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Case Report

Systemic Lupus Erythematosus and as Hemoglobinopathy: An Unusual Association about a Case in Lomé

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Abstract

We report a case of systemic lupus erythematosus in an adolescent with AS hemoglobinopathy, hospitalized from September 26 to October 20, 2018 in the internal medicine department of the Sylvanus Olympio teaching hospital of Lomé. It is a 17 years old girl, with a family history of sickle cell disease, hospitalized for a prolonged fever. The physical examination on admission noted: A fever at 40°C, a deterioration of the general state, a puffiness of the face with edema of the lower limbs, firm and painful adenopathies under chin and cervical, purpura, diffuse alopecia and non-deforming bilateral polyarthritis affecting the metacarpophalangeal and proximal interphalangeal joints. The biological assessment revealed an inflammatory syndrome, 24h proteinuria = 4,690 mg and hemoglobin AS, the native anti-DNA antibodies higher than 380 IU/ml. The diagnosis of systemic lupus erythematosus was made on the basis of at least 4 of the 11 criteria of the American College of Rheumatology 1997. The clinical manifestations of the lupus and AS hemoglobinopathy association in our patient are characterized by the absence of the major cutaneous-mucosal signs in particular the malar rush in butterfly wing, discoid lupus, photosensitivity and mouth or nasopharyngeal ulcerations.

Keywords: *Lupus; Hemoglobinopathy; Lomé; Togo*

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease characterized by a more or less disseminated damage to the connective tissue and whose causes remain unknown [1]. Its clinical manifestations are very polymorphic [2]. After the description of the first case in Africa by Basset in Senegal in 1960 [3], several cases have been reported in the same country [4,5] and in other countries of the sub-region [2,6-8], with a constantly increasing frequency probably due not only to the interest in the disease, but also to a better knowledge of the unusual and misleading forms. However, the lupus-hemoglobinopathy association is rare [9]. We report a case of this association revealed by a long-term fever in a department of internal medicine in Lomé.

Case Observation

She is a 17-year-old girl, high school student in final year class, admitted on September 26, 2018 in the internal medicine depart-

ment of the Sylvanus OLYMPIO teaching hospital of Lomé for a long-term fever (evolving for 4 months) that required unsuccessful care in health centers. In her history, she has no known tare but there is a concept of family sickle cell disease. The physical examination upon admission noted: A fever at 40°C, a blood pressure at 100/60 mmHg in both arms, a deterioration of the general state, a puffiness of the face with edemas of the lower limbs, firm and painful adenopathies under chin and neck, petechiae and bruise purpura predominant in the back, arms and legs, diffuse alopecia, pain of both hallux and non-deforming bilateral polyarthritis interesting the metacarpophalangeal and the proximal inter phalangeal joints. The additional exams had noted: lymphopenia (1,02/G L) and normocytic normochromic anemia (Hemoglobin rate 7.8 g/dl), blood sedimentation rate = 69 mm/h, C-Reactive Protein less than 6 mg/l, hypoalbuminemia (26,34 g/l) associated with a discreet polyclonal hypergammaglobulinemia (16,20 g/l) tending to form a beta2-gamma block in the electrophoresis of serum proteins, a thick negative drop, a negative tuberculin skin test (0 mm); Hbs negative antigen; a negative hepatitis C serology, a negative retroviral serology, a negative blood culture; a normal blood ionogram, a negative uroculture, blood sugar = 0.81 g/l, urea = 0.99g/l and creatinine = 21 mg/l on admission (urea = 0.17 g/l and creatinine = 7 mg/l on discharge); ASAT = 130 u/l (4N), ALAT = 47 u/(1.46N), gamma GT = 86 u/l (2N), PAL = 150 u/l, calcemia = 84 mg/l on admission and 87 mg/l on discharge and a phosphoremia = 29 mg/l, 24 proteinuria = 4690 mg to the input and 3654 mg/24 to the output, hemoglobin electrophoresis: AS heterozygous subject. Native anti DNA antibodies were greater than 380 IU/ml. There was a sinus tachycardia at 107 beats/minute and a first-degree atrioventricular block on the electrocardiogram. The chest X- ray was normal. The diagnosis of systemic lupus erythematosus was made on the basis of at least 4 of the 11 criteria of the ACR (American College of Rheumatology of 1997:

- Skin damage (purpura, alopecia)
- Joint damage (Non-deforming arthritis)
- Hematological disorders (Anemia, lymphopenia)
- Kidney damage (massive proteinuria)
- Positive native anti-DNA antibodies.

The patient received blood transfusions and a prednisone-based corticosteroid therapy (1 mg/kg/day) for 4 weeks and then a gradual reduction by 10% reduction in the previous dose every 15 days. The evolution was marked by apyrexia on the third day and a spectacular disappearance of joint pain and cervical lymphadenopathy in five days. But purpuric lesions and alopecia gradually regressed.

Figure 1: Purpura in the right leg.

Figure 2: Diffuse alopecia.

Discussion

This work allowed us to describe an observation concerning systemic lupus erythematosus in an AS hemoglobinopathy. It is an

association revealed by a prolonged fever. The occurrence of lupus in hemoglobinopathies is rare [9]. In fact, in a Senegalese series, Diop., *et al*. reported only 2 cases of sickle cell disease out of 161 cases of lupus collected in 12 years [5]. An association of sickle cell disease (SS) and systemic lupus erythematosus was also reported in Brazil in 2015 in an 8-year-old girl [10]. The female predominance of SLE is well known [2,4-8]. It has also been reported only in female subjects when associated with sickle cell disease [9,10] as in our patient. This preponderance of lupus in women is due to the role of estrogens [5]. The lupus disease occurs at a young age whether in Africa [2,4-9] or in Europe [11] even when associated with sickle cell disease [9,10]. However, cases of lupus have been reported in the elderly [4,5]. The prolonged fever revealing the disease in our patient was in the series of Diop., *et al.* [5] in 49% of the cases (79 cases out of 161), and in 10% of the cases in that of Ka., *et al* [4]. In reality, due to the preponderance sometimes of general signs, notably fever during lupus disease, many cases are treated as parasitic (malaria) and bacterial (typhoid fever) infections by inexperienced practitioners [12,13] and it is only if the fever persists that these patients are referred to internists. This is the case of our patient who dragged her fever for 4 months in health centers in our country before consulting in our service. The other manifestations of the disease found in our patient were cutaneous, articular, hematological and renal. On the cutaneous level, there was only a purpura and an alopecia. Purpuric and alopecic lesions were reported in the series of Diop., *et al.* [5] in Senegal respectively in 10% (16 cases out of 161) and 30.43% of cases (49 cases out of 161) against 13, 33% of alopecia in that of Ka., *et al.* [4] in the same country. In Togo, Kombate., *et al.* [2] and Teclessou., *et al.* [14] reported 43.75% (7 cases out of 16) and 28.44% (33 cases out of 116), respectively, of alopecia. The frequency of purpura and alopecia during lupus therefore varies from one study to another. However, the major mucocutaneous signs in this case Lupus discoid and malar rash in butterfly wing [5,14] have not been observed in our patient as well as photosensitivity and mouth or nasopharyngeal ulcerations. The joint damage found in our patient was also noted in the Togolese [2,14], Senegalese [4,5] and Beninese series [7,8]. The hematological manifestations were represented by anemia and lymphopenia in our patient, thus confirming the data in the literature [2,5,15,16]. Massive proteinuria and probable acute renal failure were the renal manifestations in our patient. The 24 hour proteinuria was significant in 41% of Diop., *et al.* patients [5] and in almost all the patients of Louzir., *et al* [17]. But our patient did not undergo a renal biopsy puncture for lack of technical platform so that we cannot attribute this glomerular nephropathy to lupus alone since sickle cell anemia can also be the cause. Therapeutically, only corticosteroid therapy was used in our patient as it is the case in some patients [5,14]. Indeed, corticosteroid therapy has an important place in the therapeutic arsenal of lupus [7]. However, the background treatment remains dominated by hydroxychloroquine [7].

Conclusion

The occurrence of lupus on sickle cell disease or sickle cell trait is rare. The epidemiological and clinical aspects of this association in our patient are similar to those of lupus occurring in a subject having AA hemoglobin apart from the absence of malar rash in butterfly wing, discoid lupus, photosensitivity and mouth or nasopharyngeal ulcers. Dare further research is needed to better understand the epidemiological and clinical aspects of this association.

Conflicts of Interest

None.

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