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ALOPECIA UNIVERSALIS: A CASE REPORT

Dermatology

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ABSTRACT

Alopecia universalis is a rare, non-scarring, autoimmune disorder. It is characterized by sudden hair loss that starts with one or more circular bald patches. It is a disease where the immune system mistakenly attacks the hair follicles. There is no cure for alopecia areata, however, some treatments may help in improving hair growth. We present the case of a 22-year-old male who presented with a complaint of hair loss since 2 years. Physical examination revealed diffuse type of non-scarring hair loss all over scalp, face, and body. Our capacity to tackle this skin disorder and improve patient outcomes will be further enhanced by ongoing research and breakthroughs in treatment techniques. Overall, this case report underscores the significance of prescribing more potent and efficacious drugs in correct dosage with immediate use after diagnosis for rapid results.

KEYWORDS

INTRODUCTION:

Alopecia universalis represents a rare and severe manifestation of alopecia areata, a non-scarring disorder causing hair loss. This condition is characterized by the total absence of hair on both the scalp and body. It occurs due to an autoimmune response, where the immune system erroneously targets and destroys the hair follicles, resulting in hair loss. [1] This case report aims to present an analysis of a specific instance of alopecia universalis.

AA prevalence is lower in adults than children, is increasing over time, and significantly differs by region[2] and its most extreme manifestation is alopecia universalis.While the precise etiology of alopecia universalis remains uncertain, there is a prevailing belief that genetic predisposition and immune system dysfunction are influential factors. The prominent characteristic of this condition is its profound hair loss, which manifests abruptly and advances swiftly. This leads to the development of smooth, hairless skin not only on the scalp but also on the face, body, eyebrows, and eyelashes. The absence of hair follicles can be verified through microscopic analysis. Additionally, certain individuals may encounter nail alterations like pitting or ridges.[3]

Alopecia areata is associated with several concurrent diseases (comorbidities) including depression, anxiety, and several autoimmune diseases including thyroid disease (hyperthyroidism, hypothyroidism, goiter ant thyroiditis), lupus erythematosus, vitiligo, psoriasis, rheumatoid arthritis and inflammatory bowel disease, which makes the treatment difficult. [3]

Treatment of alopecia totalis and alopecia universalis is often challenging and unsatisfactory. Recently, Janus kinase inhibitor has shown promising results.[4]

Hence it is essential to report such occurrences to ignite discussion and take steps towards finding better forms of therapy. Alopecia areata affects approximately 2% of the general population at some point during their lifetime. The prevalence of alopecia areata was reported to be between 0.1% to 0.2% with a life time incidence of 1.7%. Though it can occur at any age, most patients develop the condition before the age of 40 years with the with a mean age of onset between 25 and 36 years. Early-onset alopecia areata (mean age of onset between ages 5–10 years) predominantly presents as a more severe subtype, such as alopecia universalis.[5]

Several studies state that alopecia areata has a genetic basis. The prevalence of adult patients with a family history is estimated to be between 0% and 8.6%, whereas in children data between 10% and 51.6% are reported. One study found that men were more likely to have a positive family history than women. The occurrence of the disease in identical twins, siblings and families with several generations of affected individuals indicate that it is hereditary.[5]

Case Report:

A 22-year-old male patient, presented to the Out-patient Department of Dermatology SIMS Hyderabad with complaints of hair loss since 2 years. Patient had no other complaints other than hair loss which was gradual in onset.

He is unmarried and is an electrician by occupation. The patient was not suffering from other co-morbidities such as COVID, thyroid disorders, diabetes, hypertension, typhoid.

The patient first noticed a gradual increase in hair loss which later progressed to multiple patches of hair loss on scalp and involved the entire scalp in about 2 years [FIG 2,3,4]. There was loss of eyebrows, moustache, beard, including hair all over the body[FIG 1].

On Examination: The patient was suffering from diffuse type of non-scarring hair loss, all over the scalp, face, and body.

The patient was also found to have discoloration on his fingernails known as melanonychia.[FIG 6,7,8]

FIGURES:
PRESENTED AS:

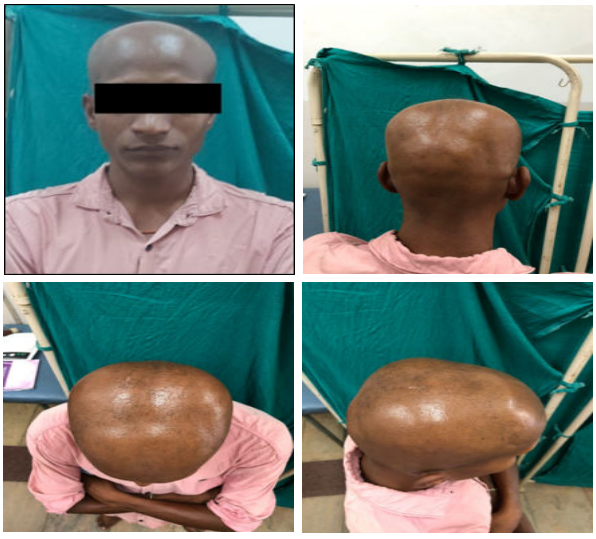

FIG:1 ,2 ,3 and 4

AFTER 6 Months of Treatment:

FIG:5

FIG:6

FIG:7

DISCUSSION:

Alopecia areata (AA) is an autoimmune disease affecting people of all ages. There is currently no cure for AA, and a highly efficacious therapy for severe AA has been elusive. Recently, scientific advances have identified the Janus kinase pathway as a target for treatment. Both Janus kinase inhibitors approved by the US Food and Drug Administration, tofacitinib and ruxolitinib, have shown promise in open-label clinical trials.[6]

AA is a straightforward clinical diagnosis to make, when it presents in its classical form with focal or multifocal patches of acute non-scarring alopecia, and a positive hair-pull test. Occasionally, a skin biopsy might be required to distinguish diffuse AA from other conditions such as female androgenetic alopecia or telogen effluvium. In these cases, AA associated with the histopathological finding of a peribulbar immune infiltrate of cytotoxic T cells (the so-called swarm of bees). However, newer non-invasive technologies aid in the diagnosis of difficult cases.[7]For the presented case we used *Tofacitinib* as studies claim that those with alopecia totalis or alopecia universalis, 77% of the subjects achieved a clinical response, with 58% of patients achieving greater than 50% change in SALT (Severity Alopecia Tool) score over 4 to 18 months of treatment. Patients with AA had a better response to *Tofacitinib* than patients with alopecia totalis or alopecia universalis (81.9% vs 59.0%). It was well tolerated, and there were no serious adverse events.[8]

Tofacitinib is an inhibitor that targets specific kinases and has the ability to selectively inhibit JAK-3, thereby blocking the receptors associated with the γ chain. The signaling pathways of interferon gamma (IFN-γ) receptors and γc family receptors are mediated by JAK1/2 and JAK 1/3, respectively. Studies conducted on mice have demonstrated that a specific subset of cytotoxic CD8+NKG2D+ T cells, which produce IFN-γ, play a crucial role in the development of AA. Furthermore, when tofacitinib is administered systemically, it has been observed to decrease the frequency of CD8+NKG2D+ T cells. As a result, it is believed that tofacitinib exerts its effects by influencing these molecular mechanisms, thereby contributing to the histological and clinical reversal of AA/AU.[9]

CONCLUSION:

In conclusion, this case report highlights the clinical presentation, diagnosis, and management of a dermatological condition called alopecia universalis. It especially emphasizes the importance and the role of the immunosuppressant drug tofacitinib in the treatment of the disease. Alopecia universalis is the rarest and the most severe form of alopecia areata. In a nutshell, the pathological basis causing manifestation of AA is such that, the CD8+NKG2D+ T cells producing IFN-γ are necessary and sufficient for its induction. Thus, it is currently classified as an autoimmune disorder.

The point of interest is that the Interferon gamma (IFN-γ) receptors and γc family receptors signal through JAK1/2 and JAK 1/3, respectively. Tofacitinib being a targeted kinase inhibitor, selectively inhibits JAK-3, thus blocking γ chain receptors. In addition, systemic tofacitinib treatment has been found to reduce the frequency of CD8+NKG2D+ T cells. Thus, it is believed that aforesaid drug exerts its effects through these molecular events and contributes to histological and clinical reversal of AA/AU.Tofacitinib when put into use, was found to be well tolerated, and there were no serious adverse events observed during the treatment of the case. Application site reactions are common.Very few AA cases treated with tofacitinib or other JAK inhibitors have been reported to date.9 Our capacity to tackle this skin disorder and improve patient outcomes will be further enhanced by ongoing research and breakthroughs in treatment techniques.

Overall, this case report underscores the significance of application of targeted and efficacious drugs in correct dosage for desired results.

REFERENCES:

1. Islam N, Leung PSC, Huntley AC, Eric Gershwin M. The autoimmune basis of alopecia areata: A comprehensive review. *Autoimmun Rev*. 2015;14(2):81-89. doi:10.1016/j.autrev. 2014.10.014
2. Lee HH, Gwillim E, Patel KR, et al. Epidemiology of alopecia areata, ophiasis, totalis, and universalis: A systematic review and meta-analysis. *J Am Acad Dermatol*. 2020;82(3):675-682. doi:10.1016/j.jaad.2019.08.032
3. Chu SY, Chen YJ, Tseng WC, et al. Comorbidity profiles among patients with alopecia areata: The importance of onset age, a nationwide population-based study. *J Am Acad Dermatol*. 2011;65(5):949-956. doi:10.1016/j.jaad.2010.08.032
4. Shin JW, Huh CH, Kim MW, et al. Comparison of the Treatment Outcome of Oral Tofacitinib with Other Conventional Therapies in Refractory Alopecia Totalis and Universalis: A Retrospective Study. *Acta Derm Venereol*. 2019;99(1):41-46. doi:10.2340/00015555-3057
5. Pratt CH, King LE, Messenger AG, Christiano AM, Sundberg JP. Alopecia areata. *Nat Rev Dis Primer*. 2017;3:17011. doi:10.1038/nrdp.2017.11
6. Liu LY, King BA. Tofacitinib for the Treatment of Severe Alopecia Areata in Adults and Adolescents. *J Investig Dermatol Symp Proc*. 2018;19(1):S18-S20. doi:10.1016/j.jisp.2017. 10.003
7. King BA, Senna MM, Ohyama M, et al. Defining Severity in Alopecia Areata: Current Perspectives and a Multidimensional Framework. *Dermatol Ther*. 2022;12(4):825-834. doi:10.1007/s13555-022-00711-3
8. Wang ECE, Christiano AM. The Changing Landscape of Alopecia Areata: The Translational Landscape. *Adv Ther*. 2017;34(7):1586-1593. doi:10.1007/s12325-017-0540-9
9. Liu LY, Craiglow BG, Dai F, King BA. Tofacitinib for the treatment of severe alopecia areata and variants: A study of 90 patients. *J Am Acad Dermatol*. 2017;76(1):22-28. doi:10.1016/j.jaad.2016.09.007
10. Erduran F, Adışen E, Burhan Aksakal A. Excellent response to tofacitinib treatment in a patient with alopecia universalis. *Acta Dermatovenerol Alp Pannonica Adriat*. 2017;26(2). doi:10.15570/actaapa.2017.15
11. Hordinsky MK. Current Treatments for Alopecia Areata. *J Investig Dermatol Symp Proc*. 2015;17(2):44-46. doi:10.1038/jidsymp.2015.41