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A STUDY OF PARAPSORIASIS

Dermatology

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ABSTRACT

Parapsoriasis is a papulosquamous disorder of unknown etiology with remissions and exacerbations.

Aim: We studied clinical and histopathological features of parapsoriasis.

Methods: This is a prospective, study of patients clinically diagnosed as parapsoriasis and confirmed by histopathology.

Results: There were 28 (15 males and 13 females) cases of parapsoriasis in the study period. The maximum number of cases, 10 (35.70%) were in their third decade of life. Pityriasis lichenoides chronica was diagnosed in 24 cases (85.71%), pityriasis lichenoides et varioliformis acuta (PLEVA) in 2 cases (7.14 %) and plaque parapsoriasis in 2 cases (7.14%). Histopathologically, parakeratosis, basal cell vacuolation, and perivascular infiltrate was seen. One case of plaque type parapsoriasis showed epidermoptropism with atypical cells suggestive of mycosis fungoides.

Conclusion: Pityriasis lichenoides chronica is a relatively common disease amongst parapsoriasis. Histopathology is a must to rule out early mycosis fungoides in plaque type of parapsoriasis

KEYWORDS

PLEVA, PLC, parapsoriasis

INTRODUCTION

Parapsoriasis describes a group of cutaneous diseases that can be characterized by scaly papules, patches that has resemblance to psoriasis. The currently accepted classification includes three entities as large plaque parapsoriasis, small plaque parapsoriasis and pityriasis lichenoides.

There is unresolved controversy as to whether or not parapsoriasis variant are either precursor to mycosis fungoides or established but early mycosis fungoides from outset.

MATERIAL AND METHOD

This study was carried out on 28 patients of out patients department of skin and venereology of tertiary care hospital Mumbai in three years.

Data of each patient was recorded on separate proforma, which includes preliminary data regarding the name, age, sex and address.

A careful history of disease was noted with special reference to duration of lesions. Sites of involvement and any exacerbating factor like throat infection, stress etc. Inquires were made about recurrences and different treatment undertaking with duration.

A thorough examination of the lesion was carried out paying special attention to the distribution of lesion and different morphology like erythema, papules, plaque, hypopigmented macules, ulceronecortic, poikilodermatic lesions were noted. Routine laboratory test were performed in all the patients. A punch biopsy was taken from early erythematous papule or plaque and was stained with hematoxylin and eosin. Different histological findings were seen carefully suspecting mycosis fungoides. The various modality of treatment were tried.

RESULT

Out of 28 patients maximum number of patients of parapsoriasis were in the age group of 21 -30 years. Youngest patient was 4 year old and oldest patient was 57 year old.

In case of pityriasis lichenoides chronica (Figure1) maximum number of patients were in the age group of 21 -30 year. Two cases of pityriasis lichenoides et varioliformis acuta were a 29 year old male and a 45 year old female.

Parapsoriasis was more common in male in second and third decade while in females is commonly seen in the third and fourth decades.

Male to female ration was nearly equal 1.15:1. Similar variations were seen in patients of pityriasis lichenoides chronica.

Most common symptom was pruritus in about 85% of patients. Both cases of pityriasis lichenoid et varioliformis acuta (Figure 2) had associated constitutional symptoms like fever and arthritis.

According to pattern of distribution, patients were classified in to 3 major groups. Largest group of 17 patients had typical lesions that involved the whole body surface more or less uniformly called diffuse distribution. A second group of 8 patients had lesions confined to the trunk, neck and only proximal part of extremities called central distribution and third group of 3 patients had lesion in an acral distribution called as peripheral distribution.

Both parapsoriasis and pityriasis lichenoides chronica 22 patients presented with erythematous papules centrally adherent scales with negative auspitz sign. All early lesions were having deep dermal tenderness.

These lesion healed as hypopigmented macules.

In two cases of pityriasis lichenoid et varioliformis acuta one patient had multiple erythematous papules, and plaques covered with central adherent scales and one had along with the above lesions, necrotic papules, which healed with scarring.

All routine laboratory investigations were normal in all patients except four patients showed high ASLO titers.

Common histopathology finding were : the epidermis- focal parakeratosis, vacuolization of the basal layer, spongiosis, extravasation of erythrocytes. Dermoepidermal junction showed mononuclear cell infiltrate. In the dermis, perivascular mononuclear cell infiltrate, edema of papillary dermis and presence of atypical cells. Histological examination revealed that findings described for acute and chronic form were seen in majority of patients but in 6 patients changes of both type co- existed.

Histological findings did not show any relevance to clinical course.

In case of plaque type parapsoriasis (Figure 3) it is very difficult to distinguish from early stage of mycosis fungoides. Hence careful histological examination was carried out with suspicion of mycosis fungoides.

We had given systemic PUVA in 8 patients and topical PUVA in three

patients. Five patients were treated with systemic antibiotics like erythromycin and tetracycline along with topical steroids and emollients. Treatment was given for 2-3 week. Two patient on antibiotics showed recurrence.

Two cases of pityraisis lichenoides et varioliformis acuta were treated with antibiotics like tetracycline for 3 weeks and later with topical steroids and emollients.

As both cases of plaque type parapsoriasis showed early changes of mycosis fungoides, they were treated with radiotherapy and chemotherapy for 6 pulses. Clinical improvement was seen in the form of flattening of lesions. There was improvement of symptoms like pruritus and burning.

Repeat biopsy after treatment showed partial response. These patients were followed up every 3-6 months for clinical and histopathologic examinations.

DISCUSSION

Out of 28 patients, maximum number of patients of parapsoriasis among adults were in the age group of 21- 30 years, while in children between 10-12 years. According to study by Wood GS et al a peak incidence was in the fifth decade, with slight male preponderance in parapsoriasis.(1)

In this study parapsoriasis was more common in males between age group 11 to 30 years, while in female between 21 to 40 years. Male: Female ration was 1.15:1 nearly equal. Study by Irena E et al including 27 parapsoriasis patients observed male outnumbered the females (Male:23, Female:4). (2)

In children with parapsoriasis, male to female ration was 1.33:1.

Median duration of disease before presentation was up to 1 year among adults and 2-3 years in children. While study of 15 parapsoriasis patients by Koh WL et al revealed median duration of disease before presentation was 5.5 months in adults and 6 months in children.

Out of 28 patients 24 were symptomatic, pruritus was the most common symptom encountered, which correlated well with the study by Koh WL et al. (3)

In this study ,all early papular lesions were deep dermal tender positive.

In parapsoriasis lesion were mainly distributed over trunk and extremities, relatively sparing face, palm soles and mucosae. In a study of 34 parapsoriasis same distribution of skin lesion was noticed by El-Darouti MA et al,. (4)

Pityriasis rosea like distribution was noted among 18 patients pityriasis lichenoides chronic.

Most common morphological presentation was erythematous papules followed by hypopigmented macules. Other presentations were erythematous plaque, poikiloderma, and necrotic papules were also encountered.

We noted different type of scaling , of which central adherent scales were commonest followed by collarette scales and diffuse powdery scales.

All routine laboratory investigations were normal in all the patients except in four patients who showed high ASLO titre.

Histological examination revealed that findings described for acute and chronic form in majority of patients were seen. However in 6 patients histopathologic changes of both acute and chronic type co existed.

In case of plaque type of parapsoriasis it is very difficult to distinguish from early stage of mycosis fungoides. On careful histological examination with suspicion of mycosis fungoides, two patients of plaque type parapsoriasis showed lymphohistiolcytic infiltrate in papillary , subpapillary dermis. Epidermotropism was present with small amount of atypical cells suggestive of mycosis fungoides.

Study by Irena E et al noted that out of 27 patients of parapsoriasis none of them progressed to mycosis fungoides on follow up for of 10 years. (2) But study by Mohammad A et al found out of 34 patients of parapsoriasis 5 turned in to mycosis fungoides on 5 year follow up**.** (4) Response to different modality of treatment was good with clearance of skin lesions with only seven patients showed recurrence. Out of 24 patients, 11 were treated with PUVA therapy (8: systemic PUVA, 3: Topical PUVA), median time to resolution was 6-10 months. Similarly Koh WL et al found median time to resolution was 8 months in adults and 21 months in children.

CONCLUSION

Clinically the presentation of parapsoriasis is similar in adults and children; however, it runs a longer course in children. In our series, postinflammatory hyperpigmentation was common, which may be explained by our patients' darker skin phototypes. Upper respiratory tract infections can be a precipitating factor. Pityriasis rosea like distribution of skin lesions were unique findings in our study. Two patient on histology turned out to be a mycosis fungoides. Hence we suggest a long term follow up of patients of parapsoriasis, clinically as well as histologically to look for development of mycosis fungoides. Essentially the treatment consists oral and topical PUVA, systemic antibiotics, emollients and topical steroids. In patients in whom there is raised ASLO titre responded early to systemic antibiotics.

Figure1- Pityriasis Lichenoides Chronica

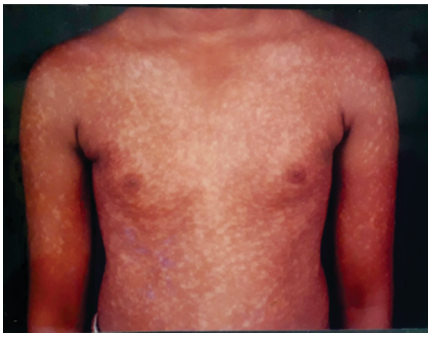


Figure 1 : Hypopigmented macules and few erythematous papules.

Figure 2- Pityriasis lichenoides et varioliformis acuta

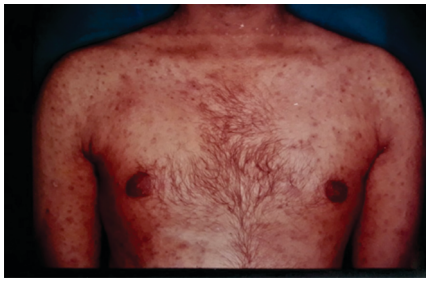


Figure 2 : variolliformis scar , erythematous papules, and necrotic papules covered with crust

Figure 3- Plaque type parapsoriasis

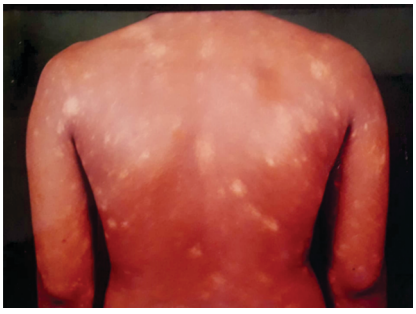
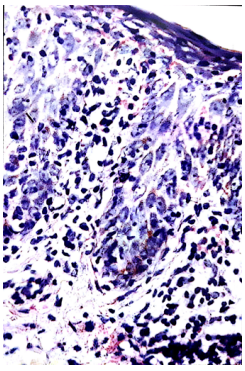


Figure 3 :Hypopigmented macules and patches

Figure 4- Histopathology of Pityriasis Lichenoides Chronica



H&E (40x)- Showing lymphocytic infiltrate in the dermis, extending into epidermis with extravasation of erythrocytes

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