How not to miss autoinflammatory diseases masquerading as urticaria.

Urticarial skin reactions are one of the most frequent problems seen by allergists and clinical immunologists in daily practice. The most common reason for recurrent wheals is spontaneous urticaria. There are, however, several less common diseases that present with urticarial rash, such as urticarial vasculitis and autoinflammatory disorders. The latter include cryopyrin-associated periodic syndrome and Schnitzler's syndrome, both rare and disabling conditions mediated by increased interleukin-1 secretion. Apart from the urticarial rash, patients are suffering from a variety of systemic symptoms including recurrent fever attacks, arthralgia or arthritis and fatigue. Autoinflammatory diseases are often associated with a diagnostic delay of many years and do not respond to antihistamines and other treatments of urticaria. Also, the chronic inflammation may lead to long-term complications such as amyloidosis. It is therefore important not to miss these diseases when diagnosing and treating patients with chronic recurrent urticarial rash. Here, we present clinical clues and tips that can help to identify autoinflammatory disorders in patients presenting with chronic urticarial rash and discuss their clinical picture and management.