Nevus depigmentosus with unilateral bluish sclera, a rare entity

Sir,

An 8-year-old boy presented with an asymptomatic depigmented patch on the right side of forehead since birth that was slowly increasing in size along with the growth of the child. There was no history of seizures, hearing defects, or growth retardation, and family history was not contributory. Developmental milestones of the child and mental IQ were normal. On examination, there was a segmental depigmented patch with serrated margin on the right side of the forehead. There was also unilateral bluish discoloration of the sclera, strabismus of left eye [Figure 1], circumscribed poliosis [Figure 2]. Systemic examination of the central nervous system, cardiovascular system, and musculoskeletal system revealed no abnormality. On wood lamps examination, there was an off-white accentuation without fluorescence. Upon diascopy there was no blending of the lesional skin colour with that of the surrounding skin. The parents were explained about the treatment options such as excimer laser, autologous epidermal grafting and suction blister grafting along with their merits and demerits.

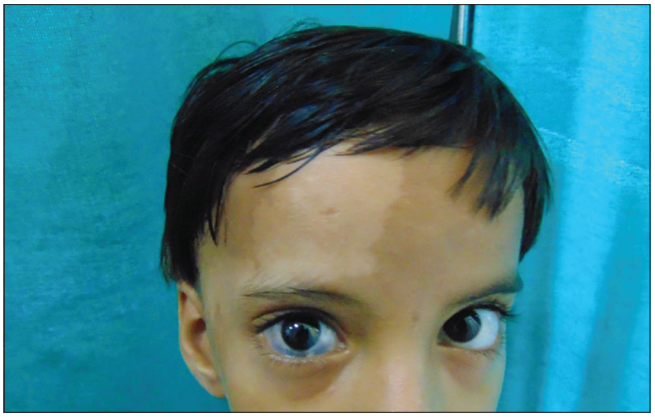

Figure 1: Nevus depigmentosus with unilateral bluish sclera

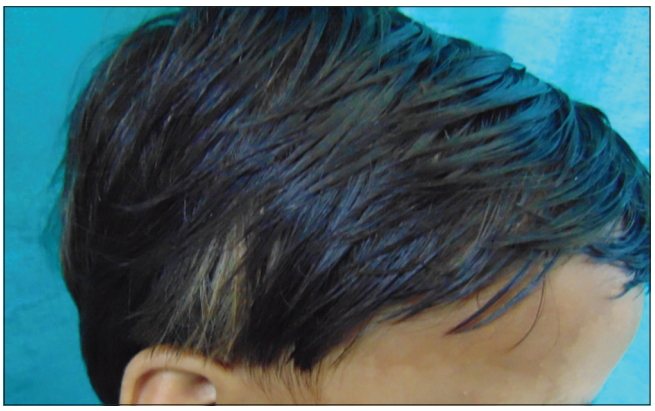

Figure 2: Poliosis

DISCUSSION

The condition was first described in 1884 by Lesser.[1] In the majority of cases of nevus depigmentosus (ND), the etiology is not known. Presumably, ND results from functional abnormalities of melanocytes. ND presents as a congenital condition or shortly thereafter leukodermic macule with regular or serrated margin that does not cross the midline and can be isolated, segmental, or generalized in distribution.

The commonly used clinical diagnostic criteria for ND are as follows:[2-4]

- Leukoderma present at birth or of an early onset
- No alteration in distribution of leukoderma throughout life
- No alteration in texture or change of sensation in the affected area
- Absence of hyperpigmented border

ND has to be differentiated from nevus anemicus, hypomelanosis of ito, vitiligo, albinism. Vitiligo is characterized by depigmented macules as a result of loss of functional cutaneous melanocytes. Lesions often show homogenous depigmentation and are well demarcated. Vitiligo lesions are well accentuated with obvious fluorescence upon Wood lamp examination. Nevus anemicus can be differentiated by diascopy where the lesion blends with the surrounding skin.

ND may have neurological, eye, hair, musculoskeletal, or internal organ involvement. The treatment is difficult and options such as 308 nm excimer laser, epidermal grafts, and suction blister grafting has been tried.

Bluish sclera, mostly bilateral, may be associated with Marfan's syndrome, Ehler Danlos syndrome, myasthenia gravis, iron deficiency anemia, blue sclera syndrome (Van der Heave syndrome), chromosomal disorders such as Turner syndrome and trisomy, osteogenesis imperfecta, Goltz syndrome, incontinenta pigmenti and minocycline induced pigmentation. Poliosis can be associated with piebaldism, Waardenburg syndrome, Alezzandrini syndrome, vitiligo, and tuberous sclerosis. We report this case as ND with unilateral bluish sclera and poliosis is a rare presentation.

Abhijeet Kumar Jha, Rajesh Sinha, Smita Prasad

Departments of Dermatology, STD and Leprosy,
All India Institute of Medical Sciences,
Patna, Bihar, India

Address for correspondence:
Dr. Abhijeet Kumar Jha,
Departments of Dermatology, STD and Leprosy,
All India Institute of Medical Sciences, Patna, Bihar, India.
E-mail: drabhijeetjha@gmail.com

REFERENCES

1. Lesser E. Anomalien der Hautfärbung. In: von Zeimssen H, editor. Handbuch der Hautkrankheiten. Leipzig, Germany: FCW Vogel; 1884. p. 161-200.
2. Shim JH, Seo SJ, Song KY, Hong CK. Development of multiple pigmented nevi within segmental nevus depigmentosus. J Korean Med Sci 2002;17:133-6.
3. Bolognia JL, Lazova R, Watsky K. The development of lentigines within segmental achromic nevi. J Am Acad Dermatol 1998;39:330-3.
4. Khumalo NP, Huson S, Burge S. Development of lentigines within naevoid hypopigmentation. Br J Dermatol 2001;144:188-9.

Access this article online

Quick Response Code:

Website: www.idoj.in

DOI: 10.4103/2229-5178.164491