*CASE REPORT*

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# SWEET'S SYNDROME

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Sweet Syndrome (also known as acute febrile neutrophilic dermatosis) is typically characterized by the acute onset of pyrexia and painful cutaneous lesions that are composed of a densedermal inflammatory infiltrate of mature neutrophils. Neutrophilia is also frequently present. Both the condition -associated symptoms and the dermatosis-related lesions promptly resolve after initiation of treatment with systemic corticosteroids.

Dr. Robert Douglas Sweet, originally described acute febrile neutrophilic dermatosis in the August-September 1964 issue of the British Journal of Dermatology. He summarized the cardinal features of "a distinctive and fairly severe illness" that had been encountered in 8 women during the 15-year period from 1949 to 1964. IN Dr. Sweet's department, the condition was originally referred to as the Gomm- Button disease "in eponymous honor of the first two patients" with the disease. Subsequently, this acute febrile neutrophilic dermatosis has become best known by the eponym "Sweet syndrome"

The prevalence of the disease is up to 9 cases per 1 million people. Scientific publications include describe several hundred cases of Sweet's syndrome [2].

Sweet's syndrome occurs in adults of any age (average age 56 years) and is not typical for children. In many patients, the onset of the disease is preceded by an acute respiratory infection of the upper respiratory tract. The paraneoplastic syndrome of Sweet's syndrome can affect the mucous membranes, tend to become chronic and is most common in men. Other associated conditions include streptococcal infection, inflammatory bowel disease, autoimmune diseases (Hashimoto's thyroiditis, Sjogren's syndrome).

More often, women between the ages of 30 and 60 are sick (80-90% of cases). However, Sweet's syndrome is also observed in young adults and children [1, 2].

This disease has three clinical forms: classical, or idiopathic, associated with cancer, and drugs [2, 4]

The classic version of Sweet's syndrome may be associated with an infection of the upper respiratory tract or gastrointestinal tract, as well as pregnancy [2, 4]..

Cases of Sweet's syndrome associated with cancer of the blood are more often associated with acute myelogenous leukemia. Among solid tumors, breast and gastrointestinal cancers are more common [4, 5].

The drug related variant of Sweet's syndrome is more likely to develop after taking granulocyte colony stimulating factor [6]. However, there is evidence of its occurrence when using other drugs, such as minocycline, hydralazine, furosemide [2].

The pathogenesis of Sweet's syndrome is influenced by many factors. It is assumed that the septic process, which is accompanied by fever and peripheral leukocytosis, is at the heart of the disease. An etiological factor may be a bacterial infection, since in most patients with the classic form of Sweet syndrome, an infection of the upper respiratory tract or tonsillitis, which occurs with fever, is observed 1-3 weeks before the development of skin manifestations. Another argument in favor of a bacterial infection is an improvement in the condition of patients with Sweet's syndrome, which developed against the background of yersiniosis when taking systemic antibiotics [2, 4, 5].

Sweet's syndrome can develop with a hypersensitivity reaction to bacterial, viral or tumor antigens. Histopathological changes and the course of the disease indicate this. This hypothesis is confirmed by a rapid decrease in the severity of symptoms and resolution of skin lesions in response to corticosteroids [5, 6].

On the other hand, cytokines can play a direct or indirect role in the development of symptoms and foci. Granulocyte-macrophage colony stimulating factor, interferon, interleukin-1, interleukin-3, interleukin-6 and interleukin-8 are considered to be potential cytokines involved in the pathogenesis of Sweet's syndrome [2, 5].

Clinical picture

The disease begins with prodromal phenomena, such as headache, arthralgia, sore throat, diarrhea, conjunctivitis, fever up to 38 -39$^0$C.

After 1-2 weeks, single or multiple elements appear on the skin of the face, upper and lower extremities, represented by red or purple papules with clear boundaries, a dense texture, merging into irregular plaques. Elements of the rash are markedly painful on palpation. Over time, plaques resolve from the center, turning into rings or arches, and increase along the periphery. Due to the strong swelling of the skin, the rashes resemble blisters. After opening the blisters, ulcers form and then the lesion resembles gangrenous pyoderma. [8]. On the skin of the lower extremities, primary morphological elements can be represented as deep nodes with severe pain on palpation. Rarely, the pathological process is common. Against the background of adequate therapy, after 5-7 days, elements of a rash are resolved. The disease can recur, with the appearance of rashes in the same places.

Foci with Sweet syndrome spill out acutely and can be painful. They have a plum color and a "juicy" look (pseudovesicular). Foci can occur on any surface of the skin, but usually appear on the head, neck, legs, arms, backs of hands and fingers

There is rarely neutrophilic infiltrate within the upper respiratory tract, lungs, liver, and brain.

Laboratory clinical analysis of blood (increased ESR, neutrophilic leukocytosis), an increase in the level of alkaline phosphatase in the blood.

Leukocyte formula > 8000 cells/ml with more than 70% polymorphonuclear nucleotides, increased ESR and alkaline phosphatase level (40%)

Pathomorphological changes are represented by significant edema of the papillary dermis, massive leukocyte infiltration of all layers of the dermis, leukoclasia around the vessels of the dermis. There is no clear picture of vacuities [10]. In subcutaneous adipose tissue - edema and neutrophilic infiltration. In the epidermis, subcorneal pustules.

A differential diagnosis must be carried out with multiform exudative erythema, gangrenous pyoderma, deep red lupus Kaposi-Irganga, erythema nodosum, urticaria, adverse drug reactions

The course and forecast

Sweet syndrome in some patients goes away on its own. Sweet Syndrome quickly responds to systemic corticosteroids. Typically, laboratory parameters are corrected within 72 hours, and skin lesions are cleansed within 3 to 9 days.

Treatment includes, first of all, systemic corticosteroid drugs (prednisone, dexamethasone). It is possible to prescribe antibiotics, potassium iodide, non-steroidal anti-inflammatory drugs, nonspecific desensitizing drugs, antihis-

tamines, multivitamins, adaptogens, immunoactive drugs. In severe cases of the disease, cytostatics, hemodez, reopoliglyukin can be used. Locally - 1-2% solutions of aniline dyes [9], topical corticosteroids. Other drugs: colchicine, dapsone, clofazimine

Clinical case of one our patient

Patient K., 56 years old, came to us with complaints of rashes on the skin, fever, headache, and malaise. Diagnosis of a guided institution: "Multiform exudative erythema."

**Medical history**. Patient considers herself sick for about a month, when for the first time rashes appeared on the skin of the chest, back, and forearm, accompanied by minor itching. 1 week before the rashes, a sore throat and joints appeared, she was not treated on her own. Patient suffers from chronic bronchitis 15 years and by hypertension 10 years.

When handling, her general condition was satisfactory. Internal organs and body systems were without a pronounced pathology. Body temperature was 37.8$^0$C.

**Local status**. The pathological process is common. On the skin of the back, shoulders, and forearms (Fig.1), there were multiple papules of bright red color, a dense consistency, merging into plaques up to 5-8 cm in diameter, with clear boundaries, painful on palpation. Some elements resembled vesicles.

**The results of laboratory studies**

Clinical blood test: red blood cells - 4,93. $10^{12}$/L, hemoglobin 152 g /L, platelets - 256. $10^9$/L, eosinophils -2%, neutrophils (segmented) - 70%), lymphocytes - 12%, monocytes - 6%, ESR -35 mm / h.

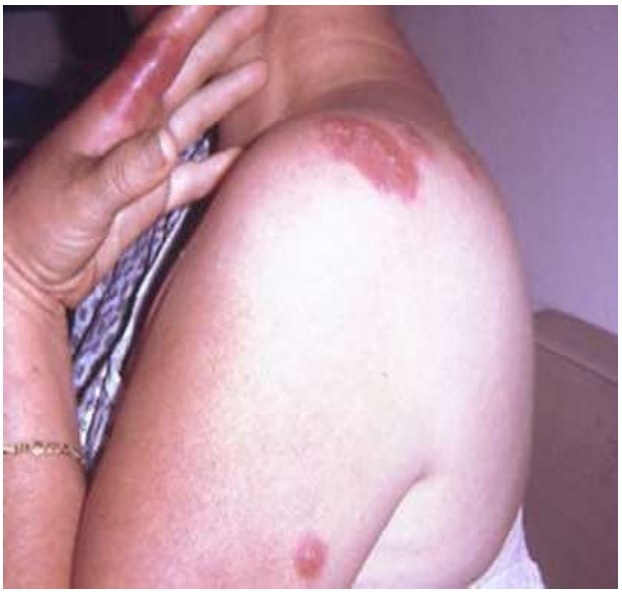

***Fig.1 Sweet's syndrome skin lesions in a woman with classical Sweet's syndrome. Cutaneous lesions of classical Sweet's syndrome on the left hand, left proximal arm and left shoulder in a 56-year-old woman with pyrexia, neutropenia, and a recent respiratory tract infection.***

Urinalysis: specific gravity - 1020, protein and glucose not detected, leukocytes - 1 -2 in the field of view, red blood cells 0 -1 in the field of view.

Biochemical blood test: bilirubin -20 ìmol /L, cholesterol - 6.5 mmol /L, total protein - 72 g /L, glucose 4.6 mmol /L, alanine aminotransferase - 61 U/L, aspartate aminotransferase -46 U /L, alkaline phosphatase - 387 U /L.

Test results for syphilis (microprecipitation reaction, enzyme immunoassay) are negative. Antibodies to HIV, HBSAg, HCVAg are negative.

Electrocardiogram: sinus rhythm correct, deviation of the electrical axis of the heart to the left, left ventricular hypertrophy.

The results of the histological examination correspond to the picture of Sweet's syndrome.

The treatment was prescribed: prednisone 25 mg per day, suprastin 1 ml intramuscularly 10 injections, asparkam 500 mg 3 times a day, topically prednisolone ointment 2 times a day.

After 7 days, the elements of the rash were almost resolved: pale pink spots with clear boundaries remained in their place. There were no fresh rashes. Body temperature returned to normal. On the 15th day, the elements of the rash resolved completely. The patient was recommended to continue taking prednisone at a dose of 20 mg per day, followed by a decrease of 2.5 per week until complete withdrawal; intake of asparkam 500 mg 3 times a day. Appointed consultation of a therapist, rheumatologist, oncologist.

Conclusion

In our clinical case, the diagnosis of Sweet's syndrome was based on the history of the disease, the clinical picture of dermatosis, the results of dermatoscopy and the course of the disease. Diagnostic difficulties are associated with the rare occurrence of Sweet's syndrome. A correctly diagnosed by a dermatologist contributes to the rapid remission of the skin process, as well as the timely contact of the patient with doctors of other specialties (therapist, infectious disease specialist, rheumatologist, and oncologist) to identify serious somatic or oncological pathology.

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