Avens Publishing Group
J Clin Investigat Dermatol
August 2020 Volume 8, Issue 2

Open Access Case Report

Journal of Clinical & Investigative Dermatology

Syphilitic Alopecia: Report of a Challenging Case and Review of the Literature

Golbert S*, Beguerie JR and Busso C

Department of dermatology, Hospital Universitario Austral, Argentina

***Address for Correspondence**

Golbert S, Department of dermatology, Hospital Universitario Austral, Buenos Aires, Argentina Av. Peron 1500, Derqui (1629) Buenos Aires Province, Argentina, Email: sgolbert@cas.austral.edu.ar

Submission: 23 April 2020

Accepted: 29 May 2020

Published: 29 August, 2020

Keywords: Secondary syphilis; Syphilitic alopecia; Neurosyphilis

Abstract

Syphilitic alopecia (SA) is a rare manifestation of syphilis, which may be the only manifestation of the disease.

Two clinical forms are described; a symptomatic alopecia and an essential alopecia.

This last clinical form is the most frequently observed, presenting as a "moth-eaten" or "patch-form" appearance, in a more diffused hair loss pattern. The term neurosyphilis refers to the infection of the Central Nervous System (CNS) by Treponema Pallidum (T. pallidum) and can occur at any time during the course of the disease.

We present the case of a 61-year-old man who was diagnosed with early neurosyphilis presenting with syphilitic alopecia, and a review of the current literature. Although there are numerous reports of individual cases of both syphilitic alopecia and neurosyphilis, to our knowledge there are only three reports that describe their simultaneous presentation. We emphasize the importance of recognizing this presentation of syphilis in clinical practice, in order to carry out a timely treatment of the patient and their contacts.

Introduction

Syphilis is a sexually transmitted disease caused by Treponema pallidum (TP), highly prevalent in Argentina. In 2018, this country reported an increase in the incidence of this infection of 55.4% in women and 36.4% in men, compared to the year 2017 [1]. However, these numbers can be underestimated since syphilis is not a mandatory reportable disease in Argentina. Syphilis presents three stages throughout its evolution: primary, secondary and tertiary. It may also present periods of latency between the secondary and tertiary stages, in which it is diagnosed only by serological tests [2]. This disease has multiple cutaneous and systemic manifestations, which depend on the stage of the disease. Syphilitic alopecia (SA) is a rare manifestation of the secondary stage that usually occurs in 2.9-11.2% of infected patients [3,4], as a single sign or associated with other manifestations. Neurosyphilis (NS) is the infection of the central nervous system (CNS) by TP and can occur at any time during the course of the disease. Although there are numerous reports of syphilitic alopecia and neurosyphilis, to our knowledge this would be the fourth case with simultaneous presentation of both signs in the same patient [5,6].

We present the case of a patient with multiple manifestations, but undiagnosed for ten years, who after consulting about his alopecia, was diagnosed with neurosyphilis.

Case Presentation

A 61-year-old male patient presented to the Dermatology Department of the Hospital Universitario Austral (HUA) with a six-month history of hair loss and pruritic eruption in the face. He had a history of multiple non specific symptoms that required evaluation of various medical specialties during the last ten years without reaching any conclusive diagnosis. Initially, these symptoms included generalized arthralgia of the cervical spine and shoulder girdle. Later on, the patient presented persistent intense asthenia, myalgia, oppressive holocranial headache, tinnitus, and decreased vision. Symptoms increased insidiously with time.

Due to persistence and aggravation of his ocular symptoms, a dilated fundus examination was performed in which papillae edema was observed with small bilateral hemorrhages. A brain magnetic resonance imaging (MRI) without gadolinium, as well as an electroencephalogram was performed, without relevant findings.

Laboratory tests showed an increased erythrocyte sedimentation rate (ESR) and a positive ANA 1/80. A lumbar puncture was performed that demonstrated discrete pleocytosis (37 leukocytes / mm3, 100% lymphocyte) and mild hyperproteinorrachia (0.55 gr / L). Venereal Disease Research Laboratory (VDRL) test and cultures on cerebrospinal fluid (CSF) were negative.

Subsequently, the patient consulted the dermatology department for scalp alopecia with papular, erythematous and pruritic lesions on the forehead, retro auricular region and on the scalp (Figures 1A,1B). Alopecia was diffuse and in patches predominantly located in the parietal region. Erythematous and asymptomatic macules were also observed in the trunk. He had no lesions in the oral or genital mucosa, palms or soles. Considering secondary syphilis as a differential

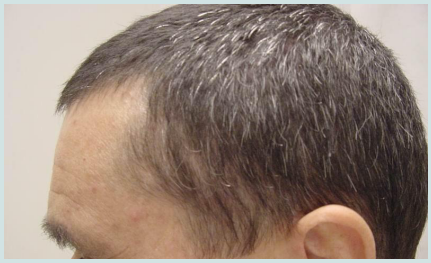

Figure 1A: Alopecic patches in temporal region of the scalp.

ISSN: 2373-1044

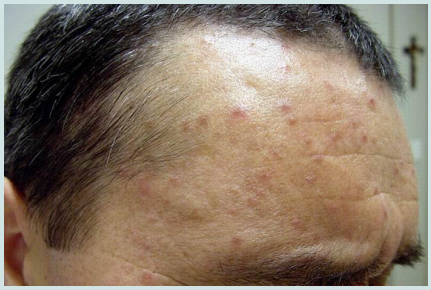

Figure 1B: Erythematous papules in forehead and retro auricular areas.

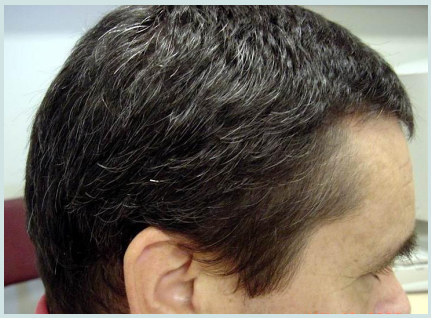

Figure 2: Three months after penicillin treatment. No alopecia is seen, nor papules in the forehead or in the retro auricular areas.

diagnosis, a quantitative VDRL was requested. Also, a skin biopsy of a papular lesion on the scalp was performed. The microscopic examination showed folliculitis with acute perifolliculitis with a neutrophilic predomination of the cells. Neutrophils compromised the follicle wall and the peripheral area. Focal rupture of the follicular wall was evident.

The VDRL test was positive (512dils) which was confirmed by fluorescent treponemal antibody absorption test (FTA abs). Consequently, diagnosis of secondary syphilis was made. Moreover, in this setting, the pathologic CSF analysis was interpreted as neurosyphilis. Diagnostic tests for human immunodeficiency virus (HIV) were non-reactive. In a new anamnesis, the patient reported a risky sexual contact four years before the onset of systemic symptoms. The diagnosis of syphilitic alopecia with neurosyphilis was reached. Treatment with penicillin G sodium 24,000,000 IU / day was initiated intravenously for 10 days.

Seven days after initiation of treatment, the patient presented slight improvement of the ocular symptoms with decreased flashes and improved vision and at day20 he reported improvement of the asthenia. Over the next three months, the facial lesions disappeared and the alopecia completely resolved (Figure 2).

Over a period of six years of follow-up, the patient remained asymptomatic with normal fundus and serum VDRL values of 2 dils.

Discussion

Alopecia is a rare clinical manifestation of secondary syphilis and its pathogenesis is not completely understood. One hypothesis is that the presence of and the inflammatory response to TP, are responsible for the loss of terminal hairs. Also, these two factors may interrupt the hair follicle cycle which results in empty follicles or broken hairs [7,8]. Another hypothesis suggests that SA is due to a telogen effluvium secondary to the inflammatory process generated by the systemic infection, rather than by a direct effect of the TP [8,9].

According to McCarthy's classification (1940), SA can be symptomatic (coexisting with other clinical manifestations); or essential (when it is the only clinical manifestation) [4,9,10]. The latter is non scarring and can present in four different clinical patterns: 1) moth-eaten, or glade like, characterized by small areas of alopecia, mainly in the parieto-occipital region, not completely devoid of hair and with poorly defined edges, this being the most frequent and considered as pathognomonic [9,11]; 2) diffuse with uniform loss and thinning of hair, similar to telogen effluvium; 3) mixed, as this patient's case; and 4) alopecia of the eyebrows: with thinning or absence of hair in the distal third section of the eyebrow [11].

A diagnostic tool rarely used for the diagnosis of SA is skin biopsy, which is usually indicated if there is a strong suspicion of another alopecia that cannot be determined clinically [12]. The main histological differential diagnosis of SA is alopecia areata (AA). In a study that compared histopathological findings between these two, the presence of peribulbar eosinophils was considered a distinctive feature present in AA. On the contrary, the absence of eosinophils, the presence of peribulbar plasma cells, and abundant lymphocytes at the isthmus, suggests SA [7]. In the present case, microscopic examination revealed a mild decreasein hair density, irregular hair shafts, and no abnormalities in the terminal: vellus ratio. There was no eosinophilicin filtrate. However, mild presence of periinfundibular inflammatory lymphocytes was observed. No specific findings in the direct immunofluorescence at the follicular basal membrane zone were found.

To our knowledge, only three cases similar to ours have been published in the literature. The first report was of a male patient with alopecia of the scalp, eyebrows and eyelashes, oral ulcers, maculopapular rash of the face and HIV infection, and rapid progression to a left ocular panuveitis [5]. The second was a woman with progressive headache and diffuse alopecia [6] and the last case, a 53-year-old woman with patchy alopecia, dysphonia, genital lesions and skin rash [13]. Although neurosyphilis can occur at any stage of the disease, it is more frequent after 2 years of infection (late neurosyphilis) [14]. In early stages, the spirochaete usually affects meninges and blood vessels, manifesting as acute or chronic meningitis and/or as auditory or ocular symptoms (iritis, retinitis or uveitis).

In our patient, blood VDRL was positive with a titer of 512 dils, but CSF VDRL was negative. So far, VDRL is the only validated test for CSF evaluation. However, it can be non-reactive in up to 50% of cases [6,15]. Consequently, this result does not exclude neurosyphilis, in symptomatic patients. In addition, in this context, the finding of pleocytosis and a high level of proteins in the CSF are also suggestive of neurosyphilis [6].

Conclusions

We present the case of a patient with rheumatologic, neurologic, ophthalmic, auditory and systemic signs and symptoms for more than ten years, with no clear and certain diagnosis reached. This

would be the fourth reported case of coexisting syphilitic alopecia and neurosyphilis.

We highlight the importance of recognizing the cutaneous manifestations of this polymorphic infection, given that it was through alopecia that the final diagnosis was reached.

References

1. Integrated surveillance bulletin: National Directorate of Epidemiology and Health Situation Analysis of the Ministry of Health of the Argentine Nation [Boletínintegrado de vigilancia-Dirección Nacional de epidemiología y Análisis de la situación de salud del Ministerio de Salud de la Nación Argentina] - N° 411 – SE 23 2018– mayo de 2018.
2. Torales G, Galarza P, Soto V. Diagnostic and Treatment Guide for Syphilis. Recommendations for health teams. Directorate of AIDS, STD, Hepatitis and TB [Guía de Diagnóstico y Tratamiento de la Sífilis. Recomendaciones para losequipos de salud. Dirección de Sida, ETS, Hepatitis y TBC]. 2019.
3. Vafaie J, Weinberg JM, Smith B, Mizuguchi RS (2005). Alopecia in association with sexually transmitted disease: a review. Cutis 76: 361-366.
4. Ye Y, Zhang X, Zhao Y, Gong Y, Yang J, et al (2014) The clinical and trichoscopic features of syphilitic alopecia. J Dermatol Case Rep 8: 78-80.
5. Wong BJ, Hong BK, Rao NA (2014) Rapid Progression of Uveitis and Alopecia Syphilitica in AIDS. JAMA Ophthalmol 132: 506 -508.
6. Cherington CC, Ressler SW (2013). 31-Year-Old Woman With Chronic Daily Headache and Alopecia. Mayo Clinic Proceedings 88: 206-210.
7. Lee JYY, Hsu ML (1991) Alopecia syphilitica, a simulator of alopecia areata: histopathology and differential diagnosis. J Cutan Pathol 18: 87-92.
8. Doche I, Hordinsky MK, Valente NYS, Romiti R, Tosti A (2017) Syphilitic Alopecia: Case Reports and Trichoscopic Findings. Skin Appendage Disord 3: 222-224.
9. Hernández-Bel P, Unamuno B, Sánchez-Carazo JL, Febrer I, Alegre V (2013) Alopecia sifilítica: presentación de 5 casos y revisión de la literatura. ActasDermo-Sifiliográficas 104: 512-517.
10. Ornelas J, Agbai ON, Kiuru M, Sivamani RK (2015) Alopecia as the Presenting Symptom of Syphilis. Dermatol Online J 21: 13030.
11. Costa MC, Peres AS, Queiróz AJR, Souza Medeiros N, Costa IMC (2015) Nonspecific diffuse alopecia as a single manifestation of syphilis infection: clinical and trichoscopic features. Int J Dermatol 57: 593-595.
12. Jordaan HF, Louw M (1995) The Moth-Eaten Alopecia of Secondary Syphilis A Histopathological Study of 12 Patients: Am J Dermatopathol 17: 158-168.
13. Hanlon CL, Galoosian A, Ali S, Edson RS (2018) Painful rash with hoarseness: an atypical presentation of syphilis. BMJ Case Rep 11: 226892.
14. Piura Y, Mina Y, Aizenstein O, Gadoth A (2019) Neurosyphilis presenting as cranial nerve palsy, an entity which is easy to miss. BMJ Case Rep 12: 226509.
15. Bansal R, Jindal N (2012) VDRL test and its interpretation. Indian J Dermatol 57: 413.