
CASE REPORT

**Acrokeratosis Paraneoplastica
(Bazex syndrome) with ear manifestation**

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Acrokeratosis paraneoplastica or Bazex Syndrome is a paraneoplastic cutaneous syndrome which is most frequently associated with squamous cell carcinoma of the upper aerodigestive tract and characterized by psoriasiform lesions. Early recognition of the condition in its first stage may allow more effective treatment of the underlying tumour to be undertaken but diagnosis of Bazex' syndrome is very difficult at this initial moment.

We report a patient with a piriform sinus tumour where the lesions at ears and face help us for the diagnosis and where the treatment of the underlying neoplasm led to disappearance of the cutaneous symptoms.

Acrokeratosis paraneoplastica is a cutaneous paraneoplastic syndrome first described by Bazex and colleagues in their report of squamous cell carcinoma of the tongue associated with a papulosquamous eruption of the extremities⁽¹⁾. At the time of this writing, more than 100 cases of this rare dermatologic syndrome, the vast majority of which occurred in Caucasian men, have been reported⁽²⁾. The most common malignancy associated with acrokeratosis paraneoplastica is squamous cell carcinoma of the upper aerodigestive tract, about 50% of which occurs in the ear, nose, or throat.

The characteristic cutaneous lesions of acrokeratosis paraneoplastica include psoriasiform and eczematous papules and poorly limited red or purple erythematous plaques of the ears, fingers, toes, and nose. The lesions are simultaneous and persistent and may progress to involve the entire surface of the hands, feet, knees, and elbows. Nail dystrophy and paronychia are common findings, and the presence of bullous lesions has been reported occasionally⁽³⁾.

We report a new case of acrokeratosis paraneoplastica with unusual involvement of the ear in a patient with a piriform sinus tumor. Chemoradiotherapy of the underlying neoplasm led to the resolution of the patient's cutaneous symptoms.

Case Report

A 58-year-old White man presented with an 8-week history of pruritus and dry patches on his ears, hands, feet, knees, and nose. He had been diagnosed as having psoriasis, and although appropriate treatment for that condition had been initiated, the lesions had not resolved. He had noticed a painless, gradually enlarging left neck mass 3 weeks before he sought medical treatment for new skin patches that had developed on his chest. This patient smoked 40 cigarettes per day and denied any significant illnesses or medical conditions except for untreated mild hypertension. His mother had died of colon carcinoma and his father of prostate cancer years ago.

Physical examination of this patient revealed violaceous and erythematous scaly plaques on the helix of the ears (figs. 1 and 2) and the tip of the nose (Fig. 2),



Figure 1: Plaques on the helix of the ears



Figure 2: Erythematous scaly plaques on the ears and the tip of the nose

as well as on the knees, elbows, and chest. Dystrophic nails with subungual hyperkeratosis and onycholysis were also noted (Fig. 3). At the otorhinolaryngologic examination, a fixed lesion of the left piriform sinus that reached the ipsilateral hemilarynx and a 3- by 3.5-



Figure 3: Dystrophic nails with subungueal hyperkeratosis and onycholysis

cm left lymph node metastasis were observed. Biopsy of the lesion at the piriform sinus was performed, and well-differentiated squamous cell carcinoma was diagnosed. The disease was staged T4, N2a, M0 (stage IV) according to the Tumor, Node and Metastasis (TNM) system of the International Union Against Cancer (UICC)⁽⁴⁾.

Chemoradiotherapeutic treatment was subsequently initiated. The chemotherapy protocol consisted of carboplatinum 130 mg and taxol 120 mg perfused over 1 hour every 7 days for 4 cycles. Intravenous dexamethasone and oral chlorpromazine were administered as antiemetics, and ranitidine and dexchlorpheniramine were also given. Concomitantly administered radiotherapy was also prescribed. The hypopharynx and cervical neck nodes were treated with 71 Gy (200 cGy/d) and the supraclavicular areas with 50 Gy. Toxicity was evaluated according to Miller's scale, and grade 4 stomatitis was noted. The cutaneous lesions had totally resolved at the conclusion of treatment.

At the patient's 12-month follow-up examination, the response was complete, and no recurrence was revealed on examination of the ears, nose, and throat or by computed tomographic scan. At the time of this writing, no new cutaneous lesions have developed, and the patient's pharynx and larynx have retained their anatomic integrity and function.

DISCUSSION

In this case report, we present an illustrative example of a paraneoplastic syndrome. The patient presented with cutaneous lesions several weeks before his pharyngeal tumor became clinically apparent. After the tumor had been diagnosed, appropriate chemoradiotherapeutic treatment of the neoplasm led to the resolution of the cutaneous condition.

Acrokeratosis paraneoplastica typically presents in 40-year-old men with cervical node metastases⁽³⁾. The most common locations of the underlying tumor are the piriform sinus, vocal cords, soft palate, tonsils, base of the tongue, lower lip, esophagus, and lungs. The cutaneous syndrome usually arises long before the clinical presentation of the underlying malignancy (average interval, 11 months)⁽²⁾.

Three stages of the cutaneous syndrome have been described⁽⁵⁾. The first stage is characterized by limited skin lesions that develop on the ears and on the tip of the nose, fingers, and toes. The findings at the initial stage of cutaneous disease are usually psoriasiform changes. The second stage of acrokeratosis paraneoplastica involves the progression of the cutaneous lesions, which eventually cover the fingers and toes and advance to the palms and soles. Nail dystrophy and paronychia are common. The characteristic lesions include psoriasiform and eczematous papules and poorly limited red or purple erythematous squamous plaques. The lesions are simultaneous and persistent. Symptoms of the underlying tumor and cervical nodes metastases are usually present at this stage. The third stage of acrokeratosis paraneoplastica is characterized by the extension of the skin lesions to the limbs, knees, elbows, and trunk; signs that are frequently associated with advanced neoplastic disease⁽⁶⁾.

The treatment of acrokeratosis paraneoplastica is that of the underlying tumor. Corticosteroids and retinoids have been shown to be of certain value, but drugs used to treat the symptoms of the disease have proven useless^(2,5). The cutaneous lesions tend to resolve when appropriate treatment of the underlying malignancy is initiated and to reappear when the tumor recurs. However, the waxing and waning of acrokera-

tosis paraneoplastica often does not correlate with the time of antineoplastic treatment and does not occur in up to 10% of the patients⁽²⁾.

CONCLUSION

Acrokeratosis paraneoplastica is a rare cutaneous syndrome that develops on the ears and elsewhere on the body before the clinical manifestation of an underlying malignancy. Clinicians must be aware of this disorder because early diagnosis may enable a more effective treatment of the primary tumor. In this article, we hope to present clinical information that obviates delay in the correct diagnosis of acrokeratosis paraneoplastica in patients with an underlying malignancy, because the cutaneous syndrome usually arises long before the clinical presentation of the neoplasm. It is especially important to consider the appearance of cutaneous lesions on the helix of the ears or on the nose, because that sign suggests acrokeratosis paraneoplastica.

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