POLICLINICO MILITARE DI ROMA "CELIO"

DIPARTIMENTO MEDICINA – UOS DERMATOLGIA e m.s.t.

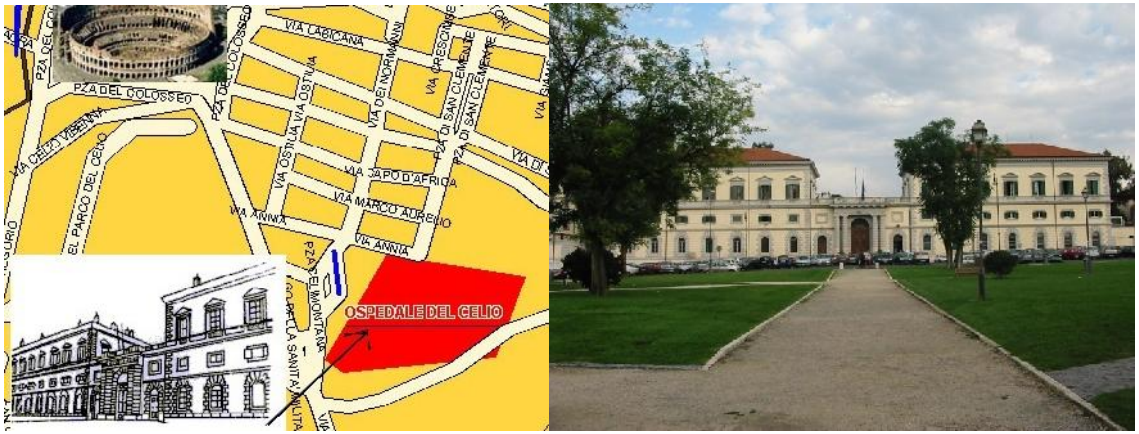
Dir. u.o.s.: Col.Med. Stefano Astorino

TECNOLOGIA E INNOVAZIONE TERAPEUTICA IN DERMATOLOGIA
dalla ricerca alla pratica clinica
3-4 Novembre 2016, Roma

4nov2016 Sessione 14.00-16.00 Il mio caso… difficile, interessante o irrisolto

"EROSIV PUSTULAR DERMATOSIS OF THE SCALP" IN A PATIENT UNDERWENT SURGERY FOR "PLEOMORPHIC DERMAL SARCOMA" / "ATIPICAL FIBROXANTHOMA": CLINICAL AND DERMATOSCOPIC FEATURES

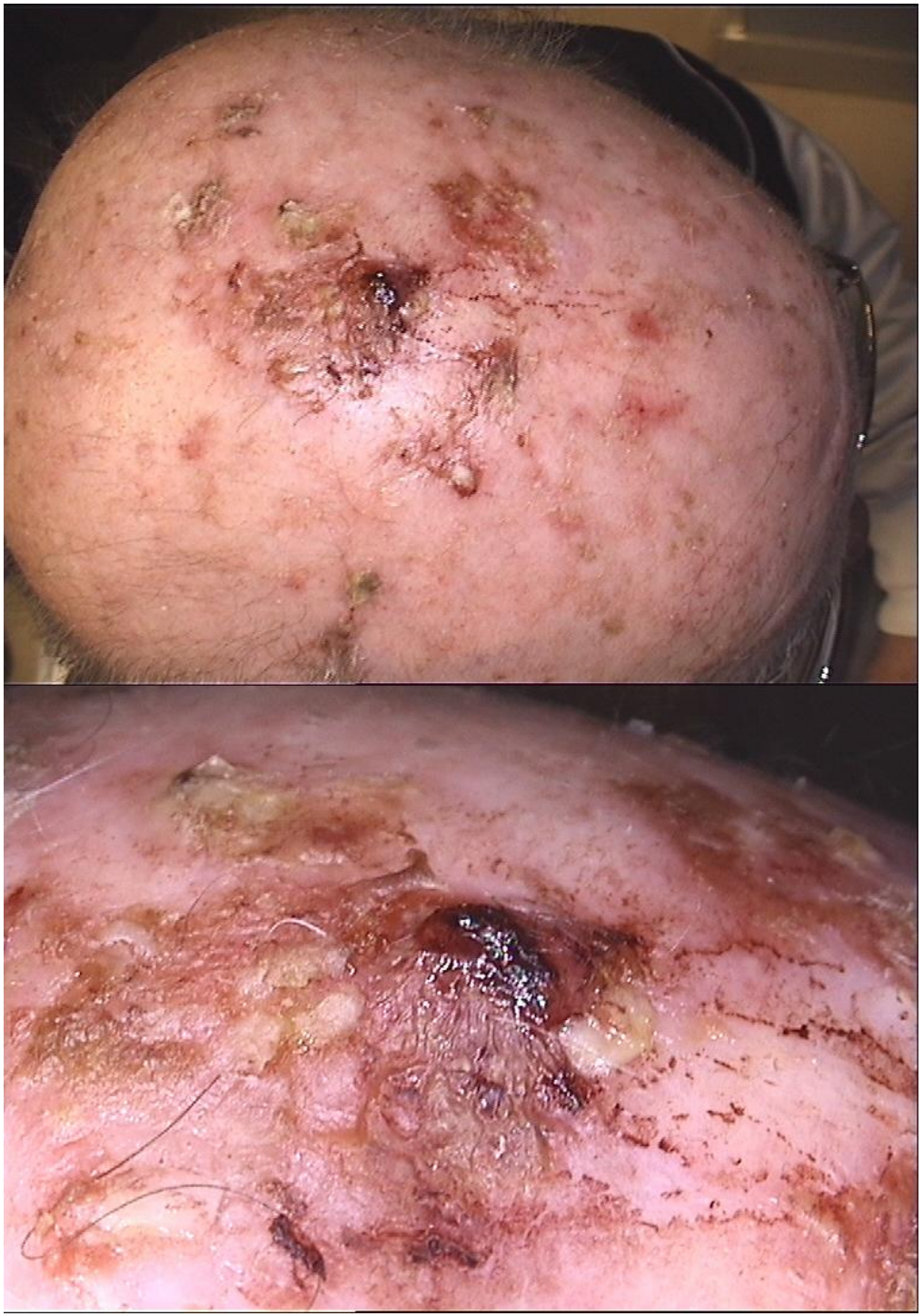
Stefano ASTORINO

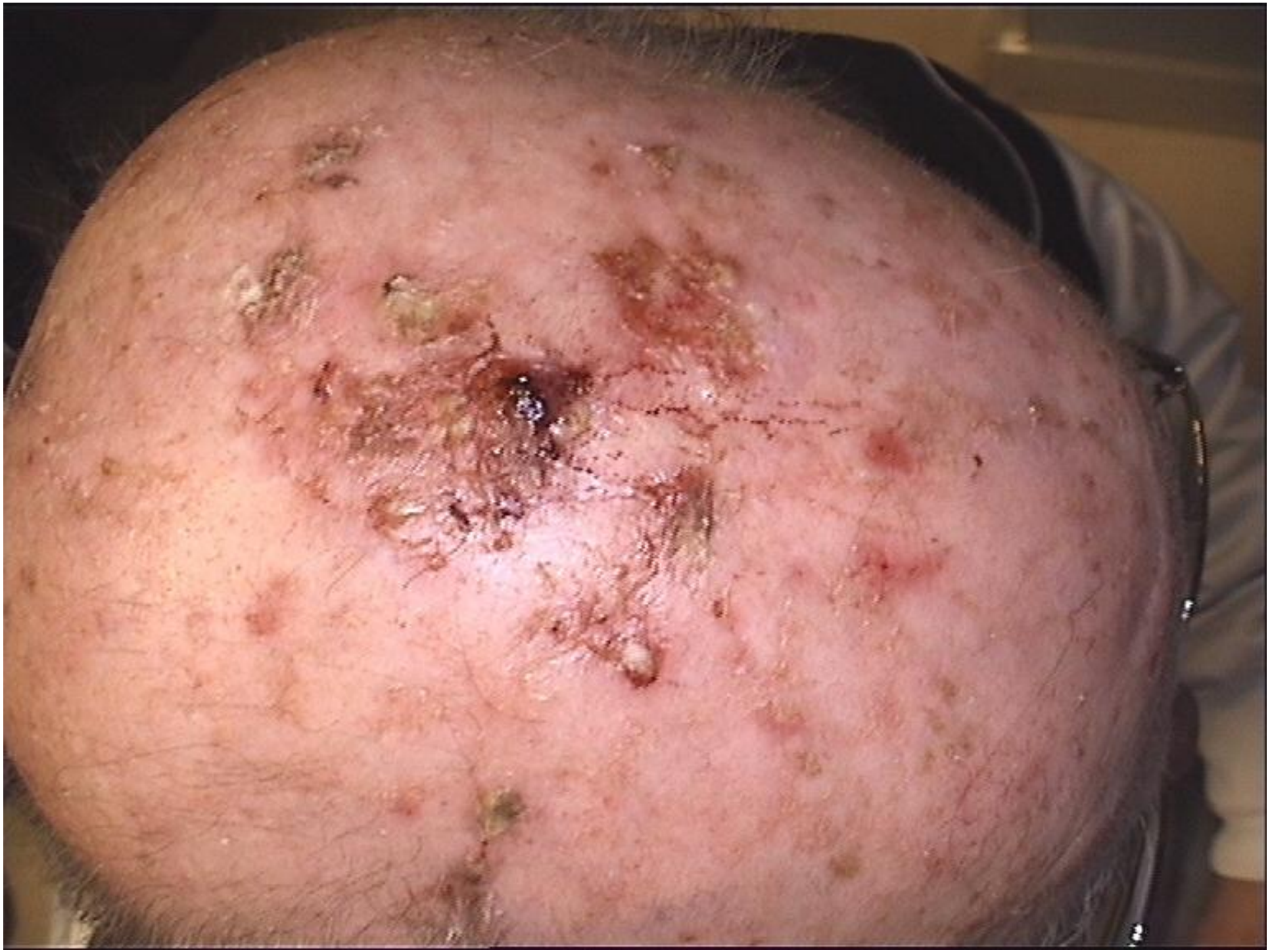


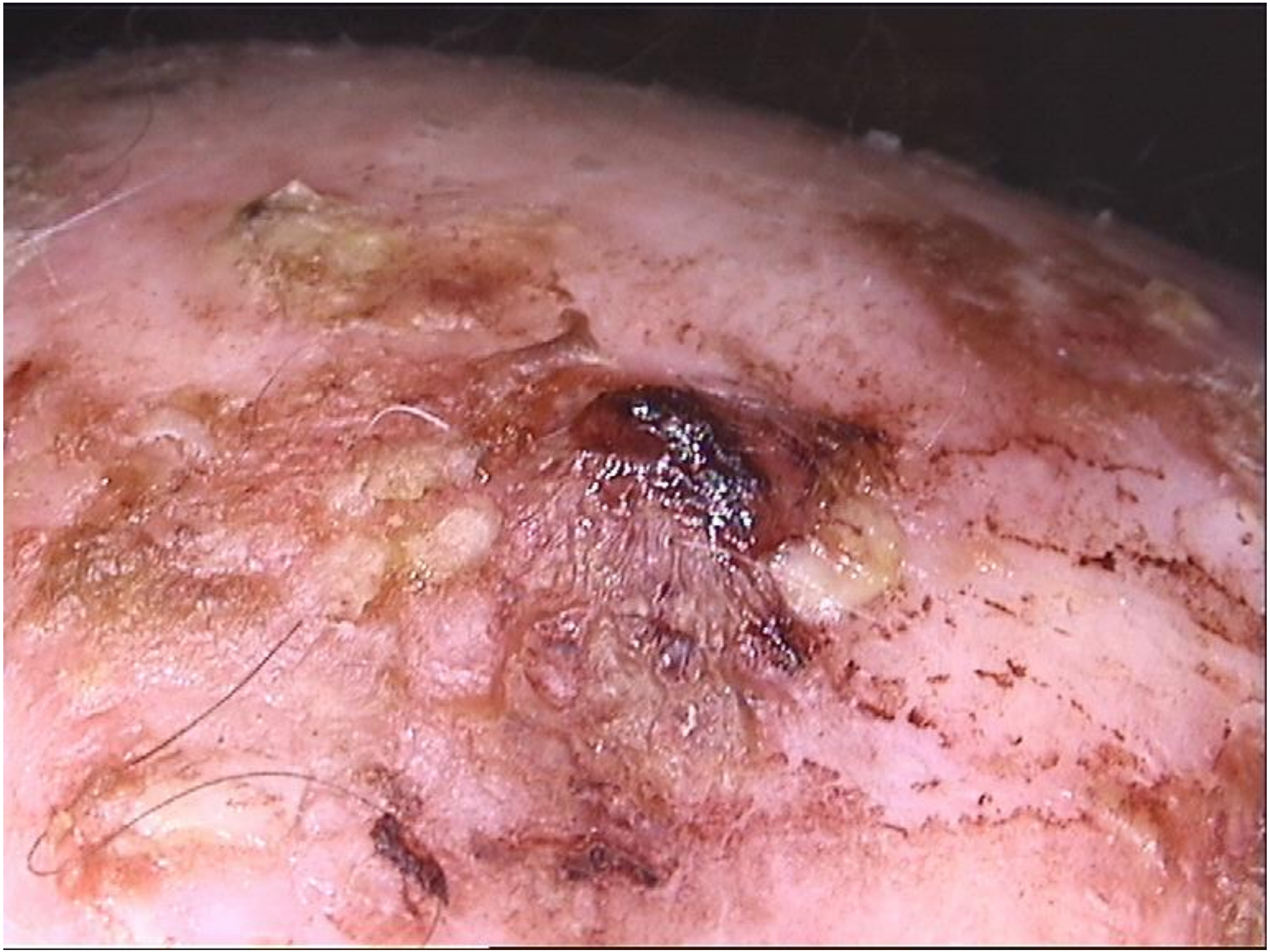
Leon's Place
Via XX Settembre, 90/94, Roma

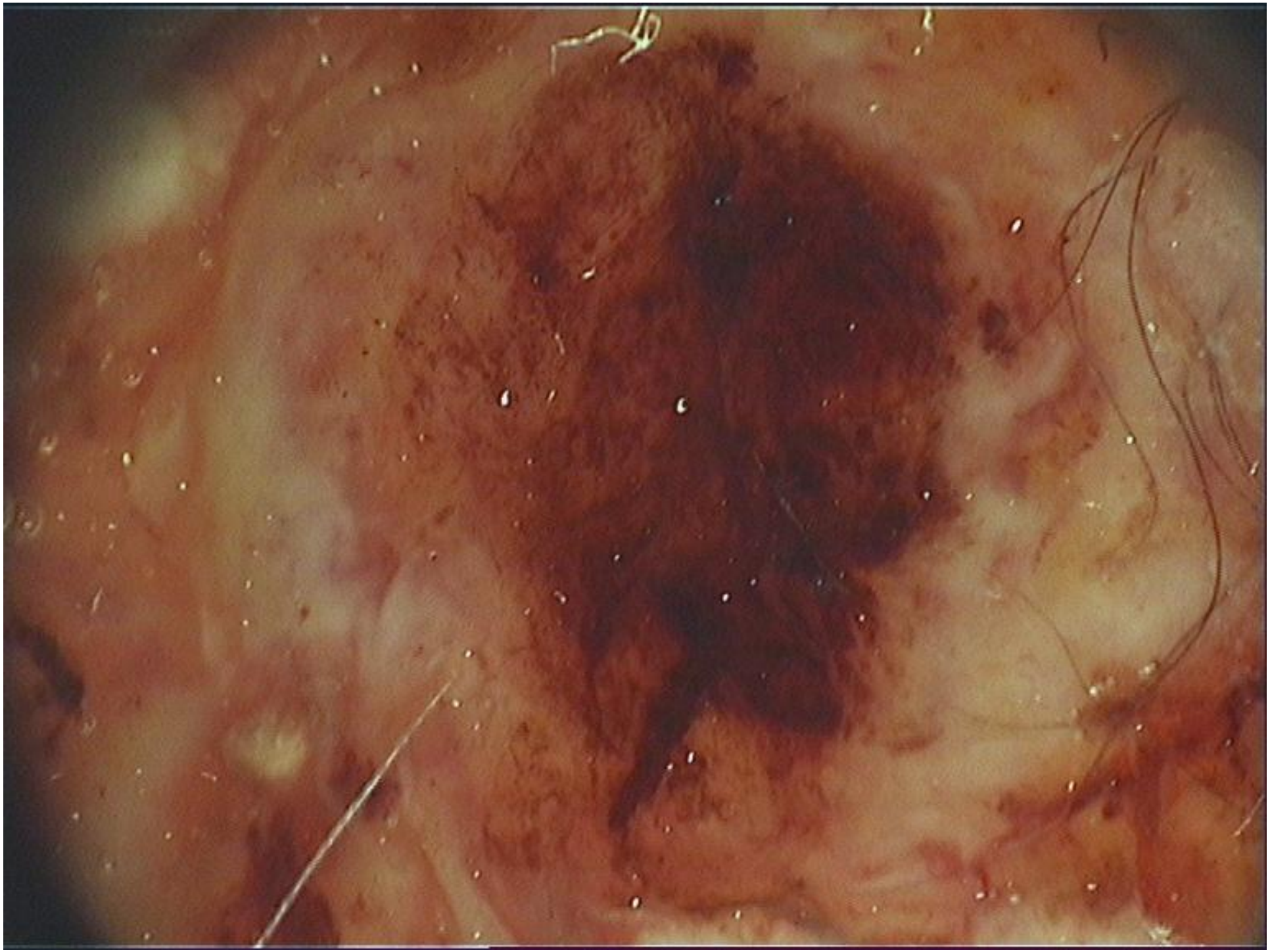
Ven. 4 Novembre 2016
h.15.20

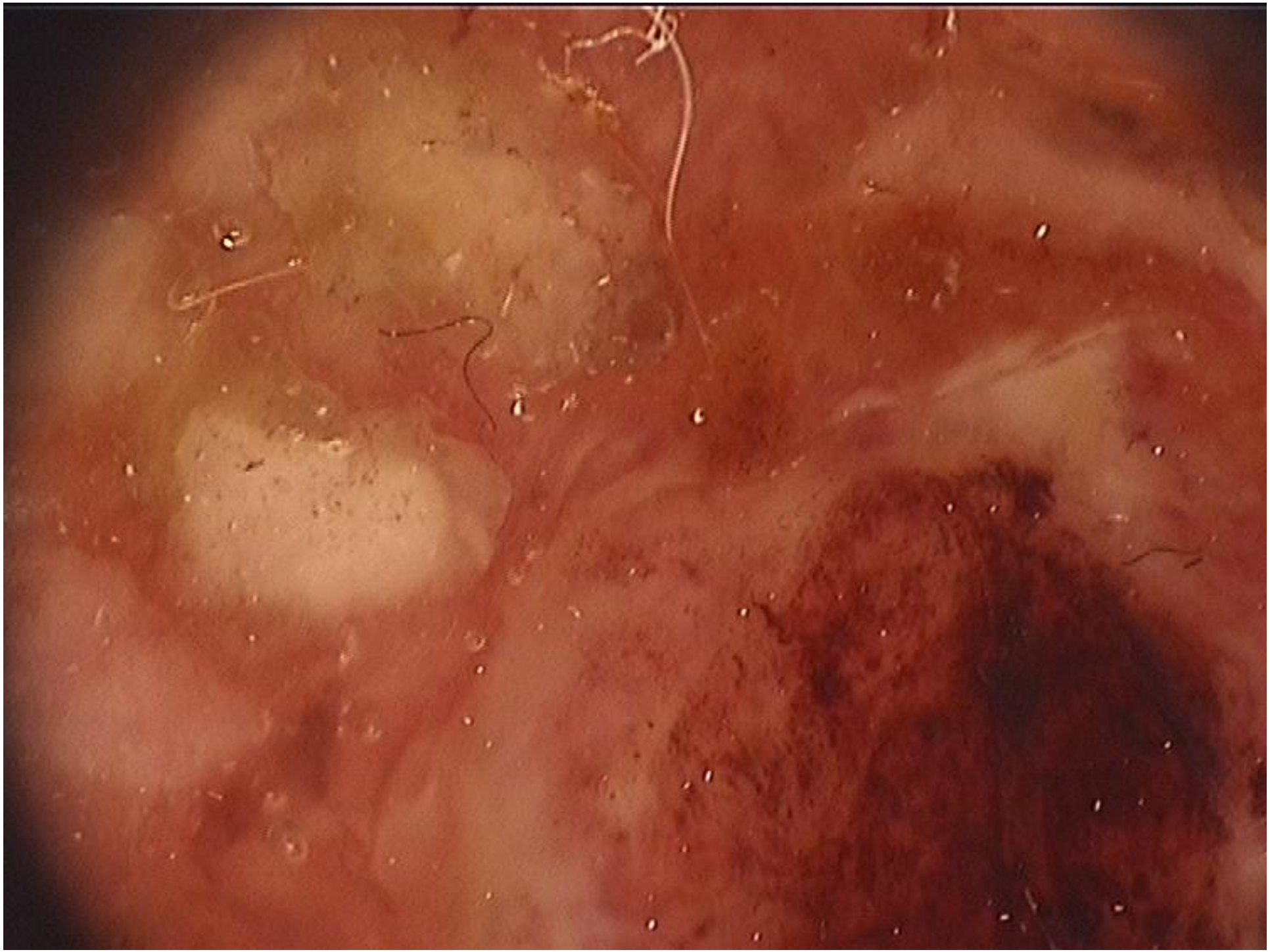
- Viene riportato il caso di un paziente di 67 anni (anamnesi positiva per recente ischemia cerebrale e per parkinsonismo in trattamento), che il 26/01/2015 giunge all'osservazione per la presenza di cheratosi attiniche in trattamento e per la riferita recente comparsa a livello del vertice del capillizio (calvo) di un nodulo ulcerato a rapida crescita (circa 2 mesi), sede di sanguinamento spontaneo

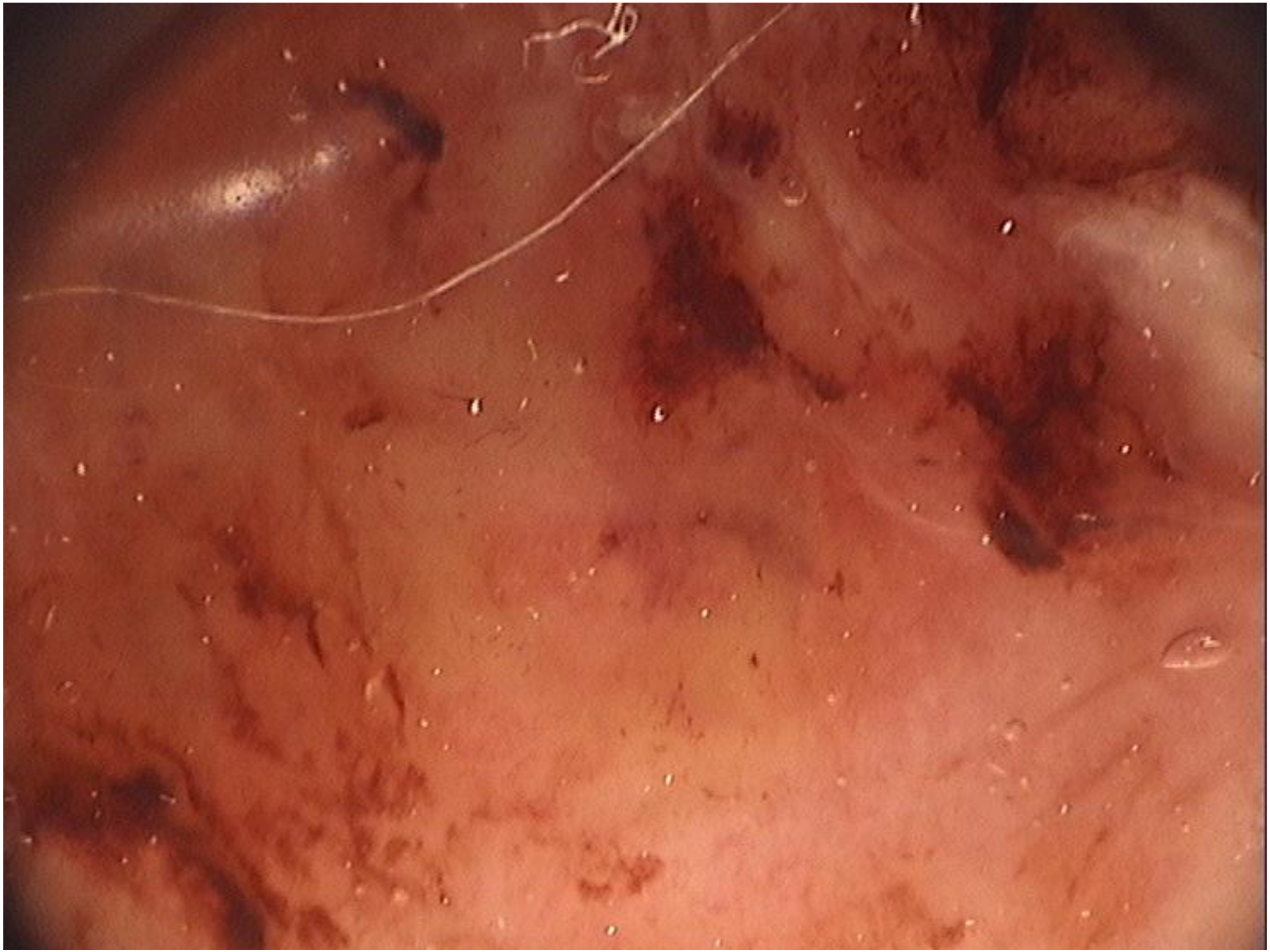








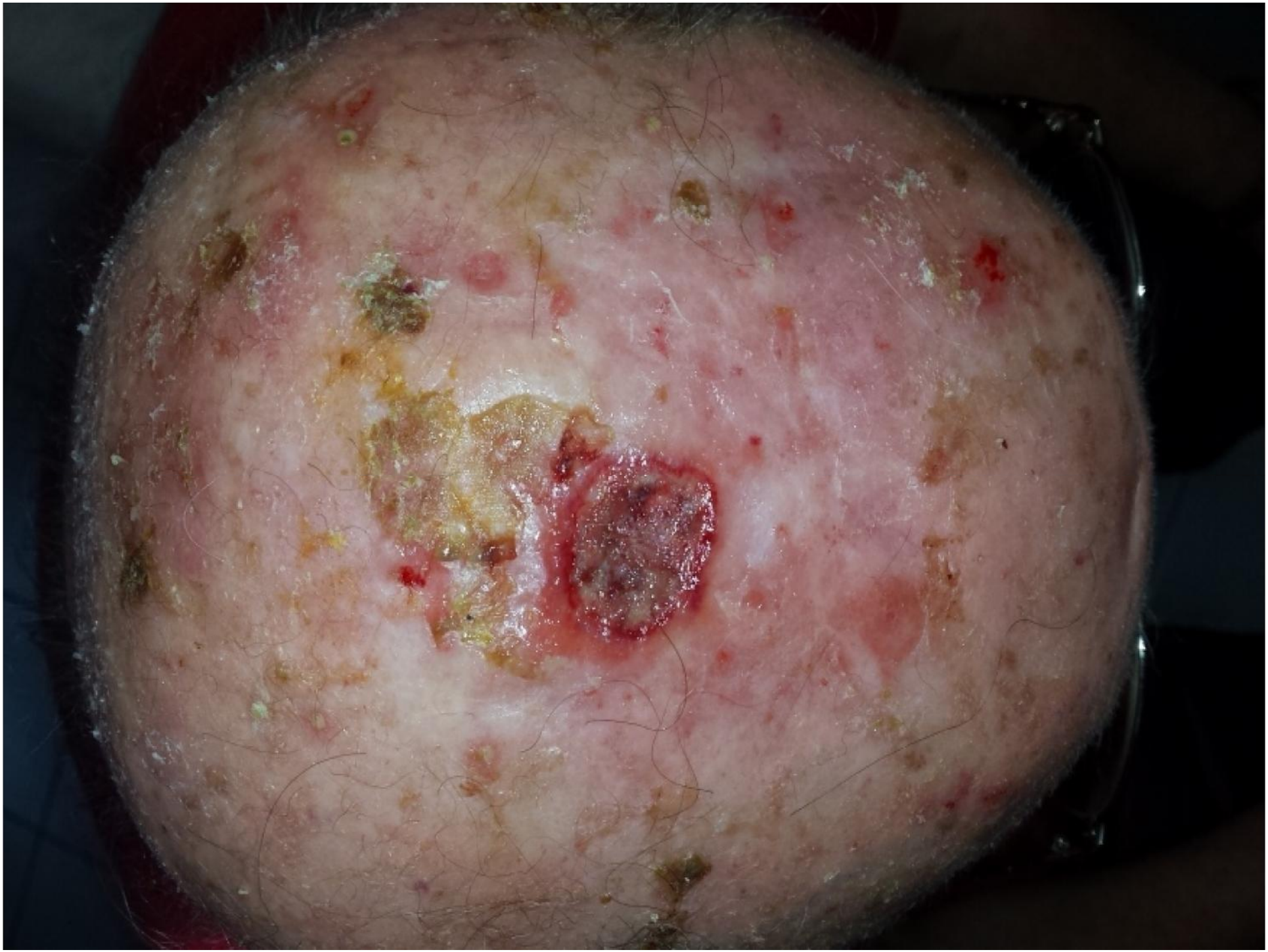


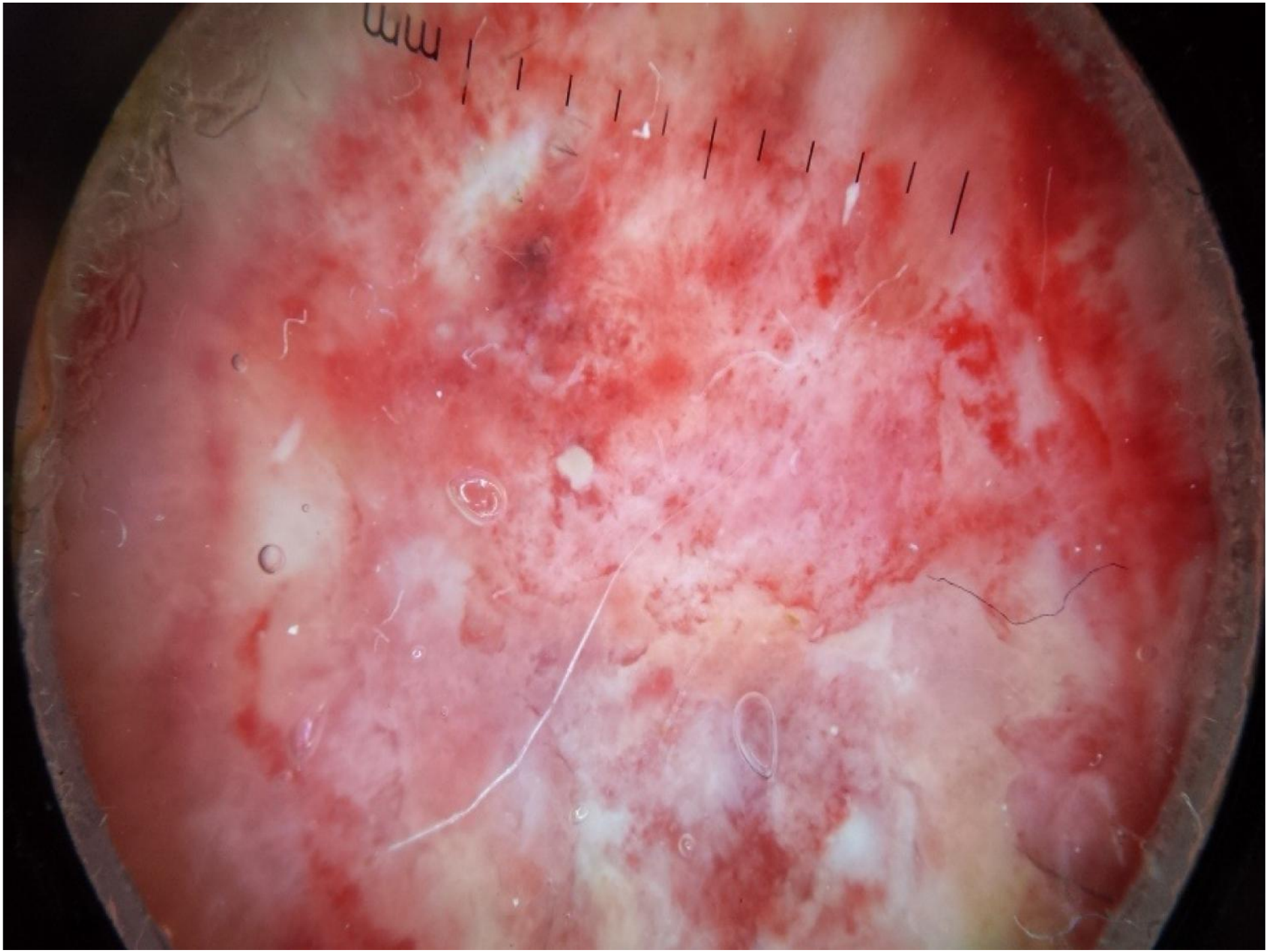


IPOTESI DIAGNOSTICHE

1. SCC ULCERATO
2. MELANOMA ACROMICO
3. BCC ACROMICO E ULCERATO
4. GRANULOMA PIOGENICO
5. ?

- Shaving-biopsy + emòstasi della base mediante DTC sulla lesione nodulare del vertice:
- R./ Diclofenac 3% gel, la sera
- R./ Azitromicina 500mg 1cpr/die x 3gg
- R./ Gentamicina crema, per medicazione locale
- Controlli ogni 1-2 settimane

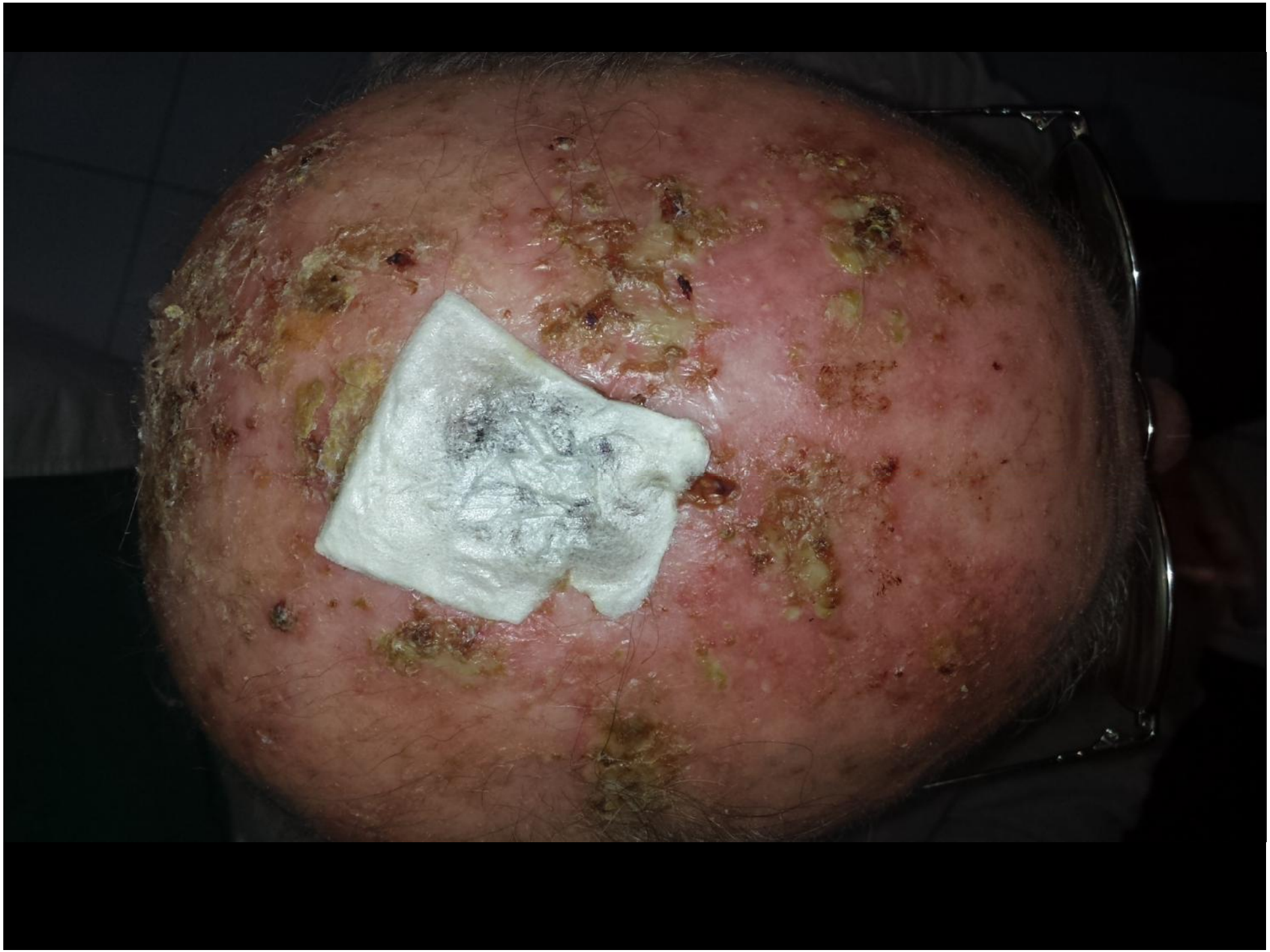


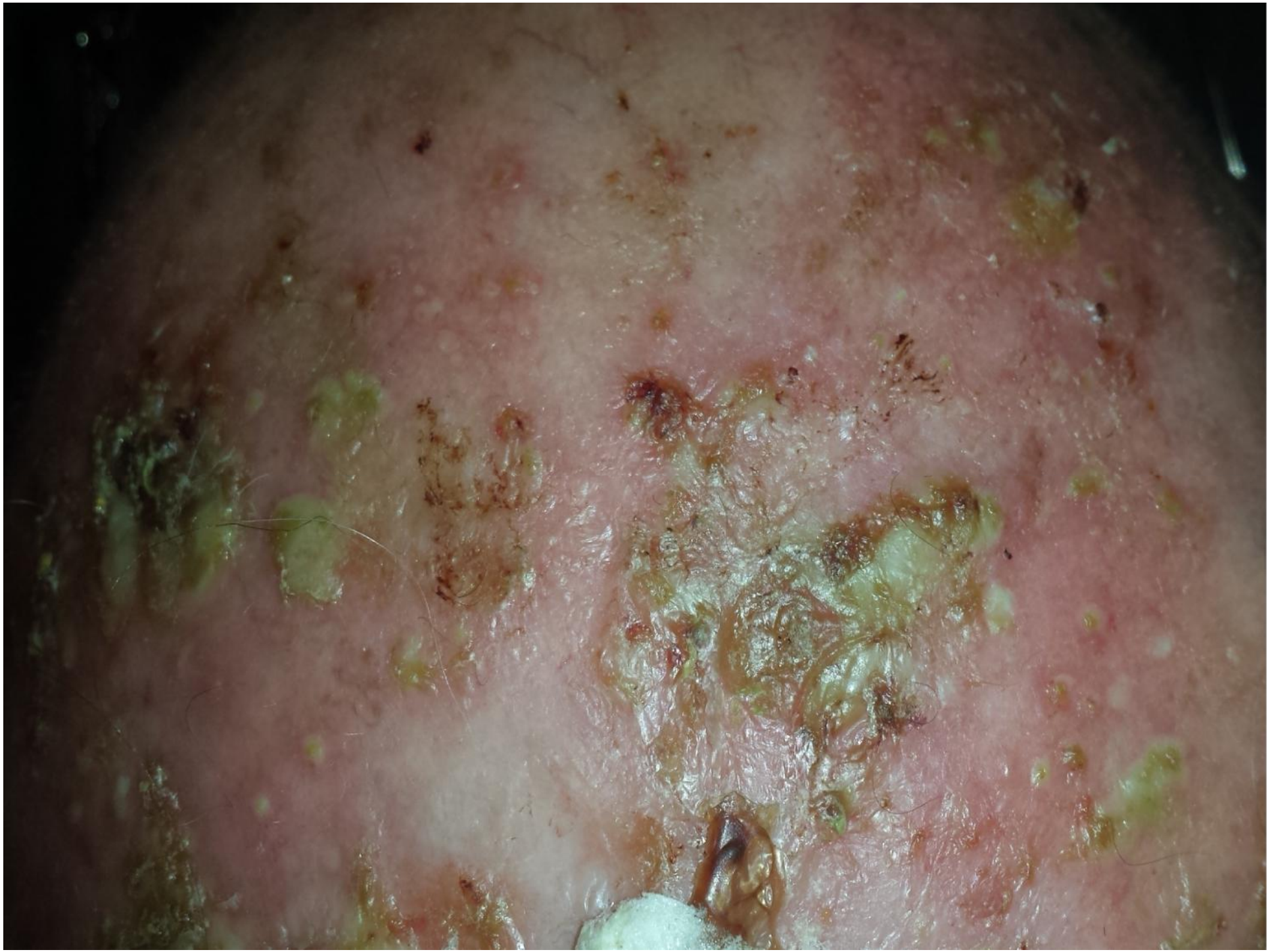


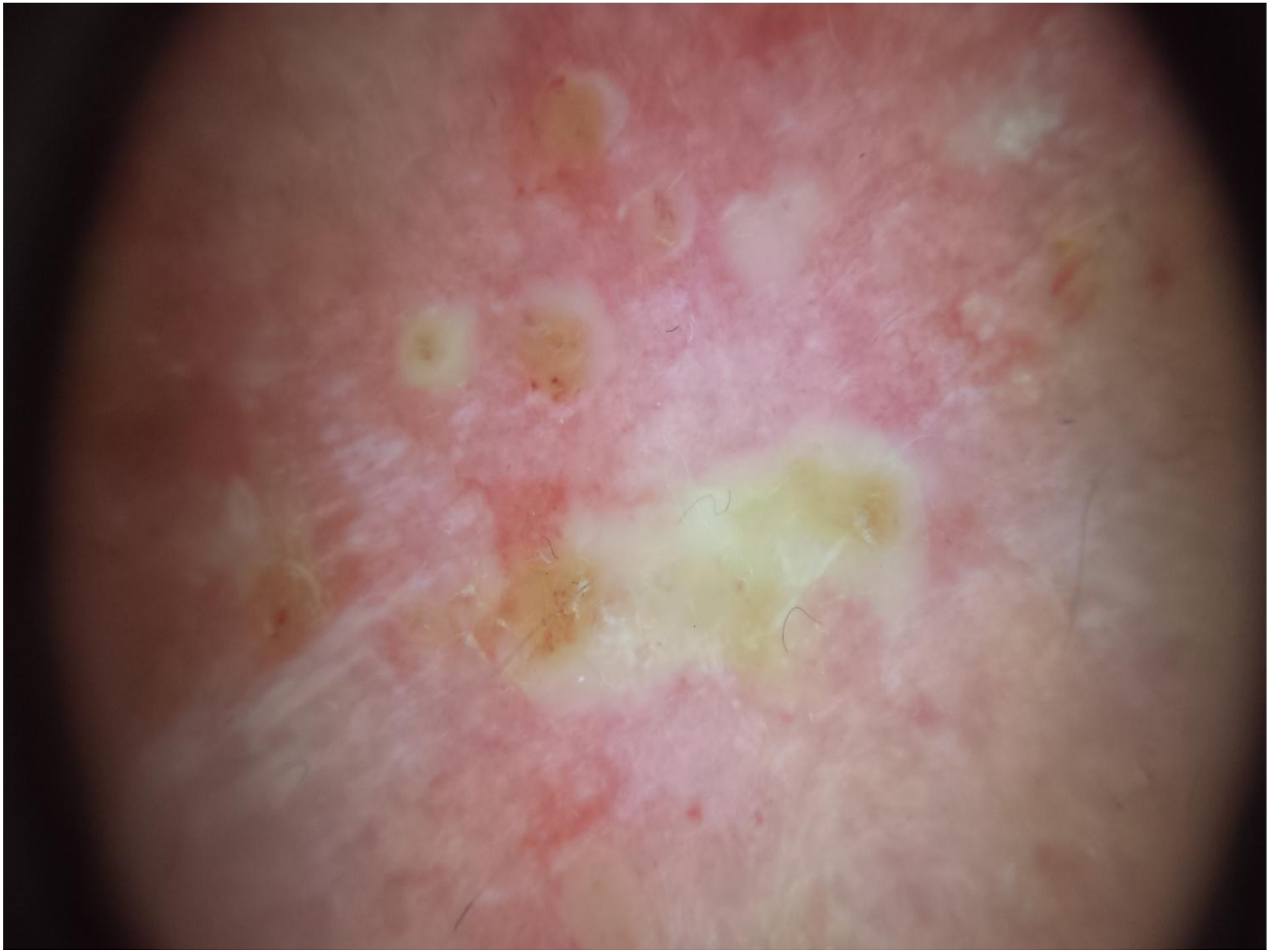
- Dermatoscopicamente la lesione presenta un pattern globale aspecifico, con assenza di pigmento, zone periferiche irregolari prive di struttura, ulcerazione, globuli rosso-lattescenti irregolarmente distribuiti e un pattern vascolare atipico con presenza di vasi puntiformi, lineari irregolari e a forcina.

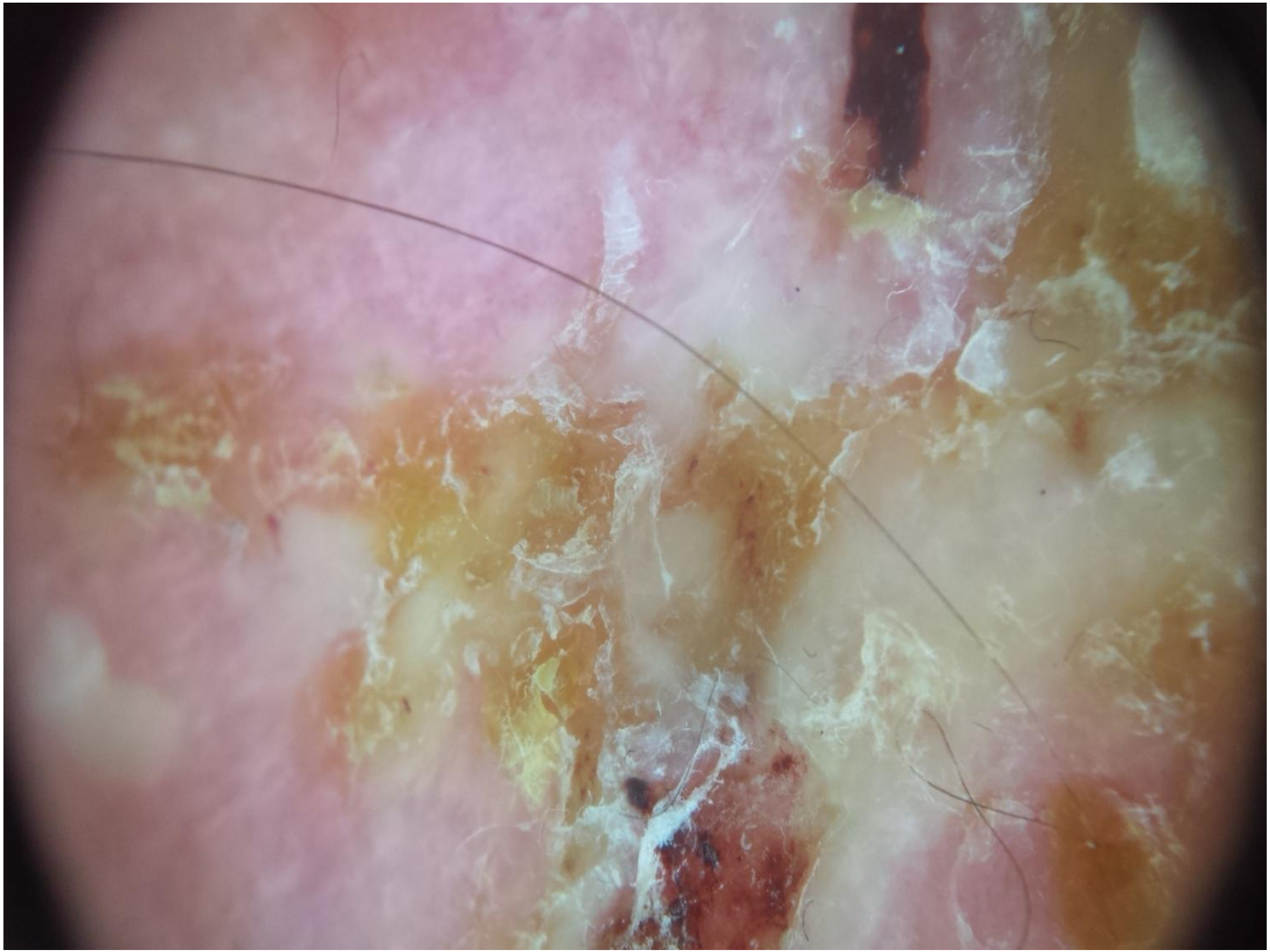
- Merc.11.2.15

- R./ ingenolo mebutato 150mcg 1v./die x 3gg

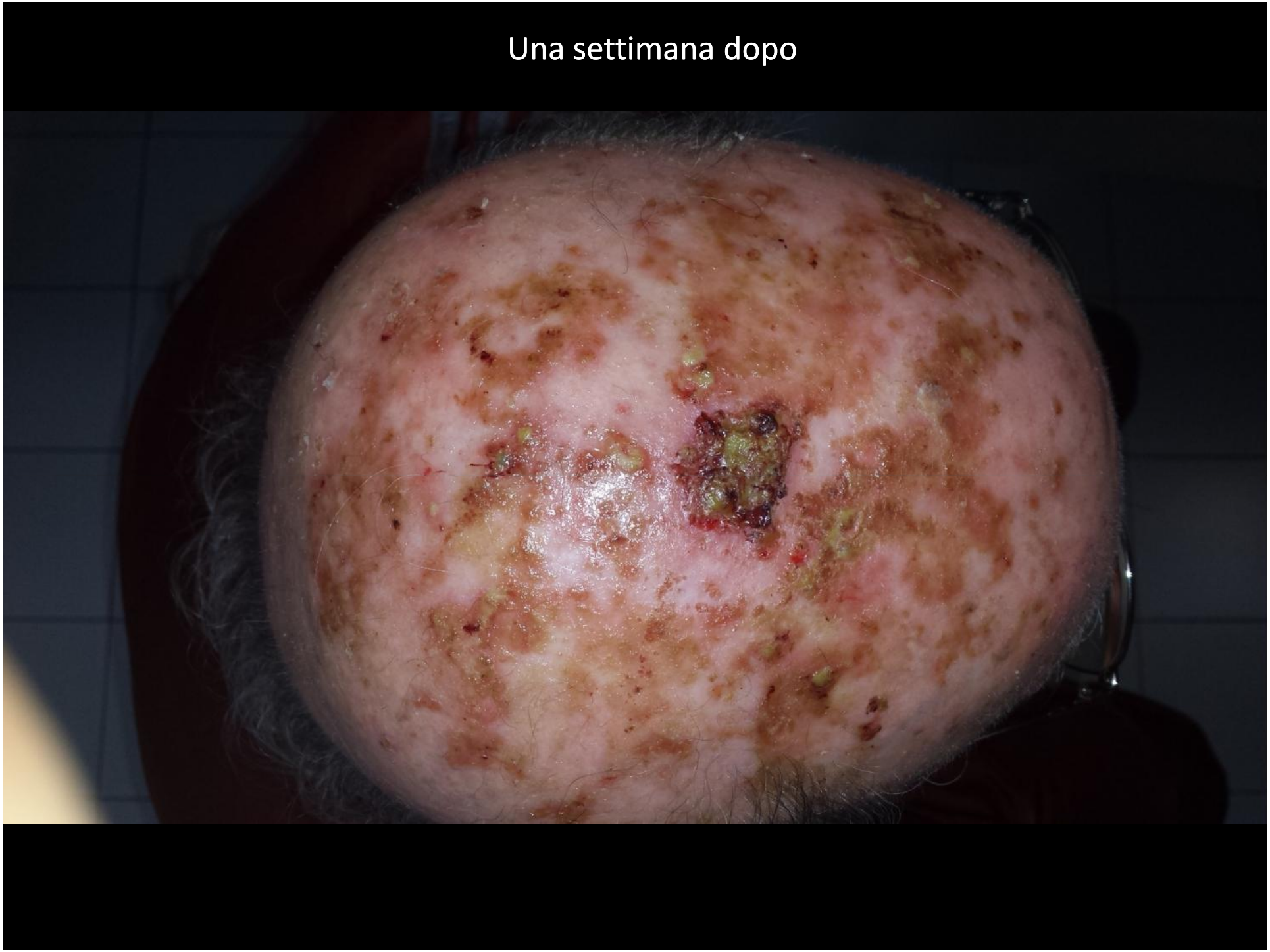








Una settimana dopo



Reparto: **DERMATOLOGIA**
Data di richiesta: 26.01.15

Medico richiedente: Dott. S. Astorino
Data di accettazione: 29.01.15

Organo/sede: vertice
Procedura: shave biopsy

Notizie cliniche: diagnosi clinica di sospetto spinalioma/cheratosi attinica ipertrofica.

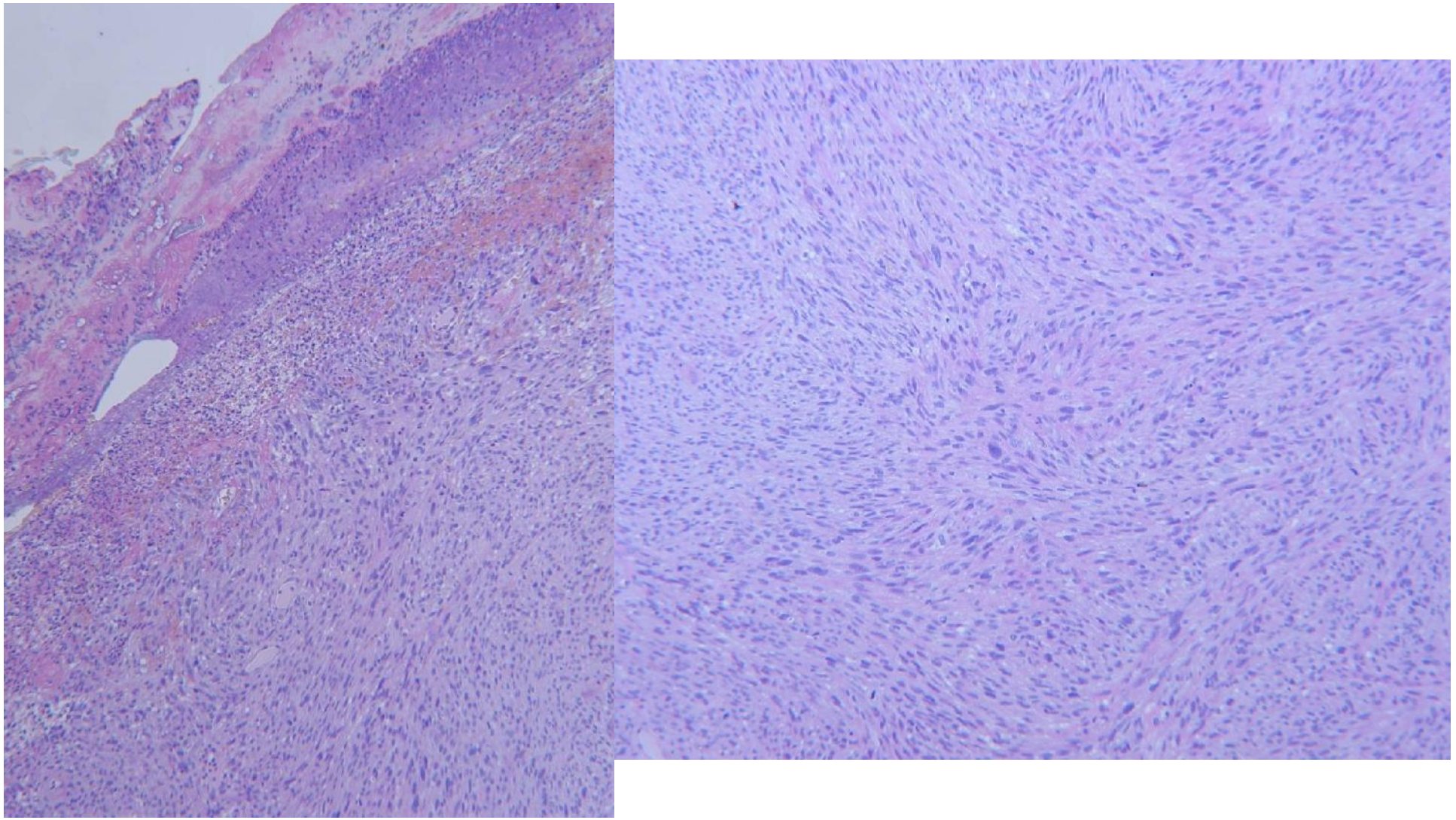
Reperto macroscopico/microscopico

Frammento non orientabile di cute delle dimensioni di cm 1,3 x 1 x 0,3 estesamente ulcerato e caratterizzato da una proliferazione dermica di cellule fusate ed epitelioidi marcatamente atipiche con disposizione fascicolata/storiforme ad attività mitotica elevata.
Il profilo immunofenotipico della neoplasia ha documentato negatività per pancitocheratina, EMA, S100, Melan-A, CD34 e desmina e positività per vimentina, CD10, actina muscolo liscia, caldesmone e fattore XIIIa.

Diagnosi istologica

Il quadro morfologico e immunofenotipico appaiono suggestivi per **sarcoma dermico pleomorfo** cfr. Am J Surg Pathol 2012 Sep;36 (9).
La lesione risulta presente a livello dei margini di resezione.

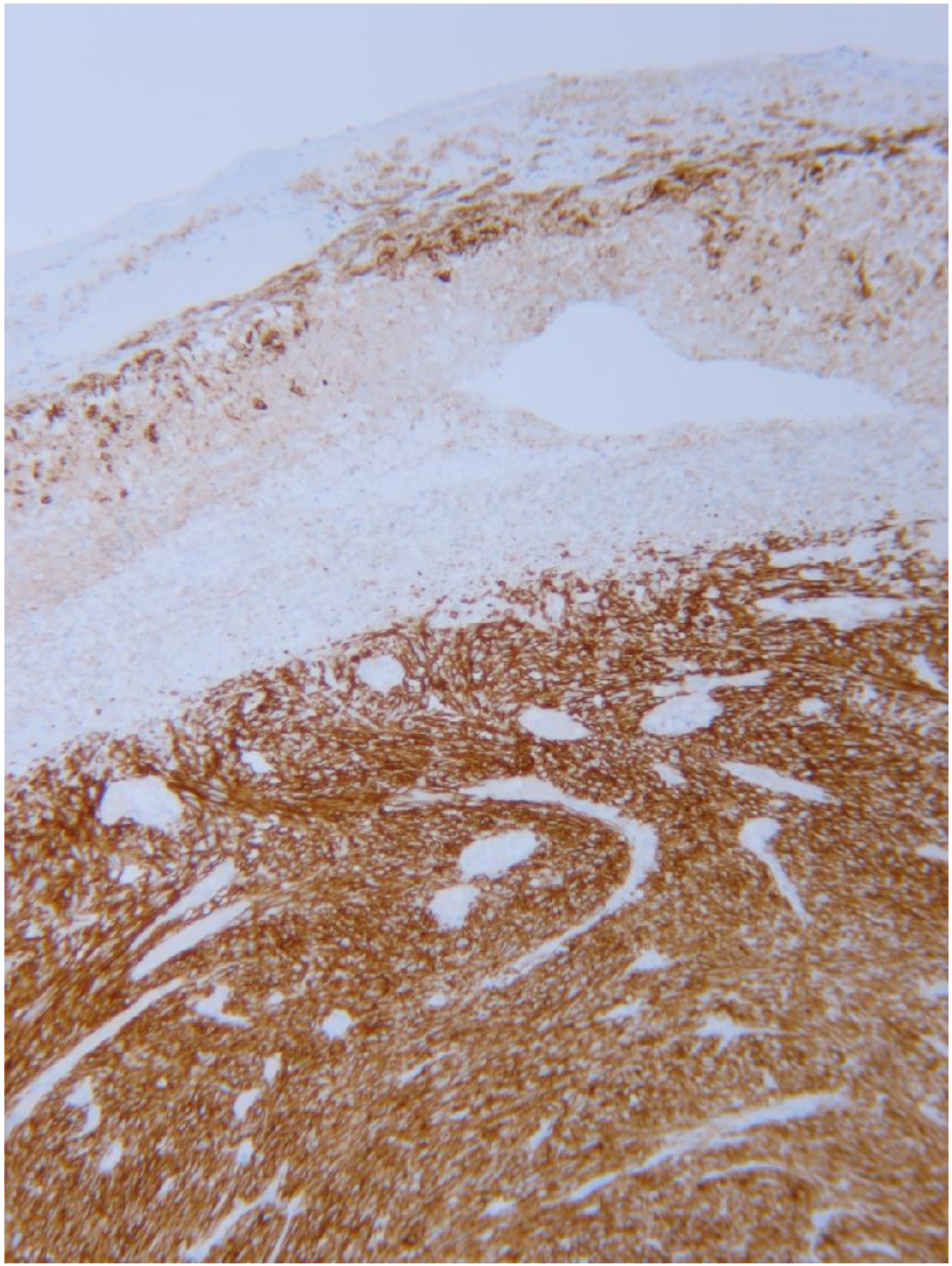
Roma, 05.03.15



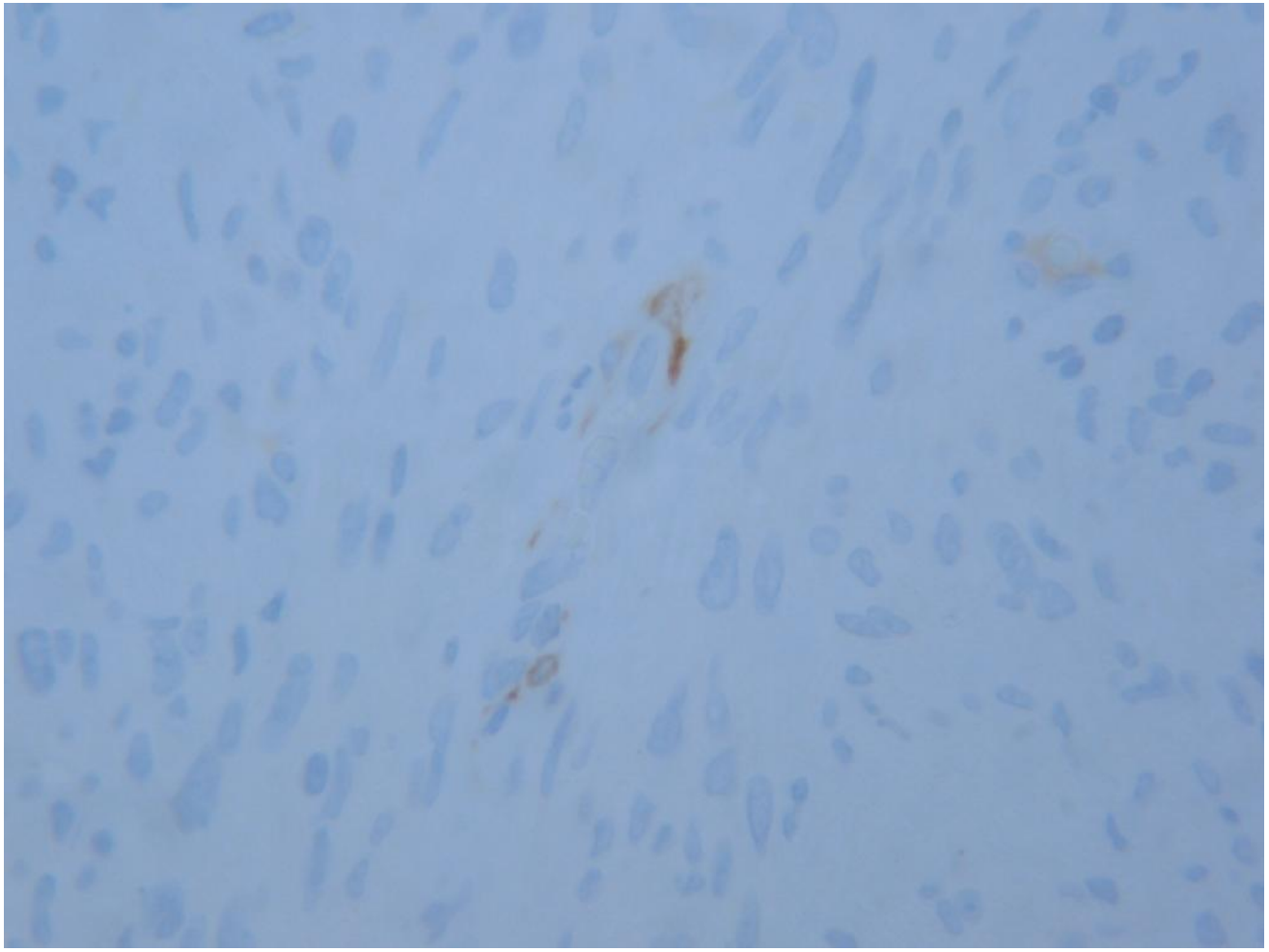
Dettaglio morfologico della lesione che ulcera la cute sovrastante ed è costituita da una proliferazione di cellule fusate e pleomorfe con marcato indice mitotico

- L’ esame istologico evidenzia una proliferazione dermica di cellule fusate epitelioidi marcatamente atipiche con disposizione fascicolata-storiforme ad elevata attività mitotica. Il profilo immunoistochimico della neoplasia ha inoltre documentato negatività per pancitocheratina, EMA, S100, Mel-A, CD34 e desmina, e positività per vimentina, CD10, actina muscolo-liscia, caldesmone e fattore XIIIa.

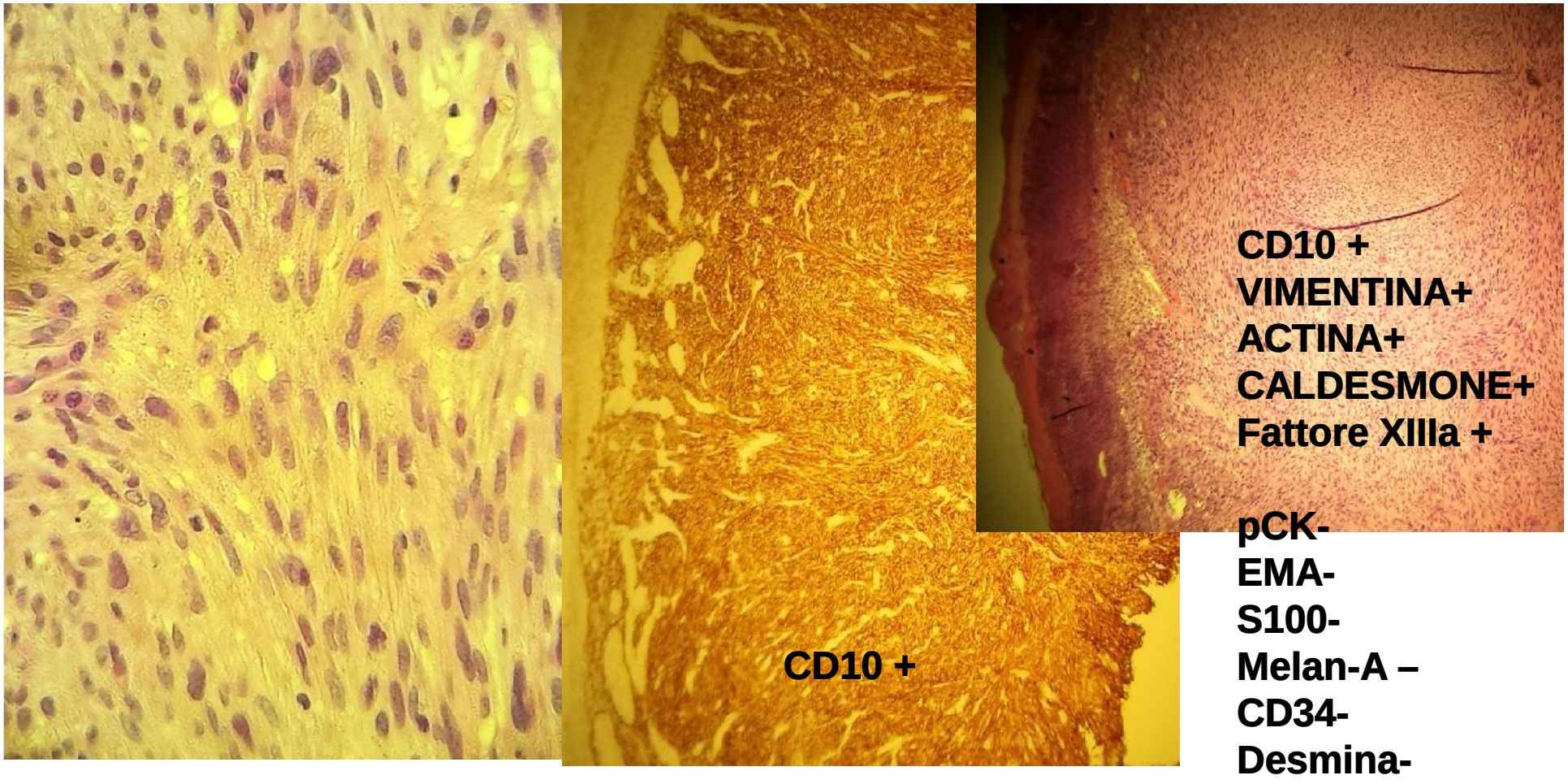
- Il quadro morfologico ed immunofenotipico permettono la diagnosi di “Sarcoma dermico pleomorfo”, entità clinico-istopatologica a differenziazione fibro-istiocitaria (precedentemente corrispondente all’ “Istiocitoma fibroso maligno”), caratterizzata da una marcata invasione del tessuto sottostante, necrosi, invasione linfovascolare e perineurale ed associata a prognosi avversa.



Positività immunoistochimica dei citoplasmi al CD10



Negatività immunoistochimica alla desmina (d.d. con leiomiosarcoma)



Zschoche C, Hamsch C, Kutzner H, et al.:

Analysis of the lymphatic vessel architecture of atypical fibroxanthoma and pleomorphic dermal sarcoma.

J Am Acad Dermatol. 2014 Oct;71(4):842-5.

- The term “pleomorphic dermal sarcoma” was introduced by Fletcher [2] and describes tumors having been referred to “cutaneous undifferentiated pleomorphic sarcomas” or “superficial malignant fibrous histiocytomas” in the past. These tumors present with a similar morphology to AFX, but in addition, show extensive invasion of deeper structures [1, 2]. Both, AFX and PDS are usually negative for cytokeratins, S100, CD34, and desmin [3].

- McCalmont TH. Correction and clarification regarding AFX and pleomorphic dermal sarcoma. Journal of cutaneous pathology. 2012; 39:8.

- L’intervento chirurgo plastico di allargamento dei margini (aprile 2015) viene effettuato con graft cutaneo autologo, che attecchisce solo parzialmente, lasciando una ulcerazione che viene trattata in vulnoterapia con medicazioni avanzate fino a settembre 2016

- TAC cranio e TB (ripetute ogni 6-8 mesi fino ad oggi): negative per lesioni metastatiche

- in data 15/06/2016 una revisione dei vetrini (n.146/15c/o S. Gallicano) dimostra un orientamento diagnostico per “Fibroxantoma atipico, varietà a cellule fusate.

- Secondo recenti vedute oncogenetiche “atipical fibroxanthoma” (AFX)e “pleomorphic dermal sarcoma” (PDS) sarebbero due varianti della stessa malattia con potenzialità maligna maggiore per il “pleomorphic dermal sarcoma” e AFX sarebbe il precursore non invasivo di PDS.

Helbig D, Ihle MA, Pütz K, Tantcheva-Poor I, Mauch C, Büttner R, Quaas A. Oncogene and therapeutic target analyses in atypical fibroxanthomas and pleomorphic dermal sarcomas. Oncotarget. 2016 Apr 19;7(16):21763-74.

Oncogene and therapeutic target analyses in atypical fibroxanthomas and pleomorphic dermal sarcomas

Doris Helbig[1,*], Michaela Angelika Ihle[2,*], Katharina Pütz[2], Iliana Tantcheva-Poor[1], Cornelia Mauch[1], Reinhard Büttner[2], Alexander Quaas[2]

[1]Department of Dermatology, University Hospital Cologne, Cologne, Germany

[2]Institute of Pathology, University Hospital Cologne, Cologne, Germany

[*]These authors have contributed equally to this work

Correspondence to: Doris Helbig, e-mail: doris.helbig@uk-koeln.de

Keywords: atypical fibroxanthoma, CDK4, CCND1, pleomorphic dermal sarcoma, TP53

Received: January 05, 2016 *Accepted: February 21, 2016* *Published: March 02, 2016*

ABSTRACT

Background: Until now, almost nothing is known about the tumorigenesis of atypical fibroxanthoma (AFX) and pleomorphic dermal sarcoma (PDS). Our hypothesis is that AFX is the non-infiltrating precursor lesion of PDS.

Materials and Methods: We performed the world-wide most comprehensive immunohistochemical and mutational analysis in well-defined AFX (n=5) and PDS (n=5).

Results: In NGS-based mutation analyses of selected regions by a 17 hotspot gene panel of 102 amplicons we could detect *TP53* mutations in all PDS as well as in the only analyzed AFX and PDS of the same patient. Besides, we detected mutations in the *CDKN2A, HRAS, KNSTRN* and *PIK3CA* genes.

Performing immunohistochemistry for CTNNB1, KIT, CDK4, c-MYC, CTLA-4, CCND1, EGFR, EPCAM, ERBB2, IMP3, INI-1, MKI67, MDM2, MET, p40, TP53, PD-L1 and SOX2 overexpression of TP53, CCND1 and CDK4 was seen in AFX as well as in PDS. IMP3 was upregulated in 2 AFX (weak staining) and 4 PDS (strong staining).

FISH analyses for the genes *FGFR1, FGFR2* and *FGFR3* revealed negative results in all tumors.

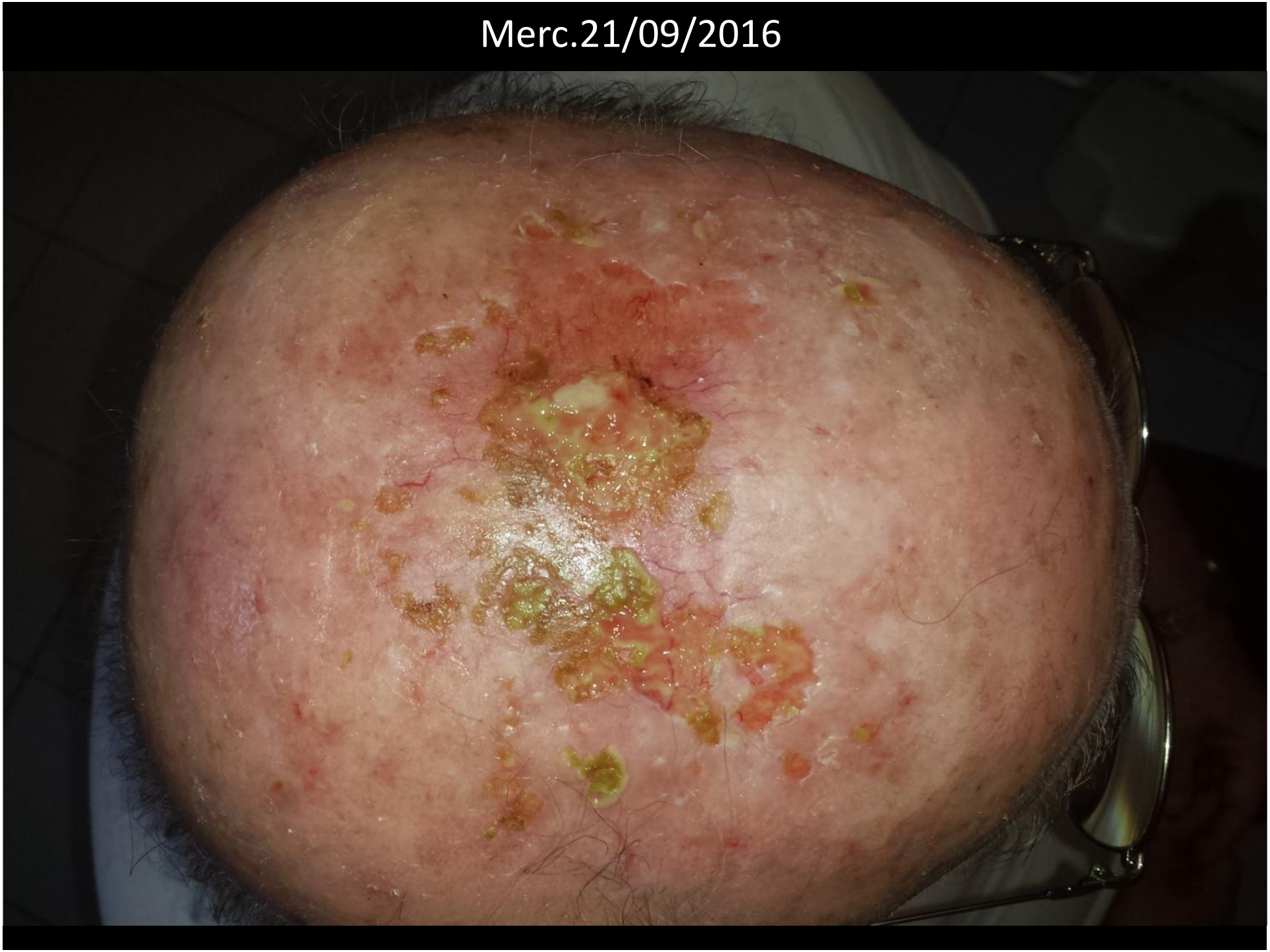
Conclusions: UV-induced *TP53* mutations as well as CCND1/CDK4 changes seem to play essential roles in tumorigenesis of PDS. Furthermore, we found some more interesting mutated genes in other oncogene pathways (activating mutations of *HRAS* and *PIK3CA*). All AFX and PDS investigated immunohistochemically presented with similar oncogene expression profiles (TP53, CCND1, CDK4 overexpression) and the single case with an AFX and PDS showed complete identical *TP53 and PIK3CA* mutation profiles in both tumors. This reinforces our hypothesis that AFX is the non-infiltrating precursor lesion of PDS.

PDS

- Tardío JC, Pinedo F, Aramburu JA, Suárez-Massa D, Pampín A, Requena L,Santonja C. Pleomorphic dermal sarcoma: a more aggressive neoplasm than previously estimated. J Cutan Pathol. 2016 Feb;43(2):101-12.

- BACKGROUND: Pleomorphic dermal sarcoma (PDS) is a rare neoplasm sharingpathological features with atypical fibroxanthoma, but adding tumor necrosis,invasion beyond superficial subcutis or vascular or perineural infiltration.Although its metastatic risk has been estimated to be less than 5%, its realoutcome is presently uncertain because of its rarity and to the lack ofhomogeneous criteria used in reported cases.
- METHODS: Retrospective clinicopathological study of 18 cases of PDS.
- RESULTS: … Three patients (20%) had local recurrences, all with incomplete primary surgicalresections. Three patients (20%) developed distant metastases …
- CONCLUSION: Our data suggest that PDS may be a more aggressive neoplasm thanpreviously estimated.

Merc.21/09/2016



EPDS

- Burton JL. Case for diagnosis: pustular dermatosis ofscalp. Br J Dermatol 1977;97(suppI15):67

- Pye RJ, Peachey RD, Burton JL. **Erosive Pustular Dermatosis of the Scalp**. Br J Dermatol. 1979 May;100(5):559-66

British Journal of Dermatology (1979) **100,** 559.

Erosive pustular dermatosis of the scalp

R.J.PYE,* R.D.G.PEACHEY AND J.L.BURTON

* St John's Hospital for Diseases of the Skin, London and Dermatology Department, Bristol Royal Infirmary, Bristol

Accepted for publication 9 September 1978

SUMMARY

We report six patients with a previously undescribed but characteristic pustular dermatosis confined to the scalp. All the patients were elderly women who developed chronic, extensive, pustular, crusted and occasionally eroded lesions of the scalp which produced scarring alopecia. Investigations were essentially negative and skin biopsies showed only non-specific changes of atrophy and chronic inflammation, sometimes with increased plasma cells in the infiltrate. The condition did not respond to antibiotics, but was suppressed by potent topical steroids.

Update on primary cicatricial alopecias

Elizabeth K. Ross, MD,[a] Eileen Tan, MD,[b] and Jerry Shapiro, MD, FRCPC[a]
Vancouver, British Columbia, and Singapore

The cicatricial alopecias encompass a diverse group of disorders characterized by permanent destruction of the hair follicle and irreversible hair loss. Destruction of the hair follicle can result from primary, folliculocentric disease or as a secondary result. This article focuses on the former, or primary cicatricial alopecias. The cause and pathogenesis of many of these disorders are largely unknown. Although unique clinicopathologic features allow for accurate diagnosis in some cases, diagnostic certainty is often elusive and reflects the limits of present understanding. Classification of the primary cicatricial alopecias on the basis of pathology provides a diagnostic and investigational framework and, it is hoped, will facilitate future enlightenment. Details of classification, etiopathogenesis, clinicopathologic features, differential diagnosis, and practical management of the primary cicatricial alopecias will be discussed. (J Am Acad Dermatol 2005;53:1-37.)

- North American Hair Research Society (NAHRS)

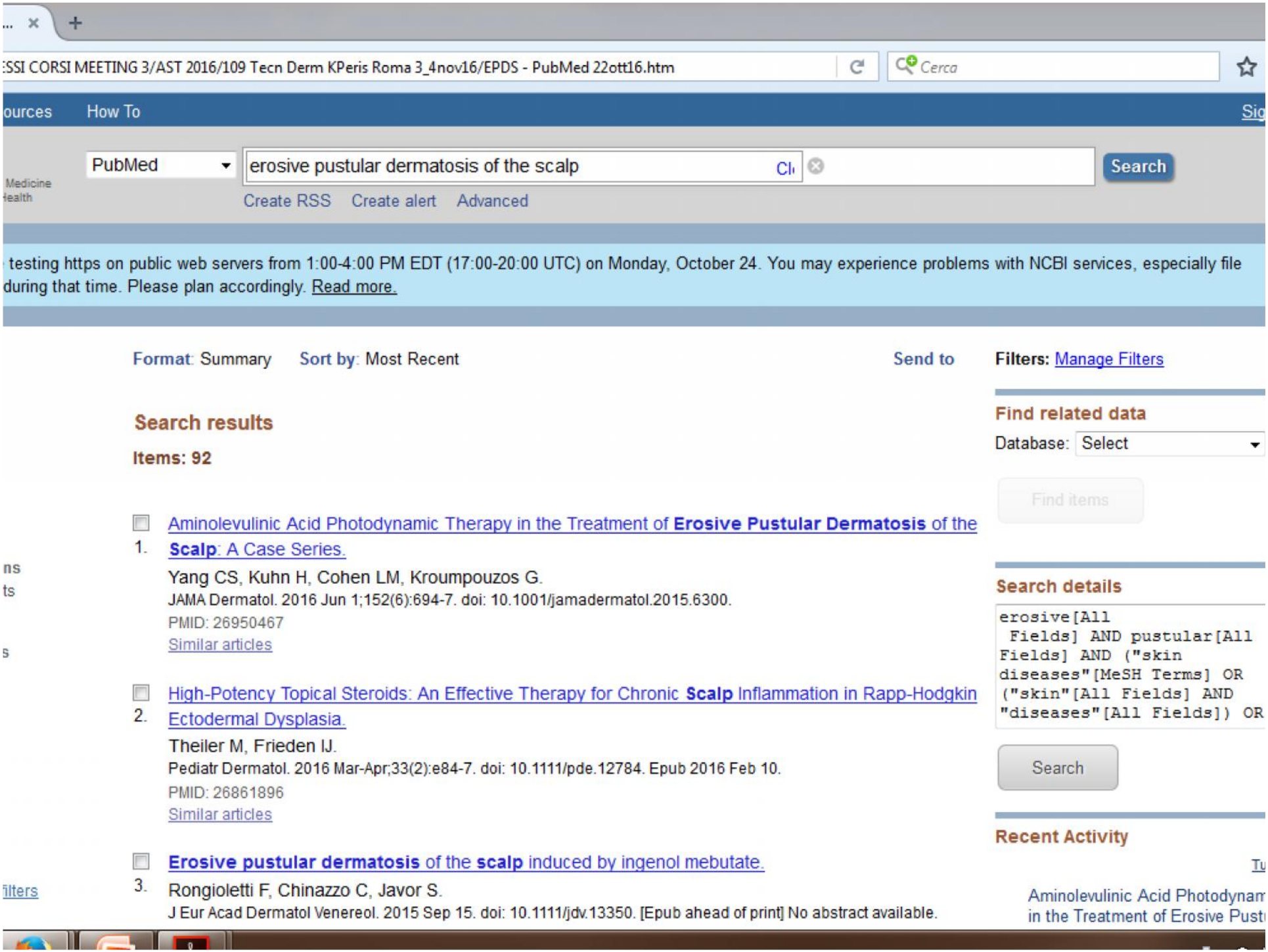
Table I. Proposed NAHRS working classification of primary cicatricial alopecia*

Lymphocytic
Chronic cutaneous lupus erythematosus
Lichen planopilaris
Classic lichen planopilaris
Frontal fibrosing alopecia
Graham-Little syndrome
Classic pseudopelade (Brocq)
Central centrifugal cicatricial alopecia
Alopecia mucinosa
Keratosis follicularis spinulosa decalvans
Neutrophilic
Folliculitis decalvans
Dissecting cellulitis/folliculitis (*perifolliculitis capitis abscedens et suffodiens*)
Mixed
Folliculitis (acne) keloidalis
Folliculitis (acne) necrotica
Erosive pustular dermatosis
Nonspecific

Table I. Risk factors/triggers of erosive pustular dermatosis of scalp

Trauma	Age, y	Sex	Time between trigger and EPDS presentation, wk	Reference
Photodynamic therapy	93	F	4	Guarneri et al[2]
CO_2 laser	63	M	4	Tavares-Bello[3]
	79	F	NR	Patton et al[4]
5% Fluorouracil cream	84	M	8	Vaccaro et al[5]
	65, 55	2 M	NR	Laffitte et al[6]
	79, 80, 85	2 F, M	NR	Patton et al[4]
Herpes zoster	71	M	7	Kim et al[7]
Radiation therapy	69	F	4	Wu et al[8]
	74	M	8	Seez et al[9]
Birth trauma	0, 0, 0, 0	3 F, M	24, 8, 0, 16	Siegel et al[1]
Liquid nitrogen	79, 79, 90, 90	4 F	NR	Patton et al[4]
Skin grafting	92	M	NR	Mehmi and Abdullah[10]
	50	F	52	Martin et al[11]
	NR	2 M, F	NR	Ena et al[12]
Surgery	53	F	6	Layton and Cunliffe[13]

CO_2, Carbon-dioxide; *EPDS*, erosive pustular dermatosis of scalp; *F*, female; *M*, male; *NR*, not reported.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testing https on public web servers from 1:00-4:00 PM EDT (17:00-20:00 UTC) on Monday, October 24. You may experience problems with NCBI services, especially file
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Items: 92
1. Aminolevulinic Acid Photodynamic Therapy in the Treatment of Erosive Pustular Dermatosis of the Scalp: A Case Series.
Yang CS, Kuhn H, Cohen LM, Kroumpouzos G.
JAMA Dermatol. 2016 Jun 1;152(6):694-7. doi: 10.1001/jamadermatol.2015.6300.
PMID: 26950467
Similar articles
2. High-Potency Topical Steroids: An Effective Therapy for Chronic Scalp Inflammation in Rapp-Hodgkin Ectodermal Dysplasia.
Theiler M, Frieden IJ.
Pediatr Dermatol. 2016 Mar-Apr;33(2):e84-7. doi: 10.1111/pde.12784. Epub 2016 Feb 10.
PMID: 26861896
Similar articles
3. Erosive pustular dermatosis of the scalp induced by ingenol mebutate.
Rongioletti F, Chinazzo C, Javor S.
J Eur Acad Dermatol Venereol. 2015 Sep 15. doi: 10.1111/jdv.13350. [Epub ahead of print] No abstract available.
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erosive[All Fields] AND pustular[All Fields] AND ("skin diseases"[MeSH Terms] OR ("skin"[All Fields] AND "diseases"[All Fields]) OR
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in the Treatment of Erosive Pust

PMID: 26373693
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4. Familial **erosive pustular dermatosis** of the **scalp** and legs successfully treated with ciclosporin.
Di Lernia V, Ricci C.
Clin Exp Dermatol. 2016 Apr;41(3):334-5. doi: 10.1111/ced.12744. Epub 2015 Aug 4. No abstract available.
PMID: 26243684
Similar articles

5. **Erosive pustular dermatosis** of the **scalp** and thigh associated with **skin** graft recipient and donor sites.
Roche-Kubler B, Monnin C, Aubin F, Dupond AS.
Eur J Dermatol. 2015 May-Jun;25(3):269-71. doi: 10.1684/ejd.2015.2528. No abstract available.
PMID: 25786713
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6. Calcipotriol/betamethasone dipropionate ointment compared with tacrolimus ointment for the treatment of **erosive pustular dermatosis** of the **scalp**: a split-lesion comparison.
Pagliarello C, Fabrizi G, Fantini C, Cortelazzi C, Boccaletti V, Annessi G, Zampetti A, Feliciani C, Di Nuzzo S.
Eur J Dermatol. 2015 Apr;25(2):206-8. doi: 10.1684/ejd.2015.2519. No abstract available.
PMID: 25786341
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7. **Erosive pustular dermatosis** of the **scalp** arising concomitantly with elevation of serum matrix metalloproteinase-3 in a patient with rheumatoid arthritis.
Aoshima M, Ito T, Tokura Y.
J Dermatol. 2015 May;42(5):540-1. doi: 10.1111/1346-8138.12815. Epub 2015 Feb 27. No abstract available.
PMID: 25728745
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8. **Erosive pustular dermatosis** of the **scalp** following topical latanoprost for androgenetic alopecia.
Vaccaro M, Barbuzza O, Borgia F, Cannavò SP.
Dermatol Ther. 2015 Mar-Apr;28(2):65-7. doi: 10.1111/dth.12197. Epub 2015 Jan 28. No abstract available.
PMID: 25627907

9. **Erosive pustular dermatosis** of the **scalp** associated with myelodysplastic syndrome.
Yu X, Liu Y, Ozukum M, Song Z.
J Eur Acad Dermatol Venereol. 2016 Feb;30(2):380-1. doi: 10.1111/jdv.12812. Epub 2014 Nov 11. No abstract available.
PMID: 25388746
Similar articles

10. **Erosive pustular dermatosis** of the **scalp** following hair transplantation.
Shahmoradi Z, Abtahi-Naeini B, Pourazizi M.
Adv Biomed Res. 2014 Aug 22;3:176. doi: 10.4103/2277-9175.139412. eCollection 2014.
PMID: 25250290 Free PMC Article
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11. **Erosive pustular dermatosis** of the **scalp** treated with 0.1% mometasone furoate cream.
Jankowski M, Skrzeczko-Kwela E, Czajkowski R.
Acta Dermatovenerol Croat. 2014;22(1):67-9. No abstract available.
PMID: 24813848
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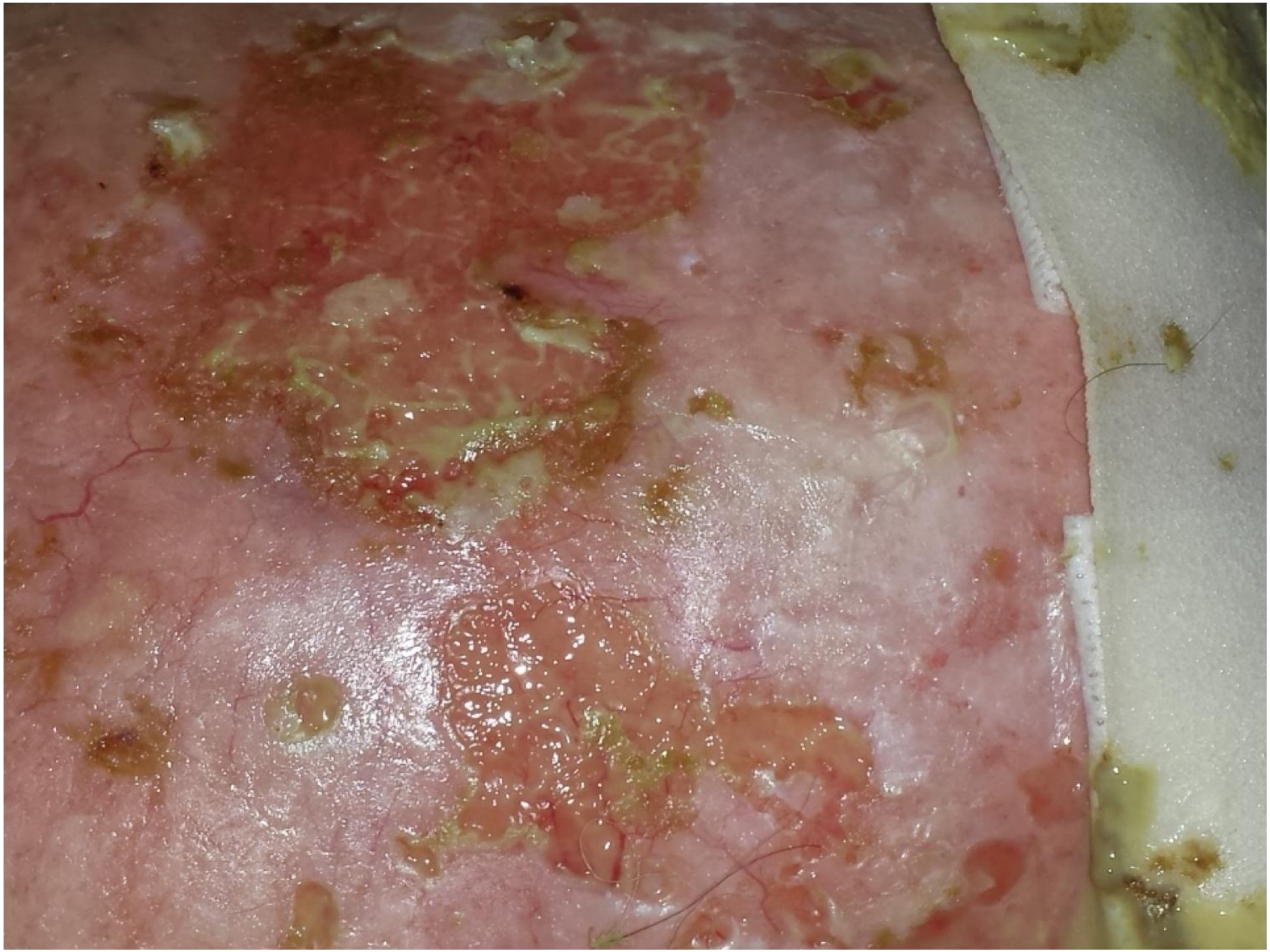

Erosive pustular dermatosis

- Erosive pustular dermatosis (also known as erosive pustular dermatosis of the scalp) is an idiopathic chronic, relapsing amicrobial pustular dermatosis of that scalp that results in cicatricial alopecia. A history of antecedent accidental or iatrogenic trauma to the affected scalp site is often elicited. Specific precipitants include minor lacerations, contusions, accidental scalping, sunburn, varicella zoster, skin grafting,radiation, synthetic hair fiber implantation, cryotherapy, topical 5% fluorouracil, topical tretinoin, topical imiquimod, topical ingenol mebuthate. The predominance of disease in the elderly has led some to postulate that chronic actinic damage to the scalp may be a predisposing factor for disease.

Erosive pustular dermatosis: clinical features.

- Erosive pustular dermatosis is an uncommon disorder that largely affects the elderly, with an apparent female predominance. In cases with known preceding trauma, onset of disease can occur contemporaneously or months to years thereafter. The characteristic lesion is a large asymptomatic, well-demarcated, boggy, superficially crusted plaque that is easily unroofed to reveal a beefy red, exudative erosion with discrete or coalescent flaccid pustules beneath. Moist erosions or crusts in the absence of pustules have also been described. Untreated lesions undergo episodic pustular flares, with slow enlargement over years.
- Cicatricial alopecia is a cardinal feature of advanced disease, the extent of which may not be fully appreciated until the lesion is healed with treatment.
- Wounds may be colonized by staphylococcal species and, less frequently, Candida.
- Aggravation of disease has been reported with attempts at reparative skin grafting and treatment of surrounding actinic keratoses.
- Development of secondary carcinoma with squamous and basal cell features has been reported in a long-standing case. Reports of a variant of erosive pustular dermatosis affecting the leg have been attributed to other diseases by some.

Erosive pustular dermatosis: Differential diagnosis

- The differential diagnosis is extensive and includes amicrobial pustulosis associated with autoimmune disease, pustular ulcerative dermatosis of the scalp (Jacyk WK. Pustular ulcerative dermatosis of the scalp. Br J Dermatol 1988;118:441-4.), pyoderma gangrenosum, pustular psoriasis, kerion, bacterial folliculitis, cicatricial pemphigoid, pemphigus vulgaris, blastomycosis-like pyoderma, erosive candidiasis of the scalp, and temporal arteritis, among other possibilities.
- Scalp involvement in amicrobial pustulosis associated with autoimmune disease, a newly recognized amicrobial chronic, relapsing intertriginous follicular and nonfollicular pustular eruption that affects young women with autoimmune disorders, is poorly characterized. However, scant reports suggest that there is some clinical, histopathologic, and treatment-response overlap with erosive pustular dermatosis that, given the added observation of erosive pustular dermatosis of the scalp in 2 patients with autoimmune disease, arguably could reflect a common disease process. Pustular ulcerative dermatosis of the scalp is a rare, noncrusted, ulcerative rather than erosive dermatosis of the vertex scalp that occurs in malnourished young male Africans, a cohort distinctly different from those with erosive pustular dermatosis.

Erosive pustular dermatosis: Pathology

- Histopathologic features of erosive pustular dermatosis are nonspecific.
- Characterization of early disease is lacking.
- Observed epidermal changes include erosion, atrophy, acanthosis, parakeratosis, and subcorneal pustules.
- A dense, chronic mixed inflammatory cell infiltrate and occasional foreign-body giant cells occupy the dermis and do not appear folliculocentric. Pilosebaceous units are variably diminished in number or are absent. Remnants of arrector pili may be seen. Findings on DIF are routinely negative.

Erosive pustular dermatosis of the scalp successfully treated with calcipotriol cream

SIR, A 95-year-old woman, resident in a home for the elderly, presented with a thickly crusted area on her frontal scalp. This area had been injured 2 years previously when she had fallen out of bed and hit her head. According to her clinical notes, following the injury she had a superficial abrasion that did not require suturing, and a haematoma. The skin initially healed but subsequently became red and irritable and broke down repeatedly. Treatment with topical antibiotics (fusidic acid cream and mupirocin ointment) and systemic antibiotics (flucloxacillin and ciprofloxacillin) was unsuccessful and the area involved extended gradually. Eventually a thick crust formed and the patient was referred for dermatological review with a suspected diagnosis of skin malignancy.

Examination showed an approximately 10 × 10 cm thickly crusted area of alopecia over the frontal scalp. Gentle removal of the crust revealed a moist, atrophic surface with prominent telangiectases, areas of erosion and superficial pustulation extending to the forehead (Fig. 1a). The rest of the skin was essentially normal, with no features of blistering disorders or psoriasis.

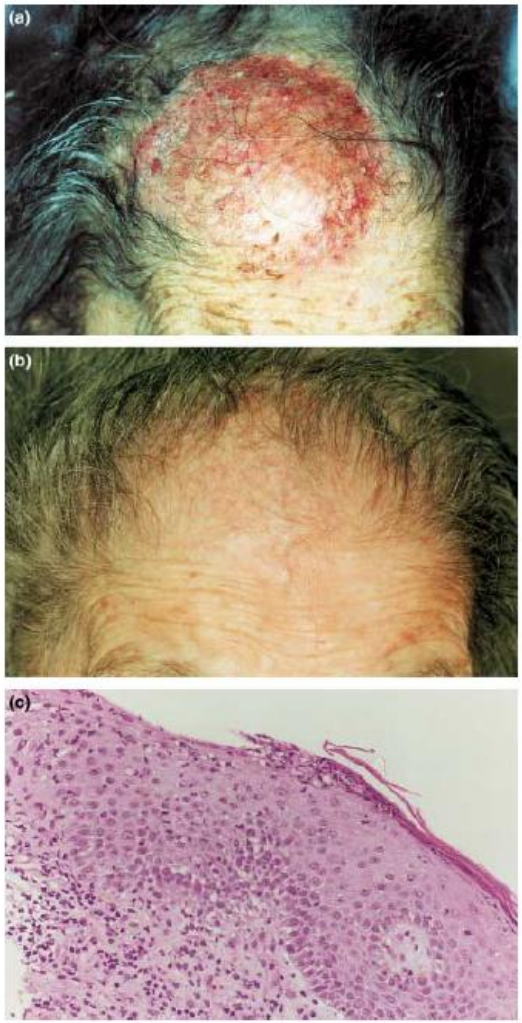

Figure 1. (a) Moist, atrophic surface with telangiectases, areas of erosion and superficial pustulation at presentation. (b) After treatment with calcipotriol cream. (c) Photomicrograph showing epidermal spongiosis, parakeratosis, ulceration, subcorneal pustulation and neutrophils in the epidermis and a mixed dermal infiltrate of lymphocytes, neutrophils and plasma cells (haematoxylin and eosin; original magnification × 80).

Skin swabs and scrapings were negative for bacteria and fungi. Histological examination of a punch biopsy specimen showed alopecia, epidermal atrophy and ulceration, spongiosis, parakeratosis, subcorneal pustulation and neutrophils in the epidermis, and a mixed dense dermal infiltrate of lymphocytes, neutrophils, plasma cells and occasional foreign body giant cells (Fig. 1c). There was no evidence of malignancy or vasculitis, and a periodic acid–Schiff stain for fungi was negative.

The clinical and histological features in this case were characteristic of erosive pustular dermatosis of the scalp. This entity was first described in 1979[1] and has been reported mainly in elderly women. In many of the published cases there was a history of antecedent local trauma and photodamage. Although the pathogenesis is not well understood, it has been proposed that the condition may represent a nonspecific inflammatory response to injury of ageing and sun-damaged scalps.[2] Cases of erosive pustular dermatosis of the scalp following surgery,[3] radiotherapy[4] and cryotherapy[5] have also been reported. The clear history of local injury in this case supports the hypothesis of trauma being important in this condition.

Potent topical corticosteroids have been reported to be helpful in the management of erosive pustular dermatosis of the scalp.[1–7] However, in view of the marked skin atrophy already evident in this patient, it was decided to avoid topical corticosteroids and instead try treating with calcipotriol cream (Daivonex® cream; 50 μg g^{-1}, 0·005% w/w; Leo Pharmaceutical Products, Copenhagen, Denmark). This was initially applied once daily and was well tolerated. The frequency of application was subsequently increased to twice daily. At review 2 months later there was remarkable improvement. The areas of erosion and pustulation had healed completely and application of calcipotriol cream was discontinued. The skin remained healed and some hair regrowth was evident when the patient was seen again after a further 3 months (Fig. 1b) and once more 9 months later.

Erosive pustular dermatosis of the scalp is uncommon (although possibly less uncommon than suggested by the relatively small number of published cases). Although the natural history of this condition has not been well documented, there is said to be little tendency for spontaneous resolution,[7] and in several of the reported cases it had been present for a number of years. In this patient the condition had been present for 2 years prior to presentation and had shown no signs of resolution before treatment with calcipotriol cream was started, suggesting that this had had a real therapeutic effect. The response to calcipotriol cream was remarkable and, to the best of my knowledge, has not been reported previously in this condition. The result appears to have been permanent and the erosive pustular dermatosis showed no signs of recurrence up to 12 months after stopping treatment.

Calcipotriol is a vitamin D_3 analogue with an excellent safety profile that has been widely used to treat psoriasis. Activity has been demonstrated in other dermatoses

Update on primary cicatricial alopecias

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The cicatricial alopecias encompass a diverse group of disorders characterized by permanent destruction of the hair follicle and irreversible hair loss. Destruction of the hair follicle can result from primary, folliculocentric disease or as a secondary result. This article focuses on the former, or primary cicatricial alopecias. The cause and pathogenesis of many of these disorders are largely unknown. Although unique clinicopathologic features allow for accurate diagnosis in some cases, diagnostic certainty is often elusive and reflects the limits of present understanding. Classification of the primary cicatricial alopecias on the basis of pathology provides a diagnostic and investigational framework and, it is hoped, will facilitate future enlightenment. Details of classification, etiopathogenesis, clinicopathologic features, differential diagnosis, and practical management of the primary cicatricial alopecias will be discussed. (J Am Acad Dermatol 2005;53:1-37.)

- North American Hair Research Society (NAHRS)

Table I. Proposed NAHRS working classification of primary cicatricial alopecia*

Lymphocytic
Chronic cutaneous lupus erythematosus
Lichen planopilaris
Classic lichen planopilaris
Frontal fibrosing alopecia
Graham-Little syndrome
Classic pseudopelade (Brocq)
Central centrifugal cicatricial alopecia
Alopecia mucinosa
Keratosis follicularis spinulosa decalvans
Neutrophilic
Folliculitis decalvans
Dissecting cellulitis/folliculitis (*perifolliculitis capitis abscedens et suffodiens*)
Mixed
Folliculitis (acne) keloidalis
Folliculitis (acne) necrotica
Erosive pustular dermatosis
Nonspecific

Table III. Documented treatments in case reports for erosive pustular dermatosis of scalp

Treatment	Strength/preparation	No. treated	Outcome	Source
Antibiotics	Gentamicin cream, amphotericin cream, bacitracin ointment	6	Little response	Pye et al[18]
	Fusidic acid cream and mupirocin ointment; amoxicillin/clavulanate	1	No response	Guarneri et al[2]
Topical steroids	Clobetasol cream, betamethasone cream, triamcinolone cream	6	Initial improvement but relapse when steroids withdrawn	Pye et al[18]
Oral steroids	Methylprednisolone 16 mg/d with taper	1	Improvement at 12 wk with residual scarring alopecia	Guarneri et al[2]
Intralesional steroids	Intralesional triamcinolone 10 mg/cc, used in conjunction with halobetasol propionate	1	Improvement when treated along with oral steroids but steroid-induced atrophy in 5 mo	Cenkowski and Silver[21]
Oral zinc sulfate	90 mg/d	1	Significant improvement after 1 wk	Ikeda et al[22]
Isotretinoin	0.3 mg/kg/d increased to 0.6 mg/kg/d, in combination with zinc sulfate	1	Significant improvement after 4 wk	Petersen and Bygum[23]
	0.75 mg/kg/d with antiseptic soaks and topical antibiotics	1	Significant improvement after 16 wk	Mastroianni et al[24]
	30 mg/d for 5 mo	1	Worsening of erosions	Laffitte et al[6]
Photodynamic therapy	Methyl 5-aminolevulinic acid cream applied to affected area and irradiated at 630 nm	1	Total of 2 treatments, 1 wk apart; marked improvement after 12 wk	Meyer et al[25]
Calcipotriol cream	50 μg/g; 0.005% daily	1	Marked improvement at 8 wk; partial hair regrowth at 12 wk	Boffa[26]
Topical tacrolimus	0.1% daily	2	Significant improvement in 2-4 wk	Laffitte et al[27]
	0.1% bid	1	Significant improvement in 1 wk; almost resolved in 8 wk	Kim et al[7]
	0.1% daily	1	Significant improvement in 16 wk; skin atrophy posttreatment	Marzano et al[28]
	0.1% bid	1	Improved noted at 4 wk; significant improvement after 16 wk	Seez et al[9]
	0.1%	1	Reversal of skin atrophy from steroids, clinical evidence of hair regrowth	Cenkowski and Silver[21]
Oral dapsone	100 mg/d	1	Little clinical improvement	Pye et al[18]

bid, Twice a day.

Erosive pustular dermatosis of the scalp: A review with a focus on dapsone therapy

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Nashville, Tennessee; and San Francisco and Mountain View, California

Background: Erosive pustular dermatosis of the scalp (EPDS) is an inflammatory disorder of unknown origin characterized by pustules, erosions, and crusting in areas of alopecia that tend to be atrophic, actinically damaged, or both. The most common treatments reported include antibiotics and topical anti-inflammatories, which can be ineffective. In the search for effective treatment for EPDS, we share our experience with topical dapsone 5% gel.

Observations: We present 4 patients with EPDS, all with classic clinical presentations and histologic findings of EPDS, who had failed a variety of treatments including oral, intralesional, or topical steroids, tacrolimus, and antibiotics. All patients demonstrated rapid improvement or resolution with topical dapsone 5% gel.

Limitations: Our experience and success with topical dapsone for EPDS is observational and not the result of a randomized controlled trial.

Conclusion: Our observations demonstrate topical dapsone 5% gel to be a novel, safe, and efficacious therapeutic alternative for mild to moderate EPDS. (J Am Acad Dermatol 2012;66:680-6.)

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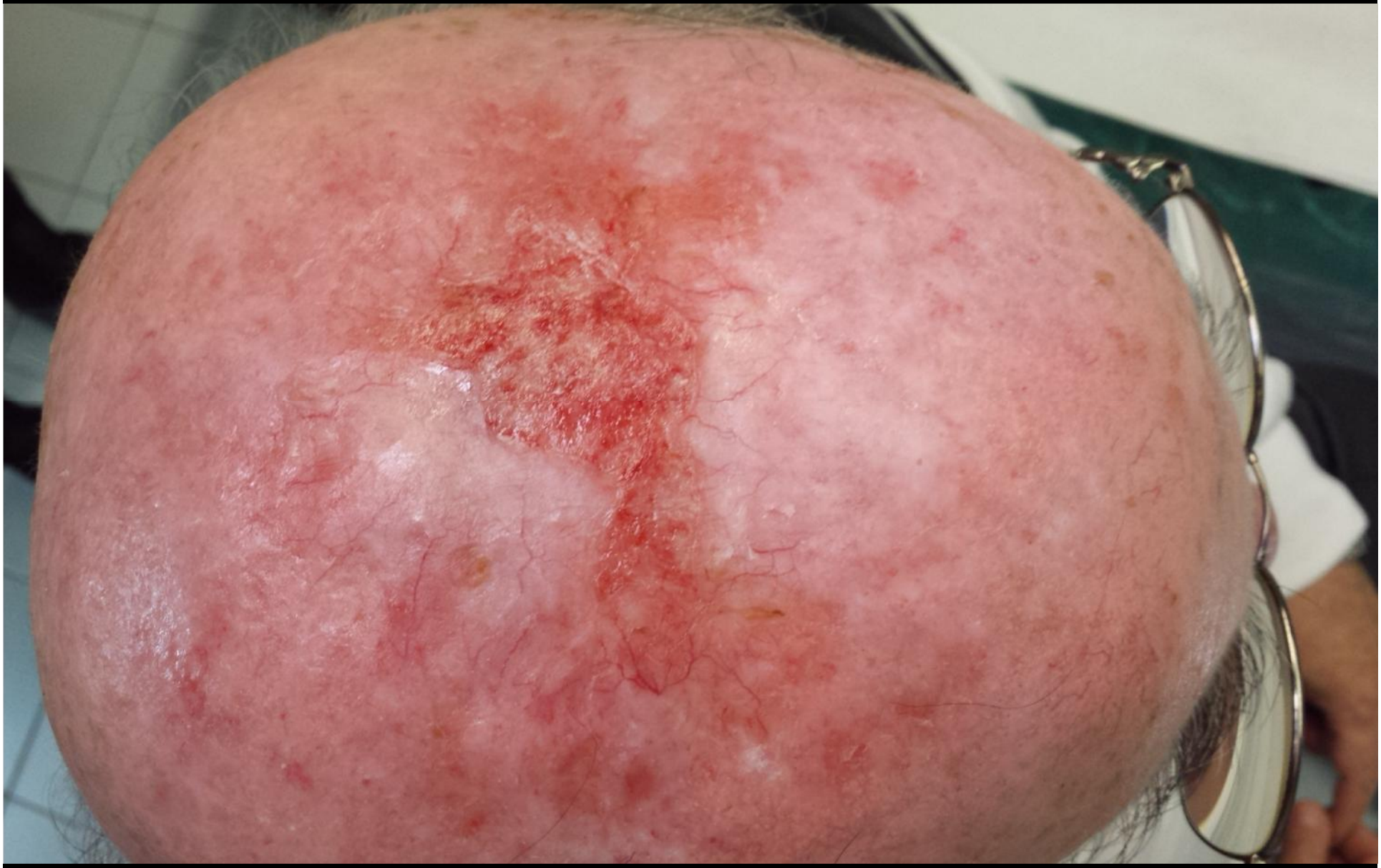


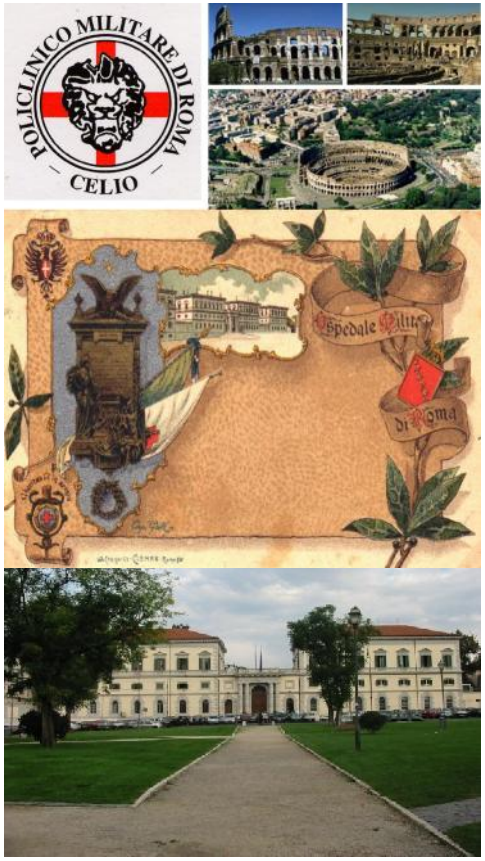
conclusioni

- Con il passare del tempo in questo paziente trattato per cheratosi attiniche e operato per AFX/PDS si accentua una dermatosi pustolosa erosiva dello scalpo , una forma particolare di alopecia cicatriziale a neutrofili (EPDS, malattia non frequente, descritta per la prima volta nel 1979 da Burton JL), forse già presente prima dell'intervento nell'area del vertice calvo, caratterizzata da pustole, erosioni e croste su cute atrofica con danno attinico e cheratosi attiniche, ad eziologia sconosciuta

- L'esame istologico dimostra una flogosi neutrofilica aspecifica
- L'esame batterioscopico e colturale dimostra la presenza di stafilococco aureo, sensibile a vari antibiotici
- Il quadro clinico è caratteristico di una dermatosi pustolosa erosiva del capillizio, (EPDS: "Erosiv pustular dermatosis of the scalp")

- La terapia cortisonica e antibiotica locale porta a un miglioramento ma non a risoluzione completa a tutt'oggi

- è discusso se le terapie per le cheratosi attiniche (in particolare ingenolo mebutato) e l'intervento chirurgico possano avere avuto un ruolo eziopatogenetico



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