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A Rare Case of Multiple Nevoid Hypertrichosis with Atrial Septal Defect

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Dear Editor:

An 18-month-old female was evaluated for localized hypertrichosis on the left infraorbital area, left upper back, and dorsal aspect of the left leg (Fig. 1). The lesions were small at birth and had enlarged as she grew up. There was no relevant family history. The patient was born by cesarean section at 37 weeks and admitted to the neonatal intensive care unit after birth with a diagnosis of respiratory distress syndrome. Ultrasonography revealed an atrial septal defect that disappeared after 1 year. The patient had normal developmental milestones. A physical examination revealed localized hypertrichosis on the left infraorbital

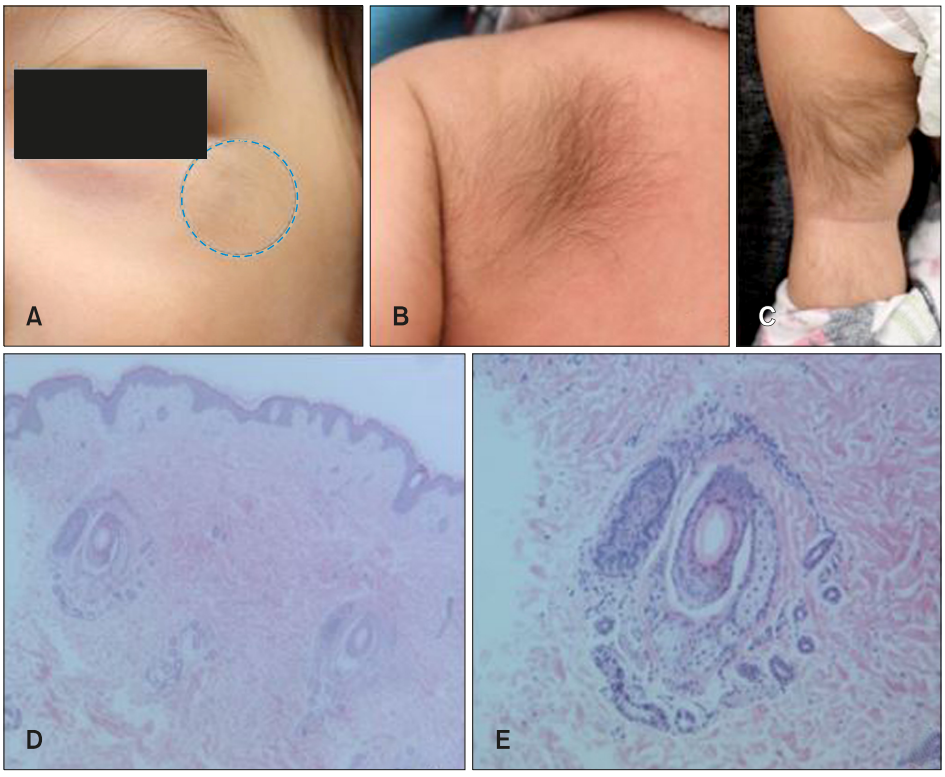

Fig. 1. (A) Localized hypertrichosis on the left periocular area (dotted circle), (B) left upper back, and (C) dorsal aspect of the left leg. Black and coarse terminal hairs about 2 to 3 cm in length were present on the back and left leg. Slight hypopigmented patches were seen on the left leg. (D) Histology revealed mild acanthosis on the epidermis and mild perivascular inflammation on the upper dermis with terminal hair follicles (H&E, ×40). (E) The same findings are shown in a magnified view (H&E, ×100). We received the patient's consent form about publishing all photographic materials.

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Table 1. Reported cases of multiple nevoid hypertrichosis

Author (year)	Sex/age (mo)	Onset	Ethnicity	Location	Associated disease	Family history	Treat-ment	Prognosis
Cox et al. (1989)[5]	Female/6	At birth	Caucasian	Upper lips, both scapulae, upper arms, buttocks, Rt. lumber region, Rt. upper thigh	Lipodystrophy	NS	None	Unknown
Rogers (1991)[6]	Female/14	At birth	Asian	Trunk, both extremities	Lipodystrophy, hypomelanosis of Ito congenital malrotation of the gut diaphragmatic hernia, focal iris colobomata, congenital lung cyst, polydactyly, partial anodontia, malalignment of some of her teeth	NS	None	Unknown
Rupert et al. (1994)[7]	Female/23	At birth	Caucasian	Rt. clavicle, Rt. shoulder, Rt. upper arm, buttock both proximal thigh	NS	NS	None	Unknown
Ballmer-Webet et al. (1996)[8]	Female/16	At birth	Caucasian	Genitalia, both shins	Hypomelanosis of Ito, follicular keratosis, dysmorphic face, salmon patch, dysplastic teeth, bilateral genu vara, pes valgus, hypoplasia of Lt. buttock, bilateral hip dislocation	NS	None	Unknown
Lestringant et al. (1997)[9]	Female/21	At birth	African	Both cheeks, back, both extremities	Hypomelanosis of Ito, dysmorphic face, digital anomalies, mental retardation, partial absence of corpus callosum	NS	None	Unknown
Chang et al. (1997)[4]	Male/21	1 yr	Asian	Chest, Lt. shoulder, Lt. upper extremity	Depigmented skin	NS	None	Remained stable for 3 yr
Dudding et al. (1998)[1]	Female/ at birth	At birth	Asian	Rt. shoulder, Rt. upper arm, Lt. axilla both buttocks and thighs	Hypomelanosis of Ito, epidermal nevus, alopecia, retinal hyperpigmentation	NS	None	Complete resolution after 2 yr
López-Barrantes et al. (2002)[10]	Female/2	At birth	Caucasian	Rt. trunk, Lt. arm, both legs	Hypomelanosis of Ito	NS	None	Unknown
Sotiriadis et al. (2009)[2]	Female/3	5 mo	Caucasian	Lt. scalp, lumbosacral lesion, both extremities	NS	NS	None	Remained stable for 2 yr
Khurana et al. (2014)[3]	Female/3	At birth	Asian	Both extremities	Hypomelanosis of Ito, nail dystrophy	NS	None	Unknown
Our case	Female/18	At birth	Asian	Lt. infraorbital area, Lt. upper back, Lt. leg	Hypopigmentation, atrial septal defect, clinodactyly	NS	None	Unknown

Rt.: right, Lt.: left, NS: not significant.

area, left upper back, and dorsal aspect of the left leg. Black and coarse terminal hairs 2 to 3 cm in length were present on the patient's back and left leg. Hypopigmented patches were observed on the left leg, but no other skin changes or tumorous lesions were observed. Clinodactyly was noted on the patient's fifth finger. A skin biopsy was performed on the patient's left upper back; histology revealed mild acanthosis and mild folliculocentric inflammation in the upper dermis with terminal hair follicles (Fig. 1). After the biopsy, the patient visited two more times but did not receive any treatment; she has since been lost to follow-up.

Nevoid hypertrichosis is a rare congenital disorder with extraordinary terminal hair growth on normally pigmented skin[1]. The involved hair may be hypopigmented with a rough texture that usually appears as a single lesion and rarely as multiple lesions[2]. Multiple nevoid hypertrichosis refers to nevoid hypertrichosis that occurs at multiple areas. It is predominant in females and presents at or soon after birth[1,3]. The cause is unclear, but concurrence of nevoid hypertrichosis in Gorlin syndrome and Aicardi syndrome resulted in possible relationship with *PTCH* gene and X chromosome, which is not clarified yet. Histopathologically, there is no characteristic feature of the disease other than the presence of terminal hairs, as seen in our case. Smooth muscle hamartoma, Becker nevus, and melanocytic nevi which can be presented as focal hypertrichosis should be differentiated and all of them are benign. Ten cases have been reported and some of them were related to accompanying abnormalities (Table 1)[1-10]. It was significant that this case showed concurrent congenital atrial septal defect which was not reported in previous report in addition to cutaneous manifestation and skeletal abnormality[1-4,11]. Moreover, it is noteworthy that majority of affected cases, including our case, have been female; this could be more than a chance association.

In conclusion, the etiology of multiple nevoid hypertrichosis is unknown, but our experience of a case of multiple nevoid hypertrichosis with other cutaneous and systemic findings (especially atrial septal defect) could clarify its origin. Herein, we report a rare case of multiple nevoid hypertrichosis with a literature review.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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