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Case presentation

A case of Graham-Little–Piccardi–Lasseur syndrome

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Abstract

Graham-Little–Piccardi–Lasseur Syndrome (GLPLS) is a variant of lichen planopilaris, which is characterized by progressive cicatricial alopecia of scalp, non-cicatricial alopecia of axillae and pubic regions, and keratosis pilaris-like follicular papules over trunk and extremities. GLPLS is a disease of unknown etiology. However, recent reports support a central role for a T-cell-mediated immune response in the pathogenesis of GLPLS. Besides, although GLPLS is believed to occur sporadically, a genetic predisposition also has been implicated in the pathogenesis. On the other hand, GLPLS typically affects middle-aged women, particularly of the postmenopausal age group. A diagnosis of GLPLS is generally apparent with the presence of characteristic findings in a postmenopausal woman. Herein, we report a case of GLPLS in a 75-year-old woman with the typical triad of alopecia of the scalp, non-cicatricial alopecia of axillae and pubis, and a follicular keratotic eruption on the trunk.

Keywords: Graham-Little–Piccardi–Lasseur syndrome, lichen planopilaris

Introduction

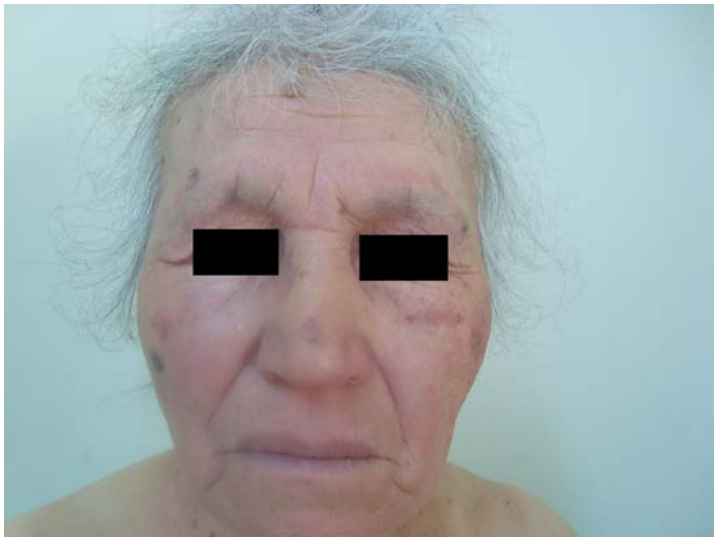
In 1914 Piccardi reported a case with progressive cicatricial scalp alopecia associated with non-cicatricial alopecia of axillary and pubic regions and a follicular lichenoid eruption on the trunk. Piccardi realized that the patient was representing a rare medical entity to which he gave the name ‘cheratosi spinulosa’ (keratotic spinulosa) [1]. In 1915, Ernst Graham-Little also published a similar case observed by Lassueur, which he called ‘folliculitis decalvans et atrophicans’ [2]. Today this condition is known as Graham-Little–Piccardi–Lasseur Syndrome (GLPLS), a rare variant of lichen pilanopilaris [3,4]. GLPLS is a rare dermatosis and usually evident in middle-aged Caucasian women [4-7]. Herein, we present a case of GLPLS with the characteristic triad of symptoms originally described by Piccardi.

Case synopsis

A 75-year-old woman was admitted to our outpatient clinic with a few-year history of scalp, axillary, and pubic hair loss. Her past medical history was remarkable for well-controlled hypertension and her family history was irrelevant. On dermatological examination, we observed hair loss of eyebrows and scalp along with follicular papules on eyebrows and hundreds of erythematous follicular papules with prominent central keratotic plugs scattered over the scalp (Figure 1,2,3). Examination also disclosed axillary and pubic hair loss and several discrete, flesh-colored minute papules grouped over the bilateral anterior pectoral regions (Figure 1,4,5).

Routine laboratory studies were within normal limits and histopathological examination of the scalp lesions demonstrated a lymphocytic infiltrate in the follicular infundibular region (Figure 6). A diagnosis of GLPLS was made based upon

history, clinical and histopathological findings. The patient was given topical corticosteroids to apply on scalp lesions, but we could not assess the outcome of the treatment, because the patient was lost to follow up.



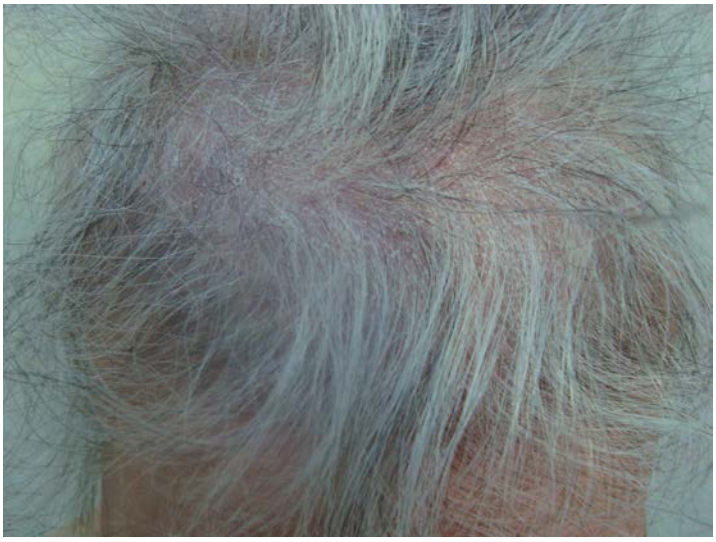
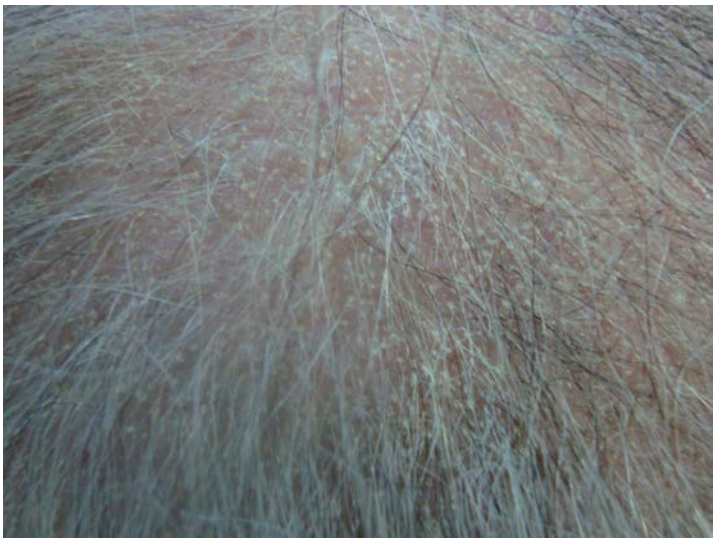


Figure 1. Hair loss of eyebrows. **Figure 2.** Numerous follicular papules with plugs of keratin in the follicular orifice and perifollicular erythema



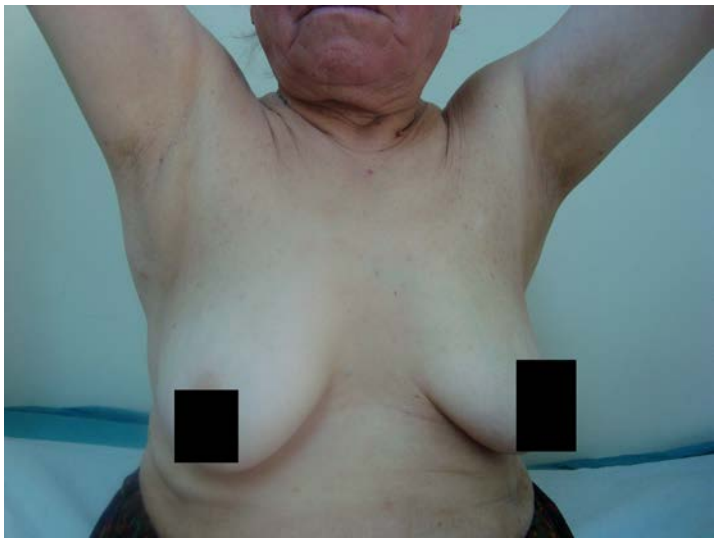
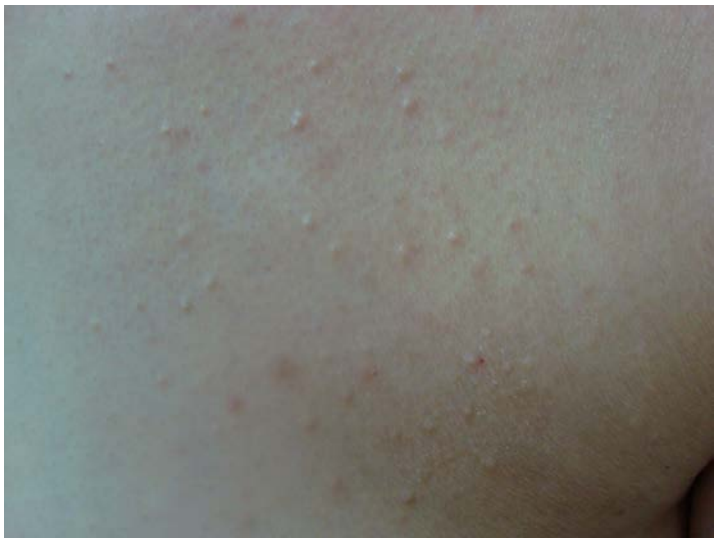


Figure 3. Follicular keratotic plugs. **Figure 4.** Bilateral axillary hair loss



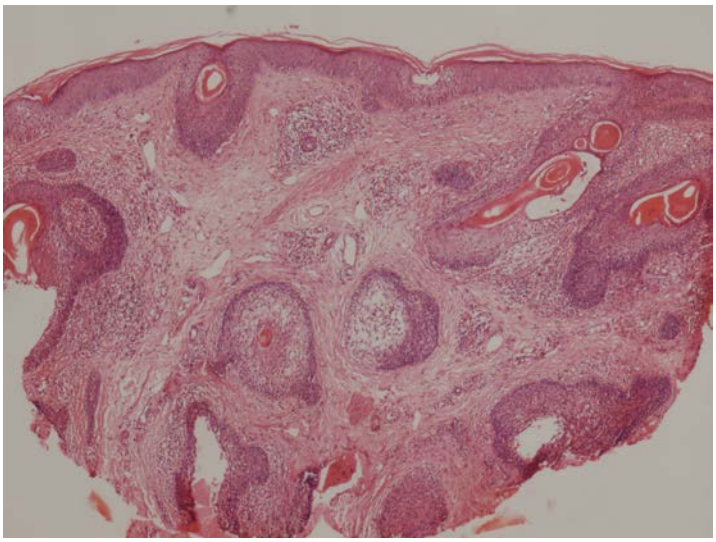


Figure 5. Firm, flesh-colored minute papules. **Figure 6.** Lymphocytic infiltrate in the follicular infundibular region (H&Ex40)

Discussion

GLPLS is a rare lichenoid dermatosis with unknown etiology. On the other hand, as it is an inflammatory dermatosis, an aberrant immune response has been implicated in its pathogenesis [4,5]. The first evidence for autoimmunity in GLPLS was documented by *Rodríguez-Bayona et al*. They demonstrated autoantibodies against the chromosomal passenger protein INCENP in a patient with GLPLS as the proof of autoantigenic response in the pathogenesis of the disease [8]. In addition, hepatitis B virus (HBV) vaccination [9], androgen insensitivity syndrome [10], hormonal dysfunctions, vitamin A deficiency, and neuropsychological stress [5] have been correlated with GLPLS. A genetic background also has been implicated in the development of GLPLS and *Viglizzo et al.* described the first familial occurrence of GLPLS in a mother and her daughter associated with HLA-DR1 antigen [11]. However, it is generally accepted that GLPLS is a sporadic disease most commonly affecting middle-aged women [4-7]. The characteristic triad of symptoms are not needed to occur simultaneously and scalp alopecia usually precedes the follicular eruption [4,5]. It has been claimed that half of the patients display symptoms of cutaneous or mucosal LP at least once in the course of the disease [12].

GLPLS is a chronic disease with a progressive course [5,13]. The scalp lesions begin as hyperkeratotic follicular papules slowly evolving into patchy atrophic alopecic areas [14]. Herein, we report a case of GLPLS in a 75-year-old woman with characteristic clinical and histopathological features who exhibits the classical triad of GPLPS.

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