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ATOPIC DERMATITIS AND PSEUDO-PSEUDO-HYPOPARATHYROIDISM

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Abstract

Pseudo-pseudohypoparathyroidism and atopic dermatitis is an extreme unusual coexistence in the same patient, who appeared with clasic bone abnormalities of hands and feet.
The incidense, clinical features and pathology are discussed.
Key words: Dermatitis, atopic, pseudohypoparathyroidism

A 43 year-old white man was admited to hospital for a recurrence of a seven year old atopic dermatitis. He complained of a generalized pruritus, loss of apetite, malaise, dysphagia and occasional vomiting. His past history was not remarkable, except for an operation 4 years ago for a repeated right shoulder dislocation. There was no family history of seizures or skeletal abnormalities. On examination he appeared pale, his weight was 49 Kg and height 162 cm. The face was broad and round, the neck short and the hands stubby. The hair was soft without brillancy. The mucosa was pale. The tongue was smooth. His skin appeared in some areas, atrophic, purplish erythematous, dry with lichenified lesions. Such areas were seen on face, neck, upper trunk, wrists, hands and in the folds of knee and elbows. His fourth right finger was relatively shorter than the other ones. There was absence of fourth knukle on the right hand (Albright's dimpling sign), shorteness in the third, fourth and fifth on right toe and in fourth and fifth on left toe (Fig. 1). Cvostek and Trousseau signs were absent. On oral examination it was noted a bad oral hygiene

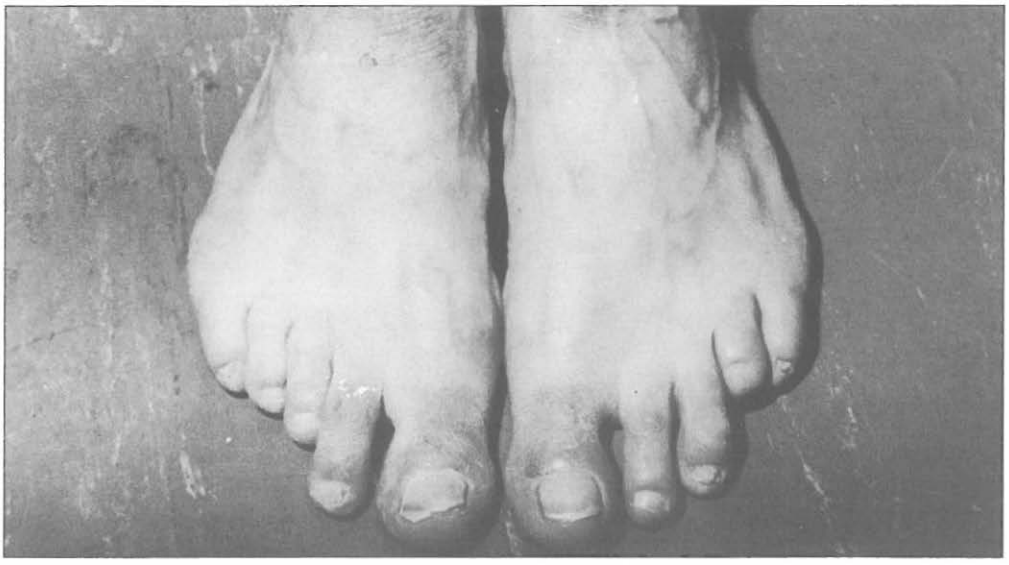

Fig. 1: Shortened third, fourth and fifth right toe and fourth and fifth left toe. Skin lesions of atopic dermatitis of both feet.

(nicotinic stomatitis, candidiasiss) and loss of many teeth. Eye examination showed no corneal or lenticular opacities. The remaining examination was not remarkable.

Laboratory findings showed hematocrete 28%, hemoglobin 8 gr %, red cell count 3,8 x $10^6/mm^3$, MCV 73 m^3, MCH 21 pg, reticulum red cells 1,6%, white cell count 72.000/mm^3, with normal diferentiation. Urinalysis were negative. Blood glucose and urea were normal. Total serum protein and protein serum elecrophoresis were normal. The serum calcium was between 8,8 and 10 mg% and the inogranic phosphate between 2,9 and 4,1 mg% whenever tested (more than six times). Alkaline phosphatase level in the serum was 9,4 U K.A. (normal for his age). The 24 hour urine calcium and phosphate were normal. The PTH of the blood was 3,95 μU/ml (reference values < 4 μ U/ml). The glucose tolerance test after 100 gr per os ingestion showed the following values: 72-92-79-65-55 gr% at times 0′-30′-60′-90′-120′ respectively. A D-xylose test was found 4,9 gr/5 h (normal values < 5 gr/5h). Serum immunoglobulins were normal, Ra-test was negative, le-cells were not found, as well as anti-DNA antibodies against gastric mucosa, thyroglobulin and adrenal tissue. The hormonal profil and the spermodiagram gave normal findings.

The x-ray studies of the wrists, long bones, hands, feet, skull and spine did not show any soft tissue calcification, or demineralization of the hand and spine. X-ray of both hands and feet showes: shortening of fourth metacarpal on right hand and first metacarpal on left hand shortening of third, fourth and fifth metatarsals on right foot and fourth and fifth metatarsals on left foot. Also it was noted shortening and cone-shaped appearance on the middle and distal phalanges of the toes. On panoramic dental x-ray the lamina dura appeared normal. X-ray study of the gastrointestinal truck and gall blader were normal, except that of persistence of the materia in glossoepiglottic incisures, simulating paralized swallow. Endoscopic examination of the upper gastrointestinal truck was normal except of a low degree of esophagitis at the end of the esophagus. Esophageal peristalsis after instrumental irritation was normal.

The mental quotient (WAIS) of speech scale was found 93 degrees and the one of practice scale 70 degrees. The above difference (23 degrees) between speech and practice scale was indicative of mental retardation.

The EEG showed low voltages, although it was well organized without facal or special pictures. The EEG was within normal limits. Skin liopsy from the left thigh surface revealed a picture of chronic dermatitis (Fig. 2).

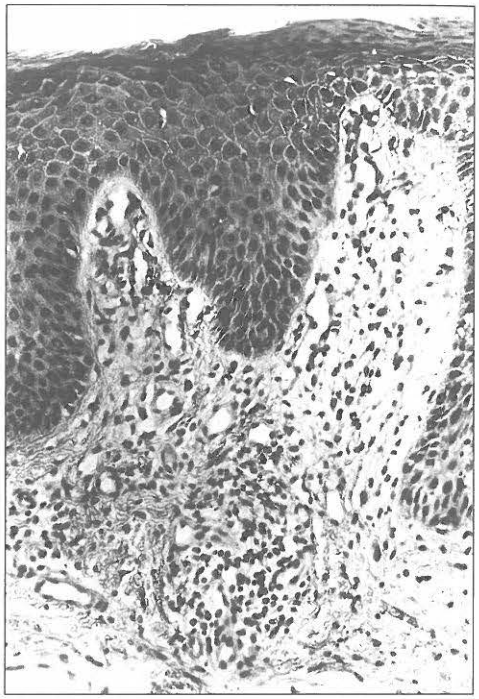

Fig. 2: Skin biopsy from the left thigh showing picture of chronic dermatitis (hematoxylin-eosin x 250).

Discussion

The term «pseudohypoparathyroidism» (PH) was first used by Albright et al (1) in 1942 to describe a clinical syndrome characterized by short stature, abnormalities of bones, mental retardation as well as biochemical and clinical features of hypoparathyroidism. A simillar, genetically related, clinical syndrome, without clinical or biohemical features of hypoparathyroidism, was also described by Albright et al (2) ten years later and named «pseudo-pseudohypoparathyroidism» (PPH). According to the classification of hypoparathyroid states by Nusynowitz et al (3), based

on physiologic principles, both the above syndromes belong to a group of diseases with deficiency of PTH sensitive adenyl cyclase - c-AMP system in bones and kidneys (4). PPH has been reported to be related with other, mainly endocrine, diseases (5). The coexistence of PPH and skin diseases, especially dermatitis, is very rare and to our knowledge only one case has been reported in the literature concerning a patient with PPH and eczema of both legs (6). We report here our recent experience of a case of PPH with extensive atopic dermatitis.

The clinical picture of PPH varies widely from a full blown syndrome (short stature, round face, short phalanges, metacarpals and metatarsals, soft tissue calcification, mental retardation, to only one short metacarpal (7). The presence in our patient of the clasic bone abnormalities of the hands and feet (8), the normal serum and urine values of calcium and phosphorus, as well as the normal blood level of the PTH, are indicative of the diagnosis of PTH.

The atopic dermatitis has been reported to be associated with clinical symptoms and signs of thyroid diseases (9,10). The incidence of the existence thyroid antibodies in the serum of patients with atopic dermatitis is a great deal of controversy, because it has been reported as increased (11) or as normal (12). A clear relation between atopic dermatitis and other endocrine diseases has not been reported up date.

The anemia of our patient may be the result of impaired intestinal absorption, because the D-xylose test showed a value which was near to the lower reference ones and the glucose tolerance test showed a flat curve. An etiologic relation between PPH and the generalized atopic dermatitis is not possible to be defined at the present.

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