Desmoplastic spindle cell squamous cell carcinoma of skin

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ABSTRACT
Desmoplastic squamous cell carcinoma (SCC) and spindle cell SCC is very rare in skin. An 86-year-old woman presented a skin tumor of face. The tumor measured 1.8 cm × 1.6 cm × 0.6 cm. An excisional biopsy was performed. Histologically, malignant spindle cells with hyperchromatic nuclei and nucleoli were seen to proliferate together with marked solar elastosis and desmoplasia. Mitotic figures were scattered. Immunohistochemically, the malignant cells were positive for pancytokeratin AE1/3, pancytokeratin WSS, cytokeratin (CK) 5/6, CK34BE12, CK14, vimentin, S100 protein, α-smooth muscle actin, p53, p63, and Ki-67 (labeling = 70%). They were negative for EMA, pancytokeratin CAM5.2, CK7, CK8, CK18, CK19, CK20, desmin, HMB45, Melan-A, SOX10, MITF, myoglobin, CEA, CA19-9 and CD34. The lesion recurred two times and showed neck lymph node metastasis during the following 2 years. The patient died of other diseases 3 years after the first presentation.

Key Words: Skin, Desmoplastic squamous cell carcinoma, Spindle cell squamous cell carcinoma, Histopathology

1. INTRODUCTION
Most of squamous cell carcinoma (SCC) of skin is ordinary SCC with keratinization and intercellular bridges. Desmoplastic SCC is a variant of cutaneous SCC, and it is very rare.[1–3] Breunonger et al.[2] reported 44 cases (7%) of desmoplastic SCC out of 594 cutaneous SCCs. They showed that the local recurrence, metastasis and mortality were higher in desmoplastic SCC than in ordinary cutaneous SCC. Velazquez et al.[3] reported 6 cases of cutaneous desmoplastic SCC, and found that they showed high recurrence, median age of 72 years, predominant occurrence of sun-exposed skin, high mortality, male predominance, predominant development of head and neck skin, high infiltrative features. Spindle cell SCC is also rare.[4–10] These two variants of skin SCC lack keratinization and intercellular bridge, thus causing diagnostic problems.[1–10] The author herein reports a case of desmoplastic SCC composed predominantly of spindle cells.

2. CASE REPORT
An 86-year-old Japanese woman consulted to our hospital because of a skin tumor of face. The tumor was reddish and measured 1.8 cm × 1.6 cm × 0.6 cm. An excisional biopsy was performed. Histologically, islands of atypical spindle cells with hyperchromatic nuclei and nucleoli were seen to proliferate in the dermis showing marked solar elastosis and desmoplasia (see Figure 1A). They were located in the marked solar elastosis (see Figure 1B) as well as in the severe desmoplasia (see Figure 1C). Mitotic figures and apoptotic bodies were scattered; the atypical cells were regarded as malignant. Although keratinization and intercellular bridges were not recognized, histological diagnosis was compatible with desmoplastic spindle cell SCC.

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An immunohistochemical study was performed with the use of Dako Envision method.\textsuperscript{11} Immunohistochemically, the malignant cells were positive for pan-cytokeratin AE1/3, pancytokeratin WSS, cytokeratin (CK) 5/6 (see Figure 2A), CK34BE12, CK14, vimentin (see Figure 2B), S100 protein (see Figure 2C), $\alpha$-smooth muscle actin (see Figure 2D), p53 (see Figure 2E), p63 (see Figure 2F), and Ki-67 (labeling = 70\%). They were negative for EMA, pan-cytokeratin CAM5.2, CK7, CK8, CK18, CK19, CK20, desmin, HMB45, Melan-A, MITF, SOX10, myoglobin, CEA, CA19-9 and CD34. No melanin pigment was detected by Mason-Fontana stain. A pathological diagnosis of definite desmoplastic spindle cell SCC was made.

The lesion recurred two times and showed neck lymph node metastasis during the following 2 years. The patient died of other diseases 3 years after the first presentation.

### 3. DISCUSSION

The tumor cells of the present case showed nuclear and cytological atypia, and invasive features. Mitotic figures and apoptotic bodies were scattered. These histological features are indicative of malignant neoplasm. Immunohistochemistry revealed p53 expression and very high Ki-67 (70\%) expression, thus confirming the diagnosis of malignant neoplasm. $P53$ seems to be mutated. In fact, the present tumor

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**Figure 1. Histological features**

A: The skin tumor showed marked desmoplasia and solar elastosis, in which atypical cells are seen. HE, ×4. B: The atypical carcinoma cells in the solar elastosis. C: The carcinoma cells in the dermis. HE, ×200. The cells are mostly spindle shaped and show marked desmoplasia. HE, ×200.

**Figure 2. Immunohistochemical features**

The carcinoma cells are positive for CD5/6 (A), vimentin (B), S100 protein (C), $\alpha$-smooth muscle antigen (D), p53 (E) and p63 (F). A-F: Immunostaining, ×200.
recurred and metastasized thereafter.

The present tumor expressed vimentin and various CKs. The present tumor is carcinoma. Atyypical fibroxanthoma and desmoplastic melanoma was excluded because the present tumor expressed various CKs and did not express HMB-45, Melan-A, SOX10, MITF. The specificity of S100 protein is low. The possibility of desmoplastic melanoma is quite unlikely. Because the present tumor expressed CKs, it is carcinoma. Because vimentin was positive, the present tumor has some features of mesenchymal differentiation. The HE histology was highly suggestive of carcinoma.

The present tumor showed no apparent differentiation. No keratinization or intercellular bridges were recognized. However, the present tumor is SCC because it was positive for p63 and high-molecular-weight cytokeratins including CK5/6 and CK 34BE12 (3–10). Differentiation into skin adnexae was not seen, and CEA and CA19-9 were negative; however the author thinks the possibility that the present case may be an undifferentiated adnexal tumors.

The present case showed extensive stromal desmoplastic reaction. This feature is compatible with desmoplastic SCC, which is very rare. Since the carcinoma cells of the present case were composed largely of spindle cells, the present case fulfills the criteria of spindle cell SCC. Namely, the present case showed dual features; desmoplastic SCC and spindle cell SCC.

Of interest, the present case showed immunoreactivity for α-smooth muscle antigen and S100 protein. Morgan et al. showed that α-smooth muscle antigen was positive in 2 out of 12 cases of spindle cell SCC. The author recently reported a vulvar spindle cell SCC with expression of S100 protein and α-smooth muscle antigen. The expression of S100 protein and α-smooth muscle antigen in this sarcomatoid carcinoma may imply that the tumor cells are so undifferentiated that the tumor cells produce these antigens. The author recently reported a case of sarcomatoid SCC of the uterine cervix and nasal cavity. The author recently reported a case of cutaneous basal cell carcinoma with dual differentiations into SCC and spindle cell SCC.

Breuninger et al. described that cutaneous desmoplastic SCC showed an aggressive course. The present case also recurred two times and lymph node metastasis was seen, being consistent with findings of Breuninger et al. However, much more studies with more cases remain to be done.

CONFLICTS OF INTEREST DISCLOSURE

The author has no conflict of interest.

REFERENCES


