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Anaphylaxis as a causative factor in loss of consciousness in an elderly woman. Mastocytosis — a case report

Abstract

A female patient, 72-years-old with recurrent systemic hypotension, was admitted to the Allergology Department. She had long been diagnosed with *urticaria pigmentosa*. She also had a history of planoepithelial vaginal cancer, mitral valvular insufficiency and osteoporosis. Recurrent episodes of anaphylaxis and mastocytosis were diagnosed at the Department of Allergology.

It is important to consider a systemic hypersensitivity reaction in differential diagnosis even in elderly patients, since it may mimic other medical emergencies. The unusual and broad spectrum of symptoms in mastocytosis usually requires the cooperation of different specialists.

Key words: mastocytosis, anaphylaxis, *urticaria pigmentosa*

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Introduction

In the case of an elderly patient with loss of consciousness, the main differential diagnosis is syncope due to heart disease, neurological disorders and miscellaneous conditions [1, 2].

Anaphylaxis is a severe, potentially life-threatening, systemic hypersensitivity reaction [3] that can mimic other medical emergencies, and thus should be considered in a differential diagnosis even in elderly patients [4].

Systemic hypotension leading to the point of unconsciousness is one of the most severe symptoms of anaphylaxis. However, in elderly patients this will not usually be considered causative when an anaphylactic reaction occurs for the first time.

In this paper, a case of recurrent systemic hypotension due to anaphylaxis in the course of mastocytosis is reported.

Case report

A female patient (JO), 72-years-old, was admitted as an outpatient to the Department of Allergology at the Medical University of Gdansk, Poland, in November 2008 because of recurrent syncopes due to hypotension and suspected of having mastocytosis. She had been diagnosed with *urticaria pigmentosa* a long time previously (Figure 1). The first skin lesions occurred when she was thirty years old. She also had a history of planoepithelial vaginal

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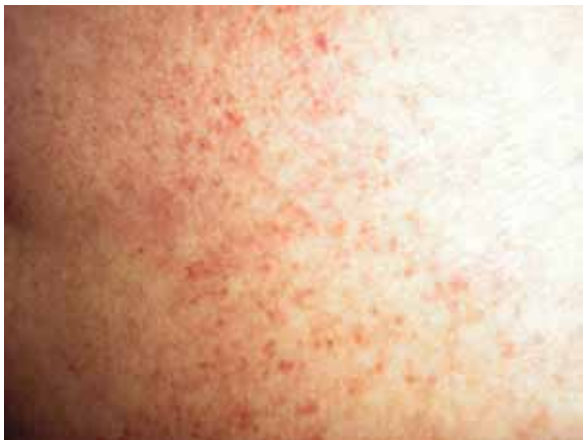

Figure 4. *Urticaria pigmentosa*

cancer (she underwent surgery and radiotherapy in 2000) and deep vein thrombosis a few years earlier.

At the time of admission she complained of a recurrent sudden weakness due to hypotension and four episodes of a loss of consciousness during the previous two years. The patient related these episodes to the intake of food (nuts, chocolate and strawberries) and drugs (antibiotics and heparin). In 1991, while visiting Italy, she developed anaphylactic shock-like symptoms after aspirin intake, although no medical documentation was available.

In September 2007 she was admitted to the Internal Diseases and Rheumatology Department in Warsaw, Poland, because of weakness and fever. At this time, an abdomen CT revealed hepato- and splenomegaly and a USG detected neck and axillary lymphadenopathy. She was seen by a haematologist but no final diagnosis was made; additional findings, however, included mitral valvular insufficiency (although not significant haemodynamically) and osteoporosis.

Finally, she was sent to the Allergology Department in Gdansk at her own request.

Consultation at the Allergology Department confirmed systemic mastocytosis. No subjective symptoms were present at the time of the consultation: cutaneous lesions presented as disseminated yellow-brown patches on the skin over her entire body; there were positive signs of Darier's disease; and the serum triptase level was highly elevated (> 200 μg/ml). Further examination included the following: bone marrow and skin biopsy, serum total and specific IgE concentration (it was not possible to perform a skin prick test because of generalized *urticaria pigmentosa*), abdominal imaging study, and the expression of mast cells CD25 and CD2. The patient was told to avoid triggering factors causing the release of mast cell mediators, such as aspirin, alcohol, some food (chocolate and nuts), rapid changes in temperature, rubbing, and exercise. She received an emergency kit — containing adrenaline for self-application, rapidly acting H1 antihistaminics and oral corticosteroids — and she was recommended to take non-sedating H1 antihistaminics as continuous prophylactic medication.

Discussion

Mastocytosis is a rare and difficult-to-diagnose group of disorders characterized by a pathological increase in mast cells in tissue, including skin, bone marrow, liver, spleen and lymph nodes. In mastocytosis, symptoms and signs of disease are typically related to the tissue mast cell burden and to the response for the release of mast cell mediators [5]. This unusual and broad spectrum of symptoms, therefore, requires the cooperation of different specialists [6]. A curative treatment for mastocytosis does not exist and therapy should be carefully considered for each patient. The management of patients within all categories of mastocytosis includes their avoidance of factors triggering acute mediator release, treatment of acute mast cell release (*i.e.* systemic hypotension), treatment of chronic mediator release, and an attempt to treat organ infiltration by mast cells [5].

Syncope is a common clinical problem. The estimated incidence of self-reported syncope is 6.2 per 1,000 person-years in the Framingam study [7]. The frequency of a fall in blood pressure on standing increases with age [8]. The prevalence of syncope in subjects aged 70 years and older is near 30% [9]. Syncope is a clinical manifestation of the interruption of global cerebral perfusion. In the elderly, especially in patients with a history of heart disease, a cardiac cause of loss of consciousness is the most likely. The most common neurogenic causes of a transient loss of consciousness are cerebrovascular disease [transient ischemic attack (TIA), or atherothrombotic or cardioembolic stroke] and epilepsy. Patients with TIA are at increased risk of stroke, cardiovascular events, and death. Transient ischemic attack and stroke must be recognized and treated urgently [10]. Neurogenic syncope could be associated with vertebrobasilar insufficiency and other abnormalities of the vertebral arteries *i.e.* arteriosclerotical plaques [11].

In anaphylactic episodes, the release of mast cell-derived mediators leads to vasodilatation, systemic hypotension and hypoperfusion of the central nervous system. In the case being described, the cause

of systemic hypotension was anaphylaxis. The reported incidence of anaphylaxis ranges from 3.2 to 20 per 100,000 head of population [12]. Determining the exact diagnosis is an important challenge, since the proper final diagnosis and treatment are guarantors of a successful outcome [2].

Mastocytosis is a rare disease, the prevalence being less than 0.01% in the whole population [6]. Anaphylaxis is a frequent event in mastocytosis due to strongly increased mediator release from the increased number of mast cell. This explains the much higher prevalence of anaphylaxis in mastocytosis than in the general population and its severe and sometimes fatal course [13]. Mastocytosis should be suspected in patients with recurrent anaphylaxis who present with syncopal or near-syncopal episodes without associated hives or angioedema [14]. Patients with mastocytosis accompanied by extensive skin disease have an increased risk of anaphylaxis [15]. A delay in establishing a diagnosis of mastocytosis can result in either late or inappropriate treatment and subsequent morbidity and mortality.

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