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ECZEMA HERPETICUM KAO KOMPLIKACIJA ERITRODERMIJSKOG PEMPHIGUS FOLIACEUS-a – PRIKAZ SLUČAJA
ECZEMA HERPETICUM AS A COMPLICATION OF ERYTHRODERMIC PEMPHIGUS FOLIACEUS – CASE REPORT

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Sažetak

Uvod. *Eritrodermijski pemfigus foliaceus predstavlja autoimunsko bulozno oboljenje uzrokovano autoantitelima na proteine dezmozoma. Oštećenje epidermalne barijere i imunosupresija su faktori rizika za razvoj eczema herpeticum.* ***Prikaz slučaja.*** *Žena, 70 godina, sa eritrodermijom kože i brojnim erozijama, ranije lečena od ekcema, a 2019. godine učinjena je histerektomija usled maligne neoplazme uterusa. Histopatološki nalaz biopsije kože leđa: hronični spongiotični dermatitis. Direktnim imunofluorescentnim pregledom uočeni su intraepidermalni, intercelularni depoziti IgG i C3. U krvi su detektovana pemfigusna antitela, eozinofilija i povišena koncentracija IgE. MSCT pretrage i tumorski markeri su bili uredni. Usled neefikasnosti terapije metilprednizolonom, ordinirana je pulsna kortikosteroidna terapija, nakon čega se uočava pojava vezikula i žućkastomrkih krusti na koži leve polovine lica. Klinički je dijagnostikovan eczema herpeticum i ordinirana parenteralna terapija aciklovir amp. (500 mg/8 h tokom 14 dana) i levofloksacin amp. (1 g tokom 10 dana) sa potpunom regresijom promena.* ***Zaključak.*** *Parenteralna i promptna terapija aciklovirom je osnova lečenja.*

Ključne reči: *pemfigus; autoimuna oboljenja; ekcem; Kapošijeva varičeliformna erupcija; dijagnoza; faktori rizika; terapija*

Abstract

Introduction. Erythrodermic pemphigus foliaceus is a rare autoimmune blistering disease characterized by autoantibodies targeting desmosomal proteins. Eczema herpeticum affects individuals with impaired skin barriers or immunosuppression, often complicating pre-existing skin conditions. **Case Report.** We present a case of a 70-year-old woman with erythroderma and skin erosions, previously treated for generalized eczema. Her medical history included hysterectomy for uterine neoplasia in 2019. Skin biopsy revealed chronic spongiotic dermatitis, and direct immunofluorescence demonstrated intracellular and intradermal IgG and C3 deposits. Serological analysis confirmed pemphigus antibodies, along with eosinophilia, and elevated IgE levels. Computed tomography and tumor markers excluded underlying malignancy. After inadequate response to methylprednisolone, the patient underwent pulse corticosteroid therapy. Subsequently, vesicles and crusts developed on her face, leading to a diagnosis of eczema herpeticum. She was treated with intravenous acyclovir (500 mg every 8 hours for 14 days) and oral levofloxacin (1 g daily for 10 days), resulting in complete resolution of the lesions. **Conclusion.** Prompt recognition and treatment of eczema herpeticum with intravenous acyclovir are essential for favorable outcomes.

Key words: Pemphigus; Autoimmune Diseases; Eczema; Kaposi Varicelliform Eruption; Diagnosis; Risk Factors; Drug Therapy

Uvod

Pemphfigus folicaeus (PF) je autoimunsko oboljenje koje se karakteriše pojavom bula i erozija na eritematoznoj osnovi od kojih su pojedine pokrivene krustama najpre na koži skalpa, lica,

Introduction

Pemphigus foliaceus (PF) is a rare autoimmune blistering disease characterized by the development of superficial blisters, erosions, scaling, and crusting on an erythema-

*gornjih delova trupa, a kasnije tokom diseminacije i na ostalim delovima tela sa terminalnim razvojem eksfolijativne eritrodermije. Zahvaćenost sluznica je veoma retka, a promene su često praćenje osećajem peckanja i bola. Pemphigus erythematosus (PE) ili Sener-Usher syndrom predstavlja podtip PF koji odlikuje pojava erozija prekrivenih ljuspama i krustom u fotoeksponiranim zonama (skalp, lice, gornji delovi trupa). Ali, istovremeno sa razvojem ovih lezija mogu biti prisutne i lezije nalik lupusu. Sve navedene promene se pogoršavaju prilikom fotoekspozicije, a 80% pacijenata sa PE mogu imati pozitivan lupus bind test [3]. Incidencija PF-a u Aziji, Evropi, Severnoj Americi i Južnoj Africi iznosi od 0,5–1/100.000, dok je značajno viša u delovima Južne Amerike i Severne Afrike [4]. Pojava bolesti u porodicama je retka, ali je uočena veća prevalencija u određenim etničkim grupama, naročito među Aškenazi Jevrejima, što je povezano sa genetskim faktorima, posebno HLA DRB1*0402 genotipom [1]. U patogenezi PF-a ulogu imaju autoantitela protiv komponenti dezmozoma, što dovodi do akantolize, formiranja subepidermalnih rascepa i bula. Kod PF-a dominantna su autoantitela za dezmoglein 1 (Dsg 1), što rezultira lezijama uglavnom na koži, dok je zahvatanje sluznica izuzetno retko zbog niske koncentracije Dsg 1 u mukoznim tkivima. Dijagnoza se potvrđuje histopatološkim pregledom, koji pokazuje supkornealnu akantolizu, direktnom imunofluorescencijom, koja otkriva intercelularne intraepidermalne depozite IgG i C3 pretežno u površinskim slojevima epidermisa i serološkim testovima koji određuju titar pemfigusnih antitela [1, 2]. Pemfigus pripada grupi dermatoza sa oštećenjem epidermalne barijere, a uobičajene komplikacije su bakterijske i virusne infekcije kože. Termin Kapošijeva varičeliformna erupcija obuhvata vezikulozne erupcije kože izazvane herpes simplex virusom tip 1 i 2, Varicela zoster virusom (VZV), Coxsackie A16 i poksvirusima kod pacijenata sa hroničnim dermatozama. Termin eczema herpeticum se odnosi na virusne superinfekcije uzrokovane HSV kod pacijenata sa atopijskim dermatitisom [5].*

Veoma retko, kao komplikacija eritrodermijskog pemfigusa, može se javiti Kapošijeva varičeliformna erupcija (eczema herpeticum). Ovo je potencijalno životno ugrožavajuće stanje koje se uglavnom javlja u detinjstvu, ali su zabeleženi slučajevi i kod odraslih. Najčešće se javlja na koži sa oštećenjem epidermalne barijere, a opisana je kao komplikacija atopijskog dermatitisa, dermatitis herpetiformis-a, mycosis fungoides-a,

tous base. The disease typically begins on the scalp, face, and upper trunk, with subsequent progression to other parts of the body, potentially culminating in exfoliative erythroderma. Mucosal involvement in PF is uncommon, and patients often report burning sensations and pain associated with skin lesions [1, 2]. A distinct variant of PE - also known as Senear-Usher syndrome - is defined by erosions covered with scales and crusts, predominantly in sun-exposed areas such as the scalp, face, and upper trunk. In some cases, discoid lupus erythematosus-like lesions may coexist. These lesions typically worsen with ultraviolet exposure, and up to 80% of patients with PE may demonstrate a positive lupus band test [3]. The disease usually manifests in individuals between the fourth and sixth decades of life. The incidence of PF ranging from 0.5 to 1 per 100,000 people in Asia, Europe, North America, and South Africa, but is significantly higher in parts of South America and North Africa [4]. While familial clustering is rare, PF shows increased prevalence among certain ethnic groups, paricularly Ashkenazi Jews, attributed to genetic predisposition - most notably the HLA DRB1*0402 genotype [1].

The pathogenesis of PF involves autoantibodies directed against desmosomal – primarily desmoglein 1 (Dsg 1) - leading to acantholysis, subepidermal cleft formation, and blisters. Due to the low concentration of Dsg 1 in mucosal epithelium, mucosal involvement is extremely rare. Diagnosis is confirmed by histopathological examination revealing subcorneal acantholysis, direct immunofluorescence showing intercellular IgG and C3 deposits in the upper epidermis, and serological assays that direct pemphigus antibodies [1, 2].

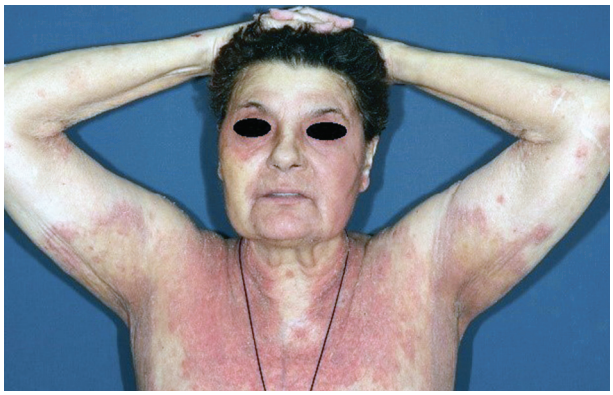
Because PF disrupts the epidermal barrier, affected individuals are susceptible to secondary skin infections, including both bacterial and viral etiologies. Kaposi's varicelliform eruption (KVE) refers to a vesiculopustular viral superinfection occurring in the context of underlying dermatoses. It is caused by viruses such as herpes simplex virus (HSV) types 1 and 2, varicella-zoster virus (VZV), Coxsackievirus A16, and various poxviruses. A specific subtype of eczema herpeticum denotes HSV infection superimposed of atopic dermatitis [4].

Darierove bolesti, Hejli-Hejli bolesti, Groverove bolesti, iritantnog kontaktnog dermatitisa i opekotina [6]. Najčešće se EH viđa kod atopijskog dermatitisa (oko 3% pacijenata), dok je u jednoj studiji opisan kod jedne trećine pacijenata sa pemphigus foliaceusom [7, 8]. Eczema herpeticum se manifestuje pojavom vezikula, hemoragičnih erozija i krusti. U slučaju sistemske infekcije, EH može dovesti do malaksalosti, groznice, keratokonjuktivitisa, encefalitisa i sepse. Promene na koži su obično lokalizovane na glavi, vratu i trupu i traju u proseku 2–6 nedelja. Terapija izbora je intravenska primena aciklovira i širokospektralnih antibiotika radi prevencije sekundarnih kožnih infekcija. Neadekvatno prepoznavanje i lečenje EH nosi rizik od smrtnog ishoda usled sistemske diseminacije, zahvatanja više organa i sepse.

Prikaz slučaja

Žena, starosne dobi od 70 godina javlja se u Kliniku u suberitrodermiji sa mnogobrojnim erozijama po koži, na koži trupa i ekstremiteta ***(Slike 1–4)****.*

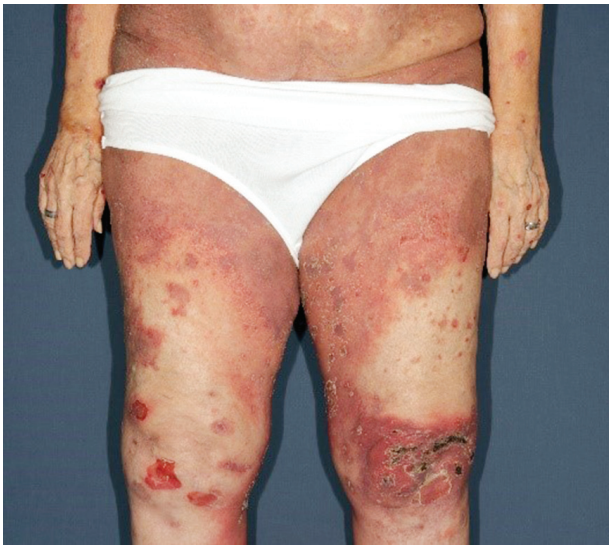
Pacijentkinja navodi da se deset godina unazad lečila od generalizovanog ekcema, koji nije



Slika 1. *Erozije sa sitnom skvamom na eritematoznoj osnovi u regijama lica, vrata, trupa i gornjih ekstremiteta*

Figure 1. Diffuse macular erythema with fine adherent scales and small skin erosions involving the face, neck, décolletage, and upper extremities.
Legenda. *Prisutne su eritematozne makule sa tankom adherentnom skvamom, u regiji lica, vrata, dekoltea i gornjih ekstremiteta, međusobno slivene sa prostorima neizmenjene kože između*
Legend. The patient presents with erythematous macules, thin adherent scales, and small erosions scattered across the décolleté, trunk, and proximal upper extremities, interspersed with areas of unaffected skin

Although extremely rare, KVE (eczema herpeticum) can complicate erythrodermic pemphigus foliaceus. This potentially life-threatening condition typically affects children but has also been documented in adults. It usually develops in areas of compromised skin barrier and has been reported in association with atopic dermatitis, dermatitis herpetiformis, mycosis fungoides, Darier disease, Hailey-Hailey disease, Grover disease, irritant contact dermatitis, and burns [6]. While EH occurs in approximately 3% of patients with atopic dermatitis, one study reported its occurrence in up to one-third of patients with PF [7, 8]. Clinically, EH presents with vesicles, hemorrhagic erosions, and crusts, predominantly affecting the head, neck, and trunk. In systemic dissemination, symptoms may include malaise, fever, keratoconjunctivitis, encephalitis, and sepsis. Lesions typically persist for 2 to 6 weeks. Intravenous acyclovir remains the treatment of choice, often combined by systemic antibiotics to prevent secondary bacterial infections. Delayed or missed diagnosis can result in severe complications, including multiorgan failure and sepsis.



Slika 2. *Erozije kože, od kojih su pojedine pokrivene krustama na koži donjih ekstremiteta*

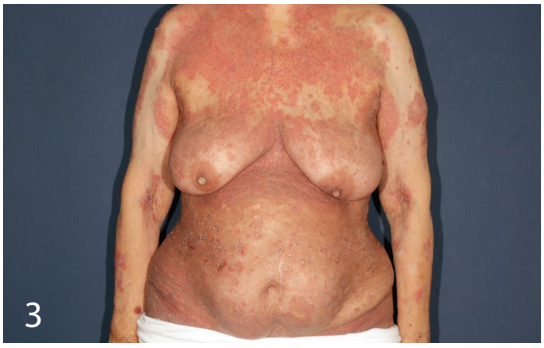
Figure 2. Erosions with scaling and crusts on an erythematous base in the lower extremities
Legenda 2. *Kod pacijentkinje se vide erozije kože sa tankom adherentnom skvamom na eritematoznoj osnovi od kojih su pojedine pokrivene krustama u regiji donjih ekstremiteta.*
Legend 2. The patient presents with erosions on an erythematous base with associated scaling and crusting on the lower extremities, with several lesions entirely covered by crusts.

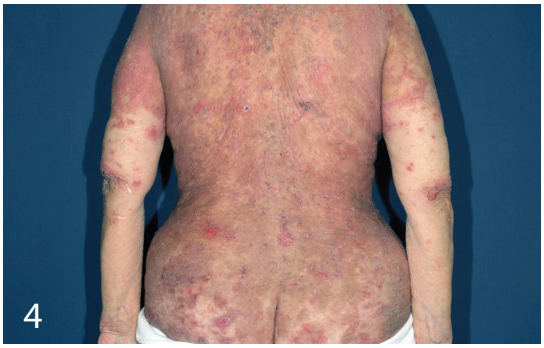
potvrđen biopsijom kože i drugim ispitivanjima. Do decembra 2023. godine lečena je antihistaminicima, sistemskim i lokalnim kortikosteroidima u drugim ustanovama, uz delimično poboljšanje. Godine 2019. učinjena je histerektomija sa adneksektomijom zbog maligne neoplazme uterusa, koja je sada u remisiji. Biopsija kože sa leđa pokazala je histopatološke nalaze epiderma sa znacima blage do umerene iregularne psorijaziformne hiperplazije sa očuvanim granularnim slojem, dok je u kornealnom sloju uočena parakeratoza, fibrinski eksudat i neutrofilne supkornealne mikropustule. U papilarnom dermisu primećen je blagi perivaskularni limfocitni i histiocitni infiltrat. Direktnom imunofluorescencijom uočeni su intraepidermalni, intercelularni depoziti IgG i C3. Indirektnom imunofluorescencijom detektovana su pemfigusna antitela u titru 1:80. Krvne analize su ukazale na povišene eozinofile (632/μL) i povećanu koncentraciju IgE (p>3.000 IU/ml), dok su ostali imunski parametri (ANA, ENA skrining) bili u granicama referentnih vrednosti. Pacijentkinja je ispitivana i u pravcu postojanja neoplazije te su učinjeni CT grudnog koša, abdomena i male karlice i tumorski markeri za tumore želuca, pluća, jetre i kolona koji su bili u referentnim vrednostima. Pacijentkinji je dijagnostikovan eritrodermijski pemphigus foliaceus, započeta sistemska kortikosteroidna terapija (1 mg/kg metilprednizolona), a usled neefikasnosti ordinirana je i pulsna kortikosteroidna terapija (1 g/dan me-

Case presentation

A 70-year-old woman presented to our clinic with suberythroderma and numerous erosions affecting the skin of the trunk and extremities **(Figures 1–4)**.

The patient had a 10-year history of presumed generalized eczema, though this diagnosis had never been confirmed by biopsy or other diagnostic procedures. Prior to December 2023, she received antihistamines and both topical and systemic corticosteroids at various institutions, resulting in only partial improvement. Her past medical history was notable for a hysterectomy with adnexectomy in 2019 due to a malignant uterine neoplasm, currently in remission. A skin biopsy was performed from a lesion on the back. Histopathological analysis revealed mild to moderate irregular psoriasiform epidermal hyperplasia and a preserved granular layer. The stratum corneum exhibited parakeratosis, fibrin exudate, and a subcorneal micropustule rich in neutrophils. A mild perivascular lymphohistiocytic infiltrate was noted in the papillary dermis. Direct immunofluorescence demonstrated intraepidermal intercellular deposits of IgG and C3, while indirect immunofluorescence revealed pemphigus antibodies at a titer of 1:80. Laboratory tests showed elevated eosinophils (632/μL) and markedly increased IgE level (p>3,000 IU/ml). Antinuclear anti-





Slika 3 i 4. *Erozije kože, od kojih su pojedine pokrivene krastama na koži grudnog koša, abdomena i leđa*

Figures 3 and 4. Erosions with scaling and crusts on an erythematous base involving the chest, abdomen, and back

Legenda. *Pacijentkinja sa izraženim erozijama, praćenim deskvamacijom i formiranjem krusta na eritematoznoj osnovi na koži grudnog koša, abdomena i leđa, uz zahvatanje značajne površine kože. Na osnovu svih do sada prezentovanih prikaza, može se zaključiti da pacijentkinja ispoljava stanje suberitrodermije kože.*

Legend. The patient presents with extensive erosions accompanied by scaling and crusting on an erythematous base, visible on the chest, abdomen, and back, covering a large skin surface area. The overall clinical appearance is consistent with suberythroderma.

tilprednizolona tokom tri uzastopna dana). Tokom hospitalizacije, pacijentkinji nisu ordinirani lekovi koji smanjuju potrebu za kortikosteroidima. Nakon završene pulsne kortikosteroidne terapije dolazi do pojave vezikula i žućkastosmeđih krusti u regiji leve periokularne regije, a zatim i leve perioralne regije i duž leve polovine lica ***(Slike 5 i 6).***

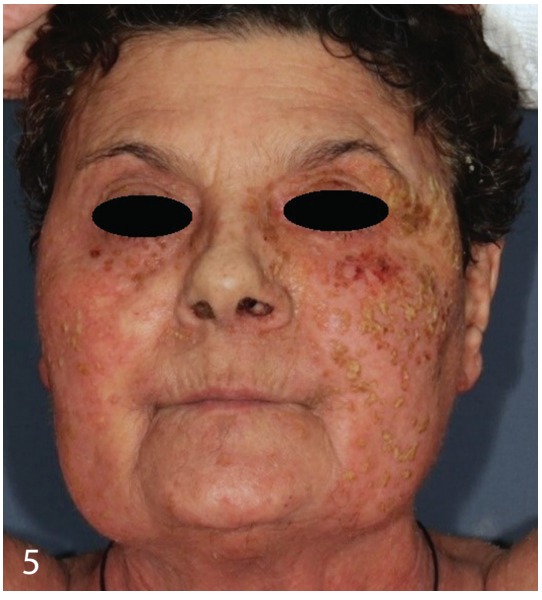
Na osnovu kliničke slike postavljena je dijagnoza Kapošijeve varičeliformne erupcije i odmah je započeta intravenska antivirusna (aciklovir amp. 500 mg/8 h tokom 14 dana) i antibiotska (levofloksacin amp. 1 g/dan) terapija tokom 10 dana sa potpunom regresijom promena na koži ***(Slika 7)****.*

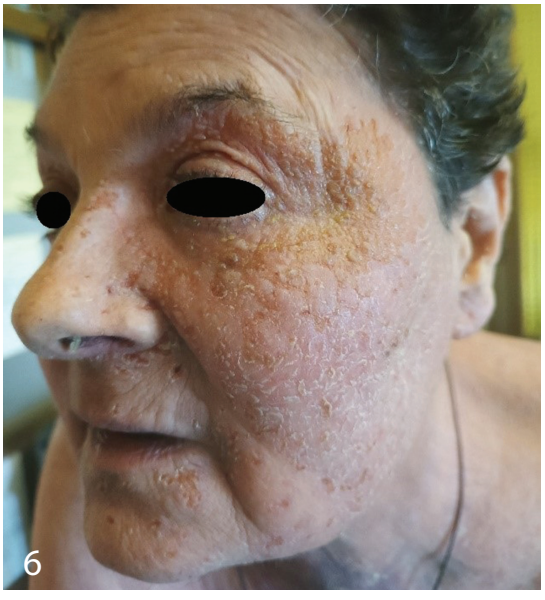
Diskusija

Pemfigus pripada grupi autoimunih oboljenja koja nastaju kao posledica stvaranja specifičnih autoantitela na komponente dezmozoma. U patogenezi bolesti dominantna autoantitela se stvaraju na proteine dezmoglein 1 i 3, sa posledičnim razvojem subepidermalnog rascepa i bula [3, 4]. Eritrodermijski pemfigus predstavlja podtip pemphigusa foliaceusa koji se karakteriše prisustvom specifičnih autoantitela pretežno usmerenih na dezmoglein 1 koji se eksprimira u keratinizujućim tkivima (koža) čime se objašnjava pojava plikova, erozija i krusti na koži uz očuvanost sluznice. Kli-

bodies (ANA) and extractable nuclear antigen (ENA) screening were within normal limits. A thorough malignancy workup - including CT scans of the chest, abdomen, and pelvis, as well as tumor markers for gastric, lung, liver, and colon cancer – yielded no evidence of malignancy. Based on clinical, histopathological, immunofluorescence, and serological findings, the patient was diagnosed with erythrodermic pemphigus foliaceus. Systemic corticosteroid therapy was initiated with methylprednisolone at 1 mg/kg. Due to inadequate response, the patient received pulse corticosteroid therapy (methylprednisolone 1 g/day for 3 consecutive days). No steroid-sparing therapy was initiated during hospitalization. On the 10th day of hospitalization, following pulse therapy, the patient developed vesicles and golden-brown crusts localized to the left periocular and perioral regions, later spreading to the left hemiface **(Figures 5 and 6)**.

Based on the clinical features, a diagnosis of Kaposi's varicelliform eruption (eczema herpeticum) was made. Intravenous antiviral therapy was initiated promptly with acyclovir ampoules at 500 mg every 8 hours, and continued for 14 days. Simultaneously, intrave-





Slike 5 i 6. *Vezikule i žućkasto-smeđe kruste u regiji leve polovine lica*

Figures 5 and 6. Vesicles and golden-brown crusts localized on the left side of the face
Legenda. *Na slikama se uočavaju vezikule i žućkasto-smeđe kruste u regiji leve polovine lica, što klinički ukazuje na* eczema herpeticum.
Legend. Vesicular lesions with yellowish-brown crusts are present on the left side of the face, representing a clinical picture consistent with eczema herpeticum.

nički se može manifestovati eritrodermijom, ali to nije uvek slučaj [7]. U seriji slučajeva od šest pacijenata sa eritrodermičnim PF-om, 80% je inicijalno ispoljavalo eritrodermu [7]. Retko se javlja kod dece. Zbog narušenog integriteta epidermalne barijere, uobičajene komplikacije pemfigusa su bakterijske i virusne infekcije kože. Jedna od retkih komplikacija je Kapošijeva varičeliformna erupcija, potencijalno fatalna reaktivacija herpes simpleks virusa tipa 1 i 2. Obično se javlja u detinjstvu, verovatno zato što je EH najčešća komplikacija atopijskog dermatitisa kod dece, iako su zabeleženi slučajevi i u starijim uzrasnim grupama [9]. U literaturi je identifikovano nekoliko faktora rizika za razvoj EH: oštećenje epidermalne barijere, imunosupresija, dizbioza kožne flore i stariji uzrast [8, 9]. Kod naše pacijentkinje prisutno je nekoliko navedenih faktora rizika. Eczema herpeticum je češća kod stanja koja uključuju oštećenje epidermalne barijere. Iako se EH najčešće viđa kao komplikacija atopijskog dermatitisa (3% pacijenata), može se javiti i kao komplikacija: ihtioze, opekotina, Sezarijevog sindroma, Darierove bolesti, pemfigusa, benignog porodičnog pemfigusa i kontaktnog dermatitisa [6–8]. Kada se razmatra imunosupresija kao potencijal-

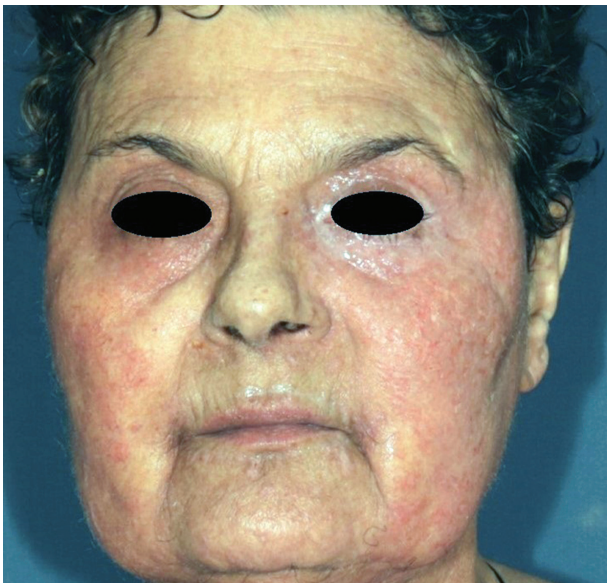


***Figure 7.** Potpuna regresija ranije prisutnih kožnih promena na levoj polovini lica*

Figure 7. Complete regression of the facial skin lesions
***Legenda.** Uočava se kompletna regresija ranije prisutnih*
Legend. Complete regression of the previously observed skin lesions on the left side of the face.

nous antibiotic therapy with levofloxacin was administered at 1 mg/kg for 10 days to prevent secondary bacterial infection. This dual therapy resulted in complete regression of the lesions **(Figure 7)**.

Discussion

Pemphigus encompasses a group of autoimmune blistering diseases caused by pathogenic autoantibodies against desmosomal proteins, primarily desmoglein 1 and 3. The binding of these antibodies to their respective targets results in formation of subepidermal clefts and bullae. Erythrodermic pemphigus foliaceus is a rare subtype characterized by autoantibodies targeting desmoglein 1, which is predominantly expressed in the superficial layers of keratinized skin. As a result, patients present with bullae, erosions, and crusting of the skin, typically without mucosal involvement [1, 2]. Although PF can present with erythroderma, it is not a universal feature. In a case series of six patients with erythrodermic PF, 80% initially presented with erythroderma [7]. The disease is rare in children. A major concern in pemphigus is the compromised epidermal barrier, which predisposed patients to secondary bacterial and viral infections. Among these, Kaposi's varicelliform eruption (KVE) – also known as eczema herpeticum (EH) when caused by herpes simplex virus types 1 and 2 (HSV) – represents a potentially life-threatening viral complication. KVE is most frequently seen in pediatric patients with atopic dermatitis, but it can also occur in older age groups [9]. Several risk factors for EH have been identified in the literature, including impaired skin barrier function, immunosuppressive therapy, dysbiosis of skin flora, and advanced age [8, 9]. Our patient presented with multiple such risk factors. Eczema herpeticum is more common in conditions that inherently involve epidermal barrier damage. Although EH is most commonly associated with atopic dermatitis, affecting approximately 3% of patients, it has also been reported in other conditions such as ichthyosis, burns, Sezary syndrome, Darier disease, pemphigus, benign familial pemphigus, and contact dermatitis [6–8]. In a case series of 12 patients with autoimmune blistering dermatoses complicated by EH, three had not received immunosuppressive therapy, specifically those with linear IgA der-

ni faktor rizika, u seriji slučajeva od 12 pacijenata sa autoimunim buloznim dermatozama koji su razvili EH, kod njih troje nije korišćena imunosupresivna terapija, posebno kod pacijenata sa linearnom IgA dermatozom. Kod pacijenata sa pemfigusom razvoj ove komplikacije bio je povezan sa imunosupresijom izazvanom kortikosteroidima, bilo kao monoterapijom ili u kombinaciji sa drugim imunosupresivima [10]. Međutim, u metaanalizi 100 pacijenata, 76% njih nije primilo sistemsku terapiju kortikosteroidima najmanje četiri nedelje pre razvoja EH, što ukazuje da je sistemska imunosupresija samo jedan od faktora rizika [6]. O značaju lokalne imunosupresije kao faktora rizika za razvoj EH govori retrospektivna japanska studija koja pokazuje češću pojavu EH kao komplikacije AD kod pacijenata na lokalnoj kortikosteroidnoj terapiji u odnosu na lokalnu primenu inhibitora kalcineurina, ali pokazuje i korelaciju duže primene lokalnih kortikosteroida sa većim rizikom za razvoj EH (Kapošijeva varičeliformna erupcija je urgentno, životno ugrožavajuće stanje koje zahteva hitno lečenje. Mortalitet kod imunokompetentnih odraslih iznosi 10%, ali može dostići i 50% kod imunokompromitovanih osoba [12, 13]. Dijagnoza se često može postaviti na osnovu anamneze i kliničke slike. Za potvrdu se mogu koristiti Tzanckov bris i bris kože. Lečenje treba započeti odmah. Najzastupljeniji lek, a ujedno i prva linija terapije jeste aciklovir koji se primenjuje intravenski u dozi od 1,5 g dnevno [8]. Uz primarnu antivirusnu terapiju neophodno je sprovesti i intravensku širokospektralnu antibiotsku terapiju u cilju prevencije sekundarne bakterijske infekcije kože. Prema literaturi, propisana terapija obično dovodi do regresije promena na koži nakon 10–14 dana. Međutim, u nekim slučajevima može doći do sistemske diseminacije, zahvatanja više organa, sepse i smrtnog ishoda. S obzirom na to da se imunosupresivni lekovi koriste u lečenju PF-a i drugih dermatoza sa oštećenjem epidermalne barijere, preporučuje se da se u teškim slučajevima EH imunosupresivi (MTX, mikofenolat mofetil i ciklosporin) obustave, a ako je moguće, smanji i doza sistemskih kortikosteroida [14].

Zaključak

Kod hroničnih dermatoza koje u osnovi imaju oštećenje epidermalne barijere, a prisutni su i faktori rizika kao što su starija životna dob i imunosupresija, uz pojavu tipičnih kožnih promena, treba razmišljati u pravcu mogućeg ra-

matosis. The development of this complication in patients with pemphigus was associated with immunosuppression from corticosteroids, either as monotherapy or combined with other immunosuppressants [10]. Notably, a meta-analysis of 100 patients found that 76% had not received systemic corticosteroids within four weeks prior to developing EH, indicating that systemic immunosuppression is just one of the risk factors [6]. Local immunosuppression also plays a role, although a retrospective Japanese study found no statistically significant difference in EH incidence between patients using topical calcineurin inhibitors and those using topical corticosteroids in the treatment of atopic dermatitis [11].

KVE is considered a dermatologic emergency. In immunocompetent adults, the mortality rate is approximately 10%, but this can rise to 50% in immunocompromised individuals [12, 13]. Diagnosis is primarily clinical, based on the medical history and clinical presentation. Supportive diagnostic tools include the Tzanck smear and skin swab. Treatment must be initiated promptly Intravenous acyclovir remains the first-line therapy, typically administered at 1.5 g per day [8]. In addition to antiviral therapy, systemic broad-spectrum intravenous antibiotics are often necessary to prevent or treat secondary bacterial infections. In most cases, skin lesions regress within 10 to 14 days of treatment initiation. However, in severe cases, systemic viral dissemination, multi-organ involvement, sepsis, and death can occur. In patients receiving immunosuppressive therapy, particularly those with PF and other dermatoses with epidermal barrier damage, it is recommended to discontinue or reduce immunosuppressive agents, such as methotrexate, mycophenolate mofetil, or cyclosporine [14].

Conclusion

In chronic dermatoses characterized by impaired epidermal barrier function, particularly in the presence of risk factors such as advanced age and immunosuppression, the possibility of eczema herpeticum should be considered when typical skin lesions appear. Early recognition and prompt initiation of antiviral therapy, primarily intravenous acyclovir, in combination with antibiotics, are essential to prevent systemic dissemination, secondary

zvoja eczema herpeticum, a zatim i primeniti intravenski aciklovir i antibiotike u cilju sprečavanja moguće sistemske diseminacije, sekundarnih infekcija kože, sepse i smrti.

Skraćenice

EH – eczema herpeticum
PF – Pemphigus foliaceus
PE – Pemphigus erythematosus
VZV – Varicela zoster virus
SLE – Sistemski eritemski lupus
CT – Kompjuterizovana tomografija
IgE – Imunoglobulin E
IgA – Imunoglobulin A
MTX – Metotreksat
H – Čas
G – Gram
mg/kg – Miligram po kilogramu
μL – Miktolitar
IU/ml – Internacionalnih jedinica po mililitru
Dsg 1 – Dezmoglein 1
DIF – Direktna imunofluorescencija

infections, sepsis, and potentially fatal outcomes.

Abbreviations

EH – Eczema herpeticum
PF – Pemphigus foliaceus
PE – Pemphigus erythematosus
KVE – Kaposi's varicelliform eruption
HSV – Herpes simplex virus
VZV – Varicella-zoster virus
SLE – Systemic lupus erythematosus
CT – Computer tomography
IgE – Immunoglobulin E
IgA – Immunoglobulin A
MTX – Methotrexate
H – Hour
G – Gram
mg/kg – Milligram per kilogram
μL – Microliter
IU/ml – International units per milliliter
Dsg 1 – Desmoglein 1
DIF – Direct immunofluorescence

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