Sir,
An 8-year-old girl child was brought to the outpatient department with complaints of rough, coarse, and easily breakable hair never growing beyond a length [Figure 1]. She was school going and had no history or symptoms suggestive of physical or mental developmental delay or abnormality.

On examination, hair of the entire scalp was of different lengths, was standing out, and was brittle. Along the scalp margin, follicular keratotic papules suggestive of keratosis pilaris were seen [Figure 2]. Hair of the eyebrows was sparse and of the same nature as the scalp hair [Figure 3]. Follicular keratotic papules as those over the scalp margins were seen over the upper and lower extremities too. Nail plates were thinned out. Teeth were normal. There was a large nevus achromicus extending from the right jaw to lower down the neck [Figure 4]. Her height and weight for age were normal.

The child was investigated. Light microscopy of hair shaft was consistent with trichorrhexis nodosa [Figure 5]. Trichoscopy of scalp and eyebrows revealed both normal and curled hair of varying thickness with normal color [Figure 6] which is nonspecific for trichorrhexis nodosa. Skin biopsy of scalp and eyebrows was deferred as:
1. The patient had a hair shaft anomaly easily diagnosable with light microscopy
2. The patient was a child and an invasive procedure of lesser diagnostic value was best not done
3. The procedure would invariably leave a scar on the eyebrow and scalp.

Iron deficiency and hypothyroidism were ruled out. Liver and renal function tests and urine analysis were normal. Quantitative analysis showed no argininosuccinic aciduria. Hair microscopy of parents, younger brother, maternal, and paternal grandparents was unremarkable.

A course of biotin 10 mg with other essential micronutrients daily for 3 months showed no improvement.

The child was thus diagnosed to have isolated primary congenital trichorrhexis nodosa with nevus achromicus. Trichorrhexis nodosa is an acquired or congenital hair shaft disorder where the hair is rough, dry, coarse, and easily breakable.[1] It is acquired when there is extreme or continuous physical and chemical damage to the hair shaft.[1] When congenital, it is autosomal dominant and associated with aminoacidurias and mental retardation.[2]

It has also been reported in association with biotinidase deficiency, underlying systemic illnesses, endocrinopathies such as hypothyroidism, malnutrition, especially iron deficiency anemia and ectodermal dysplasias.[2] It has also been reported with Netherton syndrome.[3]

Hair microscopy of trichorrhexis nodosa shows nodes in the hair shaft where the hair splits, giving a brush-like appearance.[1,5] The hair shafts of our patient showed nodes going in for splitting.

When congenital, it is commonly associated with argininosuccinic aciduria leading to mental retardation and developmental abnormalities.[1,6] The child did not have any growth and developmental abnormalities suggestive of malnutrition, aminoacidurias, endocrinopathies, or any systemic illnesses. Investigatory evidence was also in favor of the above.

The absence of improvement with biotin supplementation ruled out biotinidase deficiency.

Trichoscopy of trichorrhexis nodosa, according to literature, shows hair shafts that are broken or breaking off and other signs of damage if, of acquired type.[1,5] Trichoscopy in our patient showed abnormally curled up hair shaft not completely out which has not been reported until now.

It is felt that the occurrence of nevus achromicus with TN could be coincidental. Reports of similar cases, if any, in future may prove or disprove an association.

Hence, we report this case of primary congenital trichorrhexis nodosa alone with neither family history nor underlying associated conditions, with a nevus achromicus.
To the best of our knowledge, such a case has not been reported in the literature until now.

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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.

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**Conflicts of interest**

There are no conflicts of interest.

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