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# A Rare Case Report Of Bazex-Dupré-Christol Syndrome

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***Abstract-***
*Bazex-Dupré-Christol syndrome (BDCS) is a rare autosomal dominant genetic disorder characterized by skin and hair abnormalities, including multiple basal cell carcinomas (BCCs), palmoplantar keratoderma, follicular papules, and alopecia.[1] Mutations in the ACTRT1 gene are implicated in its pathogenesis, although the exact mechanisms remain unclear. The disorder's clinical features often manifest early, with skin cancers primarily appearing on sun-exposed areas, posing significant risks for metastasis if not promptly managed. Additional features include facial keratotic lesions, skin atrophy, and sebaceous hyperplasia. Differential diagnosis includes Gorlin syndrome.[3] Due to its impact on physical appearance and risk of skin cancers, BDCS necessitates vigilant dermatologic surveillance, early intervention, sun protection, and psychosocial support. Awareness among clinicians and genetic counseling are essential for effective management of affected families.[4]*

***Keywords:*** *Bazex-Dupré-Christol syndrome, basal cell carcinoma, ACTRT1 gene, palmoplantar keratoderma, follicular papules, alopecia, genetic disorder, dermatology, skin cancer, surveillance, genetic counseling*

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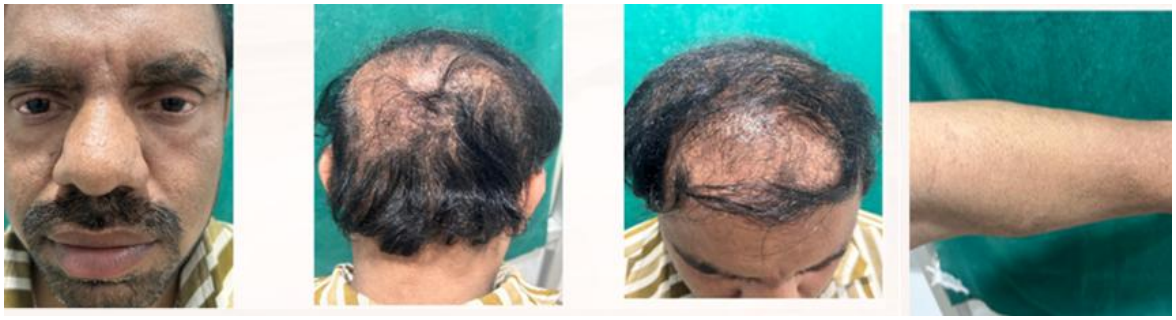
## I. Intoduction –

Bazex-Dupré-Christol syndrome (BDCS) is a rare hereditary dermatological disorder characterized by a combination of skin, hair, and facial abnormalities, along with a predisposition to developing multiple basal cell carcinomas (BCCs).[2] It is inherited in an autosomal dominant pattern, meaning that only one mutated copy of the responsible gene is sufficient to cause the condition. Although its exact prevalence is unknown, BDCS typically manifests early in life, often during adolescence or young adulthood.[1] The syndrome is notably associated with mutations in the ACTRT1 gene, which contribute to the pathogenesis of the disorder. The clinical features include facial keratotic lesions, follicular papules, alopecia, palmoplantar keratoderma, and an increased risk of skin cancer, primarily on sun-exposed areas.[4] Due to its genetic and dermatological complexity, early diagnosis and vigilant management are essential to improve patient outcomes.

## II. Case Report:

A 34 years old male presented with asymptomatic lesion over the forearm, arms since childhood. history of multiple BCCs on neck, face,c/o loss of hair since 5 years recurrent soreness of the scalp and sparse hair growth affecting the scalp, eyebrows and eyelashes. His family history was unremarkable

On Examination – There was hypotrichosis predominantly affecting the scalp over frontal, vertex and parietal area, eyebrows and Sparse hair present over the eyelashes , presence of broadened nasal bridge. (Fig. 1,2,3). The hairs of the scalp showed an admixture of irregularly curly hairs intermingled with normal hairs. Multiple icepick and box scar which are follicularly oriented and follicular atrophoderma present over the extensor aspect of forearm(Fig 4)



(Fig:1,2,3,4)

## III. Discussion:

Bazex-Dupré-Christol syndrome (BDCS) is a rare genetic disorder that primarily affects the skin and hair, with some associated risks for skin cancer. It is inherited in an autosomal dominant pattern, meaning only one copy of the altered gene is sufficient to cause the disorder.[1] Mutations in the **ACTRT1** gene have been identified as a cause of BDCS, though the precise genetic mechanism may vary. Skin Abnormalities such as Basal Cell Carcinomas (BCCs)[4] One hallmark of BDCS is the development of multiple basal cell carcinomas, often manifesting at an early age—sometimes during adolescence or young adulthood. These skin cancers are typically found on sun-exposed areas like the face, neck, and hands but can also appear elsewhere. They tend to be numerous and may recur or metastasize if not properly managed.[7]

Palmoplantar Keratoderma: Thickening and hyperkeratosis on the palms and soles are common, often leading to discomfort or increased susceptibility to infections.[3] Facial Features: Patients may have follicular papules and keratotic lesions around the nose, cheeks, and forehead.[4] Hair Abnormalities: Alopecia: Sparse or absent scalp hair, sometimes congenital or appearing early in childhood. The eyebrows and eyelashes may also be affected.Other Features:

Some patients develop cutaneous atrophoderma—areas of skin with thinning and scarring.There might be a mild sebaceous hyperplasia.[5] Differential Diagnosis with multiple basal cell carcinomas such as Gorlin syndrome (nevoid basal cell carcinoma syndrome) [6]

## IV. Conclusion –

Since it is a dermatological condition with unknown incidence .It has psychosocial impact of having multiple skin cancers and their treatment. It is important for the dermatologist to know about the condition and its treatment.With vigilant monitoring and treatment, individuals can manage skin lesions effectively. Early detection of skin cancers is vital to prevent extensive tissue destruction or metastasis. Regular dermatologic surveillance for early detection and treatment of skin cancers and sun protection. Cosmetic treatment for skin lesions and hair abnormalities .Genetic counseling for affected families.

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