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KARCINOM DŽINOVSKIH BAZALNIH ČELIJA – LEZIJA SLIČNA PEČURKAMA NA REBARNOM LUKU

Apstrakt: Karcinomi džinovskih bazalnih ćelija (GBCC) su retke neoplastične lezije sa incidencom manjom od 0,5% i destruktivnim sposobnostima većim od njihovih prekursora. Gljivasti rast kožnih tumora, koji je 1806. opisao Albert, još uvek je retka pojava. GBCC sa makroskopskim karakteristikama nalik pečurkama jedva da su prijavljeni. Predstavljamo GBCC nalik pečurki, uspešno lečen širokom hirurškom ekscizijom.

U našu kliniku je primljen muškarac, star 85 godina sa progresivno rastućom lezijom kože. Došao je da traži medicinsku negu sa krvarećom bolnom lezijom kože. Tumor se iz male papule razvio u egzofitnu pedukulisanu leziju sa smrdljivom, ulcerisanom, eksudativnom površinom. Fizikalnim pregledom utvrđena je pečurkasta egzofitna masa na pečurkama 7,5x1,5 cm sa valjanim ivicama i delimičnom nekrozom kože, znaci sekundarne infekcije i krvarenja. Lezija je hirurški uklonjena. Histopatologija je pokazala ćelije bazalnog karcinoma (BCC). Dva meseca posle operacije nije bilo znakova recidiva tumora i sekundarnih metastaza bez potrebe za dodatnim lečenjem.

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GBCC se još uvek mogu identifikovati u 21. veku. Nastaje usled zanemarivanja lezije, neupadljive lokalizacije obično prekrivene odećom. Morfologija slična pečurkama je retka u BCC. Lečenje je uglavnom hirurška ekscizija, sa visokom stopom uspeha i poboljšanjem kvaliteta života.

Ključne reči: karcinom bazalnih ćelija, dermatološka hirurgija, rekonstruktivna hirurgija

Uvod:

Karcinomi bazalnih ćelija (BCC) su najčešće maligne neoplazme kože, sa manifestacijama sličnim drugim tumorima bazalnog epitelnog porekla. Obično se smatraju sporo napredujućim, indolentnim i benignim tumorima. S druge strane, džinovski BCC (GBCC) su kategorizovani kao lezije sa više od 5 cm mereno na najvećem prečniku, agresivne su, sa lokalnom destrukcijom tkiva, većim rizikom od metastatskog potencijala i lošom prognozom, u poređenju sa konvencionalnim BCC (1).

Faktori rizika za razvoj ove vrste kožnih neoplazmi su nizak socio-ekonomski status, zanemarivanje lezije i imunosupresija. (1–3) Međutim, u literaturi su retko prijavljeni egzofitni, vegetirajući GBCC nalik pečurkama. (4) Diferencijalna dijagnoza obuhvata melanome, karcinome skvamoznih ćelija, cilindrome (3, 4, 12). Arai et al. su opisali dva slučaja ogromnog tumora adneksa kože sa morfologijom nalik pečurki. (5) Adekvatna diferencijacija između ovih lezija je ključna zbog različitog ponašanja, evolucije, metastatskog potencijala i protokola lečenja.

Iako se većina BCC-a razvija na delovima tela izloženim suncu (6), skoro 10–15% BCC-a evoluiraju na neuobičajenim anatomskim mestima, često prekrivenim odećom (7).

Prikaz slučaja:

Na našu kliniku je primljen muškarac, star 85 godina, sa lezijom koja ne zarasta, krvareći, koja se nalazi u nivou donje torakalne aperture i desnog hipohondralnog dela stomaka bočno od 7. rebrene hrskavice (Sl. 1).

Usled dugotrajne infekcije urinarnog trakta, dijagnostikovao je maligni tumor desnog bubrega. Nakon toga, kod pacijenta je učinjena desna kompletna nefrektomija 2018. godine, nakon čega je usledio nekomplikovani postoperativni tok. Nisu prijavljena druga značajna medicinska stanja, nije imao alergije na lekove i negativnu porodičnu anamnezu malignih bolesti.

Lezija je opisana kao sporo napredujuća, koja je u početku napredovala od male papule do egzofitnog, pedukulisanog tumora tokom nekoliko godina, sa epizodama krvarenja i na kraju znacima nekroze i sekundarne infekcije. Pacijent je odlučio da potraži lećenje nakon što je primetio povećanje krvarenja i iscedak neprijatnog mirisa iz lezije.

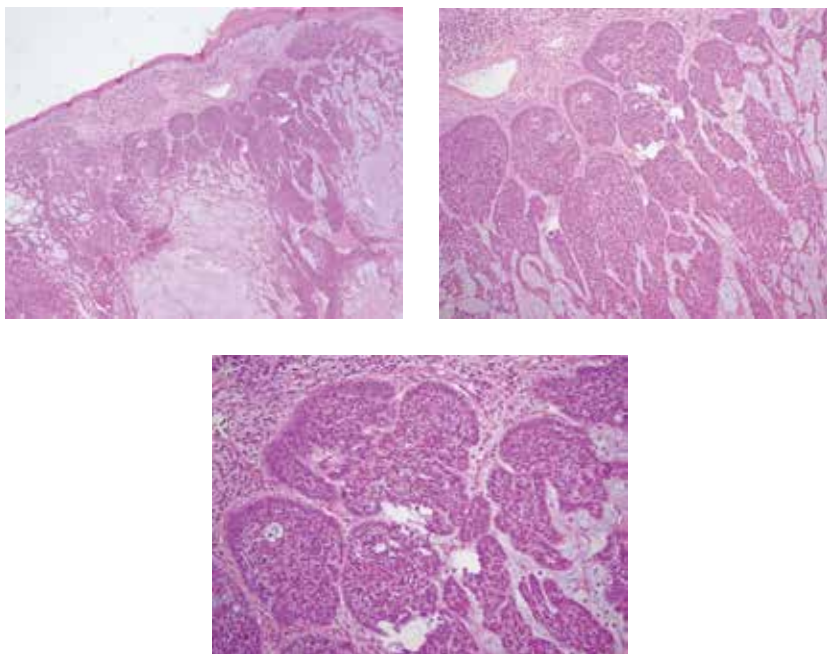
Tip kože pacijenata je bio Fitzpatrick tip 2. Nije imao istoriju intenzivnog izlaganja suncu.

Fizikalnim pregledom utvrđena je egzofitna masa 7,5x1,5cm, sa gnojnim eksudatom neprijatnog mirisa. Makroskopske karakteristike tumora uključivale su veliko lobulisano nekrotično tkivo sa uvijenim ivicama, adherirani hemoragični eksudat i perilezijsku sklerotičnu morfologiju kože (Fig. 1).



Uzorak za hiruršku resekciju se sastojao od kože i potkožnog tkiva, 72,51x15 mm, sa beličasto-braon čvorom dimenzija 61x52x30 mm. Histopatološke, mikroskopske karakteristike otkrile su nodularni tip karcinoma bazalnih ćelija sa neoplastičnim obalama i palisadnim ćelijama. Retrakcija mukopolisaharida strome je primećena mikroskopski unutar neoplastičnih ćelijskih obala. Tumorske ćelije su pokazale hiperhromatska jezgra, sa lošim sadržajem i zapreminom citoplazme. Tumor je infiltrirao duboki korijum.

Slika 1. Klinički prikaz lezije sa šablonom rasta nalik pečurkama



Slika 2A. H&E x40 Karcinom bazalnih ćelija nodularnog tipa, 50x39 mm (220x220 DPI)

Slika 2B. H&E x100 Karcinom bazalnih ćelija nodularnog tipa, 49x39 mm (220x220 DPI)

Slika 2C. H&E x200 Sve slike označavaju nodularni tip karcinoma bazalnih ćelija, 53x38 mm (220x220 DPI)

*Slika 2A. H&E x40 Slika 2B. H&E x100, Slika 2C. H&E x200
Sve slike označavaju nodularni tip karcinoma bazalnih ćelija.*

Obavljen je kompletan laboratorijski pregled (uključujući hematološka, biohemijska, serološka, koagulaciona i infektivna ispitivanja) koji je pokazao normalne rezultate.

Na prijemu je urađen bris površinske rane, koji je otkrio sekundarnu infekciju *Pseudomonas aeruginosa*, koja je lečena intravenskim Cefepimom. Nisu zabeleženi znaci metastatskog širenja. Snimanje (uključujući i rendgenski snimak abdomena i *CT grudnog koša, abdomen i karlica*) nisu pokazali susednu infiltraciju mekog tkiva, niti zahvaćenost grudnog koša i kosti.

Nakon adekvatne preoperativne pripreme, pacijent je podvrgnut širokoj lokalnoj eksciziji i uklanjanju tumorske mase sa negativne hirurške margine preko 5 mm i potpuno zatvaranje rane. Postoperativno praćenje je pokazalo potpuno, primarno zarastanje rane, sa optimalnim poravnanjem ivica rane. A dvomesečno praćenje je dokumentovalo zadovoljavajuće estetske rezultate, bez komplikacija ili znakova recidiva tumorske mase (slika 3).



Direktno zatvaranje defekta, praćenje dva meseca nakon operacije – linearni ožiljak, 65x39 mm (96x96 DPI)

Slika 3. Direktno zatvaranje defekta, praćenje dva meseca nakon operacije – linearni ožiljak

Diskusija:

GBCC se javlja tipično kod starijih muških pacijenata, sa najvećom incidencijom u 7. deceniji, obično se nalazi na leđima, licu i gornjim ekstremitetima. U našem slučaju se nalazio između trbušnog zida i grudnog koša. GBCC se generalno definiše kao lezija preko 5 cm mereno u njenom najvećem prečniku (1). Najčešći histološki podtip je nodularni BCC. Obično se predstavlja kao lokalizovana egzofitna masa na površini kože i češća je od karcinoma skvamoznih ćelija (SCC). Neophodno je razlikovati BCC od SCC, jer SCC ima veći rizik od recidiva i metastatskog potencijala. Druga opisana diferencijalna dijagnoza su melanomi i tumor adneksa kože (4, 5). Štaviše, obrazac rasta sličan pečurkama opisali su za cilindrom Vollina et al. (12).

Iako ne postoje jasne smernice o preciznom metodu lečenja, agresivan pristup nudi najbolji ishod lečenja (5). Potencijalni izazovi se javljaju zbog veličine tumora i često je nemoguće izvršiti zatvaranje defekta bez naknadnog funkcionalnog i kozmetičkog oštećenja (6). Optimalni tretman GBCC se sastoji od široke lokalne ekscizije sa histološki potvrđenim marginama bez tumora, često praćenim adjuvansom terapija (8). Radikalna hirurška ekscizija ostaje poznati zlatni standard, ali nekoliko studija izveštava o mogućnostima nehirurškog lečenja (uključujući radioterapiju, vismodegib, lokalni imikvimod, acitrecin) za GBCC (9). Karcinom bazalnih ćelija, u našem slučaju, bio je sporo rastuća neoplazma u prirodi, bez promene u obrascu rasta, što sugerise da je gigantski BCC rezultat nemara u traženju ranog medicinskog lečenja. Hronično zanemarivanje, infekcija, nekroza tkiva i krvarenje doprinose većoj verovatnoći neuspešnog ishoda lečenja (9, 10).

Često za džinovske BCC, pokrivanje defekta kože nakon ekscizije pomoću kožnog reznja ili različitih kožnih transplantata može biti neophodno (11). Kao što se vidi u našem slučaju, defekt kože je zatvoren direktno, uz ekstenzivno podrivanje tkiva.

Opisan je jedinstven slučaj džinovske BCC nalik na pečurku u punoj remisiji nakon potpune ekscizije i zatvaranja. BCC imaju tendenciju da budu indolentni, bezbolni tumori, što može dozvoliti pacijentima da ignorišu leziju, što dovodi do zanemarenog BCC-a, koji može postati velika masa nalik na gljivicu, sa rizikom od krvarenja, infekcije, širenja metastaza i loših rezultata lečenja.

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A GIANT BASAL CELL CARCINOMA – A MUSHROOM-LIKE LESION ON THE RIB CAGE

Abstract: Giant basal cell carcinomas (GBCCs) are rare neoplastic lesions with an incidence less than 0,5% and destructive capabilities higher than their precursors. Mushroom-like growth of cutaneous tumors described in 1806. by Albert, is still a rare phenomenon. GBCC's with mushroom-like macroscopic characteristics were hardly reported. We present a mushroom-like GBCC, treated successfully with wide surgical excision.

An 85-year-old male with a progressively growing skin lesion was admitted to our clinic. He came seeking medical care with a bleeding painful skin lesion. The tumor developed from a small papule into an exophytic pedunculated lesion with a fetid, ulcerated, exudative surface. Physical examination revealed 7x5x1.5cm mushroom-like, exophytic, pedunculated mass with rolled borders and partial cutaneous necrosis, signs of secondary infection and bleeding. The lesion was surgically removed. Histopathology showed basal carcinoma cell (BCC). Two months post-operatively there was no signs of tumor recurrence and secondary metastases without a need for additional treatment.

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GBCCs can still be identified in the 21st century. It arises due to neglecting of the lesion, inconspicuous localization usually covered by clothing. Mushroom-like morphology is rare in BCCs. Treatment is usually surgical excision, with a high success rate and an improvement in quality of life.

Keywords: Carcinomabasalcell, Cutaneoussurgery, Reconstructive surgical procedure

Introduction:

Basal cell carcinomas (BCCs) are the most common skin malignant neoplasms, with manifestations similar to other tumors of basal epithelial origin. They are typically considered slow-progressing, indolent and benign tumors. On the other hand, giant BCCs (GBCCs), are categorized as lesions with more than 5cm measured at its largest diameter, are aggressive, with local tissue destruction, higher risk of metastatic potential and poor prognosis, compared with conventional BCCs. (1)

Risk factors for development of these type of cutaneous neoplasms include low socio-economic status, neglect of the lesion and immunosuppression. (1-3) However, exophytic, vegetating, mushroom-like GBCCs have rarely been reported in the literature. (4) Differential diagnosis includes melanomas, squamous cell carcinomas, cylindroma. (3,4,12) Moreover, Arai et al. described two cases of giant skin adnexal tumor with a mushroom-like morphology. (5) Adequate differentiation between these lesions is crucial due to different behaviour, evolution, metastatic potential and treatment protocol.

Although most BCCs develop in photo-exposed areas, (6) almost 10-15% of BCCs evolve in uncommon anatomical sites, often covered by clothes. (7)

Case report:

An 85-year-old male patient was admitted to our Clinic, with a non-healing, bleeding lesion, located at the level of the inferior thoracic aperture and right hypochondriac region of the abdomen laterally from the 7th thoracic costal cartilage (Fig. 1)

Due to a long-term infection of the urinary tract, a malignant tumor of the right kidney was diagnosed. Afterward, the patient underwent a right complete nephrectomy in 2018. followed by an uncomplicated postoperative course. No other significant medical conditions were reported, he had no drug allergies and a negative family history of malignant disease.

The lesion was described as slow-progressing, initially progressing from a small papule to an exophytic, pedunculated tumor over the course of several years, with episodes of bleeding and eventually signs of necrosis and secondary infection. The patient decided to seek treatment after noticing an increase in bleeding and foul-smelling discharge from the lesion

The patient's skin type was Fitzpatrick type 2. He had no history of intense sun exposure.

Physical examination revealed a 7x5x1,5cm exophytic mass, with a purulent, foul-smelling exudate. Macroscopic characteristics of the tumor included large lobulated necrotic tissue with rolled borders, adhered haemorrhagic exudate and perilesional sclerotic cutaneous morphology. (Fig. 1)



The surgical resection specimen was comprised of cutaneous and subcutaneous tissue, 72x51x15mm, with whitish-brown node measuring 61x52x30mm. Histopathological, microscopic characteristics revealed nodular type of basal cell carcinoma with neoplastic shores and palisade cells. Stromal mucopolysaccharide retraction was observed microscopically within neoplastic cellular shores. Tumor cells showed hyperchromatic nuclei, with poor cytoplasmic content and volume. The tumor infiltrated the deep corium.

Figure 1. Clinical presentation of the lesion with a mushroom-like growth pattern

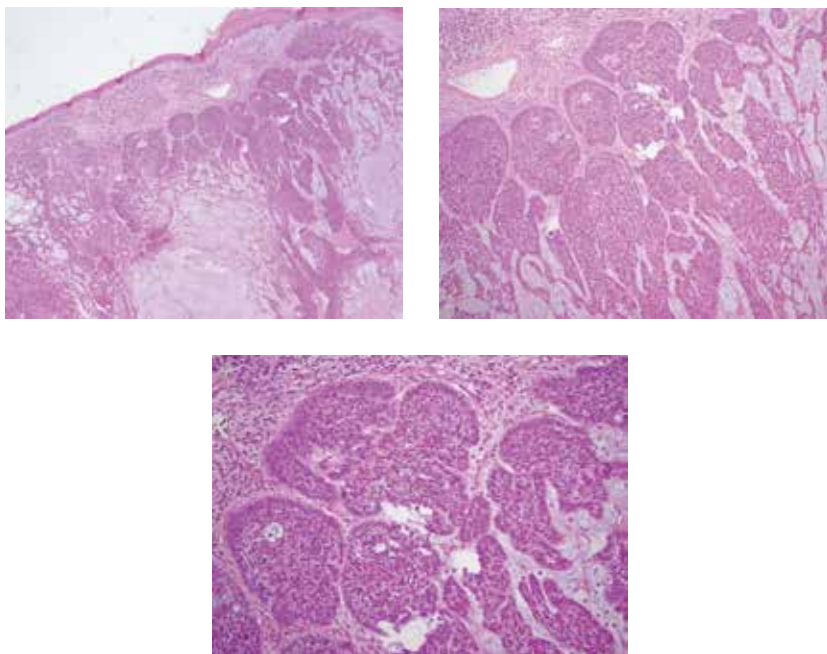


Figure 2A. H&E x40 Basal cell carcinoma nodular type. 50x39mm (220 x 220 DPI)

Figure 2B. H&E x100 Basal cell carcinoma nodular type. 49x39mm (220 x 220 DPI)

Figure 2C. H&E x200 All figures denote basal cell carcinoma nodular type. 53x38mm (220 x 220 DPI)

*Figure 2A. H&E x40 Figure 2B. H&E x100 Figure 2C. H&E x200
All figures denote basal cell carcinoma nodular type.*

Complete laboratory examination (including hematological, biochemical, serological, coagulation and infectious testing) was conducted and showed normal results.

Superficial wound swab was performed on admission, which revealed a secondary *Pseudomonas aeruginosa* infection, which was treated with intravenous Cefepime. No signs of metastatic spread were noted. Imaging (including both abdominal X-ray and *CT of the chest/abdomen and pelvis*) showed no adjacent soft tissue infiltration, and no involvement in thoracic cavity and bone

After adequate preoperative preparation, the patient underwent a wide local excision and removal of the tumor mass with (negative surgical margins of over 5mm and complete wound closure. Postoperative follow-up showed complete, primary wound healing, with optimal alignment of wound edges. A bi-monthly follow-up documented satisfactory aesthetic results, with no complications or signs of recurrence of the tumor mass. (Fig. 3).



Defect closure directly, follow up two months after surgery-linear scar

Figure 3. Defect closure directly, follow up two months after surgery-linear scar

Discussion:

GBCC arises typically in elderly male patients, with highest incidence in the 7th decade, usually located on the back, face and upper extremities. In our case, it was located on the abdominal wall. GBCC is generally defined as a lesion over 5cm measured at its greatest diameter. (1) The most common histological subtype being nodular BCC. It is usually presented as a localized exophytic mass on the surface of the skin, and is more common than squamous cell carcinoma (SCC). It is necessary to differentiate BCC from SCC, since SCC has a higher risk of recurrence and metastatic potential. Another described differential diagnoses are melanomas and skin adnexal tumor (4,5). Furthermore, the mushroom-like growth pattern has been described for cylindroma by Wollina et al. (12)

Although there are no clear guidelines on the precise method of treatment, aggressive management offers the best treatment outcome. (5) Potential challenges occur due to tumor size and it is often impossible to perform closure of defect without subsequent functional and cosmetic damage (6) Optimal management of GBCC consists of wide local excision with histologically confirmed tumor-free margins, frequently followed with adjuvant therapy. (8) Radical surgical excision remains a known gold standard, but several studies reporting nonsurgical treatment options (including radiotherapy, vismodegib, topical imiquimod, acitrecin) for GBC-Cs. (9) The basal cell carcinoma, in our case, was slow growing neoplasm in nature, with no change in growth pattern, thereby suggesting that giant BCC was the result of negligence in looking for early medical treatment. Chronic neglect, infection,

tissue necrosis and bleeding all contribute to a higher probability of an unsuccessful treatment outcome (9,10)

Often for giant BCCs, covering the skin defect after excision using a cutaneous flap or various skin grafts may often be essential. (11) As seen in our case, the skin defect was closed directly with extensive tissue undermining.

A unique case of a giant mushroom-like BCC in full remission after complete excision and closure has been described. BCCs have a tendency to be indolent, painless tumors, which may permit patients to ignore the lesion, resulting in neglected BCC, which can become a large fungus-like mass, with a risk of bleeding, infection, metastatic spread and poor treatment outcomes.

Conflicts of interest: There are no financial conflicts of interest to disclose.

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