

Chronic Mucocutaneous Candidiasis and Alopecia Areata as Cutaneous Expressions of Autoimmune Polyglandular Syndrome Type I

I. Carboni, R. Soda, L. Bianchi and S. Chimenti

Department of Dermatology, "Tor Vergata" University of Rome, Ospedale S. Eugenio P.le dell'Umanesimo, IT-00144 Rome, Italy. E-mail chimenti@uniroma2.it

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Sir,

Autoimmune polyglandular syndromes (APS) are hereditary disorders classified into APS types I, II and III in accordance with several endocrine and non-endocrine failures or defects (1). Chronic mucocutaneous candidiasis, alopecia areata and vitiligo variously complete the clinical picture of these syndromes (1).

A case of APS type I showing chronic mucocutaneous candidiasis and alopecia areata is described, with these skin disorders outlined as cutaneous typical expressions of this syndrome.

CASE REPORT

Since the age of 8 years, a 37-year-old man had observed the onset of onychodystrophies, first affecting his fingernails, later involving the toenails (Fig. 1). Five years later, a primary adrenal insufficiency occurred, presenting with melanoderma, fatigue, vomiting, weight loss and blood hypotension. Furthermore, primary hypoparathyroidism and insulin-dependent diabetes mellitus worsened the previous endocrinological dysfunctions. Alopecia areata either of the occipital area (Fig. 1) or of the eyelashes and recurrent oral candidiasis arose 10 years after onset of the adrenal failure.

At the time of our observation, nail biopsy and mucosal cultures were positive for candida infection, supporting the diagnosis of chronic mucocutaneous candidiasis involving nails and oral mucosa. Immunological

investigations did not display non-organ-specific auto-antibodies or autoantibodies to adrenal gland, thyroid, parathyroid and gastric parietal cells. Furthermore, autoantibodies to steroidogenic enzymes, such as steroid 17 α -hydroxylase, cytochrome P450 side-chain cleavage enzyme and steroid 21-hydroxylase, were not detected using immunoprecipitation assay. Only anti-insulin and anti-pancreatic islet cell autoantibodies were found. The patient is receiving replacement therapies for endocrinological failures and systemic anti-fungal therapy (itraconazole 200 mg/day for 2 months) resulting in a substantial improvement of candida infections.

DISCUSSION

In our patient, chronic mucocutaneous candidiasis, Addison's disease, primary hypoparathyroidism, insulin-dependent diabetes mellitus and alopecia areata confirmed the diagnosis of APS type I. This syndrome is a rare condition first presenting in childhood and defined by chronic mucocutaneous candidiasis, primary hypoparathyroidism and autoimmune adrenal insufficiency (2). It is an autosomal-recessive inherited disorder caused by mutations in a single gene named AIRE (autoimmune regulator) coding for a protein that is thought to act as a transcriptional factor based on sequence homologies and transient expression assays in cell lines (3). Several additional dysfunctions, including different autoimmune endocrinopathies (gonadal failure, insulin-dependent diabetes mellitus and thyroid diseases), gastrointestinal diseases (chronic atrophic gastritis, pernicious anaemia and malabsorption), chronic active hepatitis, ectodermal dystrophies and keratopathy have been variously associated with this syndrome (2, 4). Cutaneous signs of APS type I are usually represented by chronic mucocutaneous candidiasis, alopecia areata and vitiligo (5).

Chronic mucocutaneous candidiasis, characterized by chronic and recurrent candida infections of nails, skin and mucous membranes, has been considered a significant diagnostic criterion of the syndrome occurring as first clinical manifestation in 73–78% of the cases described (1, 6). The recurrence and resistance to topical anti-fungal drugs, typical of these infections, have been variously referred to different cell-mediated anomalies detected during chronic mucocutaneous candidiasis (7, 8).

Alopecia areata is reported in about 30% of patients with APS type I involving scalp, eyelashes, eyebrows,



Fig. 1. Onychodystrophy of the fingernails and occipital alopecia areata.

axilla and pubis and is likely to be related to an autoimmune pathogenesis (4, 6). Recently, a new autoantigen related to APS I, tyrosine hydroxylase, has been identified in patients affected by alopecia areata (9).

Vitiligo, observed in 8–13% of APS I patients, appears in childhood and is typically associated with the presence of autoantibodies directed against the SOX9 and SOX10 autoantigens (1, 10).

The high incidence of chronic mucocutaneous candidiasis in APS type I, as first presenting feature of the syndrome, and its absence in APS types II and III underline the relevance of chronic mucocutaneous candidiasis as early diagnostic criteria for APS type I. In light of this view, and related to the subsequent onset of the different signs that complete the syndrome, patients affected by chronic mucocutaneous candidiasis in early childhood should have a long-term monitoring dermatological and endocrinological follow-up in order to detect whether, other than the immunological defects commonly associated with this infection, an early diagnosis of APS type I can be suspected and a correct therapy can be established (5, 8).

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A Case of Localized Acne Following Radiation Therapy

Ji youn Song, Seog Jun Ha, Chung Won Kim and Hyung Ok Kim

Department of Dermatology, College of Medicine, The Catholic University of Korea, 505, Banpo-Dong, Seocho-Gu, Seoul, 137-040, Korea. E-mail: knderma@cmc.cuk.ac.kr

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Sir,

Acute and chronic radiodermatitis are well-known, frequent complications of radiation therapy. However, acne eruption localized to the radiation field is an unusual late sequelae of radiation therapy. We describe here a case of localized acne eruption on the neck of a patient who received radiation therapy for laryngeal carcinoma.

CASE REPORT

A 51-year-old man presented with an acne eruption on his neck for 15 days. Four months previously he noticed hoarseness. Laryngoscopic examination showed an exophytic mass on the right true vocal cord and subsequent endoscopic biopsy revealed squamous cell carcinoma of the larynx. The patient received three courses of cisplatin–sodium thiosulfate chemotherapy and concurrent radiotherapy. Initially, 6 MV radiation therapy in a total dose of 5000 cGY was applied to the entire neck, followed by a

booster irradiation of 2200 cGY to the glottic region. A total dose of 7200 cGY in 40 fractions over 74 days was therefore applied on a 12 × 7 cm rectangular area of his neck. He did not have a past history of acne and had not been using steroid or any topical preparation.

On physical examination, there was a sharply defined area of acne eruption consisting of erythema, follicular papules, pustules and black comedones in the anterior neck. The lesion corresponded to the portal field of radiation (Fig. 1) and the density of acne was more compact on the area of the booster irradiation. Biopsy showed open and closed comedones containing keratin plugs with flattened follicular epithelial lining and a few atrophic sebaceous glands. He was treated with retinoic acid gel 0.01% for 2 months and showed improvement.

DISCUSSION

Radiation-induced acne eruptions have been reported following several types of radiotherapy, including super-