Urticarial vasculitis: a brief essay on the importance of observation

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Dear Editor,

A patient came into my office who looked sad and tired, but also strong and determined, given that she had gotten up at dawn to travel more than 200 km from her little town, Carnaúba dos Dantas, Rio Grande do Norte, to the state capital, Natal, to find answers and a diagnosis that, until then, no one had been able to give her. “How can I help you?” I asked. She replied, “Doctor, you are my last hope! I have a serious skin problem, I have been to four doctors and none of them wanted to look at me!” That's right, she reported that no doctor she sought out wanted to get up and have her undress so they could assess her skin lesions and perform something fundamental to our profession – a physical examination.

During the anamnesis, she reported having been diagnosed with an “allergy” approximately six months ago and that since then she had been taking high daily doses of corticosteroids and sedative antihistamines, which prevented her from continuing to work as an artisan due to intense drowsiness. During this period, she gained eight kilograms and began experiencing muscle weakness and hypertensive spikes.

During the examination, I realized that the lesions were probably compatible with a clinical picture of urticarial vasculitis (UV). I took something very simple that we can all have in the office, a glass microscope slide, and confirmed that the lesions did not disappear under pressure – a simple test for differential diagnosis between urticaria and UV.

I requested tests and a biopsy and, due to adverse effects from prolonged use, I began tapering her off corticosteroids. In a return visit, the tests showed normal levels of complement proteins (C3, C4, and C1q) and the biopsy confirmed vasculitis. Thus, I was faced with a diagnosis of normocomplementemic UV.

UV is a rare cutaneous vasculitis of small vessels characterized by recurrent episodes of papule-like lesions that tend to last for more than 24 hours and are accompanied by residual ecchymotic postinflammatory hyperpigmentation. The histopathological pattern is that of leukocytoclastic vasculitis, consisting of fibrinoid necrosis of the dermal vessel walls and neutrophil-rich perivascular inflammatory infiltrates. Although its etiopathogenesis remains unclear, UV is now considered to be caused by immune complexes that activate the complement cascade, leading to the exaggerated production of anaphylatoxins, which are responsible for the recruitment and activation of neutrophils. Based on serum complement levels, this condition can be categorized as normocomplementemic UV or hypocomplementemic UV (low levels of C1q and C4 and variably decreased levels of C3), the latter being associated with circulating anti-C1q autoantibodies and possible extracutaneous manifestations. Important differential diagnoses must be considered and excluded, such as bullous pemphigoid, Henoch-Schönlein purpura, lupus erythematosus tumidus, Wells syndrome, erythema multiforme, cutaneous mastocytosis, cryopyrin-associated periodic syndromes, etc.[1]

Although UV is mainly idiopathic, it may be associated with medication use, malignancy, autoimmunity, or infection. In some situations it can be difficult to treat, and treatment should be guided by the severity of cutaneous and systemic involvement. Corticosteroids are effective for cutaneous symptoms in most patients with UV, but long-term administration can lead to potentially serious adverse effects. Adding immunomodulatory or immunosuppressive agents often facilitates the corticosteroid tapering process and improves therapeutic efficacy. To date, there is no consensus on the best medication, with recommendations primarily based on case reports and retrospective studies.[2]

In the present case, an immunosuppressant was prescribed, in addition to joint monitoring with an endocrinologist. After six months of monitoring, the patient’s symptoms were in clinical remission and she had lost weight. She no longer had hypertensive spikes and was able to return to her handicrafts, which are her family’s source of income. However, the course of UV can be long-lasting and difficult to control clinically.

What we must immediately ask is why the value of such a crucial step in our daily medical practice – the “gaze” in all its dimensions – has been lost.

Whenever I see clinical cases like this, I am reminded of Samuel Luke Fildes’ (1843-1927) *The Doctor* (1887, The Tate Britain, London), an often-cited painting in discussions of the strengths or weaknesses of the medical profession.

This iconic work portrays a doctor on a house call for the child of an impoverished worker. The child's improvised bed consists of two chairs. The house's interior is humble and befitting that of a worker. The central figure is the imposing doctor, observing his patient, while in the background the father rests his hand on the shoulder of his wife, who is in a position of supplication or prayer. Fildes' skillful use of light and perspective focuses attention on the doctor's intent gaze. Other details include a pestle, cup, and spoon, suggesting that the doctor prepared a potion or poultice to apply to the sick child. However, evidence of then-current equipment, such as a stethoscope or a thermometer, is lacking. It is likely that by this time a doctor would have been using biomedical practices, which rely on quantitative and measurable evidence rather than qualitative observation and judgment.[3]

The painting prompts us to reflect on the current medical practice. Indeed, its essence is "patient-centeredness," an essential characteristic of the contemporary doctor-patient relationship and an important consideration for all physicians.

Much time has passed since the artist brought this scene to life, but its strong expression of the doctor-patient relationship continues to stir up deep feelings that surround the search for a cure. Although this piece was created in a completely different medical-social context, the ideal of healing and the desire to be cured is a perennial relationship that overcomes time and external changes, which gives this work a timeless quality.

Inspired by the painting "The Doctor", writer and thinker Rubem Alves (1933-2014) wrote *O Médico*, in which he states: *"I loved this painting the first time I saw it, without understanding it. Perhaps this is the reason why, as a young man, for many years, I dreamed of being a doctor. I loved the beauty of the image of a lonely man, fighting against death. When faced with death, we are all lonely. We love the doctor not for his knowledge or his power, but for the human solidarity that is revealed in his meditative waiting. And all of his failures (for are they not all doomed to lose the final battle?) will be forgiven if, in our helplessness, we realize that he silently remains and meditates together with us"* (translated from the original Portuguese).[4]

The Doctor (1887, The Tate Britain, Londres), by Samuel Luke Fildes (1843-1927)

Doctors, who are now pressured to work exhaustively to earn a decent salary, may be forgetting the importance and beauty of the gaze that Fildes represented and the need, as Rubem Alves argues, to *remain*, in the best sense of the word, with patients in their helplessness.

References

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