible discussion of starting immunosuppressive treatment after puncture.

3. Discussion

The patient who arrived in the emergency room with renal failure of unknown origin associated with sepsis on a femoral catheter in the left foot to be dialyzed had no neurological signs at the start of her treatment. Who during his hospitalization began presented other symptoms such as: repeated tonic clonic seizures, pruritus, hyperthermia, abdominal pain and persistent postprandial vomiting with associated weight loss. The psychological aspect was also an important element. Because being far from the family environment and a lack of emotional support especially towards his biological family, adding to this the problem of accommodation contributed to having some difficulties in complying with treatment. The diagnosis of lupus nephropathy was made possible thanks to the etiological examinations which were carried out during his hospital stay, thus allowing adequate treatment. Lupus nephropathy is the kidney damage caused by systemic lupus erythematosus. Lupus is an autoimmune disease (dysregulation of the immune system with the development of antibodies directed against the patient's cells) mainly affecting young women, but can also affect men or children.

People with black skin have an increased risk of developing kidney damage from lupus. This damage can be present from the start of the disease, or appear secondarily during follow-up. It most often affects the skin or joints, and can be complicated by kidney damage. Our observation confirms this hypothesis. The kidney damage of lupus is not painful, and generally does not change the quantity or color of urine. It can occur during a skin or joint outbreak of the disease, but can also occur in isolation.

It can manifest itself by the appearance of edema (swelling of the feet), headaches (related to the appearance or worsening of high blood pressure), or severe fatigue, or be discovered after a vein thrombosis.

It can also occur without the patient feeling any particular symptoms, and must therefore be checked regularly by the doctor in charge of monitoring lupus, using the urine test strip (search for proteins, blood or leukocytes in the urine), or urinary laboratory tests (proteinuria, presence of red blood cells or white blood cells on ECBU) or blood (increase in blood creatinine). In our observation, an increase in creatinine was noted and at ECBU, we found a recurrent Klebsiella urinary infection responding well to Tazocillin.

Systemic lupus erythematosus is an autoimmune disease predominantly affecting women 9 times out of 10, its occurrence in men remaining rare [1]. Re-

garding neurolupus, the overall prevalence of these manifestations varies significantly between studies, ranging from 20% to 97% [2], highlighting the diagnostic difficulty and heterogeneity of these manifestations. As for the association of chronic inflammatory demyelinating polyradiculoneuropathy with systemic lupus erythematosus, very few cases are described in the literature [3].

In addition to treatments dictated by the clinic (steroids, antihypertensives, lipid-lowering agents, ACE inhibitors, etc.), the introduction of cytotoxic drugs has made it possible for several decades to transform the prognosis of patients with NL. Treatment is dictated by histology, the degree of activity of the lesions, renal function, taking into account the toxicity of the treatment. The treatment regimens are valid for both children and adults [7].

Transplantation was late attempted and studied in SLE. In the short term, graft survival is similar for patients with or without SLE, for identical populations in terms of donor types. A French study confirms these results with graft survival of 83% at one year and 69% at five years in the lupus group, compared to 82.5% and 70% in the non-lupus group. Patients with SLE can therefore benefit from a kidney transplant just as well as other patients. Current recommendations suggest transplantation three to six months after establishment of end-stage renal disease (ESRD), because there is potential for recovery during this time. Note that our patient was not transplanted after PBR. Baseline disease activity is very often less in ESRD patients, with or without dialysis, allowing a reduction in corticosteroid therapy. Although rare, a few cases of recurrence of NL have been described in transplanted patients, estimated at 2% of patients [8]. Graft losses due to recurrence of NL represent less than 5% of total graft failures [9].

4. Conclusion

Lupus nephropathy was discovered in our patient as part of an exploration of chronic renal failure of unknown origin, hence the importance of systematic research for urinary abnormalities and by a histological examination. This approach will make it possible to determine the patient's prognosis and above all to order the appropriate treatment for lupus patients. Even after remission, a recurrence of NL is always to be feared and patients must be regularly monitored.

Conflicts of Interest

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

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