

Fuchs' Syndrome (Stevens-Johnson Syndrome Without Skin Involvement) in an Adult Male – A Case Report and General Characteristics of the Sporadically Diagnosed Disease

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SUMMARY Fuchs' syndrome (Stevens-Johnson syndrome without skin involvement) is a sporadically diagnosed disease. Most authors consider it to be a pure mucosal variant of Stevens-Johnson syndrome; however, some consider the syndrome a separate entity. The complete absence of cutaneous symptoms may be the reason that not all cases of Fuchs' syndrome are diagnosed and properly classified.

The authors describe a case of a 22-year-old patient suffering only from mucosal symptoms, diagnosed as Fuchs' syndrome from the context of the tests performed. A *Mycoplasma pneumoniae* infection triggered the disease onset. *Mycoplasma* infection, as a trigger factor of Fuchs' syndrome in adults, has so far been described in only a few isolated cases worldwide.

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INTRODUCTION

The term Fuchs' syndrome (FS) is used for two completely different diseases. Firstly, it is the conventional term for a chronic inflammatory disease of the eye, usually affecting young adults as unilateral anterior uveitis. Although the etiopathogenesis of this eye disease is not completely understood, a significant proportion is clearly attributable to genetic factors (1). Association with the polymorphism of cytotoxic T antigen (cytotoxic T cell antigen – CTLA) 4 was demonstrated, and recently also a polymorphism of interleukin-23 receptor (IL23R), which is associated with a number of other immune mediated diseases (inflammatory bowel diseases, psoriasis, Behcet's disease) (1).

In dermatology, the term Fuchs' syndrome sometimes also indicates a pure mucosal variant of Stevens-Johnson syndrome (SJS) (2). It is a rare disease that can often be found under the name "Stevens-Johnson Syndrome without skin lesions" or "Atypical Stevens-Johnson Syndrome" in the literature, because many authors consider the disease a separate entity (3). It is usually reported in children and adolescents, but very rarely in adults. We present a rare association of *Mycoplasma pneumoniae* infection and FS in an adult male, successfully managed with targeted antibiotic therapy and symptomatic anti-inflammatory treatment.

CASE REPORT

A 22-year-old patient was admitted to the Dermatology clinic because of gradually progressing mucosal disease. The first symptoms manifested in the form of dysuria that was diagnosed as prostatitis by a urologist, and the patient was recommended a therapy of fluoroquinolone antibiotics for seven days. During this time, however, dysuria persisted; the patient also developed redness and tenderness of the right eye. On the fifth day of ofloxacin therapy the patient noticed the emergence of vesicles on the buccal mucosa and the glans penis. The following day there was an alteration in his general condition, with fever, chills, and coughing with expectoration of yellowish sputum. The patient was hospitalized with a persistent fever (38.1°C); subjectively he complained of soreness in the area of the manifestations on the oral mucosa when eating, of dysuria, and also of back pain localized in the lumbar spine. In the clinical findings on admission numerous erosions and erythema of the buccal mucosa and mucosa of the glans penis (Figs. 1 and 2) were the dominant concern. The physical examination also confirmed noticeable redness of the right eye.

The ophthalmologist subsequently diagnosed the patient with bilateral conjunctivitis. Other medical history data were not significant; the patient had never been seriously ill, did not use any long-term medication, and did not report any risky sexual intercourse or allergies. Initial laboratory tests found elevated inflammatory markers (CRP and sedimentation rate). Urinalysis (biochemistry and sediment) revealed no pathology and neither did a number of culture tests (throat swab, nasal, oral, urethral swabs, swab from the surface of the glans penis). All the tests for sexually transmitted diseases were also negative (HIV, gonorrhoea, syphilis, *Chlamydia* infection).



Figure 1. Painful erosion on the oral mucosa.



Figure 2. Balanitis with erythema and superficial erosions.

regression of mucosal changes, with significant subjective improvement. There was no recurrence of the symptoms after several months of follow-up.

DISCUSSION

Stevens-Johnson syndrome (SJS) belongs to the spectrum of diseases affecting the skin and mucous membranes, together with erythema multiforme, toxic epidermal necrolysis (TEN), and transitional variant of SJS/TEN (2,4).

Although the pathophysiological mechanisms of this group of diseases are not entirely clear, a hyperergic response of cytotoxic T-lymphocytes is considered a common denominator, which subsequently causes apoptosis of keratinocytes (2,4). Some authors classify SJS, SJS/TEN and TEN in adult patients separately as severe bullous drug reactions, because the triggering element in most cases is medication, unlike in children. In children, the triggering factor for SJS and SJS/TEN is usually infection, mainly *Mycoplasma pneumoniae* and herpes simplex virus (2,5,6).

The atypical variant of SJS is rarely reported. It is characterized by the complete absence of skin symptoms and with two or more mucosal sites affected. Most commonly, the disease affects the mucosa of the mouth, genitalia, and conjunctiva. This disease can be found in the literature under various above-mentioned synonyms. From our perspective, the discussion about whether FS (SJS without skin lesions) is a variant of SJS or a separate entity is merely academic. Despite the fact that virtually all the literature sources available for pediatric and adult patients show a close association with mycoplasma infection, the fact that a greater incidence of FS induced by drugs has not been recorded in adults indicates it should be treated as a separate clinical entity.

Mycoplasma are the smallest free-living microorganisms. After an incubation period of one to four weeks, about 10% of patients develop pneumonia; in other cases, the infection is manifested by the upper respiratory tract becoming affected in the form of cough, rhinitis, and pharyngitis. The patient will rarely develop arthritis, encephalitis, hemolytic anemia, and SJS (4,7). *Mycoplasma* infections are mainly diagnosed with serological tests, possibly in combination with a polymerase chain reaction (4,7).

The actual diagnosis of FS (SJS without skin lesions) is difficult due to an almost complete absence of cutaneous symptoms. The most common symptoms of FS include erythema, erosion, and ulceration of the oral mucosa, or on the genitals. It is also often associated with conjunctivitis. In addition to mucosal involvement, most patients also have, to a greater or

lesser degree, the general symptoms resulting from the ongoing mycoplasma infection. To determine the correct diagnosis and subsequent targeted therapy, it is important to get a detailed medical history and perform the tests designed to detect triggering agents.

Mycoplasma pneumoniae infection is a triggering factor in the vast majority of cases of FS in children and even in adults (4,6-10). Given the minimal number of reported cases of FS in adults, however, this information cannot yet be generalized. Based on the literature up to August 2012 – the association of *Mycoplasma* infection and FS in adult patients was reported in only seven patients worldwide. The drug etiology, unlike the classical variant of SJS, has not yet been described. The question is whether the low number of published cases is due to the rare incidence of FS in adults, or whether that fact also contributed significantly to inaccurate diagnosis.

CONCLUSION

Due to the absence of skin symptoms, the patient often seeks other specialists, particularly dentists, ophthalmologists, gynecologists, or urologists. Differential diagnosis of these mucosal manifestations is quite broad and includes aphthous stomatitis and infections, as well as various manifestations of contact hypersensitivity presenting as stomatitis, balanoposthitis, vulvovaginitis, or conjunctivitis. Very similar mucosal lesions may also be part of various manifestations of autoimmune diseases. It is therefore possible that many cases of FS are treated under other diagnoses and thus escape attention.

FS therapy can be divided into causal and symptomatic. Macrolide antibiotics or doxycycline are recommended for mycoplasma infection. Symptomatic therapy involves the use of solutions and gargles with antiseptic, analgesic, and anti-inflammatory effects, and gels with corticosteroids in the oral cavity. In the case of eye mucosa damage, the use of combined topical preparations containing corticosteroids and antibacterial substances is recommended.

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