Generalized benign acanthosis nigricans in an infant

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ABSTRACT

The generalized form of acanthosis nigricans, especially in infants, is extremely rare. Herein we report a 1-year-old female child who developed generalized acanthosis nigricans without any evidence of internal malignancy or endocrine disorder. This case is being reported for its rarity.

Key words: Benign acanthosis nigricans, generalized type, infant

INTRODUCTION

Acanthosis nigricans (AN) is characterized by symmetric, hyperpigmented, velvety thickening of the skin predominantly over the flexors. The word AN (acantho, Greek for thorn and nigricans, Latin for becoming black) was first proposed by Unna in 1890.[1] AN is usually benign in children, and the generalized form is very rare in this age group. Malignant AN secondary to an internal malignancy is seen over 40 years of age. It is of sudden onset and is rapidly progressive. Lesions are more severe and extensive and may be accompanied by skin tags, multiple seborrheic keratoses or tripe palms.[2]

CASE REPORT

A 1-year-old female child born of non-consanguineous marriage with normal growth and developmental milestones presented to us with progressive generalized darkening of the skin predominantly over the flexors. Her mother had first noticed the darkening around the axillae and groin at about 2 months of age. The darkening gradually progressed to involve the other body parts, including the palms and soles. The skin progressively became more hyperpigmented and thickened over the flexors. There was no family history of similar disease. There was no history of drug intake, polyuria, polydipsia and weight loss.

Cutaneous examination revealed generalized hyperpigmentation and thickening excluding the face [Figure 1]. The skin was more hyperpigmented, thickened, velvety and rugosed over the neck, axillae, cubital fossae, wrists, groin, buttocks, knees and ankles. She also had tripe palms and plantar keratoderma [Figure 2]. The mucosae, nails, hair and scalp were free of any lesions. Systemic examination was uneventful.

Routine laboratory tests including serum biochemistry panel, urinalysis, thyroid profile, growth hormone, insulin level and oral glucose tolerance test were within normal limits. Chest X-ray and ultrasonography of the abdomen revealed no abnormality.

Histopathology of the skin revealed hyperkeratosis, papillomatosis and mild acanthosis. The dermal papillae showed upward finger-like projections and the valleys between the papillae were filled with keratotic material. The dermis and the subcutis were absolutely normal [Figure 3]. Based on the clinical and histological findings, a diagnosis of “generalised benign acanthosis nigricans” was made. The infant was prescribed emollients, topical retinoids and urea-containing keratolytics.

DISCUSSION

AN can be broadly classified into type I, type II and type III.[3]

Type I or malignant AN may either precede, accompany or follow internal cancer. Most cases are associated with adenocarcinoma of the
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Gastrointestinal tract, lung and breast. Age of onset more than 40 years, sudden onset, rapid progression, mucosal involvement and tripe palms are clinical indicators of malignancy. Tripe palms (acanthosis palmaris) are characterized by thickened velvety palms with prominent dermatoglyphics.

Type II or familial AN is inherited in an autosomal-dominant manner, which presents at birth or may develop during childhood.

Type III or AN associated with obesity, insulin-resistant states and endocrinopathy is the most common type. Various endocrinopathies include diabetes mellitus, acromegaly, Addison’s disease, Cushing syndrome, hypothyroidism, Stein-Leventhal syndrome and hypogonadal syndrome. Insulin-resistant states are lipoatrophic diabetes, leprechaunism, pinealoma and acral hypertrophy syndrome-type A and type B syndrome.

Syndromic AN is associated with various congenital syndromes like Bloom syndrome, Crouzon disease, Rud syndrome, Prader-Willi syndrome, ataxia telangiectasia, Costello syndrome, Mental Retardation, Overgrowth, Remarkable Face, Acanthosis Nigricans syndrome (MORFAN) and Wilson’s disease.

Drugs causing AN are nicotinic acid, diethyl stilbesterol, testosterone, oral contraceptives, systemic corticosteroids and fusidic acid.

AN is likely caused by factors that stimulate epidermal keratinocyte and dermal fibroblast proliferation. In the benign form, the factor is probably insulin or an insulin-like growth factor that incites the epidermal cell propagation. Other proposed mediators include other tyrosine kinase receptors, epidermal growth factor receptor or fibroblast growth factor receptor. In malignant AN, the stimulating factor is hypothesized to be a substance secreted either by the tumor or in response to the tumor. Transforming growth factor-alfa is structurally similar to epidermal growth factor and is a likely candidate.\textsuperscript{[4]}

Differential diagnoses considered in our case were Addison’s disease, hemochromatosis and epidermolytic hyperkeratosis (EHK). Absence of systemic symptoms and mucosal involvement ruled out Addison’s disease and hemochromatosis. Absence of bullous lesions and typical histopathology of AN ruled out EHK.

Consistent with our case, histopathology of acanthosis nigricans (AN) reveals hyperkeratosis, papillomatosis but only slight, irregular acanthosis and usually no hyperpigmentation.\textsuperscript{[5]}

Treatment is directed towards the etiology. Weight reduction, withdrawal of offending drugs, treatment of endocrinopathies and removal of underlying tumor may lead to improvement. Keratolytics including retinoid, calcipotriol, oral retinoids, CO\textsubscript{2}
laser and long-pulsed alexandrite laser may be successful in individual cases.

In conclusion, the generalized form and palmoplantar involvement, although indicators of malignant AN, can represent benign AN also, especially in the childhood period. The rarity of documentation of its occurrence in the literature prompted this report.

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