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CASE REPORT

Leukocytoclastic vasculitis mistaken for chronic idiopathic urticaria

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ABSTRACT

Chronic urticaria is a commom health problem. Though not a rare disease, only part of these patients are correctly diagnosed and even fewer of them evaluated and treated in a proper way. Herein described, a case report of an old female with such diagnosis since 8 years ago. She suffered from multiple episodes of diffuse and itching hives and plaques evaluated by dermatologists and allergy specialists along these past years. Skin lesions of long lasting nature and pettechial appearance made this diagnosis reappraised and a skin biopsy was ordered. Vasculitis was shown. The bottom line: leukocytoclastic vasculitis mistaken for chronic urticaria is something to be considered when evaluating an apparent simple case of chronic idiopathic urticaria.

Key words: urticaria, cutaneous leukocytoclastic vasculitis, differential diagnosis

INTRODUCTION

Chronic urticaria is a commom health problem. Some surveys estimate 0.5% of population suffers from it. Although not a rare disease, only part of these patients are correctly diagnosed and even fewer of them treated in a proper way.[1]

The term “chronic” refers to symptoms lasting more than six weeks. It affects mainly women in their third and forth decades of life and it is related to major compromise in quality of life1.

Even though regarded as idiophatic in ninety percent of cases, the knowledge landscape has evolved in recent years and, lately, forty percent of chronic urticaria cases are considered possibly having an autoimmune substrate.[2] One-tenth of such cases are in fact vasculitis masquerading as simple urticaria.

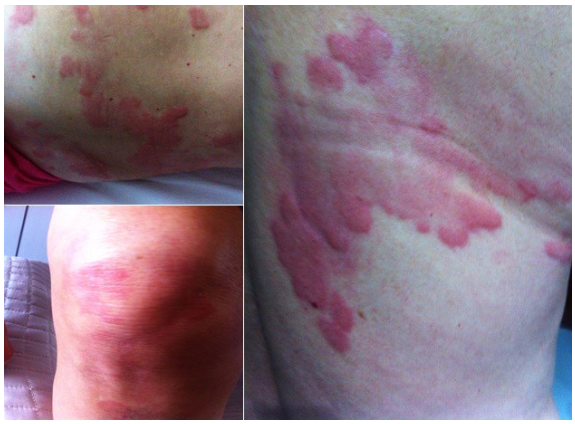

Figure 1, Diffuse and long-lasting hives and plaques on the abodmen, knees, and back..

CASE REPORT

A 69-year-old woman presented to the clinic complaining of itching and sometimes burning diffuse hives and plaques of long-lasting nature (Fig. 1). It was in her own words a flare from a recurring disease that led her to frequent outpatient visits to dermatologists and allergy specialists in the last 8 years. She´s been receiving multiple prescriptions of anti-histamine therapy these years long, but remission of such flares usually occurred only after a corticosteroid course. Some of these lesions definitely persisted more than 24 hours, in a fixed rather than migratory pattern and one of them had a pettechial appearance. She denied constitutional symptoms such as fever or weight loss, even malaise, but itching and sometimes burning sensation were really annoying and mental status examination revealed an anxious old woman.

As she portend a clinical diagnosis of chronic urticaria, Urticaria Severity Score (USS) was applied and the result was 59/93 points, which means a severe disease.[3]

A skin biopsy was ordered with a prior hypotheses formulated of urticarial vasculitis. Pathology report described small vessels in the superficial dermis with their walls infiltrated by eosinophils, neutrophils and

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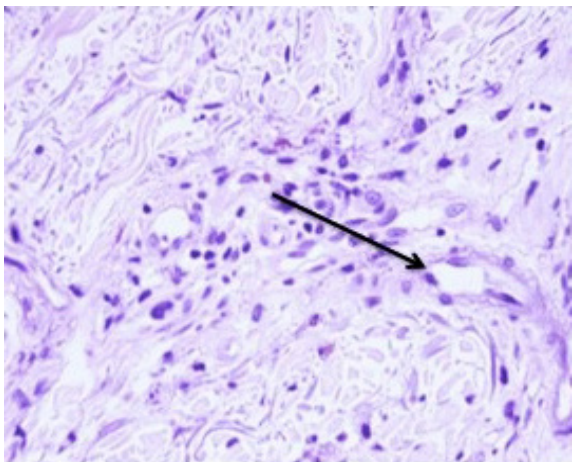

Figure 2, Histopathology of leukocytoclastic vasculitis with vascular damage promoted by nuclear debris from infiltrating neutrophils..

fibrin debris (Fig. 2). In summary, a pattern of leukocytoclastic vasculitis is shown.

Laboratory tests were done and also some images, with the aim of excluding an autoimmune substrate or an internal disease. ANCA, hepatitis C and B, HIV were all negative. Eritrocyte sedimentation rate was 47 mm (elevated) and ANA was positive, its titer 1:80 with a nuclear fine speckled pattern, but anti-DNA double strand, anti-RO, anti-LA were all negative and C3, C4 and CH50 returned normal results too. Anti-thyroid peroxidase antibodies were negative. Mammogram, abdominal ultrasound and chest CT were normal as well a colonoscopy.

Due to an adverse event to oral colchicine, with rash exacerbation, and a skin-limited vasculitis, she received a prescription of topical dapsone 5% twice a day, obtaining good control of skin lesions two months later (USS was reapplied, with a 17/93 score).[3,4]

Even though an apparent early and initial good recovery, medication was suspended by the patient and some weeks later she reported a flare of her disease, promptly remitting with oral prednisone 5mg twice a day and topical dapsone.

DISCUSSION

Urticarial vasculitis is the main differential diagnosis of chronic urticaria. A Medline search was done (via PubMed) for the terms "vasculitis" and "chronic urticaria", filtering "case report" and "last 10 years", and it returned 17 results with 10 related cases (Table 1).

Most of the time, this is a skin-limited vasculitis and very uncommonly constitutes manifestation of internal disease such as malignancies, systemic erythematosus lupus, Sjögren, ANCA-positive vasculitis, viral hepatitis or HIV.[5] Notwithstanding that, it is recommended a proper screening for such conditions.

As previously mentioned, in recent years the knowledge landscape has evolved and it is estimated that 40% of urticaria cases may portend an autoimmune substrate.[2] This seems to be a strong argument to defend a multidisciplinary evaluation in this disease, mainly in its chronic forms, involving internists earlier in the case approach.

As shown in table 1, some cases of urticarial vasculitis are related to infection (hepatitis C), malignancies and autoimmune diseases. In regards to laboratory results, it would be appropriate to emphasize the role of complement evaluation in this disease: low complement is related to organ damage, mainly kidney disease and possibly multi-organ involvement, leading to a more aggressive clinical course. Such cases tend to be selected for publication.[6,7,8,10,12,15]

Despite not having uniform agreement of its value, some immunologists suggest autologous serum skin test in patients with chronic urticaria. It consists in the injection of autologous patient serum collected during a disease flare into clinically normal skin. A positive test elicits an immediate wheal and allegedly relates to circulating autoantibodies to IgE or IgE receptors in mast cells ou basophils. Some case series suggest it can predict response to therapy with anti-IgE drug omalizumab.[11]

Even though considered rare entities, some forms of urticarial vasculitis are worth to mention: IgG4-related disease and Schnitzler syndrome.

IgG4-related disease is an immune-mediated condition of unknown etiology that can also simulate malignancy by organ infiltration with IgG4 positive plasma cells (an inflammatory pseudotumor). It is usually associated with elevated serum IgG4 concentration, can cause pancreatitis or lymphadenopathy, affecting mainly middle-aged or older individuals.[13]

Schnitzler syndrome is another form of urticarial vasculitis, characterized by chronic urticaria and IgM or IgG gammopathy. It usually affects male patients and is often accompanied by recurrent fever.[14]

CONCLUSION

In summary, urticarial vasculitis is not an unusual entity, possibly mistaken for chronic urticaria in daily medical practice. Clinical evaluation and treatment differ and the backbone of corticosteroid therapy should not be neglected, even though corticosteroid sparing therapies such as colchicine, hydroxychloroquine and dapsone may be successfully used.

Low complement signalizes to a possibly multisystem and more aggressive disease subtype.

CONFLICT OF INTEREST

None.

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Table 1. Summary of search strategy and results

Database	**Keywords**			**Filters**	**Results**	**Related**
Medline (via PUBMED)	"chronic urticaria" and "vasculitis"			"case report" and "10 years"	17	10
Author	Patient sex	Age	Laboratory	Related disease	Organ damage	Therapy
Salim SA et al, 2018[6]	Female	31	Low complement, low C1q		Glomerulonephritis	Cyclophosphamide and rituximabe, followed by maintenance therapy with corticosteroid and mycophenolate mofetil.
Sjöwall C et al, 2015[7]	Female	65	Low complement and anti-C1q antibodies.	Gastric carcinoid three years earlier.	Pleuritis and reduced diffusion capacity. Pericarditis.	
Kervarrec T et al, 2015[8]	Female	63	Low complement and anti-C1q antibodies.		Pericardial tamponade, pneumonitis and bulloous skin lesions.	
Kassim JM et al, 2015[9]	Female	72		Chronic lymphocytic leukaemia.		Chemotherapy.
Park C et al, 2014[10]	Male	32	Low complement, antinuclear antibodies (ANA) titer 1:40, speckled type.		Cardiac valvulopathy, arthritis and glomerulonephritis.	Corticosteroids, cyclophosphamide and mycophenolate mofetil.
Díez LS et al, 2013[11]	Three female patients	51, 54, 28	All patients had a positive autologous serum skin test.		Angioedema.	Disease refractory to corticosteroids, hydroxychloroquine azathioprine, colchicine and dapsone. Omalizumabe-responsive.
Pinto-Almeida T et al, 2013[12]	Female	49	Low complement.	Chronic hepatitis C and cryoglobulinemia.		Hepatitis C therapy.
Wakamatsu R et al, 2011[13]	Female	58	High IgG4, low complement and positive reumatoid factor.		Intra-abdominal lymph node swelling and hepatosplenomegaly	Symptoms subsided on prednisolone 15mg once a day.
Carlesimo M et al, 2010[14]			Monoclonal IgM and IgA gammopathy.	Schnitzler syndrome.		
Balsam et al, 2008[15]	Female	23	Low complement and low C1q.		Glomerulonephritis.	Corticosteroids and cyclophosphamide led to skin lesions resolution. Kidney failure responsive to plasmapheresis.

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