ORIGINAL RESEARCH PAPER

Dermatology

KEY WORDS:

# BAZEX SYNDROME– A RARE CASE REPORT

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ABSTRACT

Bazex-Dupre-Christol syndrome is a rarely reported genodermatoses consisting mainly of follicular atrophoderma with multiple basal cell epitheliomas along with other dermatological associations. Here we report a patient with classical findings of this condition along with Hidradenitis suppurativa and acne conglobata.

## INTRODUCTION:

Bazex syndrome is also known as Bazex-Dupre-Christol syndrome. It is a rare, X-linked dominant genodermatoses. The disease usually manifests in adolescence or early childhood. It is characterised by follicular atrophoderma occurring mainly on the extremities, congenital [1]hypotrichosis, localized or generalised hypohidrosis or anhidrosis and multiple basal cell carcinomas with an early onset.

## CASE REPORT:

A 20-year female patient presented to the skin OPD with complaints of multiple swellings over the face for duration of 1 week. She was apparently normal at the age of 4, after which she developed a swelling on the forehead which was then excised.Over the next few years, she developed multiple swellings over the face and trunk.The latest swelling was excised on the nape of neck 3 months back. She gave a positive family history of atopy.History of learning difficulty was also present. Examination revealed an atrophic and depigmented area with a rolled out border over the right side of the forehead marking the area of first lesion which was excised. There were multiple cystic swellings and ice-pick scars over her face. Prominent follicular ostia were also noted over the extensor aspects of the forearms and legs. Severe scaring with sinuses was noted over bilateral axillae evident of Hidradenitis suppurativa associated with marked pigmentation.

3.5mm punch biopsy was taken from lesion over right side of the forehead. Histopathological study showed islands basaloid cells within the dermis, arranged in a palisading pattern around a core of degenerated cells.

## DISCUSSION:

Follicular atrophoderma with Basal cell carcinoma or Bazex-Dupre-Christol syndrome is a rare [2]X-linked dominant genodermatoses with an early clinical manifestation varying in severity between males and females; with the condition being more severe in the male population. It has classically been reported with the [3]triad of clinical findings consisting of follicular atrophoderma, early onset basal cell epithelioma and hypotrichosis. Other features that have been reported in association with Bazex syndrome include hypohidrosis, hyperpigmentation, milia, comedones, and atopic diathesis and Keratosis pilaris. [4]Hair shaft anomalies have also been reported with Bazex syndrome. Dilated hair follicle ostia appearing as ice pick scars is characteristic of Follicular atrophoderma which in Bazex syndrome appears first over the dorsa of hands &feet, and the extensor aspects of elbows and knees.

The patient may experience a generalized reduction of sweating. The hypotrichosis that has been described usually affects the body and scalp with or without involvement of eyebrows and eyelashes. Most notably, the patient will develop multiple basal cell epitheliomas exhibiting an aggressive behaviour, by the third decade, with a predilection to occur over the face area. Hidradenitis suppurativa has been known to occur in patients with Bazex syndrome. Our patient too had signs of follicular occlusion which included Hidradenitis suppurativa of the axilla with acne conglobata. The differentials include follicular occlusion triad, Gorlin's syndrome, Rombo syndrome and Generalized basaloid follicular hamartoma. Management of the condition mainly involves early detection and treatment of the Basal cell epitheliomas with symptomatic treatment care and careful monitoring for possible complications.

## CONCLUSION:

Basex Dupre Christol syndrome or Follicular atrophoderma with basal cell carcinoma syndrome is a rare genodermatoses with very few reports in literature. To our best knowledge this happens to be the second reported case of Bazex syndrome in India[5].

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**CONFLICT OF INTEREST:** None

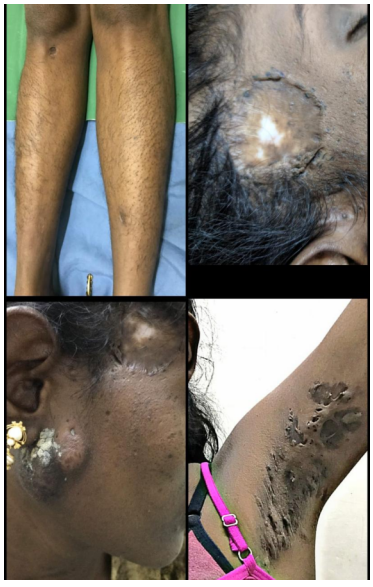

**FIGURE 1**: clinical image showing the follicular prominence of bilateral legs, hypopigmented and atrophic lesion with rolled out borders over the forehead, cystic lesions over the face and severe scarring over the axilla

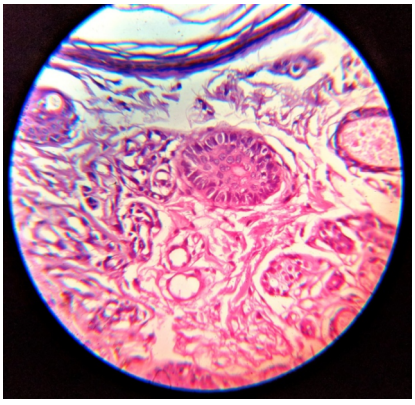

**FIGURE 2:** Histopathological examination under high power showing islands basaloid cells within the dermis, arranged in a palisading pattern around a core of degenerated cells.