3. Acquired cold urticaria

Cold urticaria is characterized by the appearance of wheals or angioedema after exposure to cold (eg, cold weather, cold sweat, swimming in cold water, air conditioning, cold food or drinks). In most patients, no triggering agent is found, although in some cases the condition has been associated with viral or bacterial infections; leukocytoclastic vasculitis; malignancies; autoimmune, hematologic, or thyroid diseases; or drug ingestion.

Cold urticaria was first described in 1866 and accounts for 2% to 3% of physical urticaria. It can also be associated with other forms of physical urticaria, such as dermographism (21%), heat urticaria (10%), and cholinergic urticaria (8%). Familial syndromes (autosomal dominant) that manifest in childhood with cold urticaria have been described, and have been classified as hereditary periodic fever syndromes.

Although most cases of cold urticaria are idiopathic, a small percentage are secondary to infection with various viruses or bacteria, such as Epstein-Barr virus, cytomegalovirus, hepatitis B, hepatitis C, HIV, rubella, Mycoplasma, Treponema pallidum, Helicobacter pylori, and Toxoplasma gondii. In our patient, serology results were negative for all of these conditions. Doeglas et al, in a study of 39 patients with cold urticaria, found no significant relationship between incidence of cold urticaria and infection with Epstein-Barr virus. They did find, however, that other infections, such as those due to Mycoplasma, cytomegalovirus, or herpes simplex virus, were more common in patients with cold urticaria.

Diagnosis and prognosis

Diagnosis is confirmed by a provocation test: applying an ice cube to normal skin and checking for wheal formation. The piece of ice is held against the forearm, typically for 3 to 5 minutes. A positive result is a specific-looking raised wheal, which might be in the shape of the ice cube or might radiate from the area of contact. Some authors suggest that an early response to this test (less than 3 minutes) could be related to the possibility of a severe systemic reaction with hypotension and shock. Wanderer and Hoffman classified the condition into 3 groups according to the severity of the clinical manifestations: type I (localized lesions); type II (systemic phenomenon without symptoms of hypotension); and type III (systemic phenomenon with hypotension or shock), which can be life-threatening. Another test that has been used to diagnose this disease is to immerse the hand in water at a temperature of 10°C for 5 minutes; however, some patients have experienced systemic reactions and this test should be used with caution.

The prognosis of this disease is quite variable; there is no method to predict the evolution of acquired cold urticaria. It has been shown that the course of cold urticaria is considerably shorter (8 to 9 weeks) when associated with a viral infection compared with the mean duration of 6 to 9 years for idiopathic disease. General practitioners should consider consulting with a urticaria specialist (ie, an allergist or dermatologist) if patients present severe symptoms or atypical manifestations.

Treatment

Treatment is mainly based on prevention—patients should avoid contact with cold water and avoid eating very cold food until their symptoms have resolved spontaneously.

For our patient, cetirizine (10 mg/d) was prescribed to minimize symptoms, which was effective and did not cause any adverse effects. There have been reports of cases treated with desloratadine (5 to 10 mg/d), cyproheptadine, doxepin, terbutaline, histamine (H2) blockers (eg, cimetidine), and leukotriene antagonists (eg, montelukast). Cold urticaria can result in potentially serious, or even fatal, systemic reactions such as anaphylactic shock; therefore, affected individuals might have to carry an injectable form of epinephrine to use in the event of a serious reaction. Aquatic activities should be avoided; if that is not possible, patients should take prophylactic therapy and make sure that these activities take place under the supervision of a person trained in administering epinephrine.