Case report:

Keratosis Follicularis Spinulosa Decalvans in a female child- a rare presentation

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Abstract:

Congenital alopecia universalis is a very rare presentation. A 6 year old girl came to us with total alopecia and multiple horny keratosis pilaris like skin lesions all over the body. The alopecia was mostly non-scarring with a few patches of scarring over the scalp. Histology from scalp revealed follicular plugging with perifollicular infiltrate of lymphocytes and plasma cells. The case was diagnosed as Keratosis follicularis spinulosa decalvans. This is very rare and even rarer in females.

Keywords: keratosis pilaris; scarring alopecia; non-scarring alopecia

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Introduction: Keratosis follicularis spinulosa decalvans is a rare genodermatosis that affects predominantly males. It appears in infancy or childhood, and is characterized by diffuse follicular keratotic papules associated with progressive cicatricial alopecia of the scalp, eyebrows and eyelashes. Family history is often positive. We report a case of KFSD in a female child.

Case-report: A 6 year old girl presented with alopecia and multiple keratosis pilaris like lesions to our outdoor. Her mother complained that she did not develop any hair since birth. Eventually she developed multiple blackish and skin coloured horny lesions all over her body. Mother also informed that since last 4 years a few hairs are appearing over the scalp followed by quick shedding. On examination multiple keratotic papules with follicular plugging were present all over the body [Figure 1], including eyebrows and scalp [Figure 2]. Except a few brittle hairs on the crown area hair was absent all over the scalp, eyelids and body. There were a few small patches of non-scarring alopecia over the scalp [Figure 3].

Palms, soles, nail and mucosa were unaffected. Family history was unremarkable with no history of consanguinity. Her twin sister was unaffected. There was no history of photophobia, no evidence of physical or mental retardation. Eye examination was unremarkable. Scalp biopsy revealed multiple follicular plugging with perifollicular lymphocytes and plasma cells [Figure 4] with absence of hair. A diagnosis of keratosis follicularis spinulosa decalvans was made.

Discussion: The entity Keratosis follicularis spinulosa decalvans was described by Siemens in 1926. It is a rare X-linked disorder. It begins in infancy with keratosis pilaris localized on the face,

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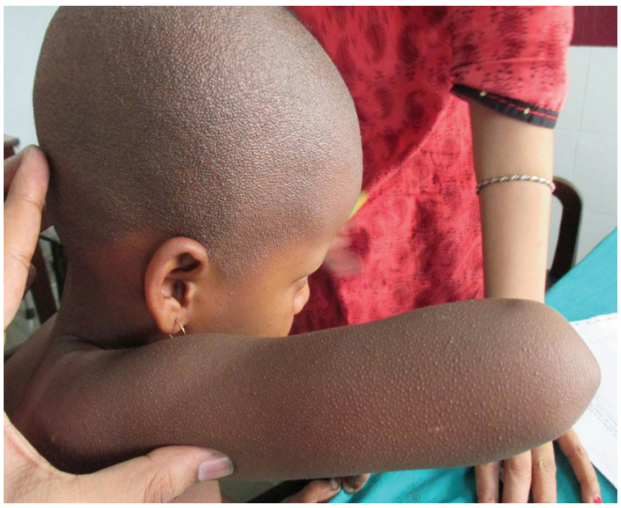

Figure.1: - multiple keratotic papules with follicular plugging were present all over the body

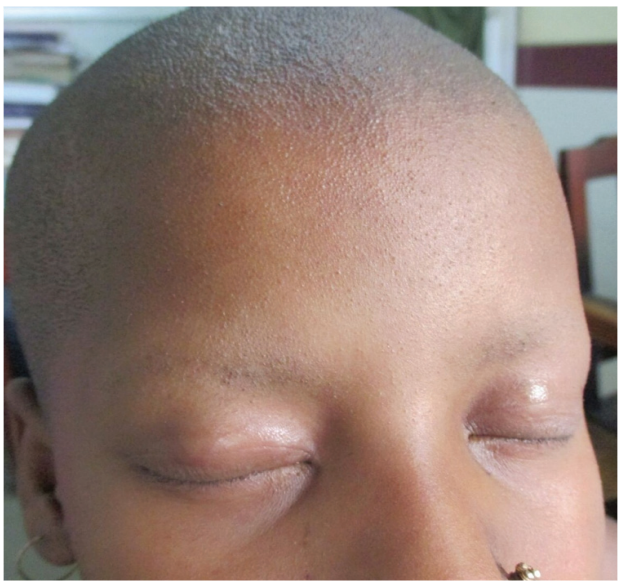

Figure.2- keratotic papules over eyebrows and scalp

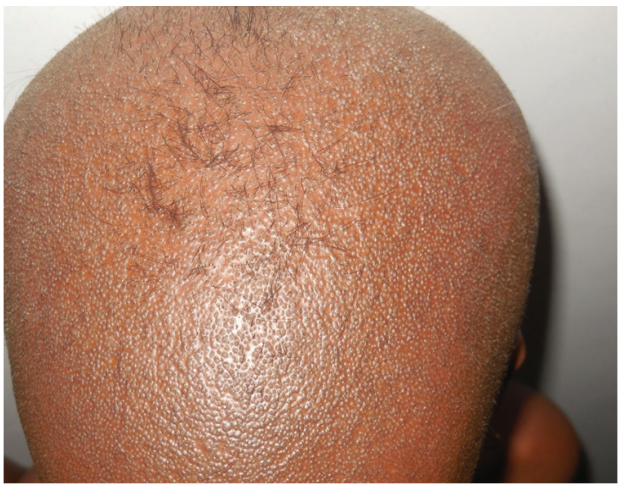

Figure.3-few small patches of non-scarring alopecia over the scalp

then evolves to more diffuse involvement. The locus of mutation in KFSD is Xp22.13–22.2.[2] But some cases may be sporadic, autosomal dominant or X-linked recessive.[3]

KFSD belongs to the family of Atrophic pilar keratosis. This comprises of pillar keratosis face atrophying, atrophodermiavermiculata and keratosis follicularis spinulosa decalvans. The basic pathogenesis of this family is follicular hyperkeratosis with inflammation followed by atrophy, the sites being different.[4]

KFSD may be associated with palmo-plantar hyperkeratosis, photophobia, corneal dystrophy and atopy. Other conditions reported to be associated with it are facial erythema, leukonychia and woolly hair.[5,6]

We kept ichthyosis follicularis alopecia photophobia (IFAP) syndrome, monilithrex, congenital atrichia with popular lesions, Graham Little PiccardiLasseur syndrome (GLPLS) and Lichen spinulosaas differentials. IFAP was ruled out due to the absence of photophobia and corneal abnormality and by the presence of scarring alopecia. Monilithrex was ruled out by finding of normal hair structure in trichoscopy. GLPLS is characterised by keratotic follicular papules over body, scarring alopecia of scalp and non scarring alopecia of axillae. It was ruled out by biopsy. Lichen spinulosa also presents with keratotic papules but they are focal and localised to acral areas. There were no milia like lesions on body nor white hypopigmented streaks on scalp. In congenital atrichia with papular lesions there is total absence of hair follicle on histology with no inflammatory cells. Follicular cysts containing keratin are found. In this patient histology showed the presence of follicular plugging with perifollicular lymphocytes and plasma cells. This confirmed the diagnosis of Keratosis follicularis spinulosa decalvans.

The treatment of KFSD is symptomatic. Topical keratolytics, emollients and Vitamin A derivatives have shown some improvement.[7]

The use of isotretinoin in the active stage can give better result.[8,9]

We started the child on oral isotretinoin 5 mg per day. After 8 weeks mother stated that the shedding of hair has stopped.

Conclusion: KFSD is a rare entity, rarely seen in females and is often a clinical challenge.

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