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Case Report

Case reports on erosive pustular dermatosis of scalp: a cross sectional study at a tertiary care centre

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ABSTRACT

Erosive pustular dermatosis of the scalp (EPDS) has been reported as a rare chronic and progressive non-infectious inflammatory condition of the actinically damaged scalp characterized by crusts, erosions and pustules. It tends to occur on the scalp of older people who have usually been treated for diffuse actinic damage and actinic keratoses. The list of possible differential diagnosis is long and includes tinea capitis, folliculitis decalvans, neutrophilic dermatosis, autoimmune vesiculobullous disorders, malignancy etc. Notoriously difficult to treat, these cases tend to be chronic and progressive. These patients often present after repeated treatment to various scalp dermatosis that doesn't result in clearing instead persistence and perpetuation of the process. A cross sectional prospective study was conducted in department of dermatology in a tertiary care centre in which six cases of EPDS attending outpatient department from 2017 to 2019 were included. EPDS is more common but under recognized condition, hence less cases are reported till date. Increased awareness of and proper diagnosis of this condition changes the approach towards these patients, protects the scalp from involuntary insults and other precipitating factors that hinders with a better outcome.

Keywords: Erosive pustular dermatosis of scalp, Inflammatory condition, Non-infectious, Chronic, Progressive, Notorious

INTRODUCTION

Erosive pustular dermatosis is a non-infectious inflammatory disorder first described by Burton, Pye and Peachey in 1979 characterised by sterile pustules, erosions and crusted lesions over scalp with progressive scarring alopecia.[1,2] Erosive pustular dermatosis of scalp (EPDS) is a rarely reported entity but the condition might be under recognized. About 150 cases has been reported so far.[3] It is encountered in both the sexes, with a female predominance of approximately 2:1.[4] Most common predisposing factors being trauma, surgery, skin grafting, thermal burn, cryotherapy and sunburn.[5]

We analyzed our data of dermatology of patients attending outdoor department in a tertiary care centre from 2017 to 2019 in which six cases of EPDS were included.

CASE REPORT

Case 1

A 35 year old female presented with pustules and erosions covered with crusts over the scalp associated with burning pain and mild itching since last 2 months (Figure 1 A and B). She consulted various private practitioners and every time broad spectrum antibiotics

were prescribed. Patient didn't notice any improvement and further worsening of the condition continued. Haematological investigations showed raised total leucocyte count with other parameters within normal limits. Pus and blood culture showed no growth. Histopathological examination revealed an ulcerated epidermis with parakeratosis and serum - crust and a mixed dense dermal infiltrate consisting of lymphocytes, neutrophils and plasma cells (Figure 2 A and B). Special staining and cultures for bacteria and fungi along with direct immunofluorescence study was also negative.

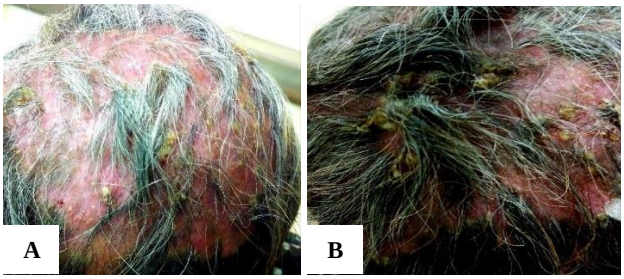

Figure 1 (A and B): Clinical image showing extensive pustules and eroisons covered with crusts along the scalp.

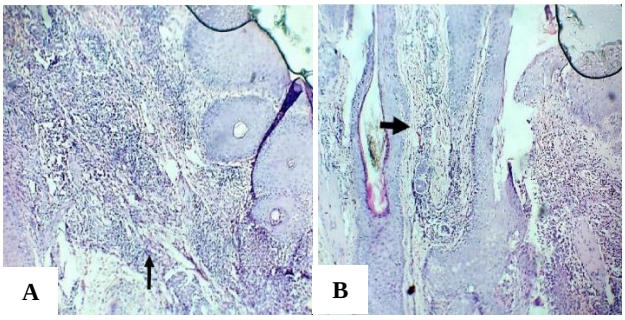

Figure 2 (A and B): Epidermal erosion with mixed dense dermal infiltrate consisting of lymphocytes, neutrophils and plasma cells.

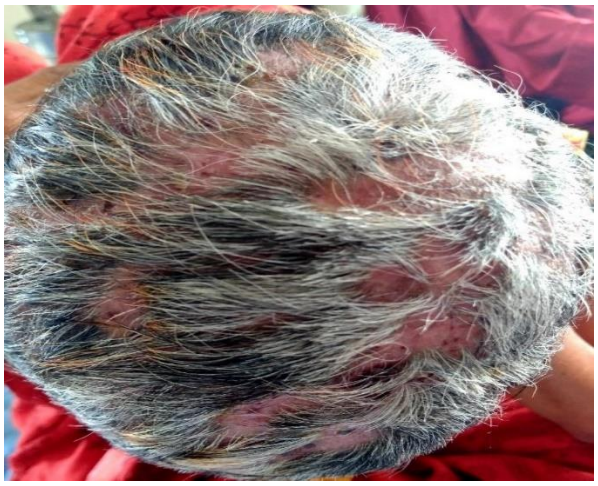

Figure 3: Post treatment with topical steroid showing resolution of initial lesions.

The clinical and histopathological findings were consistent with the diagnosis of erosive pustular dermatosis of scalp. Treatment with clobetasol propionate 0.05% twice daily application was started with marked improvement seen after 10 days, was continued for 2 weeks followed by night time application for next 2 weeks. A maintenance therapy with topical tacrolimus 0.1% was initiated without clinical relapse after 3 months (Figure 3).

Case 2

A 50 year old female presented with extensive erosions and crusts with pustules over the scalp associated with pain since 4 months with few areas of scalp showing cicatricial alopecia (Figure 4 A and B). Patient gives history of being treated with some oral and topical medications records of which are not available. Scalp dermoscopy showed a very atrophic and thin scalp, with lack of follicular ostia, sparse hair and evident superficial blood vessels. Haematological and histopathological investigations gave no conclusive evidence. Treatment with pasitrex C (clobetasol 0.05% + calcipotriol 0.005%) ointment was initiated and continued for 2 weeks and response was seen in 7 days. Later on, patient was maintained on topical calcipotriol cream that showed remarkable response and partial hair growth was seen after 8 weeks (Figure 5).

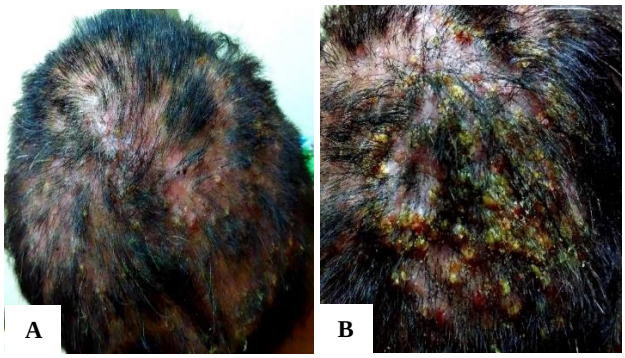

Figure 4 (A and B): Extensive erosions and crusts with pustules over the scalp few areas of scalp showing cicatricial alopecia.

Case 3

A 78 year old female presented with nonhealing ulcer with ill-defined margins and covered with crusts over scalp associated with erosion and crusts. This site had biopsy showing nonspecific findings in the form of mixed inflammatory infiltrate without any evidence of malignancy in November 2018 and symptomatic treatment was prescribed. Patient again presented after 2 months with more extensive involvement and rebiopsy was done that showed mixed dense dermal infiltrate consisting of lymphocytes and neutrophils. All other investigations were inconclusive. This is when EPDS was thought about and topical betamethasone dipropionate 0.025% cream was prescribed for 2 weeks and steps for

proper wound care was followed. Erosions, crusts or granulation tissue was not appreciated after 15 days with marked improvement over the site. At present patient is in remission and maintained on topical tacrolimus 0.1% ointment.

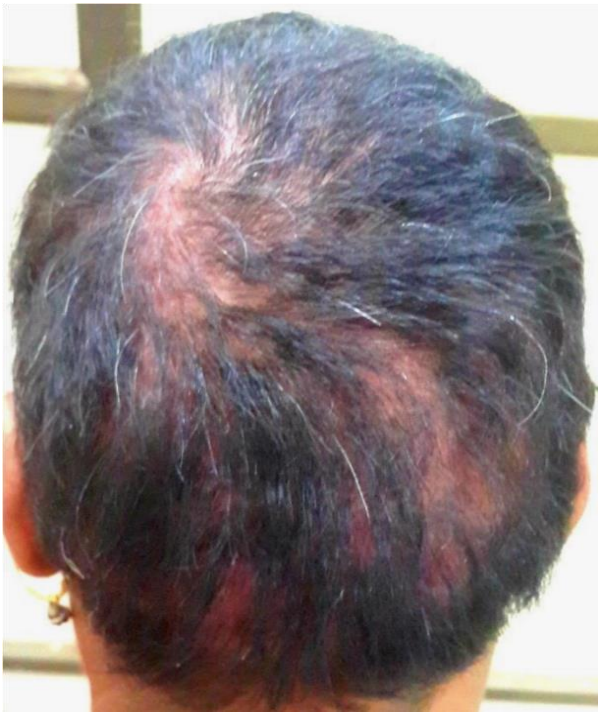

Figure 5: Post treatment with steroid and calcipotriol showing improvement and initiation of hair growth.

Case 4

A 52-year-old male presented in the OPD with coin sized white hyperkeratotic plaque with erythematous base on vertex of scalp in November 2017 and biopsy findings were suggestive of actinic keratosis and was treated repeatedly with cryotherapy. After few months, patient presented with confluent erythema with few areas of hyperkeratotic lesions. These hyperkeratotic lesions were treated with cryotherapy but this time lesions didn't respond and erythema kept on increasing over other areas of scalp. Considering it as an inflammatory process, he was started on clobetasol propionate 0.05% ointment for 2 weeks. On follow up after two weeks, erythema and hyperkeratotic lesions decreased and condition continued improving. After 4 months complete clearance was seen and patient was maintained on topical tacrolimus 0.1% ointment.

Case 5

A 65-year-old male presented in the OPD with pustular lesions all over scalp associated with mild pain and burning sensation since last 20 days. He was prescribed broad spectrum oral and topical antibiotics but lesions continued getting worse with formation of crusts and scarring over few areas. Patient was taking medications for thyroid since 10 years. There was past history of herpes zoster opthalmicus before 1.5 month and oral acyclovir was prescribed for 5 days. Haematological investigations showed mild leucocytosis with raised Erythrocyte sedimentation rate and C-reactive protein. At present biopsy showed subcorneal pustule containing neutrophils. Pus culture showed no growth. Patient was started with topical halobetasol dipropionate 0.025% for 2 weeks and responded within 5 days, hence a diagnosis of erosive pustular dermatosis of scalp was made.

Case 6

A 30-year-old male presented in the OPD with pustules over scalp involving vertex and bilateral parietal areas associated with pain on-off since last 2 months. Initially antibiotics were given and had no satisfactory outcome. Biopsy and dermoscopy findings being nonspecific and haematological investigations were within normal limits. Pus culture of the lesion showed absence of any bacteria or fungi. EPDS was considered and topical steroid trial in the form of clobetasol propionate 0.05% was started and dramatic response was seen in 1 week. Topical steroid was slowly tapered and patient was maintained in remission on topical tacrolimus 0.01% ointment later on.

DISCUSSION

EPDS is a rare inflammatory disease of unknown etiology usually occuring in elderly. Most common site being scalp but lesions can also be presented over legs. A history of trauma to the affected area can usually be established. Cases of erosive pustulosis of scalp have been reported following skin grafting, radiation, cryotherapy or topical chemotherapy in various studies.[6-8] In our study EPDS was found associated with actinic keratosis, autoimmune and idiopathic causes.[9,10]

The findings of case reports in our study synchronizes with various other case reports done on EPDS. Carol drucker conducted similar study and found that EPDS is an under-recognized condition and have been associated with actinic keratosis and various other preceding traumas.[11] Equal number of male and female patients were seen in our study contradictory to the study by Kristina and georgi showing female predominance. Although Elderly patients were being most commonly affected with an exceptions in our study in which two young patients presented with pustules, erosions and crusts over scalp and later on EPDS was diagnosed.

Patient commonly presented with pustules, erosions and crusts over areas of scalp with few cases already showing progression to cicatricial alopecia. The pustule are usually sterile but they can be secondarily colonized by bacteria after the condition has developed. The etiology and exact pathogenesis remains unclear that impedes proper categorization but some authors claim it to be a disease of inflammatory dermatoses while other speculates it as a result of immunological dysfunction.[12]

Histology is not diagnostic and plays supportive role showing varied presentations ranging from hyperkeratosis, parakeratosis, atrophy, edema and subcorneal pustules to mixed inflammatory infiltrate and plasma cells in upper dermis. Phagocytosis and giant cells can be seen around hair shafts in deep dermis with follicle destruction. Granulation tissue changes can also be seen.

Various treatment modalities have been tried ranging from topical steroids to topical tacrolimus, topical calcipotriol, acitretin etc showing varied results.[13,14,15,16] Many clinical trials on topical dapsone and oral cyclosporine are going on and surgical procedures have been tried in recalcitrant cases in which removal of chronic granulation tissue is done and biofilms are helpful for final wound closure that can have promising results in the future.[17,18] Irrespective of the newer drugs, topical steroids remain mainstay first line of treatment of choice and other drugs could be added as adjunctives.

CONCLUSION

EPDS is more common but under recognized condition and following conditions like malignancy, infection, neutrophilic dermatosis and autoimmune blistering disorder must be excluded before considering the diagnosis of EPDS. It has no pathognomonic laboratory or histologic findings and thus dermatologists plays crucial role to clinch the diagnosis and to initiate timely treatment to prevent further progression.

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