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McDuffy's Hypocomplementemic Urticarial Vasculitis: A New Observation

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ABSTRACT

We report a 14 years-old- girl case, with no medical history, admitted to our department for exploration of paroxysmal abdominal pain lasting for a year without transit disorder. Clinical examination showed a sensitive abdomen with presence of multiple urticarial lesions. Laboratory examination revealed a hypocomplementemia: CH50 = 22.6UI / ml (N: 41.68-95, 06UI/ml), C3 = 0. 25 g/l (N: 0.74-1.43), C4 = 0.02 g/l (N: 0.14-0.34). The C1-Inhibitor rate and function dosage were normal and anti-C1q antibody was positive in 107 kU/l (N < 20). The diagnosis of hypocomplementemic urticarial Vasculitis has been retained and the patient was treated with the prednisone at a dose of 1 mg/kg per day with good clinical evolution.

BACKGROUND:

Hypocomplementemic urticarial vasculitis is a rare disease. It is characterized by recurrent urticaria associated with hypocomplementemia and the presence of antibodies directed against the C1q complement fraction [1]. We report a new case of hypocomplementemic urticarial vasculitis.

CASE-REPORT:

A 14-year-old girl with no medical history presented to the internal medicine department for exploration of paroxysmal abdominal pain evolving for a year. These pains occurred in paroxysms and were not rhythmic by meals. The patient had no transitory or rectal bleeding disorders.

She was hospitalized, three months before in visceral surgery department for intense right iliac fossa' pain that suggested appendicitis. The operative report showed a healthy appendix and the anatomical examination was without abnormality.

During the interrogation, she reported recurrent urticaria in the abdomen and two lower limbs. Its appearance was concomitant with pain and it resists to anti-histhaminics. She also had polyarthralgia of inflammatory schedule affecting both small and large joints.

The physical examination showed no fever. The patient was agitated because of pain. The abdomen was sensitive with diffuse urticarial lesions as shown in (Figure 1). Blood pressure was at 120/80 mm Hg and cardiac auscultation was normal. Neurological and osteoarticular examination were without abnormalities.

Blood count showed white blood cells at 5600 cells / mm3 with lymphopenia at 900 cells / mm3. C-reactive protein was at 34mg / l (Normal value <8mg/l), serum creatinine was 54µmol / l and sedimentation rate was at 8mm at the first hour. Amylase and lipase dosage were normal. Protein electrophoresis, liver function, and thyroid status were without abnormalities. The cytobacteriological examination of the urine showed no hematuria and the 24-hour proteinuria was negative.

The abdominal computed tomography was performed showing a regular concentric thickening extended to the entire ileum with a peritoneal effusion of low abundance. Cardiac ultrasound was normal.

The biopsy of the cutaneous lesions showed a leukocytoclastic vasculitis.

In order to support the diagnosis, hepatitis B and C virus serology was done and negative. Antinuclear antibodies, anti-native DNA, anticytoplasm of neutrophils and anticardiolipins were negative. The search for rheumatoid factor and cryoglobulinemia was negative.

In addition, there was a hypocomplementemia: CH50 = 22.6UI / ml (N: 41.68-95.06UI / ml), C3 = 0.25g / l (N: 0.74-1.43), C4 = 0, 02 g / l (N: 0.14-0.34). The weight and function of inhibitor C1 were normal.

Hypocomplementemic vasculitis was suspected. The anti-C1q antibody test was positive at 107 kU / l (N <20) confirming the diagnosis.

The patient was treated with prednisone at a dose of 1 mg/kg per day combined with an antihistaminic treatment with a good clinical and biological evolution.

There was no recurrence on corticosteroid dose depression. The current decline is one year.

DISCUSSION

Hypocomplementemic urticarial vasculitis syndrome (HUVS), or McDuffie syndrome, is a rare disease process that was first described by McDuffie et al1 in 1973. It occurs in adults aged from 35 to 45 years with a female predominance. It is characterized by complement activation, decreased C1q and presence of C1q precipitins [1-2]

Urticaria is the revealing sign in most cases. Skin lesions usually evolve in daily paroxysms for several weeks. It predominates on limbs and trunk but can touch the face, the palms of the hands and the plants of the feet. Abdominal pain is observed in 40% of cases and is contemporaneous with urticarial lesions as in our patient case. Joint manifestations are almost constant (83%) (arthralgia or arthritis) [3]. Renal involvement is observed in 25% of cases. Proteinuria is the most common sign of the disease. These may be minimal glomerular lesions, mesangial proliferative glomerulonephritis, rarely membranoproliferative nephropathy [4]

Pulmonary involvement is observed in 39% of cases and is often associated with pericarditis. Ocular involvement is reported in 32% of cases, mainly uveitis, episcleritis or conjunctivitis and exceptionally retinal vasculitis [5].

Diagnostic criteria for this condition have been proposed by Schwartz et al and are shown in Table 1 [6].

Table 1: Diagnostic criteria for hypocomplementemic urticarial vasculitis syndrome.

Major Criteria[a]	Urticaria> 6 months
	Hypocomplementemia
Minor Criteria[b]	Dermalvasculitis (by biopsy)
	Arthralgia or arthritis
	Uveitis or episcleritis
	Glomerulonephritis
	Recurrent abdominal pain
	Positive C1q precipitin test

a All criteria required for diagnosis.

b Two criteria required for diagnosis.

The exclusion criteria are as follows:

• significant cryoglobulinemia;

• presence of anti- DNA antibodies ;

• high rate of antinuclear antibodies;

• hereditary Complement' deficits;

• inhibitor of C1 protein deficiency.

Our patient had two major criteria and three minor criteria confirming the diagnosis. Exclusion criteria were absent. The main differential diagnosis is systemic lupus erythematosus (SLE), especially with joint and renal involvement and the presence of anti-C1q antibodies in 28-47% of SLE cases. Nevertheless, there are never enough criteria to retain the diagnosis of SLE[7,8]

Our patient didn’t have enough criteria to retain SLE which made this differential diagnosis easily eliminated.

Treatment of hypocomplementemic urticarial vasculitis remains poorly codified. Urticaria is little releaved by antihistaminic treatment as in our patient. Colchicine and nonsteroidal anti-inflammatory drugs remain also ineffective. Corticosteroids still are the most effective treatment with doses ranging from 0.5 to 1 mg/kg/day. Corticodependence is frequently observed leading to the addition of immunosuppressive therapy such as azathioprine, cyclophosphamide or ciclosporin [9].

Our patient had cutaneous manifestations and abdominal pain. She had no other systemic manifestations. The corticosteroid treatment was effective and the patient has not relapsed during corticosteroid depression.

CONCLUSION:

There is no specific treatment for HUVS and no consensus to an effective therapeutic regimen has been established.

Antihistamines may provide temporary relief. Nonsteroidal anti-inflammatory agents for symptomatic relief of joint pain may be helpful. Thus, treatment decisions in HUVS must be individualized according to the patient's clinical status.

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Figure

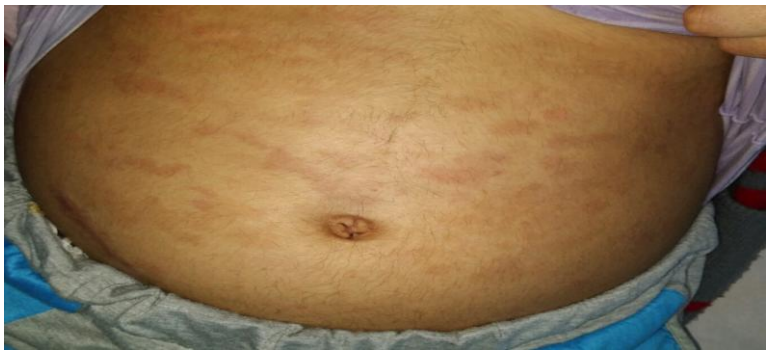

Figure 1: Active raised, erythematous eruption of wheals affecting abdomen.