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. Pruritus is uncommon. 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
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.

References


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. In the Mediterranean area, the classic form is mainly seen on the lower limbs of the elderly. 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. 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.

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.