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SHARP Syndrome

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Abstract **Case Report**

SHARP syndrome or mixed connectivity is a syndrome associating signs of systemic lupus erythematosus, scleroderma, polymyositis /dermatomyositis, rheumatoid arthris, associated with a high level of anti-RNP antibodies. The clinicobiological diagnosis is easy with the dosage of the technical progress the prognosis is poor linked to multiple complications. The literature reports that mixed connectivitis do not appear to be more common in the black population. We report a polymorphic case in Mali diagnosed and followed up with a fatal outcome after a few months.

Keywords: SHARP syndrome, clinicobiological, connectivitis.

INTRODUCTION

By definition, connectivitis is a systemic condition (even if the initial attack affects only one organ) with an autoimmune component attested by the presence of at least one auto-antibody, and by definition the qualifier of connectivitis "Undifferentiated" which is applied to them implies an opposition with the characterized and defined forms of diseases that meet recognized and validated classification criteria (lupus, etc.) [1].

First described in 1972, SHARP syndrome is an autoimmune disease whose symptoms are considered to overlap with those of various inflammatory forms of autoimmune connective tissue disease: systemic lupus erythematosus scleroderma Sjögrene syndrome inflammatory myositis (dermatomyositis and polymyositis) [1-3]. The diseases most frequently involved are lupus scleroderma and rheumatoid arthritis. People with SHARP syndrome show symptoms of some of these conditions; in many cases, the characteristic symptoms of one of these diseases, particularly lupus or scleroderma, eventually outweigh the others [4].

OBSERVATION

Ms. T F, 40, Moorish, Malian, accountant, resident in VI, was hospitalized in Poly arthralgia. Interrogation and physical examination found sickle cell trait, family history (brothers and sisters). Polyarthralgia of the large and small joints of the inflammatory type, of moderate intensity, calmed by taking analgesics, sclerodactyly, alopecia with erythematous lesions of the photo exposed parts (trunk, back, neck, elbow folds, ears), ulcerations of the oral mucosa, generalized edema against a background of unquantified fever, permanent Physical asthenia.

Paraclinical assessment including: Blood count found: Anemia at 7.1g / dl Normocytic VGM 84 fl. Hypochromia TCHM 27pg Aregenerative Reticulocytes 36400 / mm3 normal platelets and leukocytes, CRP at 12.5 mg / l (N <5) positive, ESR: 1 hour 18/2 hour 38.

Renal function renal failure probably functional creatinine 258Umol clearance 27.79ml / min, proteinuria of 7g per 24h.

Liver function test elevated transaminases ASAT: 112 ALAT: 21. Normal hemostasis assessment, absence of circulating anticoagulant.

Electrophoresis, immunoelectrophoresis and weight determination of immunoglobulins Polyclonal increase in IGG to 25.7 g/l (N <16); VDRL-TPHA

serology positive, HBs antigen negative; Rheumatoid factor: negative.

Auto Antibody: Anti-nuclear Ab SM / RNP positive; Ac Anti SC70 Positive; Anti CCP Ab: Negative.

Arterial Doppler ultrasound of the lower limbs: complete and recent cruoric thrombosis of the entire deep venous network of the left lower limb. The head of the thrombus is located in the common iliac vein. The left internal saphenous superficial venous network is flexible and permeable.

The diagnosis of systemic lupus erythematosus was made due to: Photosensitivity, oral ulceration, polyarthralgia; pleural fluid effusion syndrome. 24H proteinuria: 7g / 24H, Normochromic Normocytic anemia, AC anti SM positive.

Scleroderma in front of a sclerotic modification of the skin which is stretched at the peri inguinal level, Sclerodactyly, AC anti SCL70 positive.

Treatment

Ivermectin 03mg tablet 05cp per bone in single dose, Prednisone 05mg tablet 02 tablets per os / day, Potassium 600mg LP tablet 01 oral / day, Omeprazole 20mg tablet 01 tablet oral/day, Calcium100mg-vitamin D 880 IU 01 tablet oral/12 hours, Hydroxychloroquine 200mg tablet 01 tablet per os/12 hours, Low sodium diet, Calciparin 500 IU O1 dose in SC / 12 hours.

Five days after hospitalization, the patient developed tonicoclinic seizures lasting a few minutes with post-critical amnesia, recovery of consciousness occurring after 20 minutes. The death was declared a few days later.

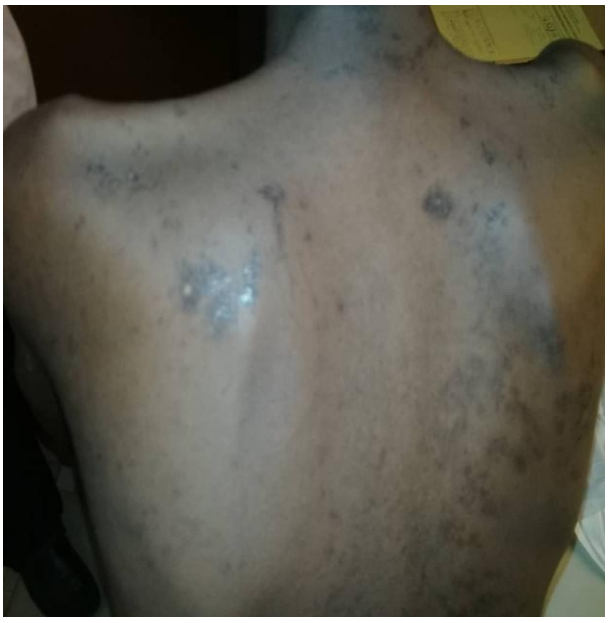

Photo-1: Dermatological lesions on the back

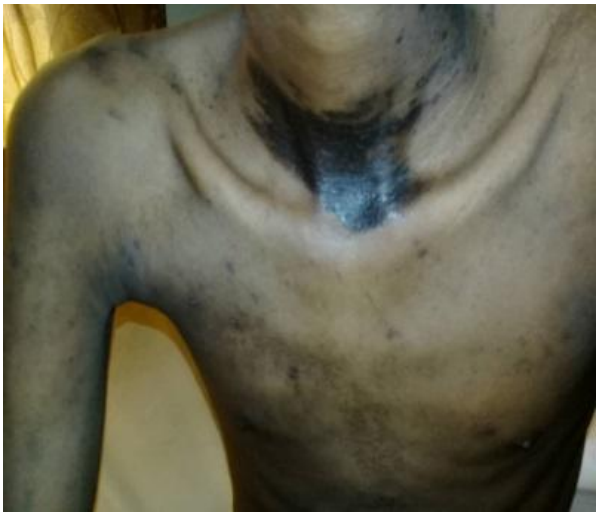

Photo-2: Dermatological lesions on the thorax

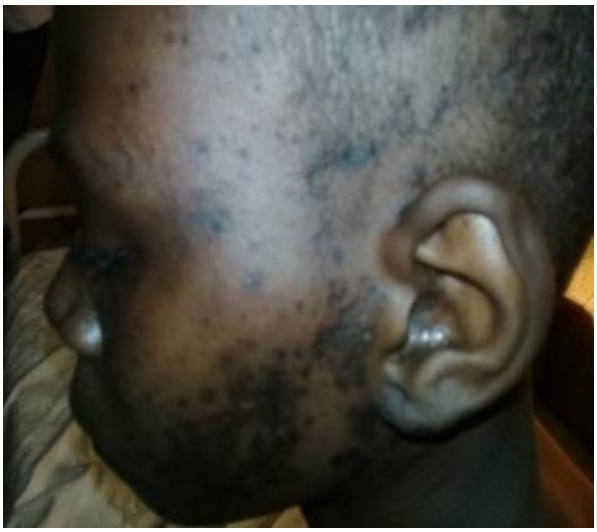

Photo-3: Dermatological lesions on the head

DISCUSSION

From an epidemiological point of view. Mixed connectivitis predominantly affects women (female / male sex ratio of 16 to 79) [2, 3]. The majority of cases occur between the second and third decade of life in our observation it is a woman in her forties. According to the clinical symptomatology of onset

Mixed connectivitis can manifest itself immediately with cardinal signs of SLE, SSc, RA and PM / DM or occur sequentially over time. Our observation initially reveals SLE, scleroderma or RA. In the event of joint damage. Joint involvement is the initial presenting sign of Sharp syndrome in 55% of Case.

The clinical picture is very close to that observed in early stage rheumatoid arthritis, which seems to be the case in our observation where the reason for consultation was polyarthralgia. At the mucous cutaneous level

Attacks resembling lupus manifestations associate malar rash, discoid plaques, alopecia, genital ulcers. Scleroderma-like manifestations include sclerodactyly, and sometimes scleroderma-like involvement of the skin that is more extensive.

In this observation we found sclerodactyly, alopecia, and photosensitivity. Other lung damage Lung involvement is common, up to 85% of patients, often asymptomatic [5]. They can reflect pleural damage (pleurisy, pleural thickening) in this observation we had pleural effusion syndrome. Neurological impairment.

Ten percent of patients with mixed connectivitis have a neurological manifestation [7]. The nervous system can be central or peripheral. These may be headache, comitiality, focal deficits, spinal cord involvement, meningitis aseptic, and disturbance of higher functions. A few cases of transverse myelitis indicative of the condition have been reported [8]. In our clinical case there was a comitiality.

Renal impairment: Renal involvement affects 10 to 20% of patients depending on the series [9]. In this study, the ratio of the level of anti-RNP / anti-Sm antibodies is inversely related to the frequency of renal impairment defined by proteinuria of at least 1.5 g / 24 h and impaired renal function. This results in the concept of renal protection of anti- (UI) -RNP antibodies.

We have during this obtained proteinuria at 7g/24 h followed by anuria after a few days of evolution.

From a biological point of view

General biology: The biological signs are not very specific and can be seen in other connectivities. They associate an inflammatory syndrome (elevation VS, PCR), anemia in 75% of cases, Sometimes secondary to hemolysis, leukopenia / lymphopenia in 75% of cases which tends to be correlated with the degree of disease activity [5].

Our patient presented with a high sedimentation rate; Elevated CRP Anemia at 7G / dl Normochromic Normocytic Hemoglobin;

Immunological assessment: AC anti SCL70 positive; AC anti SM positive; Proteinuria: 7 G / 24h; U1- RNP: From the support point of view.

The evolution

It was marked by the occurrence of neurolupus, deep vein thrombosis and survival of less than one year, all of which seem to agree with the data in the literature.

CONCLUSION

Mixed connectivitis is a systemic autoimmune disease which, by definition, incorporates clinical signs and symptoms of other connectivities (SLE, SCL, PR and DM / PM).

It is associated with often high titers of antibodies against ribonucleoproteins (anti-RNP), a prerequisite for diagnosis. The prognosis is poor with high mortality partly linked to complications, pulmonary hypertension, and proliferative vasculopathy.

REFERENCES

1. Sharp GC, Irvin WS, Tan EM, Gould RG, Holman H. R Mixed connective tissue disease--an apparently distinct rheumatic disease syndrome associated with a specific antibody to an extractable nuclear antigen (ENA). Am J Med. 1972;52: 148-59.
2. Rev Med Suisse 2203;vol -1.22995
3. Frances C, Barete S, Piette 1. C. Manifestations dermatologiques du lupus. Rev Med, Interne. 2008;29:701-9.
4. Amoura Z, Koutouzov S, Chabre H, Cacoub P, Amoura L, Musset 1. Presence of antinucleosome autoantibodies in a restricted set of connective tissue diseases: antinucleosome antibodies of the IgG3 subclass are rnarkers of renal pathogenicity in systemic lupus erythematosus. Arthritis Rheum. 2000;43:76-84.
5. Sullivan WD, Hurst DJ, Harmon CE, Esther JH, Agia GA, Maltby JD. A prospective evaluation emphasizing pulmonary involvement in patients with mixed connective tissue disease. Medicine (Baltimore). 1984;63:92-107.
6. Marshall 1B, Ravendhran N, Sharp GC. Liver disease in mixed connective tissue disease. Arch Intem Med. 1983; 143:1817-8.
7. Weiss TD, Nelson 1S, Wooisey RM, Zuckner 1, Baldassare AR. Transverse myelitis in mixed connective tissue disease. Arthritis Rheum. 1978; 21:982-6.
8. Bhinder S, Harbour K, Majithia V. Transverse myelitis, a rare neurological manifestation of mixed connective tissue disease--a case report and a review of literature. Clin Rheumatol. 2007;26:445-7.
9. Kitridou RC, Akmal M, Turkel SB, Ehresmann GR, Quismorio FP, Jr. Massry SG. Renal involvement in mixed connective tissue disease: a longitudinal clinicopathologic study. Sernin Arthritis Rheum. 1986;16:135-45.