Squamous Cell Carcinoma Arising from Chronic Ulcerative Lesion in a Patient with Disabling Pansclerotic Morphea

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Authors report herein a case of a 27-year-old male patient who had been suffering from chronic ulcerative dermatitis with scar-like changes and successive involvement of the ankles, limb folds, nape and abdomen, and no tendency to heal from age 4. At the age 14, an immunologic study showed a selective IgA deficiency with partial T-lymphocyte inactivation. Ten years later, at age 24, he showed a severe form of morphea over a generalized area and disabling joint contractures, and was diagnosed as disabling pansclerotic morphea with an IgA value that returned to a near normal level. At the age 27, an adult-fist, 7x6x4cm sized, squamous cell carcinoma with an easy bleeding tendency like an overgrowing granulation tissue vascular tumor had developed on the chronic ulcerative lesion on the posterior aspect of the right ankle for 2 months. Aggressive metastatic lesion occurred on the right popliteal area 3 months later. He died 1 month thereafter.
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Disabling pansclerotic morphea, a severe form of generalized morphea, has a relentless disabling course, producing marked muscle contractures and joint deformities due to sclerosis of the skin and the deeper structures[1].

Squamous cell carcinoma(SCC) arising from chronic ulceration of morpheic lesions is very rare. Only a few cases were reported in literatures[2-5] but they were aggressive in nature and had a high frequency of metastasis.

Our patient is a man aged 27 in whom a SCC developed in the chronic ulcer of the right ankle in association with scleroderma for a long duration. Rapid metastasis occurred on the right popliteal area 3 months later and he died 1 month thereafter.

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REPORT OF A CASE

A 27-year-old man visited our department for the evaluation and treatment of a malodorous and exophytic mass on the posterior aspect of his right ankle. The mass had developed on the chronic ulcerative lesion 2 months before and had rapidly grown to be 7x6x4cm in size(Fig. 1-A).

He had a history of suffering from chronic recurrent ulceration with scar-like changes successively on ankles, limb folds, nape and abdomen from age 4. Since then he received various kinds of treatment such as antibiotics, steroids and other modalities but the ulceration waxed and waned.

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Regardless of his continuous treatment, complete remission of skin lesions did not occur. His father had psoriasis and died of lung carcinoma. The family hitory of other members was non-contributory. The disease entity was not established at age 14, although immunologic results revealed a selective IgA deficiency with partial T-lymphocyte inactivation[6]. There were progressive ulcerative patches, and nonpliable thickening, taut and shiny skin on the belt line of the abdomen, knees, ankles, axillae and nape.

Ten years later at age 24, a follow-up visit showed a severe form of morphea over the generalized area and disabling joint contractures involving the knees, elbows and ankles(Fig. 2-A), and was diagnosed as disabling pansclerotic morphea[7], with an IgA value that returned to a near normal level. Scar-like lesions with alopecia of the posterior scalp, and ivory colored guttate morpheic lesions with confluent plaques surrounded by lilac ring on the anterior chest, abdomen, and both arms were seen(Fig. 2-B). Also, the crusted ulcerations on both ankle joints with surrounding telangiectasia were seen(Fig. 2-C). Sclerotic changes on the back and contractures of both knee joints were prominent and stiffness of the Achilles tendon and muscle wasting of the thighs, legs and feet disabled him for ambulation. Laboratory tests, including a complete blood count, urinalysis, liver and renal function tests, antinuclear antibody, HLA typing, and chromosomal study showed no significant results. A chest X-ray, upper gastrointestinal series, and electromyography were also proved to be unremarkable. Lumbar spine view showed kyphoscoliosis.

At the next visit, three years later, at age 27, a tumorous lesion had been growing for 2 months on the chronic ulcerative lesion of the right ankle which had persisted for over 23 years. This new lesion had rapidly grown to be an adult-fist, 7x6x4cm sized, semigloboid mass which had a tendency to bleed easily like a granulation tissue or vascular tumor. Regional lymph node was not palpable. No significant findings were found by routine laboratory studies. Biopsy obtained from the tumor revealed irregular downward proliferation of atypical squamous cells with mitoses and dyskeratotic changes, and a minimal degree of infiltration of small lymphocytes at the tumor base (Fig. 1-B). Therefore, it was diagnosed as SCC of grade II or III. A wide excision with a skin graft was performed.

Concomitantly, he complained of a dull pain on a swollen area of the right popliteal fossa(Fig. 3-A), which lesion was considered to be either an abscess or a metastatic focus. The biopsy revealed that keratinization was almost or completely absent and nearly all tumor cells were atypical and devoid of intercellular bridges(Fig. 3-B). It was suggestive of SCC with a high malignant potential. We recommended amputation of the involved thigh, but he refused.

Three months later, he developed an 4x5x2cm sized mass, bulged out from the biopsy site of the right popliteal area(Fig. 4-A). A wide excision was done, and showed similar histopathologic findings to the original lesion. Routine laboratory studies and computerized tomography of pelvis and abdomen were unremarkable. No abnormalities were found with immunoelectrophoresis. The helper T-cell count was 53% and the suppressor T-cell count was 29% by flowcytometer activated cell scanner and sorter. Mantoux and 1% dinitrochlorobenzene(DNCB) tests were negative. Multi-CMI test revealed decreased response. Simple X-rays showed irregular mottled radiolucencies on the distal part of the right femur. Bone scan with Tc^{99m} Magnesium Diphosphate showed hot areas on the right knee, ankle, and femur, suggestive of bone metastases. Yet he still refused any management.

One month later, he revisited our hospital which was untreatable with a three times larger mass, measuring approximately 14x10x8cm(Fig. 4-B). It was irregularly surfaced with a beef-like consistency on the right popliteal fossa. He died a few days later.

SCC arising from an area of chronic ulcer is well recongnized for its aggressive nature and high frequency of metastasis[8]. The Marjolin ulcer among precancerous lesions has a high metastatic potential, depending on the site by 8-20%[5,9]. High risk groups were presented as male patients younger than 60 years of age and patients of either sex with carcinoma of the lower extremity[10]. At the time of diagnosis, nine(81.8%) of the 11 metastasizing tumors had been present for less than one year. Thereafter, all patients with metastases had died[9]. Metastases may occur early in the disease course: 2.5% had been noted for less than

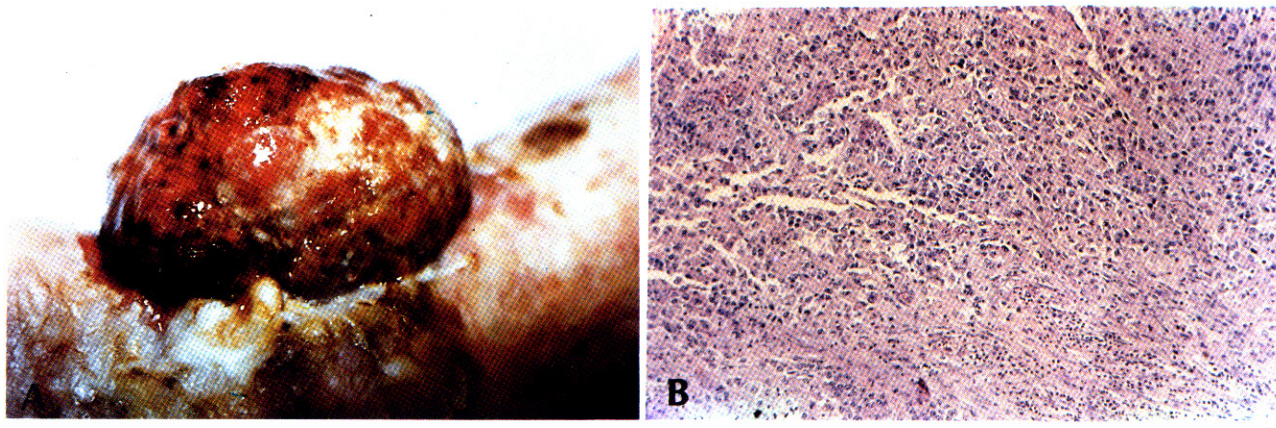

Fig. 1. A : A semigloboid, exophytic mass 7x6x4cm sized on the posterior aspect of the right ankle.
B : The biopsy reveals irregular downward proliferation of atypical squamous cells(H & E, ×100).

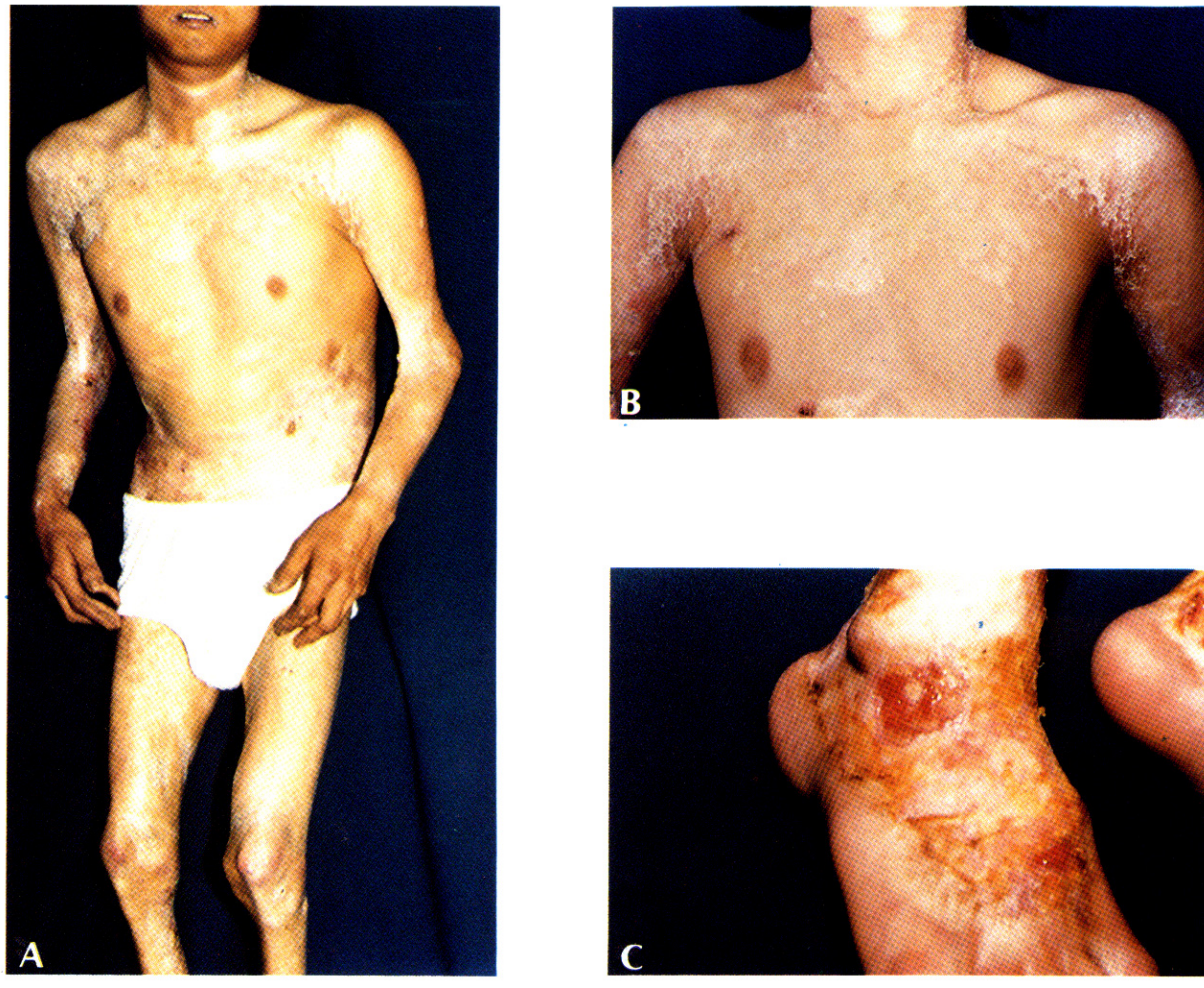

Fig. 2. A : General appearance showing the extensive sclerotic changes with deformities of extremities at age 24.
B : Ivory-colored guttate morpheic lesions on the anterior chest.
C : Ulcerations of both ankles and right dorsal foot at age 24.

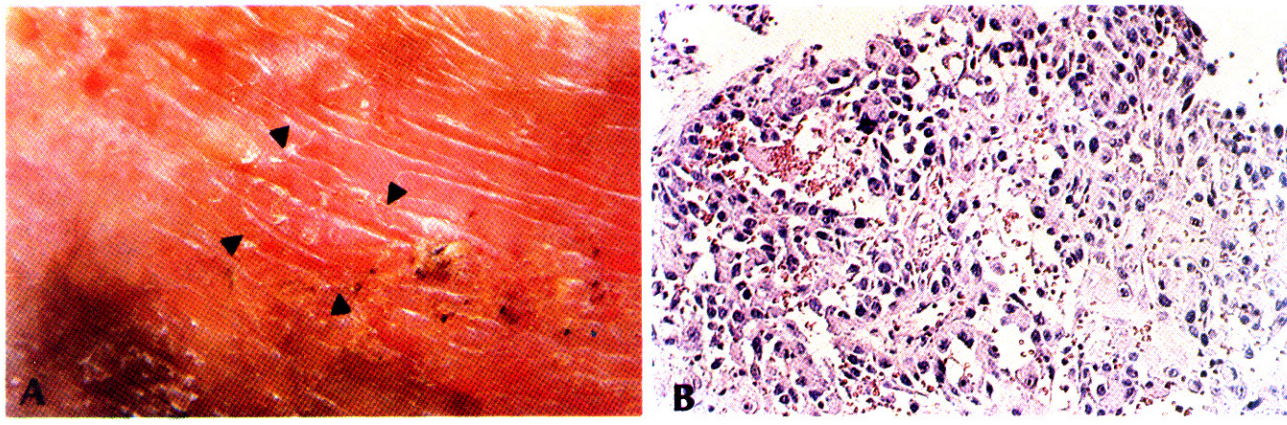

Fig. 3. A : The painful area on the right popliteal fossa(arrow-head).
B : The biopsy of right popliteal fossa reveals that keratinization is almost absent and all tumor cells are atypical and devoid of intercellular bridges(H & E, ×200).

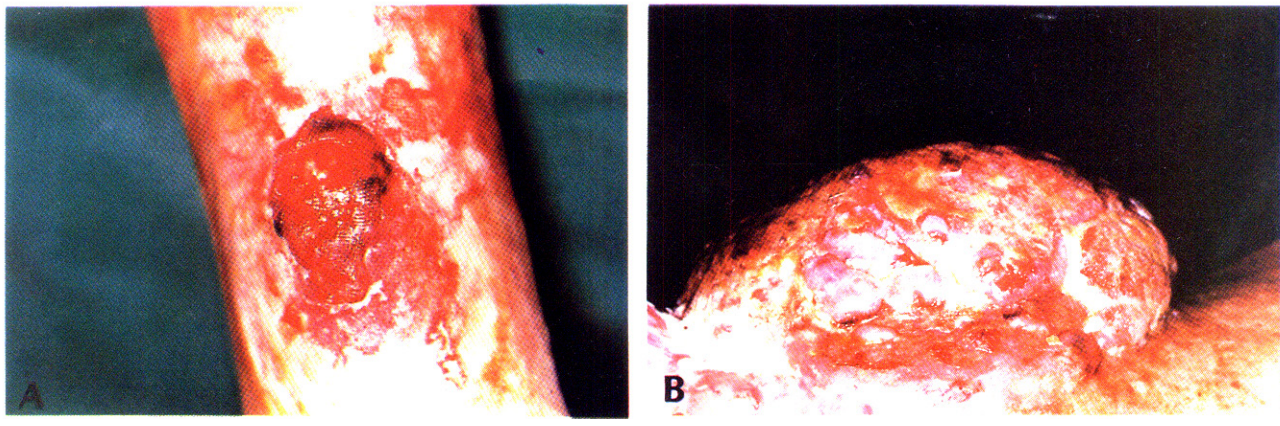

Fig. 4. Metastatic lesion of right popliteal fossa.
A : A pliable, 4x4x2cm sized mass bulged out about 3 months after biopsy.
B : A 14x10x8cm sized, rapidly enlarged mass about 1 month after mass excision.

one month, 37.8% for less than six months, and 68.1% for less than one year. This indicates that such spreading does not occur only in long neglected or inadequately treated instances[11].

Tumor behavior correlated best with the level of dermal invasion and the vertical tumor thickness. All tumors that proved fatal were at least 10mm in thickness, with extension into subcutaneous tissue or deeper structures[8]. Crawley et al.[12] suggested that patients who developed recurrence, metastasis, and death from carcinoma had borderline or low immunocompetence. Host response can be judged by the degree of infiltration of small lymphocytes[12]. Biopsy findings in our case showed minimal infiltration of small lymphocytes at the margin of the tumor.

Since age 4, our patient suffered from chronic recurrent ulceration with scar-like sclerotic changes on both ankles. Twenty years later, at age 24, he showed a severe form of morphea over a generalized area and disabling joint contractures, which was diagnosed as disabling pansclerotic morphea. Rapidly progressive SCC developed finally on the ulcer area of the posterior aspect of the right ankle at age 27. Unfortunately, he did not recognize his growing mass as being a matter of importance and wasted 2 months before seeking aid at our department of dermatology. We thought retrospectively that the malignant change had already begun before the exophytic mass stage and metastasis had already begun with tenderness on the right popliteal fossa. Its metastasis was probably hematogenous

spread. Unfortunately, the tumor growth and metastasis were too rapid and he died about 4 months after diagnosis of SCC.

The mechanism of the development of carcinoma in our case would be assumed to have various underlying causative factors. The case demostrated a decreased ability to be sensitized to DNCB and decreased delayed hypersensitivity responses to various intradermal antigens. Since childhood, there was a general weakness. Steroids were used at irregular intervals for a long period of time. Tests done at the age of 14 had revealed immunologically abnormal findings : selective IgA deficiency with partial T-cell inactivation[6]. The immunologically inapparent alterations might have facilitated and enhanced the carcinoma. Otherwise, normal immune surveillance mechanism could not react directly with the tumor specific antigens resulting from the obliteration of the lymphatics in the sclerotic lesions subjacent to the chronic ulcer[13].

The development of SCC in scleroderma appears to be very rare. Only four cases in the literatures were found[2-5]. Our case is very similar to Michalowski et al.'s one[2] in terms of clinical morphology and disease course. All cases including ours have in common the chronic ulcer of the lower legs or feet for over 20 years.

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