**INTRODUCTION**

Woolly hair is a rare congenital disorder marked by extreme kinkiness in Caucasians. The diameter of hair is 0.5 cm with normal hair growth rate, but anagen phase is truncated, so the hair does not grow long. The term woolly hair nevus was first used by Wise in April 1927.[1] Woolly hair nevus is a rare, nongenetically determined condition in which unruly and tightly curled hair is localized on one or several areas of the scalp. The abnormal hair might be slightly light in color. It is grouped under hair shaft disorder without increased fragility. We report a case of woolly hair nevus with linear epidermal nevus.

**CASE REPORT**

A 6-year-old girl born to nonconsanguineous parents presented with abnormal patch of hair since 4 years. Her parents noticed circumscribed patch of curling and coiling of hair along with change in texture over the vertex of scalp. Birth and development history were normal. Family history was negative for such condition.

On examination, there was solitary circumscribed patch of size 5 cm × 6 cm over vertex [Figure 1]. Hair over the patch had an altered texture, light in color, thinner, and tightly coiled. The hair in other areas of scalp was normal. Skin over the patch was normal. Linear epidermal nevus was present over the chin extending to the neck there [Figure 2] was no evidence of palmoplantar keratoderma and precocious puberty. Based on the above findings, we arrived at a diagnosis of woolly hair nevus.

**Investigations**

Light microscopy of hair revealed irregularities of the cuticle and twisting of hair [Figure 3]. Dermoscopy revealed snake crawl appearance and knotting of hair [Figure 4]. Diagnosis of woolly hair nevus was made.
DISCUSSION

Woolly hair was first discovered in a European family by Gossage in 1907. It can be either sporadic or genetic. Hutchinson et al classified woolly hair into (1) autosomal dominant, (2) autosomal recessive, (3) symmetrical circumscribed allotrichia, and (4) woolly hair nevus. Autosomal recessive form is associated with Naxos disease and Carvajal syndrome.

Woolly hair nevus occurs sporadically with no sex predilection. It is further classified by Pot into three types: Type 1 – Without any skin or scalp changes; Type 2 – Associated with linear verrucous epidermal nevus; and Type 3 – Acquired progressive kinking of hair. It is associated with linear verrucous epidermal nevus in more than half of the cases. In addition, it can also be associated with melanocytic nevi, café au lait macules, Mongolian spots, and precocious puberty. Ocular abnormalities include persistent pupillary membrane and retinal defects. Light microscopy shows twisted hair with reduced diameter and loss of cuticle. Electron microscopy shows triangular cross-sections with transverse grooves. These findings help differentiate woolly hair from pili torti, monolethrix or trichorrhexis nodosa, and Menkes syndrome.

Medusa head is the term given to trichoscopy findings of woolly hair nevus.

We report this rare case of woolly hair nevus because of its rare occurrence, its association with linear epidermal nevus, and crawling snake appearance in dermoscopy findings.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have
given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Wise F. Woolly hair nevus. A peculiar form of birthmark of the hair on the scalp, hitherto undescribed, with report of two cases. Med J Rec 1927;125:545-7.
2. Grant PW. A case of woolly hair naevus. Arch Dis Child 1960;35:512-4.
3. Hutchinson PE, Cairns RJ, Wells RS. Woolly hair. Clinical and general aspects. Trans St Johns Hosp Dermatol Soc 1974;60:160-77.
4. Post CF. Woolly hair nevus; report of a case. AMA Arch Derm 1958;78:488-9.
5. Thomas J, Sindhu BK, Dinesh DK, Parimalam K. Type S woolly hair nevus. Indian J Pediatr Dermatol 2014;15:27-9.
6. Venugopal V, Karthikeyan S, Gnanaraj P, Narasimhan M. Woolly hair nevus: A rare entity. Int J Trichology 2012;4:42-3.
7. Goldin HM, Bronson DM, Fretzin DF. Woolly-hair nevus: A case report and study by scanning electron microscopy. Pediatr Dermatol 1984;2:41-4.