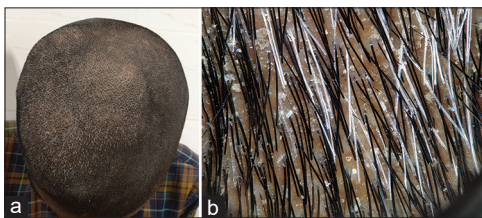


## Circumscribed Area of Leukotrichia over Plexiform Neurofibroma: An Important Clue or Mere Coincidence?

Sir,  
Plexiform neurofibroma is an uncommon cutaneous lesion of neurofibromatosis type 1. It usually presents as boggy swelling predominantly over craniofacial region.<sup>[1]</sup> When plexiform neurofibroma occurs in isolation it can possess a diagnostic challenge. Overlying hyperpigmentation, hypertrichosis, café-a-lait macules sometimes give important clue to the diagnosis of underlying plexiform neurofibroma [Table 1]. Rarely leukotrichia can be present over the surface. We present a case of plexiform neurofibroma over scalp with circumscribed area of leukotrichia over the lesion.

A 19-year-old male presented with a swelling over right fronto-parietal scalp since 4 years. It was progressively increasing in size, not associated with any pain or itching. He also noticed progressive greying of hairs over the lesion for last 3 years. Swelling was 12.5 cm × 10 cm in size, soft to firm, boggy on palpation [Figure 1a]. There were significant number of white hairs admixed with normal pigmented hairs over the swelling. The extent of leukotrichia was limited to the margin of the lesion. Dermatoscopy of scalp showed mixture of white and black hair [Figure 1b]. He



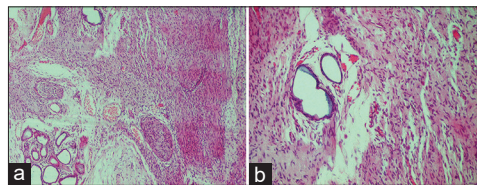
**Figure 1:** (a) Ill-defined swelling over fronto-parietal scalp with partial leukotrichia limited to the lesion. (b) Dermatoscopy picture (10x) showing white hairs admixed with normo-pigmented hairs over scalp

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had one similar swelling over left ankle, multiple skin colored nodules over back and freckling over bilateral palm. He had positive family history of similar lesions in his younger sibling. Histopathology from incisional biopsy specimen from the lesion showed interlacing bundles of elongated spindle cells with wavy dark nuclei in deep dermis with small amount of mucin separating the cells [Figure 2a and b]. The histopathological feature was suggestive of plexiform neurofibroma.

Plexiform neurofibromas are benign tumors that originate from nerve sheath cells of peripheral nerves and usually involve multiple fascicles. They are uncommon and occur almost exclusively in association with neurofibromatosis-I, in about 30% patients.<sup>[1]</sup> Well-defined localized patch of white hair or “poliosis circumscripta” classically is known to occur in the setting of several genetic syndromes including piebaldism, Waardenburg syndrome, tuberous sclerosis. In those cases, lesions are usually seen at median or paramedian location, commonly known as “white fore locks”.<sup>[2]</sup> Leukotrichia associated with neurofibromatosis is rare. To our knowledge five cases have been reported with such finding.<sup>[3]</sup> Among other lesions, congenital melanocytic nevus over scalp, blue nevus has also been found to be associated with



**Figure 2:** (a) Shows dermis with interlacing bundles of elongated spindle cells with wavy dark nuclei (H and E ×50). (b) Shows spindle cells with wavy serpentine nuclei around the adnexal structure with small amount of mucin separating the cells (H and E ×125)

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**Table 1: Cutaneous lesions over the skin of an underlying plexiform neurofibroma**

Author, year	Site of plexiform neurofibroma	Number of cases	Overlying skin changes
Neri I, 2017 <sup>[3]</sup>	Scalp	Two	Leukotrichia
Ettl A <i>et al.</i> , 1996 <sup>[6]</sup>	Face	Two	Hyperpigmentation and hypertrichosis
Khandpur <i>et al.</i> <sup>[7]</sup>	Back	Multiple	Hypopigmented macules
Choudhary SV <i>et al.</i> , 2017 <sup>[8]</sup>	Right shoulder to upper three fourth of arm	Single	Café-au-lait macule
Singh <i>et al.</i> , 2018 <sup>[9]</sup>	Back	One	Becker's nevus
Schaffer JV <i>et al.</i> <sup>[10]</sup>	Trunk	One	Hyperpigmentation and hypertrichosis
Khachemoune A <i>et al.</i> <sup>[11]</sup>	Thigh	Single	Café-au-lait macule

leukotrichia rarely.<sup>[4,5]</sup> The hypothesis behind leukotrichia has not been completely understood, it is probably due to an autoimmune response in which the T cells cytotoxic to neurofibroma might cross-react with surrounding hair bulb melanocytes, resulting in their destruction with subsequent whitish discoloration of the hair.<sup>[5]</sup> It is important to know this association as it can give an important clue in diagnosis of underlying plexiform neurofibroma.

### **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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