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Rapidly Developed Multiple Face and Neck Skin Cancers in a Patient with Sjögren's Syndrome: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Male, 76
Final Diagnosis: Skin cancer
Symptoms: Skin
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Unknown ethiology
Background: Sjögren's syndrome is a chronic, systemic disorder of an autoimmune nature, and its primary etiopathogenetic events are not known. Previous studies have found elevated incidence of malignancies in patients with primary Sjögren's syndrome. However, there are few reports regarding the association of Sjögren's syndrome with skin cancers, especially with multiple skin cancers developed within a short time.
Case Report: We reported an unusual case of a patient with primary Sjögren's syndrome who suffered from rapidly developed facial and neck skin cancers within two years.
Conclusions: Sjögren's syndrome associated with skin cancer is rare. Our case report suggests that Sjögren's syndrome patients require continuous follow-up with conventional cancer examination, including skin biopsy for suspected skin lesions.

MeSH Keywords: Head and Neck Neoplasms • Sjogren's Syndrome • Skin Neoplasms • Autoimmune Diseases

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/902828

Background

Sjögren's syndrome is the second most common systemic autoimmune disease, following rheumatoid arthritis. The prevalence of Sjögren's syndrome is about 0.5% in the general population, and may be higher especially in perimenopausal women; the ratio of women to men is 9: 1) [1,2]. Sjögren's syndrome is characterized by lymphocytic infiltration of the exocrine glands, predominantly the salivary and lacrimal glands, leading to symptoms of dryness and systemic manifestations in 30–40% of afflicted patients [3,4]. Meanwhile, the lymphocytes also produce inflammatory cytokines and autoantibodies against two ribonucleoproteinic complexes named Ro/SSA and La/SSA [3,4]. Other systems or organs, such as musculoskeletal system, lungs, liver, skin, and kidneys, can also be involved [5]. Previous studies have found elevated incidence of hematologic malignancies in patients with Sjögren's syndrome [6].

The common skin symptoms of Sjögren's syndrome are dry skin and Raynaud's phenomenon, which affected 55% and 35% of primary Sjögren's syndrome patients, respectively [7,8]. However, there are few reports regarding the association of Sjögren's syndrome with skin cancers, especially the development of multiple skin cancers. Herein, we reported an unusual case of a patient with Sjögren's syndrome who suffered from rapidly developed facial and neck skin cancers.

Case Report

A-76-year-old male was diagnosed with Sjögren's syndrome with the initial presentation of dry mouth, intermittent fever, and severe oral ulcer. Since his diagnosis of ten years, his condition was under regular control with drug therapy. He did not have history of other systematic disease or major surgeries.

In January 2014, he visited our hospital for a skin mass (4×3 cm) on his left cheek (Figure 1A). A skin biopsy was performed and the result of the pathological examination was moderated differential squamous cell carcinoma. We widely excised the skin tumor on his left cheek and applied split thickness skin grafts (Figure 1B). Post-operative pathological examination confirmed that the surgical margin of the excised tumor was free from tumor cells and the pathological stage was pT-2Nx. The graft took and the wound healed well.

In March 2014, he found two mass on the anterior aspect of his neck and occipital area of scalp, respectively. He received excision of the neck tumor (Figure 1C) and scalp. The result of scalp pathological report was moderated differential squamous cell carcinoma (T1Nx). The surgical margin of excised scalp was tumor-cell free and the wound healed well. The neck tumor pathological result was also confirmed as moderated differential squamous cell carcinoma (T1Nx). The peripheral margin was free of tumor-cells and the deep margin was invasive squamous cell carcinoma. He received wide excision and split-thickness skin grafts of the neck tumor (Figure 1D), and the pathological diagnosis was stitches granuloma and no residual tumor cells.

In November 2015, he found a skin mass (4.5×3.5 cm) over his left cheek again, adjacent to the previous operated skin tumor (Figure 1E). Pathological examination following skin biopsy revealed squamous cell carcinoma with moderate differentiation. Therefore, he underwent wide excision and split thickness skin grafts again (Figure 1F). Post-operative pathological examination also confirmed the surgical margin of the excised tumor free from tumor cells (pT1pNx). The graft took and the wound healed well. The patient was under stable post-operative condition until the writing of this manuscript.

Discussion

Cutaneous squamous cell carcinoma (cSCC) is a common skin cancer. However, the exact incidence of cSCC remains unclear and the rat of metastasis is very low (2–5%). Once distant metastasis has occurred, the median survival of cSCC patients is less than two years [8]. The risk of skin neoplasm has been noted to be significantly associated with autoimmune connective tissue diseases, including lupus erythematosus, scleroderma, dermatomyositis, and Sjögren's syndrome [9]. The mortality rate of Sjögren's syndrome is highly correlated with the serological profile of patient, such as low levels of C4, C3, CH50, and cryoglobulinaemia, which implied the worse prognosis [10,11]. Primary treatment of cSCC is to complete removal with cosmetic results, and surgery is the most common and effective treatment [9].

A cohort study reported 112 patients with primary Sjögren's syndrome of which 25 them developed various types of cancer, the major cancer was non-Hodgkin's lymphoma (44%), followed by breast cancer (12%), and melanoma (4%). There was a significantly elevated incidence of all cancer types in Sjögren's syndrome patients compared with the general population (2.63-fold) and an extreme high risk for developing non-Hodgkin's lymphoma (37.46-fold). The most interesting finding was that 9 of 112 (8%) Sjögren's syndrome patients developed more than one cancer and the cancer types were non-Hodgkin's lymphoma, bilateral renal granular cell carcinoma, lung cancer, and cervical cancer. The secondary tumor was found between three to fifteen years after the first tumor was diagnosed [12]. A nationwide population cohort study analyzed 7,852 Sjögren's syndrome patients in Taiwan and found that, overall, Sjögren's syndrome patients did not have a higher prevalence of cancer. However, female Sjögren's

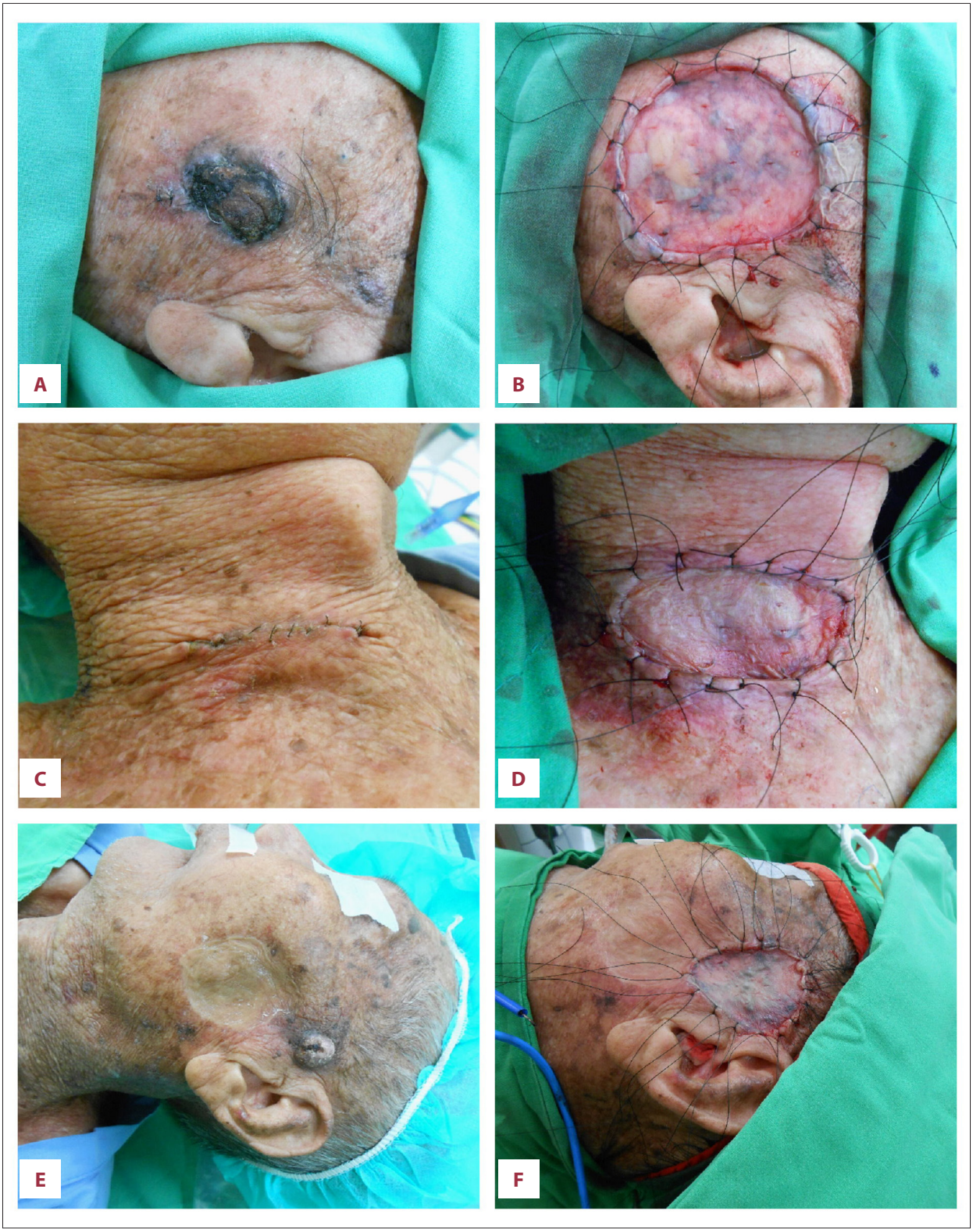

Figure 1. The patient with Sjögren's syndrome underwent three wide excisions with split thickness skin graft for squamous cell carcinoma: once on the left cheek in January 2014, preoperatively (**A**) and postoperatively (**B**); once on the neck in March 2014, preoperatively (**C**) and postoperatively (**D**); and again on the left cheek in November 2015, preoperatively (**E**) and postoperatively (**F**).

syndrome patients had a higher risk for developing non-Hodgkin's lymphoma (7.08-fold), multiple myeloma (6.09-fold) and thyroid gland cancer (2.56-fold), but lower risk of colon cancer (0.22-fold). Male Sjögren's syndrome patients had a higher risk of developing non-Hodgkin's lymphoma (3.1-fold) [13]. The major frequency of systemic manifestations in Sjögren's syndrome was arthralgia/arthritis and Raynaud's phenomenon, and Sjögren's syndrome patients had a higher risk of developing cancer, especially non-Hodgkin's lymphoma [14]. Sjögren's syndrome patients have high cancer incidence, and based on this report, while a patient developing multiple skin cancers is unusual, it requires follow-up cancer examination, including skin biopsy.

Conclusions

In our report, a male Sjögren's syndrome patient had surgical resection to remove facial cancer on three separate occasions and the post-operative pathological examination on each occasion confirmed that the surgical margin was free from tumor cells. However, secondary tumor (neck and skin cancer) developed within three months. This report is a reminder that Sjögren's syndrome patients cannot be ruled out for cancer and secondary tumor development. According to our observations, follow-up cancer-associated examinations should include skin biopsy.

Conflicts of interest

No potential conflicts of interest were disclosed.

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