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Cutaneous Spindle Cell Squamous Cell Carcinoma

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Dear Editor:

Spindle cell squamous cell carcinoma (SpSCC) is a rare variant of squamous cell carcinoma (SCC). Despite having clinical attributes that overlap conventional SCC, these lesions are pathologically distinct and are comprised of spindled cells that infiltrate the dermis as single cells or cohesive nests; that lack features of keratinization. SpSCC most commonly occurs in the larynx, and less often in the hypopharynx, oropharynx, and nasal cavity[1]. It is extremely rare to find SpSCC confined to the cutaneous lesion, and only three cases have previously been reported in Korea.

A 73-year-old woman presented with a 1-year history of a pruritic, solitary mass on the face. Physical examination revealed an irregular-shaped, erythematous, firm, fixed mass with a yellowish crust on her left upper perioral area. The mass was 1.3 cm in diameter and was slowly expanding (Fig. 1). The patient had a history of a wide excision of basal cell carcinoma on her nose 2 years previously, and no relapse has been observed.

Histologic examination of the lesion showed spindle-shaped cells with hyperchromatic nuclei infiltrating the dermis in a whorl-like pattern (Fig. 2). Furthermore, immunohistochemical investigation revealed the specimen was positive for cytokeratins (34βE12, CK5/6, AE1/3) and vimentin, but was negative for smooth muscle actin (SMA), S-100, and CD68. Through histologic features and immunohistochemical evaluation, she was diagnosed with SpSCC. The lesion was surgically removed 13 months previously and no relapse has been observed.

Cutaneous SpSCC presents as a raised or exophytic nodule accompanied by spontaneous bleeding and central ulceration similar to conventional SCC[2]. It is usually confined to sun-damaged sites such as the head, neck, chest, and upper extremities or areas that have received prior ionizing radiation[2,3]. Although the rarity of these tumors makes difficult to study patient survival rates, the literature suggests an overall prognosis similar to that of conventional SCC. SpSCC must be distinguished from other spindle cell lesions such as atypical fibroxanthoma (AFX), spindle cell/desmoplastic melanoma, cutaneous leiomyosarcoma, and scars. Immunohistochemistry has become the essential method in the pathologic workup of these tumors. The spindle cells in SpSCC stained positively for

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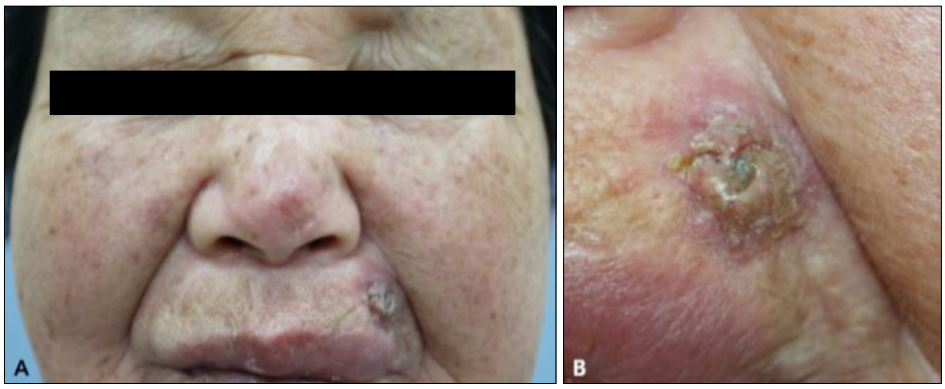

Fig. 1. (A, B) Solitary, 1.3 cm-sized, irregular-shaped, erythematous, firm mass with a yellowish crust on the left upper perioral area.

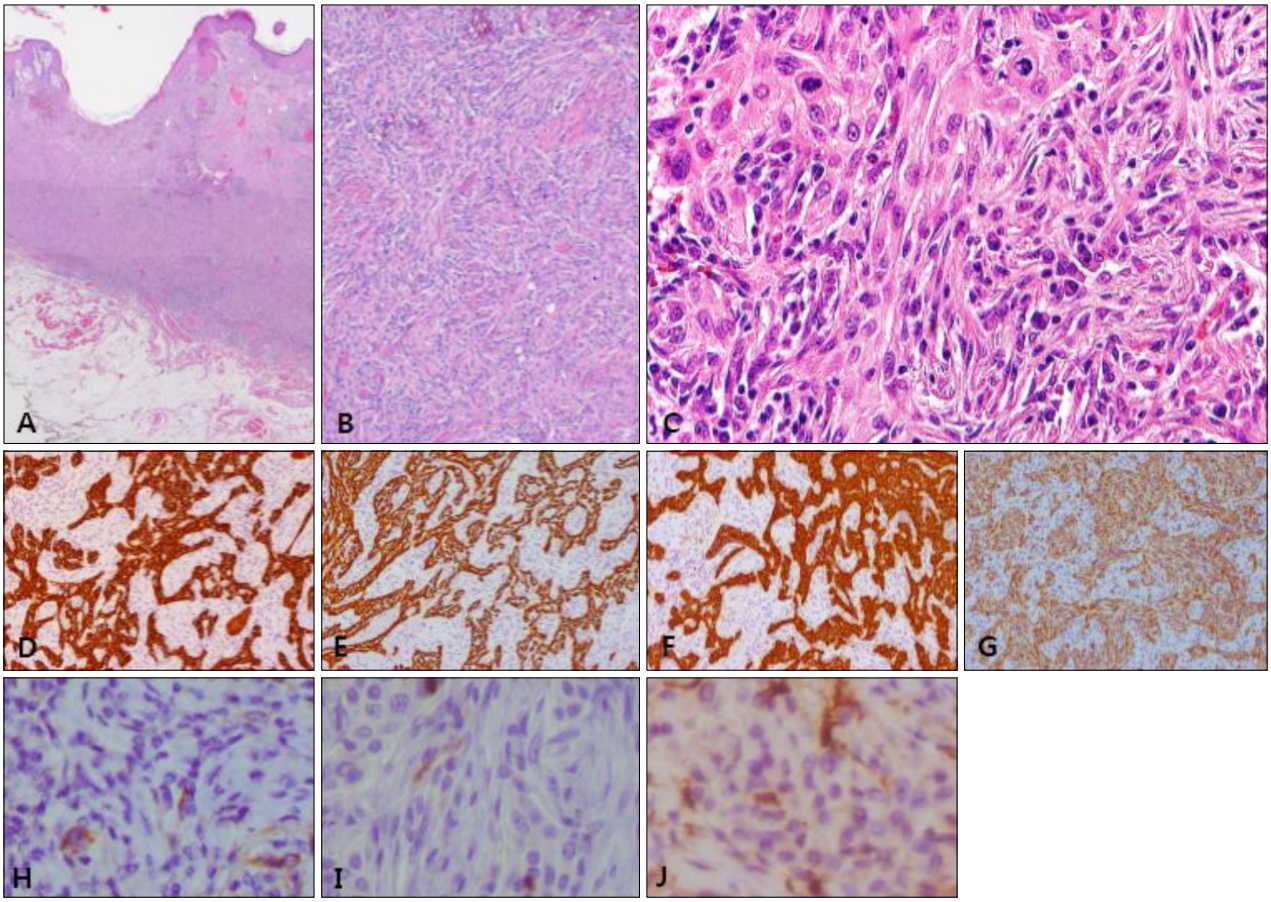

Fig. 2. (A) Diffuse dermal infiltrates of spindle cells (H&E, ×40). (B, C) Numerous spindle cells in a whorl-like pattern and atypical mitotic figures (H&E; B: ×200, C: ×400). (D) Positive immunostaining for 34βE12, (E) CK5/6, (F) AE1/3, and (G) vimentin (D~G: ×200). (H) Negative immunostaining for smooth muscle actin, (I) S-100, and (J) CD68 (H~J: ×400).

one or more of the cytokeratins (34βE12, AE1/3, cam 5.2, low molecular weight keratin) and the mesenchymal marker, vimentin. Each of these neoplasms stained negatively for S-100, CD68, and SMA; whereas AFX showed positivity for CD68, but lacked keratin and S-100. The spindle cell melanomas showed a connection with the epithelium that was contiguous with similar junctional cells possessing cytoplasmic melanin and S-100 immunostaining, but lacking keratins. The leiomyosarcomas showed positivity with SMA and vimentin, and were negative for keratins and S-100. The scars were negative for all immunohistochemical stains[4].

Cutaneous SpSCC is so rare subtype of SCC that pathophysiology, prognostic factors, and metastatic risk are not

well-established. It is known that SpSCCs that are associated with radiation, burn scars, and immunosuppression may correlate with aggressive clinical courses[5]. Further studies of this rare entity are needed to establish its biological behavior, and the accumulation of more case reports will aid in determining a precise prognosis.

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Bilateral Segmental Neurofibromatosis on the Face

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Dear Editor:

Neurofibromatosis is a heterogeneous disorder clinically characterized by the presence of neurofibromas, multiple café-au-lait spots, intertriginous freckles, and Lisch nodules[1].

A 62-year-old woman presented with diffusely scattered papules on the forehead that had been present for 10 years. She had no family history of neurofibromatosis. On physical examination, asymptomatic, soft, flesh-colored papules of 3～5 mm diameter were observed to be distributed over the ophthalmic branch of the right and left trigeminal nerves (Fig. 1A, B). There were no other abnormalities in any other body region. Her general physical examination revealed a normal status, including intelligence, speech, auditory function, and visual acuity. Histopathological examination showed a well-circumscribed tumor in the dermis with a normal overlying epidermis. The tumor consisted of loosely spaced spindle-shaped cells and wavy collagenous strands in the myxoid stroma. Nuclear pleomorphism and mitoses were not observed (Fig. 1C). Overall, the features were consistent with a neurofibroma. The papules on the forehead were excised for cosmetic reason.

Segmental neurofibromatosis is a rare form of neurofibromatosis that is characterized by cafe-au-lait macules and neurofibromas, or only neurofibromas, distributed in only one dermatome, and less commonly in two or more dermatomes[1]. Segmental neurofibromatosis was categorized into four subtypes by Roth et al. in 1987: a true segmental form (type I), a localized form with deep involve-

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