

Grahams Little Piccardi Lasseur Syndrome - A Rare Case Report with Review of Literature

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Abstract: *Grahams Little PiccardiLassueursyndrome is a rare type of Lichenplanopilaris, presents with a triad of Scarring Alopecia in the scalp, non scarring alopecia in axilla and pubic region, and follicular spinous papules in chest, abdomen and extremities. Commonly seen in adults between 40 and 70 yrs with female predominance of 4 times than males. Lichenplanopilaris is a rare variant of follicular lichen planus and is divided into Classical lichenplanopilaris, Frontal fibrosing alopecia and Grahams little piccardiLasseur syndrome. Lichen planopilaris shows the positive hair pull test for anagen hairs. Its etiology is unknown but mostly associated with defective cell mediated immunity. Diagnosis made by both clinical examination and histopathology. The definite treatment is not clear. Lichenplanopilaris with early stage of presentation can be treated with retinoids, Tofacitinib, Hydroxychloroquine, corticosteroids, cyclosporine, Anti Malarials and PUVA therapy. If scarring occurs, it becomes difficult to treat and only symptomatic management can be done and alopecia can not be reversed.*

Keywords: Grahams Little PiccardiLassueur syndrome, Lichenplanopilaris, Follicular lichen planus, Hydroxychloroquine

1. Introduction

Grahams Little PiccardiLassueur syndrome (GLPLS) is a rare type of Lichenplanopilaris^(1,2) presenting with a triad of Scarring Alopecia in the scalp, non scarring alopecia in axilla and pubic region, and follicular spinous papules in trunk and extremities. Commonly seen between 40 and 70 years with female predominance than males. Lichenplanopilaris has three variants namely Classical lichenplanopilaris, Frontal fibrosing alopecia⁽¹⁴⁾ and Grahams Little PiccardiLasseur Syndrome. Its etiology is unclear. Diagnosis made by both clinical examination and histopathology. There is no definitive treatment. Main aim of treatment lies in blocking the disease progress. If scarring occurs, it becomes difficult to treat and alopecia can not be reversed.

2. Case Report

A 40 year old female presented to the hospital with complaints of loss of hair over the scalp and axilla for 1 year which was insidious in onset and gradually progressed to the present stage. History of black discoloration of skin seen in axilla and breast x 6 months which is associated with itching. History of severe anxiety regarding scalp lesions. No history of any recent illness or stress. Not associated with any comorbidities. On examination, cicatricial alopecia seen over scalp (figure 1), mainly involving vertex with sparing of some tufts of normal hair. Non cicatricial alopecia seen over the axilla (figure 2). Ill defined patchy hyperpigmentation surrounded by spinous follicular papules (figure 3) seen over axilla, mammary and infra mammary region. Nails, oral mucosa and genitalia are normal. The diagnosis was made clinically and confirmed by biopsy (figure 4) which showed lichenoid Interface dermatitis with perifollicular lymphocytic infiltrates. Routine investigations like complete blood count, Random blood

sugar, Renal, Liver and Thyroid workups were normal. Patient was counselled about her condition. She was started on T. Hydroxychloroquine.

3. Clinical Pictures



Figure 1: Lichen planopilaris



Figure 2: Non scarring Alopecia in axilla



Figure 3: Spinous papules

Histopathology:

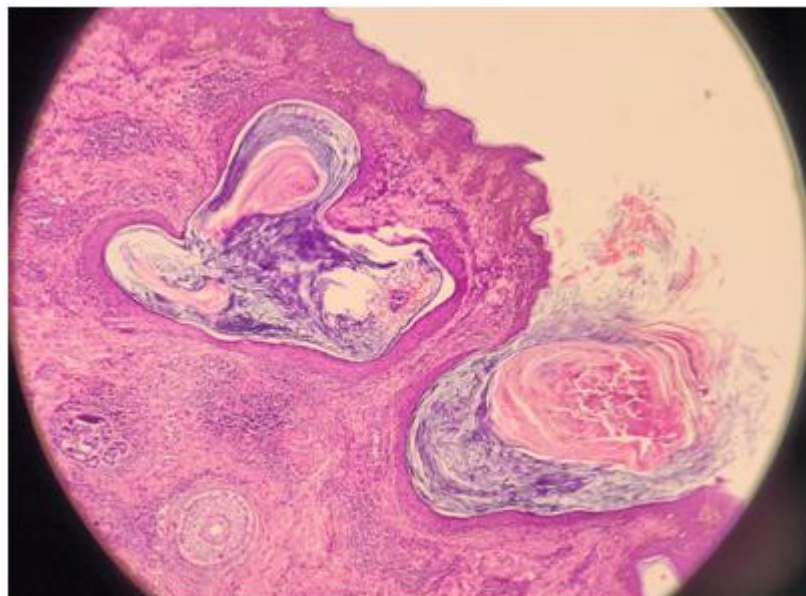


Figure 4: Histopathology shows Lichenoid Interface dermatitis with perifollicular lymphocytic infiltrates - suggestive of Lichenplanopilaris

4. Discussion

Grahams Little PiccardiLassueur Syndrome ^(1, 2) was First reported by Piccardi in 1913. Later, Grahams Little published a similar observation in a patient referred by Lassueur in 1915. Grahams Little PiccardiLassueur syndrome is a rare variant of Lichenplanopilaris ⁽³⁾ presenting with a triad of Scarring Alopecia of scalp, non scarring alopecia of axilla and pubic region, and follicular spinous papules in chest, abdomen and extremities. Commonly seen between 40 and 70 years of age ⁽²⁾. In about 50 cases reported in literature, they are mostly sporadic. Some cases with genetic predisposition ^(5, 6) has been observed. Incidence is 4 times ⁽⁷⁾ more common in females than males. Few rare cases affecting males ⁽⁸⁾ has been reported. Lichenplanopilaris (LPP) also known as lichen follicularis ⁽⁴⁾ is a rare type of follicular lichen planus affecting the hair follicles. LPP has three variants namely Classical lichenplanopilaris, Grahams little piccardiLassueur

syndrome and Frontal fibrosing alopecia. Lichen planopilaris shows the positive hair pull test for anagen hairs. Its etiology is unknown but commonly associated with defective cell mediated immunity. Some studies showed its association with Hep - B vaccination ⁽⁹⁾, Androgen Insensitivity Syndrome ⁽¹⁰⁾, Auto antibodies against the Chromosomal passenger protein INCENP ⁽¹¹⁾, HLA DR1 ⁽⁶⁾, Stress, Menopause and Vitamin A deficiency ⁽⁵⁾ and also with reduced expression of Peroxisome Proliferator Activated Receptor (PPAR) ⁽¹²⁾. On dermoscopy, shows perifollicular erythema and scaling in early stages, which progress to loss of follicular openings later. Biopsy from early lesions shows features of lichenoid interface dermatitis with perifollicular lymphocytic infiltrates ^(13, 14) in infundibulum and isthmus (collar like) with vacuolar changes seen in outer hair root sheath. Late lesions show fibrosis of the dermis with concentric perifollicular fibrosis and replacing pilosebaceous tracts with fibrous strands. Course of the disease is chronic and progressive.

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Lichenplanopilaris should be differentiated from other scarring alopecia like Discoid lupus erythematosus, Pseudopalaed of brocq, Folliculitis decalvens, Frontal fibrosing alopecia, secondary syphilis. The definite treatment is not clear and is quite challenging. Main aim is to halt the disease progress. If scarring occurs, it becomes difficult to treat and only symptomatic management can be done and alopecia can not be reversed. Some studies shows good response to Cyclosporine A (4mg/kg/day)⁽¹⁵⁾, Hydroxychloroquine⁽¹⁷⁾, Tofacitinib⁽¹⁸⁾, Pioglitazone⁽¹⁶⁾, Anti Malarials, Corticosteroids, Retinoids, PUVA therapy and excimer laser (308nm)⁽¹⁹⁾.

5. Conclusion

In late stages if scarring occurs, it becomes difficult to treat. So this case is reported because of its rarity and to have extensive knowledge about this condition and to diagnose early to prevent complications.

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