

## A Case of Transverse Hand Deficiency with Acne Inversa and Facial Freckling: A New Association or Coincidence

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### Abstract

Transverse hand deficiency is a rare congenital finding in which there is abrupt failure of formation of one or multiple bones in utero. It may be associated with other skeletal abnormalities. Here we are reporting a case of transverse hand deficiency with multiple epithelial cysts, acne inversa and facial freckling. Such associations were not reported previously.

### Back ground

Acne inversa or hidradenitis suppurativa is a severe and debilitating chronic inflammatory disorder of apocrine gland bearing areas [1]. It can present with other manifestations of follicular occlusion triad like dissecting cellulitis of scalp or acne conglobate [2]. Obesity is also commonly associated with it. Familial inheritance less commonly reported in case of acne inversa.

### Case report

40 year old female presented with a swelling and pus discharge in the left side of the neck for 1 week. On further probing she gives h/o similar current swellings in her axilla, back and neck region for over 10 years which resolve with scarring. Patient also gave history of similar swellings in her sister. She gives h/o appearance of numerous tiny brown spots over her face, which was also present in her father. Patient was having congenital deformity of left hand.

On examination she had freckles over face [Figure 1]. A tender 3 x4 cm fluctuant abscess with purulent discharge was present over the left side of neck [Figure 2]. Multiple sebaceous cysts were seen over back and upper chest. Multiple open and few polyporus comedones along with scars were present over back, axilla, and chest [Figure 3]. Left sided transverse hand deficiency was noted in the form of absence of fingers [Figure 4] on radiological evaluation all carpal bones and metacarpals were present but there was complete absence of phalanges [Figure 5] Laboratory investigations showed neutrophilia and leucocytosis. The abscess was drained and she was treated with antibiotics and analgesics.



Figure 1: Freckles and melasma like pigmentation over face



Figure 2: 3x4cm fluctuant abscess over left shoulder



**Figure 3:** Multiple polyporous comedones and epithelial cysts over back



**Figure 4:** Hand deficiency - absence of fingers in left hand



**Figure 5:** Antero-posterior and lateral view of left hand showing absence of phalanges.

## Discussion

Hidradenitis suppurativa, associated with other manifestations of follicular occlusion is reported frequently in the past. Follicular occlusion tetrad is a condition that includes hidradenitis suppurativa, acne conglobata, dissecting cellulitis of the scalp and pilonidal sinus [3]. Women are more commonly affected than males. Another triad has been described by Loo et al. in the form of Dowling Degos disease, hidradenitis suppurativa and multiple epidermal cysts [4]. There are few case reports showing coexistence of flexural reticulate pigmentation with hidradenitis suppurativa, but in this case hidradenitis suppurativa and hand deficiency syndrome was associated with familial history of multiple sebaceous cysts and facial freckling. Transverse hand deficiency comes under a group of disorders of congenital hand deformity occurring because of failure to formation of certain bones of hand [5]. These are difficult to differentiate from constriction band syndromes but nail remnant may be present in constriction band syndromes. This is a unique presentation and first of its kind as per our knowledge.

## References

1. Wollina U, Koch A, Heinig B, Kittner T, Nowak A (2013) Acne inversa (hidradenitis suppurativa): A review with a focus on pathogenesis and treatment. *Indian Dermatology Online Journal* 4: 2-11.
2. Vasanth V, Chandrashekhar BS (2014) Follicular occlusion tetrad. *Indian Dermatol Online J* 5: 491.
3. Gandhi V, Verma P, Yadav P (2013) Follicular occlusion triad associated with reticulate pigmentary disorder: Is there a genetic linkage? *Indian J Dermatol* 58: 330.
4. Loo W J, Rytina E, Todd PM (2004) Hidradenitis suppurativa, Dowling–Degos and multiple epidermal cysts: a new follicular occlusion triad. *Clin Exp Dermatol* 29: 622-624.
5. Malik S, Afzal M (2013) congenital terminal transverse deformity of upper limb: clinical and radiological findings in a sporadic case. *J Coll Physicians Surg Pak* 23: 219-20.

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