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LETTERS TO THE EDITOR

Generalized Familial Acanthosis Nigricans Associated With Hypochondroplasia

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To the Editor:

Acanthosis nigricans (AN) is a proliferative epidermal disorder expressed clinically in the form of velvety brown plaques in skin folds, although other parts of the skin and mucosa can occasionally be affected.

New associations and peculiar clinical forms have been added to classifications of the disease since Pollitzer[1] and Janovsky[2] first reported cases of AN associated with neoplasia. There are currently 8 accepted types of AN, listed by Schwartz[3] as: benign, associated with obesity (pseudoacanthosis nigricans), syndromic, paraneoplastic, acral, unilateral, drug-induced, and mixed. Generalized AN is not considered to be a specific form, but rather an extensive manifestation of other forms of the disease. The condition is mostly associated with internal malignancy in adults, but benign generalized cases have been described in childhood.[4-6]

We present a case of generalized benign familial AN associated with small stature resulting from hypochondroplasia. The patient was a 33-year-old woman who consulted for a check-up of a nevus. Examination revealed brown velvety skin coloration in the central thoracic region, on the sides of the abdomen (Figure 1), the middle of the back, the neck, and armpits. These asymptomatic lesions had been present since infancy, increasing in number until puberty and remaining stable ever since. She reported that 3 of her sisters, a niece and her oldest child have similar lesions, as probably did her late father. We had the opportunity to confirm this affirmation in 1 sister and her niece. None of them reported any illness or habitual use of medication.

We observed that all 3 patients examined presented small stature, with short limbs. The patient reported that all except 1 of her siblings were of small stature, as was 1 of her children. Radiological examinations of the patient revealed shortened long bones diagnostic of hypochondroplasia and similar results were seen in her son.

Skin biopsy revealed hyperkeratosis and papillomatosis with moderate irregular acanthosis (Figure 2).

In order to rule out any systemic disease associated with acanthosis nigricans, studies were requested for the patient, sister, and niece, including: blood tests, coagulation and general biochemistry, insulin and C-peptide, testosterone, dehydroepiandrosterone sulfate levels, and tumor markers. All the results were completely within normal ranges.

Benign familial AN is characterized by presence at birth and progression in early infancy, with cutaneous changes becoming more prominent in puberty and then stabilizing or diminishing later. Lesions tend to be located in folds, although in some cases they reach an unusual extension and intensity—probably when onset occurs early with the ensuing long period of development. Prurutis is uncommon.[7] The condition is transmitted in autosomal dominant form with variable penetrance and it is not normally associated with any endocrine or congenital abnormality.

We present a case of benign familial AN, with the classic clinical characteristics, associated with hypochondroplasia. Although we could not examine all the cases in this family, autosomal dominant inheritance appears to be present (Figure 3) as is normally the case. Benign familial AN is 1 of the usual classifications of the disease, although very few cases have been described. This could be due to the absence or minimal extent of associated symptoms and the hereditary nature of the disorder whereby patients do not consider the condition relevant or worthy of reporting to a physician. Even though there are generally no associated systemic alterations patients should be

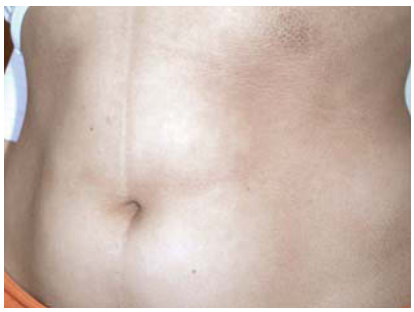

Figure 1. Brown velvety coloration along the sides of the abdomen.

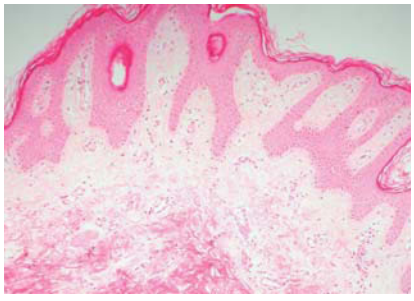

Figure 2. Epidermal hyperplasia with acanthosis, papillomatosis, and discrete hyperkeratosis compatible with acanthosis nigricans (hematoxylin-eosin × 100).

given a physical examination and a full medical history should be taken. Hyperandrogenism and insulin resistance should be ruled out whenever these appear to be present. As far as we are aware, no association with hypochondroplasia has been previously reported. The characteristic phenotype of this disease means that a general physical examination will be sufficient to guide the choice of complementary examinations given to patients.

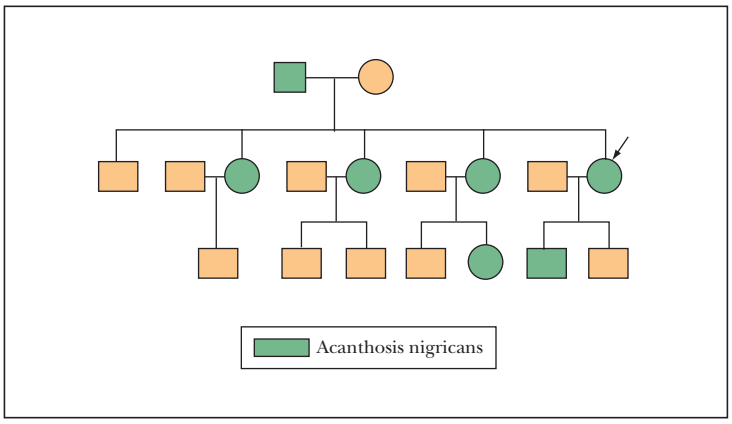

Figure 3.

References

1. Pollitzer S. Acanthosis nigricans. In: Unna PG, Morris M, Besnier E, et al, editors. International atlas of rare skin diseases. London: HK Lewis & Co; 1890. p. 1-3.
2. Janovsky V. Acanthosis nigricans. In: Unna PG, Morris M, Besnier E, et al, editors. International atlas of rare skin diseases. London: HK Lewis & Co; 1890. p. 4-5.
3. Schwartz RA. Acanthosis nigricans. J Am Acad Dermatol. 1994;31:1-19.
4. Skiljevic DS, Nikolic MM, Jakovljevic A, Dobrosavljevic DD. Generalized acanthosis nigricans in early childhood. Pediatr Dermatol. 2001;18:213-6.
5. Uyttendaele H, Koss T, Bagheri B, Scheneiderman P, Silfen ME, Gallagher MP, et al. Generalized Acanthosis Nigricans in an Otherwise Healthy Young Child. Pediatr Dermatol. 2003; 20:254-6.
6. Inamadar AC, Palit A. Generalized acanthosis nigricans in childhood. Pediatr Dermatol. 2004;21:277-9.
7. Tasjian D, Jarret M. Familial acanthosis nigricans. Arch Dermatol. 1984;120: 1351-4.

Primary Kaposi Sarcoma of the Penis in an HIV-Negative Patient

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To the Editor:

Kaposi sarcoma is a vascular tumor of multifocal origin first described by Moritz Kaposi in 1872.[1] In the Mediterranean area, the classic form is mainly seen on the lower limbs of the elderly.[2] Isolated involvement of the penis is rare, and although it is seen in AIDS patients—where it is the initial manifestation in 2-3% of cases—it is extremely uncommon in human immunodeficiency virus (HIV)-negative patients.[3] In the last 20 years, only 15 cases of immunocompetent patients with primary Kaposi sarcoma of the penis have been described in English-language journals.[4]

We present the case of an 80-year-old man, with no relevant history, presenting a rapidly growing asymptomatic tumor on the penis that had developed 2 weeks earlier. The patient had no history of local trauma, immunosuppression, intravenous drug addiction, blood transfusions, or homosexual acts. Physical examination revealed a soft pink pedunculated nodule of 10 mm in diameter on the coronal sulcus. He also presented a second clearly circumscribed painless red-violaceous lesion of 4 mm in diameter that had been present for several years (Figure 1). There was no evidence of inguinal gland involvement, hepatosplenomegaly, or other

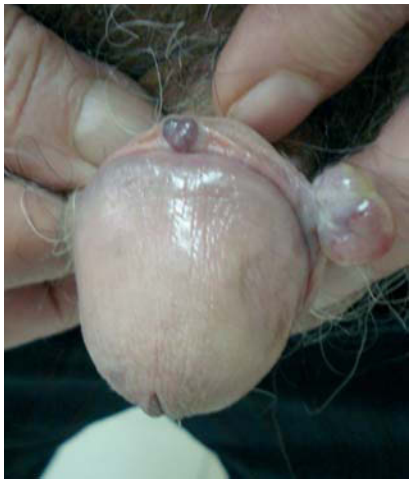

Figure 1. Two painless nodular lesions on the coronal sulcus.