Two Cases of Hypertrichosis Cubiti

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To the editor:
Hypertrichosis cubiti, also known as hairy elbows syndrome, is an uncommon form of localized congenital hypertrichosis in which an excessive amount of long, fine, lanugo-type hair is found on skin of normal texture and morphology. The hair growth follows a bilateral symmetrical distribution and affects the extensor surface of the distal third of the upper arms and the proximal region of the forearms. The condition usually appears

References
in early infancy (1-3 years). With time, the hair becomes thicker, reaching maximum thickness at age 5. It usually regresses in adolescence, but in some cases persists throughout life.\(^1\) The syndrome may present in sporadic or familial forms and its mode of inheritance is unclear. It is associated with short stature in about 50% of patients; such patients may present other malformations, the most common of which is facial asymmetry.\(^3,4\)

Two girls, aged 6 and 10 years, with the sporadic form of the syndrome were recently referred to our department by the pediatrics department. The girls were unrelated and had no relevant personal or family history. Both had suffered from the condition since the age of 2-3 years. A large amount of fine long hair was observed in both patients, dark in 1 case (Figure 1) and blond in the other (Figure 2). The hair was distributed over the distal region of the upper arms and the proximal region of the forearms. There was no excessive hair growth in any other area. The patients' height, weight, and intellectual development were normal. Blood tests (complete blood count; biochemical analysis; liver profile; thyroid function profile; and plasma cortisol, testosterone, and dehydroepiandrosterone sulfate levels) showed no abnormalities. The patients were advised to lighten or shave the area until the condition subsided in adolescence.

Hypertrichosis cubiti was first described by Beighton\(^1\) in 1970 in twin brothers belonging to an Amish family. Of the few cases reported, approximately 50% have been associated with short stature or with intrauterine growth retardation.\(^3,4\) It is in such children that other abnormalities such as facial dysmophia, abnormalities of the extremities, delays in language development, attention deficit, mental retardation, and mobility difficulties may be found.\(^3,5\) Our 2 patients presented none of these abnormalities, and the syndrome was considered a purely esthetic problem. As was observed in our patients, additional tests do not provide information of interest, and consequently, exhaustive studies are unnecessary.\(^1,2\)

Skin biopsy has been performed on only 2 occasions,\(^7,8\) together with a trichogram in 1 case.\(^7\) The trichogram showed 90% of the hair follicles to be in anagen, 9% in telogen, and 1% in catagen. This would explain the greater length of the hair, as occurs in the scalp.

Some authors suggest that the syndrome may be explained by mosaicism,\(^7\) based on the distribution of excess hair, which is confined to very localized areas and resembles that of cutaneous lesions with mosaicism; others consider it a nevoid condition of the hair follicles.\(^7,9\)

Hypertrichosis cubiti may therefore be part of a complex syndrome with varying manifestations. It is probably more common than it would appear from dermatology practice, but in cases with no associated malformations the only reason for consultation is its psychological and esthetic repercussions at a certain age. When associated with other abnormalities, these are usually identified first. In our opinion, if the condition is observed at a young age, the patient should be followed by a pediatrician, who can monitor growth and check for possible associated malformations; at later ages, as in our patients, it is advisable to reassure parents with respect to the course of the condition.

References


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Management of a Patient With Calciphylaxis and Requiring Anticoagulant Therapy

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To the Editor:

The different disorders associated with calciphylaxis include the possible relationship with oral anticoagulants, nadroparin calcium, and hypercoagulable states linked to lower concentrations of proteins S or C. The disorder most commonly associated with calciphylaxis, however, is chronic renal failure, with between 1% and 4% of these patients suffering from calciphylaxis. Calciphylaxis has also been observed in association with neoplasia, hyperthyroidism, proteinuria, rheumatoid arthritis, and alcoholic cirrhosis.

The pathogenesis remains obscure, although abnormal calcium and phosphorus metabolism (elevated calcium-phosphate product and high levels of parathyroid hormone), inflammation, and the presence of a hypercoagulable state have been observed and may lead to vascular and extravascular calcification.

The foregoing leads us to ask several questions: what attitude should be adopted in the case of a patient with calciphylaxis who requires anticoagulation therapy? What are the available antithrombotic alternatives? Which is the most recommendable option?

We present the case of a 58-year-old man with calciphylaxis who was receiving anticoagulant treatment with acenocoumarol due to ischemic heart disease that had been treated with a double coronary artery bypass graft and who had severe pulmonary hypertension, tricuspid insufficiency, and right ventricular dilation and hypokinesis. The patient visited the dermatology outpatient clinic with painful skin lesions on the legs that had appeared 10 days previously. The lesions were between 3 and 4 cm in diameter with a necrotic base and erythematous borders, and the patient was undergoing hemodialysis due to chronic renal failure. The patient presented secondary hyperthyroidism with high levels of aluminium (as a phosphorous chelating agent) but normal levels of calcium, phosphorous, and alkaline phosphates, along with anemia, high blood pressure, and dyslipidemia. The diagnosis of calciphylaxis was confirmed following a pathological study of the lesion biopsy. In patients diagnosed with calciphylaxis, it is reasonable for both oral anticoagulant therapy and therapy with both unfractionated heparin and low-molecular-weight heparin calcium (nadroparin calcium) to be omitted as they may give rise to calcium deposits. The following are proposed as recommendable alternative anticoagulant therapies: fondaparinux sodium and low-molecular-weight heparin sodium (preferably tinzaparin in patients who also present renal failure), with the clear aim of avoiding exacerbation of the calciphylaxis and the instability inherent in oral anticoagulant therapy in patients with a poor clinical prognosis.

References