Three Different Epidermal Naevi With No Organ Involvement: Sebaceous Naevus, Naevus Comedonicus and Becker's Naevus

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Sir,

Epidermal naevi (EN) are organoid naevi arising from the pluripotent germinative cells in the basal layer of the embryonic epidermis. Their location is variable, following Blaschko lines, and reflecting embryonic migration patterns of the skin (1). EN have been classified according to their predominant component, but in some naevi, the predominant tissue may vary with the evolution of the lesion, and different areas of the same lesion may show a variety of components at the same time (2, 3). This report describes a 22-year-old man with 3 different epidermal naevi (naevus sebaceous (NS), Becker's naevus and naevus comedonicus) with no organ involvement.

CASE REPORT

A 22-year-old man was admitted to outpatient clinic with some lesions on his neck and back. An ivory-coloured verrucous lesion on his neck was first noticed at 3 years of age. Some comedone-like lesions appeared on his back at 6 years of age. At 12 years of age, a giant, pigmented area, spreading to his left arm, shoulder and scapular area, emerged. He emphasized that the lesion became hairy after 2–3 years. There was no developmental problem or history of seizures. Three different EN lesions on the patient's back were noticed during dermatological examination. A giant, hairy, pigmented, dark-brown lesion spreading to the left shoulder, lateral upper extremity and scapular area was observed (Fig. 1). Brown, 1–5 mm, papular lesions with a verrucous surface and arciform shape were noted on the neck (Fig. 2). In addition, several pin-point size comedones, some of them with central plugs, were found in a 3–4 cm^2 area (Fig. 3). These lesions were localized on the right-hand side of the spine. Physical examination was normal except for the dermatological findings. Neurological examination, roentgenograms, laboratory studies, including a complete blood cell count, blood chemistries and urine analysis were normal. Histopathological examination of the papular lesions in NS revealed acanthotic and papillomatous epidermal hyperplasia. Sebaceous glands were increased in number and situated abnormally high in the dermis. Small, immature hair follicles were situated close to the sebaceous glands. Histopathological examination of the naevus comedonicus showed epidermal invagination with moderate atrophy. The invaginated wall showed acanthosis. We suggested various therapeutic options, but the patient declined treatment.

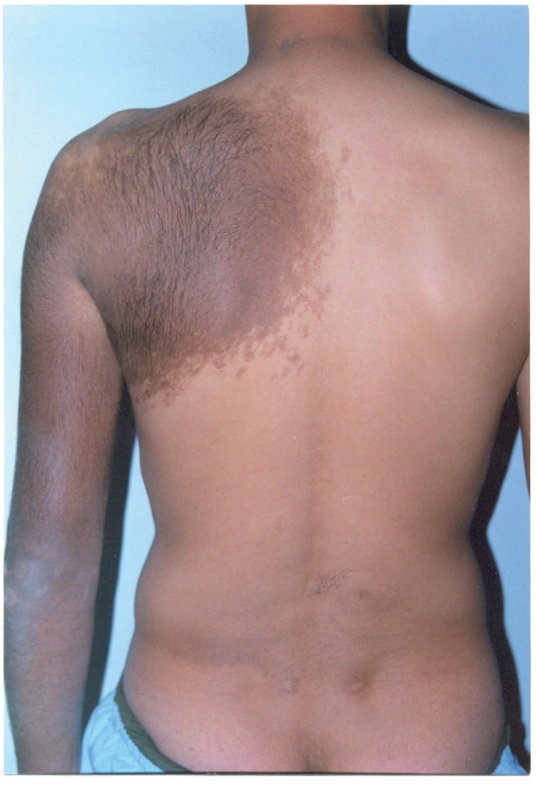

Fig. 1. Giant, hairy, pigmented, dark-brown lesion on the left shoulder, lateral upper extremity and scapular area. Becker's naevus.

DISCUSSION

The incidence of EN is approximately 1 in 1000 live newborn (1, 2). The term epidermal naevus syndrome (ENS) refers to the association of EN with abnormalities in other organ systems (3). Central nervous system, skeletal system, eyes, and oral cavity related abnormalities were reported in most. Some patients have cutaneous lesions other than EN (1–3). We did not detect any other systemic abnormality in our patient.

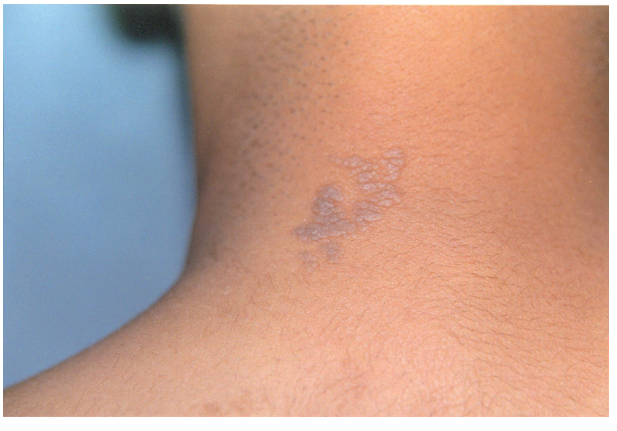

Fig. 2. Brown, 1–5 mm papular lesions with a verrucous surface in an arciform shape on the neck. Naevus sebaceous.

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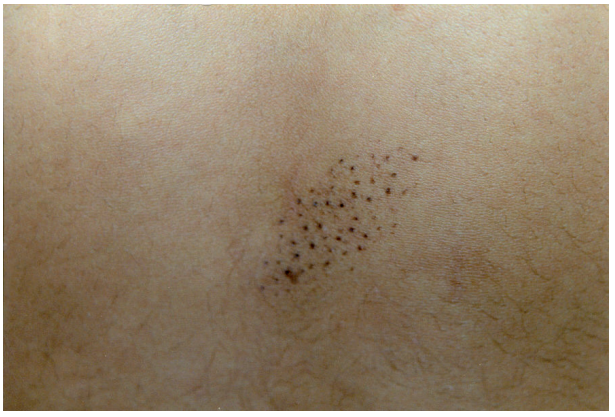

Fig. 3. Several pin-point size comedones, some with central plugs. Naevus comedonicus.

EN may involve any area of the body surface. The most commonly involved sites are the head and neck. In the present case, NS and Becker's naevus were localized on the neck and back. Naevus comedonicus was localized on the lumbosacral region. EN are often present at birth and may develop during childhood. NS and naevus comedonicus appeared first at 6 years of age in our case and Becker's naevus was observed at 12 years of age.

NS is a type of organoid naevus, characterized by yellowish-brown, raised, hairless, plaque-type lesions with a verrucous surface showing prominent proliferation of sebaceous glands and usually involving the scalp, neck and face. It is usually recognized in infancy and during childhood. At puberty, NS often thickens and develops under hormonal influences (4).

Becker's naevus, also known as "pigmented hairy epidermal naevus", is a rare disorder characterized by circumscribed light- and dark-brown hyperpigmentation with an irregular outline and hypertrichosis. Becker's naevus often localizes on the trunk or back, although it can be observed in other regions of the body. The pathogenesis of Becker's naevus is not clear. Some authors suggested that Becker's naevus is a type of hamartoma derived from the ectodermal and mesodermal tissue. The dermal component of Becker's naevus consists of numerous bundles of smooth muscle fibres unrelated to hair follicles (5). In addition, smooth muscle hamartoma is congenital, whereas Becker's naevus has its onset later in childhood and in adolescence (6). Becker's naevus can therefore be categorized as a particular type of epidermal naevus.

Naevus comedonicus is a rare, sporadic epidermal naevus, characterized by group of enlarged follicular openings with corneal plugs resembling comedones. Plugs of the comedones are removable or adherent and protrude from the skin, giving it a rough texture (1, 7). The most frequent sites of involvement are the face, chest, trunk and abdomen. Distribution of the lesions follows Blaschko lines. Naevus comedonicus may originate either through mosaicism or through influences on the juxtaepidermal mesenchymal tissue during embryogenesis. (7).

Cruz et al. (1) evaluated 443 patients with EN. They found NS in 168 (38%) patients, Becker's naevus in 31 (7%) patients, and naevus comedonicus in 5 (1%) patients. They did not detect 2 or more EN together in their series. Rogers et al. (2) reviewed 131 EN cases and found one or more systemic abnormalities with EN in 33% patients.

A variety of treatment modalities has been used with varying success. Small EN are best removed by simple excision. Cryosurgery has been attempted, with good results in some patients. Topical or intra-lesional glucocorticoids are ineffective. Some systemic drugs, such as methotrexate and etretinate, have been used successfully in resistant cases (1–3).

EN are caused by mutations that are expressed clinically as epidermal mosaicism. Mosaicism is also present in the mesodermal layer derivates, giving rise to complex syndromes. All 3 naevi in this case can be categorized as a particular type of organoid epidermal naevus and, therefore, the disorder most likely reflects mosaicism.

On the other hand, Becker's naevus appears at puberty and is correlated with an increase in both the androgen receptor and androgen receptor mRNA levels (8). NS is an androgen-sensitive tumour and may increase in size in infancy, as do the sebaceous glands, but subsequently regresses until puberty. Androgen receptor positivity is shown in sebaceous glands and eccrine glands in NS (9).

It has been suggested that both Becker's naevus and NS are stimulated by androgens. We can speculate that an increase in androgen receptor mRNA expression may result in an increase in androgen receptor proteins. Eventually, some epidermal naevi, such as Becker's naevus and NS, may be considered as a component in the spectrum of the androgen receptor hypersensitivity syndromes. There is no data about androgen receptor or androgen receptor mRNA levels in naevus comedonicus. However, naevus comedonicus shows acneiform eruption clinically and histopathologically and it can be suggested that androgen stimulation could promote their growth. Accordingly, all 3 types of EN can be accepted as androgen-stimulated naevi.

REFERENCES

1. Vidaurri-de la Cruz H, Tamayo-Sanchez L, Duran-McKinster C, de la Luz Orozco-Covarrubias M, Ruiz-Maldonado R. Epidermal nevus syndromes: clinical findings in 35 patients. Pediatr Dermatol 2004; 21: 432–439.
2. Rogers M, McCrossin I, Commens C. Epidermal nevi and the epidermal nevus syndrome. J Am Acad Dermatol 1989; 20: 476–488.
3. Hodge JA, Ray MC, Flynn KJ. The epidermal nevus syndrome. Int J Dermatol 1991; 30: 91–98.
4. Perez-Munoz MA, Hernandez Garcia MJ, Camacho F. Sebaceous naevi: a clinicopathological study. J Eur Acad Dermatol Venereol 2002; 16: 319–324.

5. Happle R, Koopman RJJ. Becker nevus syndrome. Am J Med Gene 1997; 68: 357–361.
6. Danarti R, König A, Salhi A. Becker's nevus syndrome revisited. J Am Acad Dermatol 2004; 51: 965–969.
7. Cestari TF, Rubim M, Valentini BC. Nevus comedonicus: case report and brief review of the literature. Pediatr Dermatol 1991; 8: 300–305.
8. Nirde P, Dereure O, Belon C, Lumbroso S, Guilhou JJ, Sultan C. The association of Becker nevus with hypersensitivity to androgens. Arch Dermatol 1999; 135: 12–14.
9. Hamilton KS, Johnson S, Smoller BR. The role of androgen receptors in the clinical course of nevus sebaceous of Jadassohn. Mod Pathol 2001; 14: 539–542.
10. Person JR, Bentkover S, Longcope C. Androgen receptors are increased in nevus sebaceous. J Am Acad Dermatol 1986; 15: 120–122.