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Medical Science

A CASE REPORT: GRAHAMS LITTLE PICCARDI LASSEUR SYNDROME

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ABSTRACT Graham Little-Piccardi-Lassueur syndrome (GLPLS) is also known as follicular lichen planus. It characterized by the triad of patchy cicatricial alopecia of the scalp, noncicatricial alopecia of the axilla and groin, and a follicular spinous papule on the body, scalp, or both Lichen planopilaris is a follicular type of lichen planus. There are three variants of lichen planopilaris, including GLPLS, frontal fibrosing alopecia, and classic lichen planopilaris. The onset of symptoms in GLPLS may occur in any order, although the condition often begins with the development of the hyperkeratotic papules on the trunk and extremities prior to the development of alopecia. It is four times more common in females in the age group of 30-70 years. The disease is chronic and slowly progressive and relatively rare, most reported patients with the syndrome are middle aged postmenopausal women.

KEYWORDS : Graham Little-Piccardi-Lassueur syndrome, follicular lichen planus, female, oral mucosa.

INTRODUCTION

Graham Little-Piccardi-Lassueur syndrome is a type of lichen planopilaris characterized by the triad of patchy cicatricial alopecia of the scalp, non- cicatricial alopecia of axilla and groin, and follicular spinous papule on body, scalp, or both. Onset of symptoms can occur in any order, but condition often begins with development of hyperkeratotic papules on trunk and extremities prior to development of alopecia. It is most commonly seen in females in the postmeopausal age group, than males.

CASE REPORT:

A 40-year-old female, presented with history of loss of hair over the scalp, decreased hair over limbs from two months. Also complained of small pigmented lesions over abdomen, trunk, back, and forearm.

On examination, patchy scarring alopecia over parietal, vertex area of scalp around 2×3 cm, with few residual normal looking hair present, minimal scaling.

Figure:1

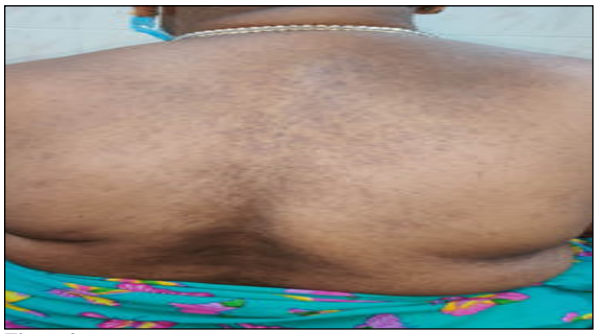

Figure 2

Axillary and pubic hair looks normal. Multiple follicular oriented papules over the abdomen, trunk, thighs, and forearm noticed.

Oral mucosa- violaceous patch on both side of buccal mucosa and tongue shows hyperpigmented patch. On dermoscopy, of scalp- cicatricial alopecia areas with perifollicular whitish-gray scaling and absence of follicular opening.

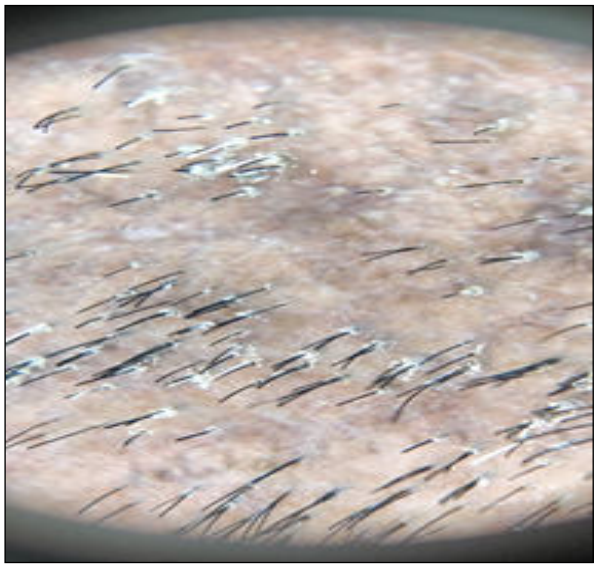

Figure : 3

On dermoscopy of body, loss of hair from follicular opening with perifollicular pigmentation and less to absent scaling. Provisional diagnosis of lichen plano- pilaris was made.

A biopsy from scalp- scarring alopecia with dermis replaced by fibrous with perifollicular and junctional lichenoid infiltrate.

Based on clincopathological corelation, diagnosis of Graham Little-Piccardi- Lassueur syndrome was made.

Patient was started on treatment and counseled for nature of condition and advised for regular follow up.

DISCUSSION

Piccardi in 1913, initially, described a case of progressive scalp cicatricial alopecia, noncicatricial alopecia in the axilla and pubic area, and follicular spinous papules on the trunk and extremities, to which he gave the name *cheratosi spinulosa* (keratotic spinulosa). Graham Little, later in 1915 published a similar case of a 55-year-old woman,

referred by Lassueur of Lausanne, Switzerland, which was described as "folliculitis decalvans et atrophicans,".

Graham Little–Piccardi-Lassueur syndrome is type of lichen planopilaris which presents in adults between 30 and 70 years and is four times more reported in females as compared to males. LPP can be subdivided into three clinical variants: classical LPP, frontal fibrosing alopecia, and Graham Little–Piccardi-Lassueur syndrome. In Graham Little-Piccardi-Lassueur syndrome, in addition to the triad, a positive pull test for anagen hair can be resent.(2)

The exact etiology is unknown, but as it is considered a variant of lichen planus, the immunological mechanism, namely cell-mediated immunity may play a major role in triggering the clinical expression of disease. Few isolated cases with a familial pattern, association with hepatitis B vaccination, and phenotypically female (genetically,XY) patient with androgen insensitivity syndrome (testicular feminization) have been documented.

Altered integrin expression, which has been shown in active LPP lesions, could explain the phenomenon of anagen hair on pull test. Our case was of middle age female who had progressive scalp cicatricial alopecia and follicular spinous papules on the trunk and extremities, third component non-scarring alopecia of axilla and pubis is not yet noticed. Which may get involved as condition get progressed or condition could be without this component. (3)

In addition oral mucosal hyperpigmentation and non-scarring alopecia over extremities were noticed. Non-scarring alopecia over the extremities may be an associated finding,but not commonly reported.

So can we consider oral mucosal pigmentation and non-scarring alopecia over extremities as new component. Histopathologically, early lesions of lichen planopilaris show perifollicular lymphocytic infiltrate at the level of the infundibulum and the isthmus, along with vacuolar changes of the outer root sheath. More developed cases show perifollicular fibrosis and epitheliala trophy at the level of the infundibulum and the isthmus giving rise to a characteristic hourglass configuration. Advanced cases show alopecia with vertically oriented elastic fibers that replace the destroyed hair follicles. Similar findings were seen in our case. This end-stage scarring alopecia with no visible hair follicle is called pseudopelade of Brocq. (2)

Treatment is less effective for such types of lichen palnopilaris as compared to lichen planus, except in the early stage. Once scarring occurs, the hair growth is theoretically impossible. Therefore, the treatment aims are to block the progression of the illness, prevent further alopecia, and symptomatic management. Various treatment modalities tried are topical, intralesional, and systemic corticosteroids, retinoids, cyclosporine, PUVA therapy, and antimalarials. In our case the patient had good symptomatic improvement with topical steroids, antihistamines, and supplements.

The course of the disease is often chronic and progressive with little potential for hair regrowth following follicular inflammation and destruction. In conclusion, this case is reported because of its rarity in presentation.(1)

CONCLUSIONS

In our case, only two components are present, scarring alopecia of scalp and follicular spinous papule on the body. Third component non-scarring alopecia of axilla and pubis is not yet noticed. Which may get involved as condition get progressed or condition could be without this component or can we consider oral mucosal pigmentation as new component.

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